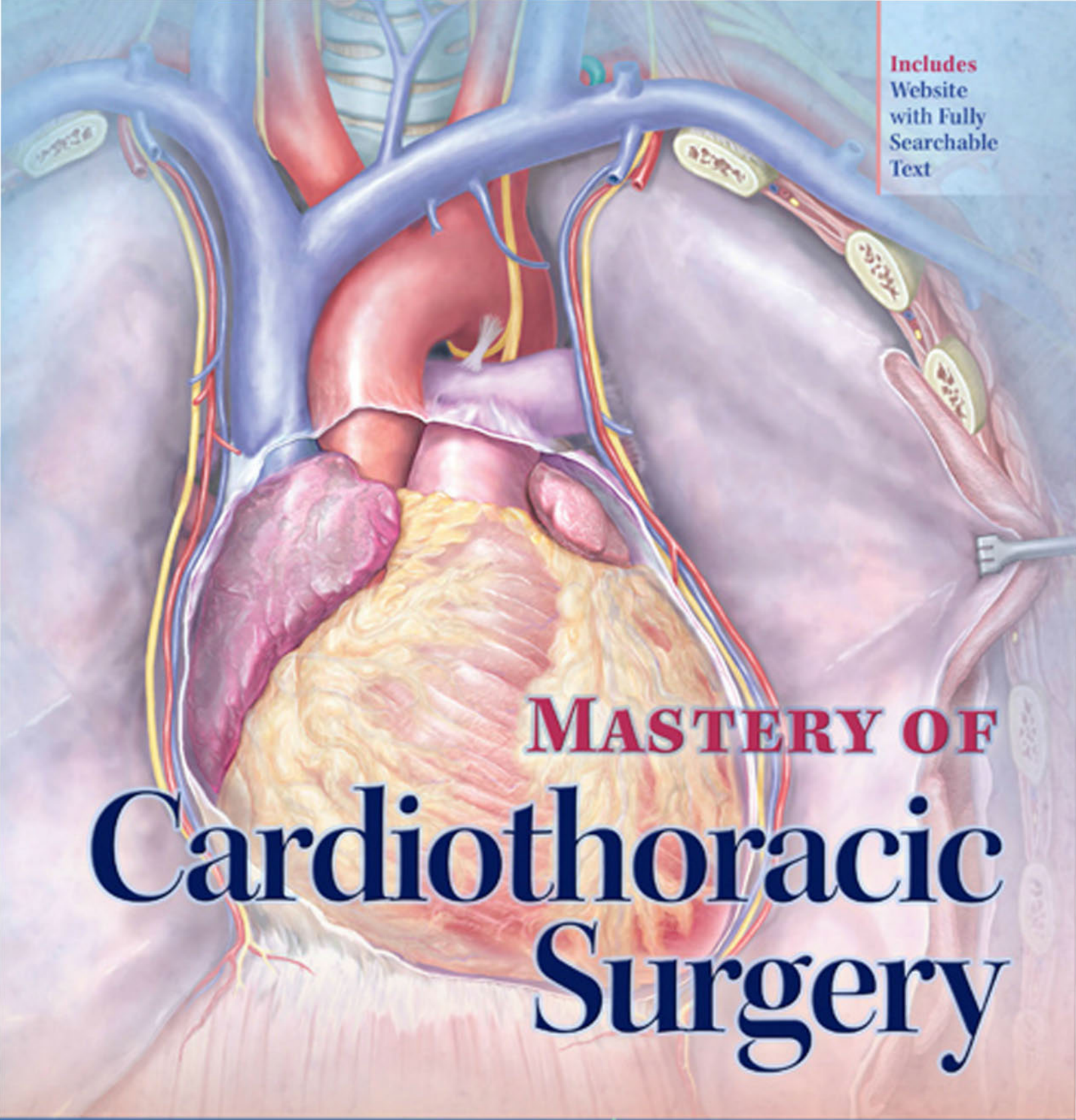


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Mastery of Cardiothoracic Surgery

THIRD EDITION

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Irving L. Kron
Thomas L. Spray

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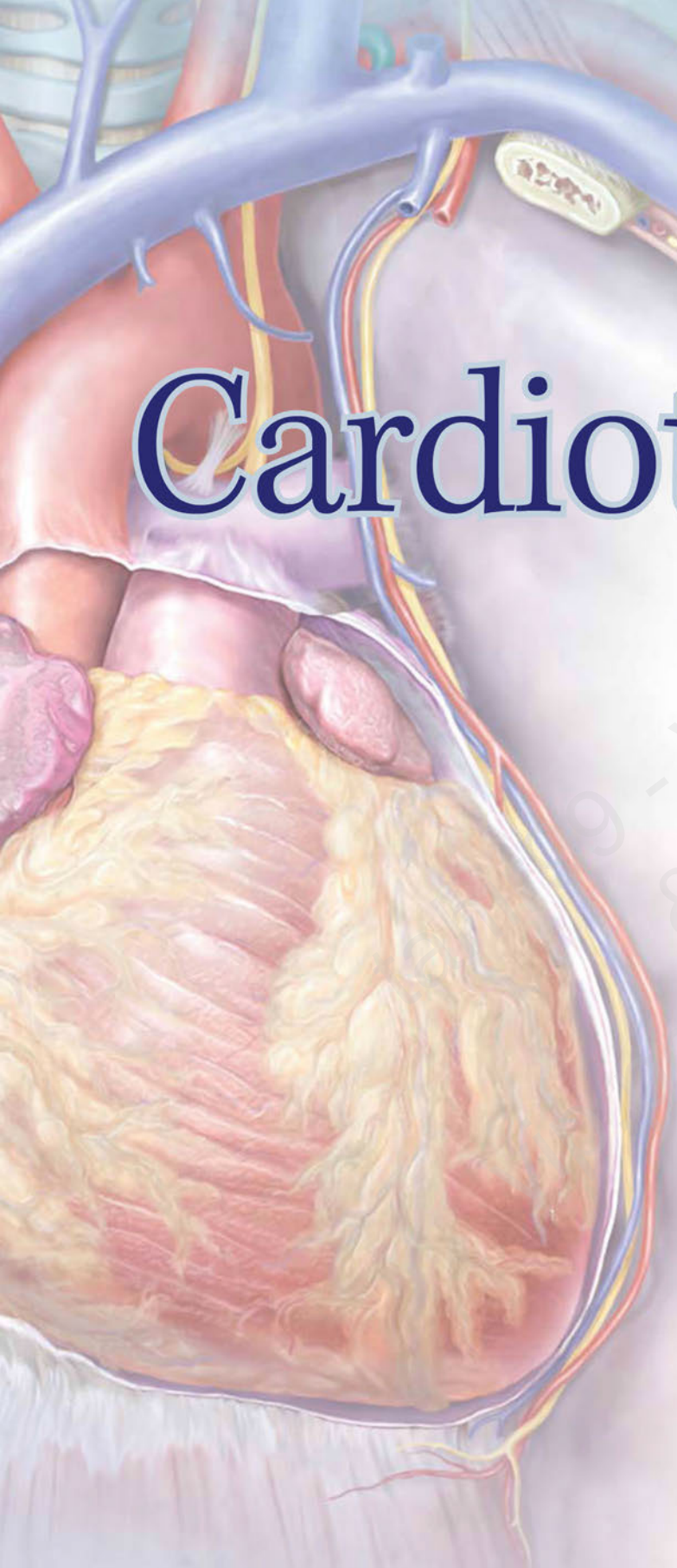
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To Lindy Lou: my partner, my friend, my strength

—LRK

I would very much like to dedicate this volume to my partners and our staff at The University of Virginia. Without their help and support, all the amazing things we are able to do for our patients would not occur. I would also like to very much thank the residents that I have had the honor of teaching over these years. Their dedication to the care of our patients makes my job the best in the world.

—ILK

To Diane.

—TLS

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Preface

Much has changed in the field of cardiothoracic surgery since the appearance of the second edition of *Mastery of Cardiothoracic Surgery*. Our field has become further fragmented (not necessarily a bad thing, by the way) with the creation of certification in congenital heart surgery following a period of additional training in a certified center. There no longer is a single textbook that covers the entire field, but we now have texts covering adult cardiac, general thoracic, and congenital cardiac. Aortic surgery has emerged as a specialty unto itself within cardiothoracic surgery and cardiac surgical education now is expected to incorporate endovascular training specifically related to the thoracic aorta, though the field of vascular surgery clearly overlaps in this area. There is now a primary certificate in vascular surgery offered in select centers, and completion of this program qualifies one to apply for entry into a residency in cardiothoracic surgery. Needless to say, those who come to cardiothoracic surgery by way of primary training in vascular surgery certainly bring a different perspective, though it remains too early to determine how these individuals will influence the specialty.

Despite the fragmentation with many programs having separate general thoracic and cardiac tracks, the specialty remains as one from the perspective of the American Board of Thoracic Surgery. The qualifying examination and the certifying examination as well as the examination given for maintenance of certification all include material from each of the specialty areas, a requirement that has engendered some controversy especially as it relates to Maintenance of Certification. It seems counterintuitive to expect a significant knowledge base relating to congenital heart surgery from a diplomate who has practiced, for example, general thoracic surgery for 10 years or more.

Bucking the trend, we have continued to include aspects of the entire specialty in *Mastery of Cardiothoracic Surgery*, with the assumption that the book will be used by trainees and practicing surgeons alike whether to review the aspects of a case prior to doing their first or as a refresher for a case that has not been done for a while. The editors have not wavered from their initial vision, and the vision of the originators of the *Mastery* series: that of having acknowledged masters of a given technique or procedure tell how they do it sharing their "tricks" and the subtleties learned from experience over the years.

The book is richly illustrated to show in detail how a procedure should be done. We have not set out to add another textbook to the canon; we have attempted to produce a volume that is complementary to the standard texts as well as to the literature. With full-text journal articles including those not yet published in hard copy just as close as the nearest computer, one has to question the relevance of a standard textbook. Yet this book provides what much of the literature does not, the "how to do it" piece. We are extremely proud of this edition as we have added a number of new authors as well as chapters, and have included several more recent procedures, including transcatheter aortic valve replacement. The editors are confident that users of this book will find each chapter of value either prior to doing a procedure or while reviewing the material prior to either the qualifying or Maintenance of Certification examinations.

Larry R. Kaiser, MD
Irving L. Kron, MD
Thomas L. Spray, MD

Acknowledgments

This book truly has been a labor of love because each of us has plenty to do in our day jobs. The task has not been easy but it is safe to say it would have been impossible without the incredible knowledge, diligence, and patience as well as support from Brendan Huffman who has been with us from the beginning of this edition. We also acknowledge the support and patience of his colleagues at Lippincott Williams & Wilkins and specifically Brian Brown who had the confidence in our abilities to see this through. It has been a pleasure working with them. We also acknowledge the outstanding quality of the work from the authors who contributed to this edition.

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Contents

I. GENERAL THORACIC SURGERY 1

Chapter 1: Endoscopy: Bronchoscopy and Esophagoscopy 2
Abbas E. Abbas

Chapter 2: Mediastinoscopy and Other Thoracic Staging Procedures 14
Michael Lanuti

Chapter 3: Thoracic Approaches 28
M. Blair Marshall

Chapter 4: Right-Sided Pulmonary Resections 40
Larry R. Kaiser

Chapter 5: Left-Sided Pulmonary Resections 53
Larry R. Kaiser

Chapter 6: Pneumonectomy 60
Robert J. Cerfolio and Ayesha S. Bryant

Chapter 7: Bronchoplastic Procedures 71
Anna Maria Ciccone, Federico Venuta, Camilla Vanni, and Erino A. Rendina

Chapter 8: Sublobar Pulmonary Resections 79
Aaron M. Cheng and Douglas E. Wood

Chapter 9: Tracheal Resection and Reconstruction 85
Joel D. Cooper

Chapter 10: Carinal Resection 100
Elizabeth A. David and Garrett L. Walsh

Chapter 11: Video-Assisted Thoracoscopic and Robotic-Assisted Pulmonary Resections 108
Eugenio Pompeo, Thomas A. D'Amico, and Abbas E. Abbas

Chapter 12: Management of Pneumothorax and Bullous Disease 116
Stephen D. Cassivi and Claude Deschamps

Chapter 13: Diagnostic Evaluation of Anterior Mediastinal Masses and Clinical and Surgical Approach to Thymic Tumors via Sternotomy 124
Usman Ahmad and Frank C. Detterbeck

Chapter 14: Transcervical Thymectomy 134
Larry R. Kaiser

Chapter 15: Resection of Posterior Mediastinal Lesions 139
Joseph B. Shrager

Chapter 16: Antireflux Surgery 147
John G. Hunter and Jeffrey S. Fronza

Chapter 17: Transhiatal Esophagectomy 157
Mark B. Orringer

Chapter 18: Thoracic Approaches to Esophagectomy 173
Shanda H. Blackmon, Wayne Hofstetter, and Stephen G. Swisher

Chapter 19: Minimally Invasive Esophageal Procedures 186
Nikiforos Ballian, Matthew J. Schuchert, and James D. Luketich

Chapter 20: Surgery for Achalasia and Other Motility Disorders 199
Farzaneh Banki and Tom R. DeMeester

Chapter 21: Esophageal Conduits and Palliative Procedures 213
Jenifer L. Marks and Ara A. Vaporciyan

Chapter 22: Excision of Esophageal Diverticula 222
Philip A. Rascoe and W. Roy Smythe

Chapter 23: Lung Transplantation 231
R. Duane Davis and Mathew G. Hartwig

Chapter 24: Thoracic Outlet Procedures 242
Robert W. Thompson and Valerie B. Emery

tahir99-VRG & vip.persianss.ir

Chapter 25: Chest Wall Resections 253
John R. Frederick and John C. Kucharczuk

Chapter 26: The Diaphragm 260
Mara B. Antonoff and Jonathan D'Cunha

Chapter 27: The Thoracic Duct and the Management of Chylothorax 274
Bradley M. Rodgers, Maxim Itkin, and John C. Kucharczuk

Chapter 28: Pericardial Procedures 289
Atasha Asmat and Nabil P. Rizk

Chapter 29: Surgical Management of Malignant Mesothelioma 296
Joseph S. Friedberg

Chapter 30: Mediastinal Lymphadenectomy 304
Boris Sepesi and David Rice

Chapter 31: Surgical Management of Empyema 313
Naveed Z. Alam and Raja M. Flores

Chapter 32: Resection of Superior Sulcus Tumors 318
Christine Lau and G. Alexander Patterson

Chapter 33: Thoracic Sympathectomy (Sympathicotomy) 328
Seth D. Force

Chapter 34: Operative Correction of Pectus Excavatum and Pectus Carinatum 333
Christopher S. Muratore, Thomas F. Tracy, and Larry R. Kaiser

Chapter 35: Repair of Paraesophageal Hernia 341
Arjun Pennathur, Matthew J. Schuchert, and James D. Luketich

II. ADULT CARDIAC SURGERY 355

A. General Considerations 356

Chapter 36: Cardiopulmonary Bypass 356
Ivan K. Crosby and Richard K. Zacour

Chapter 37: Myocardial Protection 370
Richard J. Shemin and Yousef M. Odeh

Chapter 38: Database 379
Ashok N. Babu, A. Laurie Shroyer, and Frederick L. Grover

Chapter 39: Prevention of Neurologic Injury after Coronary Artery Bypass 390
George J. Arnaoutakis and William A. Baumgartner

Chapter 40: Patient Safety 396
Thoralf M. Sundt

Chapter 41: Reoperative Cardiac Surgery—General Principles 402
Damien J. LaPar and Irving L. Kron

B. Acquired Valvular Heart Disease 407

Chapter 42: Mitral Valve Repair 407
Lawrence H. Cohn and R. Scott McClure

Chapter 43: Mitral Valve Repair—Robotic Minimally Invasive 419
Alan P. Kypson, L. Wiley Nifong, and W. Randolph Chitwood

Chapter 44: Mitral Valve Repair for Ischemic Mitral Regurgitation 431
Vincent Chan, Javier G. Castillo, and David H. Adams

Chapter 45: Percutaneous Mitral Valve Repair 440
D. Scott Lim and Gorav Ailawadi

Chapter 46: Tricuspid Valve 446
Gorav Ailawadi

Chapter 47: Pulmonary Valve Replacement 453
Damien J. LaPar, Carlos M. Mery, and James J. Gangemi

Chapter 48: Aortic Valve Replacement 458
David A. Fullerton

Chapter 49: Ross Procedure 471
John D. Oswalt

Chapter 50: Transcatheter Aortic Valve Replacement 479
Isaac George, G. Russell Reiss, and Mathew R. Williams

C. Coronary Artery Disease 493

Chapter 51: Coronary Artery Bypass Grafting using Cardiopulmonary Bypass 493
Ahmet Kilic, Chittor Sai-Sudakar, Juan Crestanello, and Robert S.D. Higgins

Chapter 52: Off-Pump Coronary Artery Bypass Surgery 507
W. Brent Keeling, Michael E. Halkos, and John D. Puskas

Chapter 53: Hybrid Coronary Artery Bypass Grafting Surgery 515
Daniel G. Cuadrado, Marzia Leacche, Ramanan Umakanthan, and John G. Byrne

D. Surgery for Heart Failure 519

Chapter 54: Left Ventricular Reconstruction 519
Lorenzo Menicanti and Serenella Castelvecchio

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Chapter 55: Ventricular Assist 532
Daniel P. Mulloy and John A. Kern

Chapter 56: Surgery for Complications of Myocardial Infarction 542
Philip W. Carrott, Timothy J. Gardner, and Irving L. Kron

E. Thoracic Aortic Disease 556

Chapter 57: Annuloaortic Ectasia 556
Tirone E. David

Chapter 58: Aortic Dissection 563
Nimesh D. Desai and Joseph E. Bavaria

Chapter 59: Descending Thoracic and Thoracoabdominal Aortic Aneurysms 575
Matthew L. Stone and John A. Kern

Chapter 60: Ascending Aortic Aneurysms 590
Matthew L. Stone and Irving L. Kron

Chapter 61: Arch Aneurysms 599
Joseph S. Coselli, Susan M. Trocciola, Kim I. de la Cruz, and Scott A. LeMaire

Chapter 62: Acute Traumatic Aortic Transection 611
Benjamin J. Pearce, Jay D. Pal, and John H. Calhoon

F. Transplantation 621

Chapter 63: Heart Transplantation 621
Christopher T. Salerno and Edward D. Verrier

Chapter 64: Heart–Lung Transplantation 629
Ahmad Y. Sheikh, Hari R. Mallidi, and Robert C. Robbins

G. Arrhythmias 638

Chapter 65: Surgical Treatment of Atrial Fibrillation 638
Sameh M. Said and Hartzell V. Schaff

Chapter 66: Minimally Invasive Treatment of Atrial Fibrillation 648
Gorav Ailawadi

H. Other Cardiac 653

Chapter 67: Surgical Treatment of Hypertrophic Cardiomyopathy 653
Hartzell V. Schaff

Chapter 68: Cardiac Tumors 664
Bo Yang, Himanshu J. Patel, Francis D. Pagani, and Richard L. Prager

Chapter 69: Acute Pulmonary Embolus 670
Salil G. Shah, Joshua N. Baker, and Thoralf M. Sundt

Chapter 70: Chronic Pulmonary Thromboembolism and Pulmonary Thromboendarterectomy 676
Michael M. Madani and Stuart W. Jamieson

III. CONGENITAL CARDIAC SURGERY 687

Chapter 71: Anatomy and Classification of Congenital Heart Disease 688
Paul M. Weinberg

Chapter 72: Echocardiographic Evaluation of Congenital Heart Disease 700
Meryl S. Cohen and Jack Rychik

Chapter 73: Cardiac Magnetic Resonance in Congenital Heart Disease 716
Mark A. Fogel and Yoav Dori

Chapter 74: Hemodynamic Assessment and Transcatheter Therapy for Congenital Heart Disease 746
Matthew J. Gillespie and Jonathan J. Rome

Chapter 75: Palliative Operations for Congenital Heart Disease 764
Hyde M. Russell and Carl L. Backer

Chapter 76: Anomalies of Systemic Venous Drainage 780
Sanjiv K. Gandhi and Ralph D. Siewers

Chapter 77: Patent Ductus Arteriosus 788
William M. DeCampli

Chapter 78: Vascular Rings, Slings, and Other Arch Anomalies 797
Christopher E. Mascio and Erle H. Austin

Chapter 79: Atrial Septal Defects 816
Richard D. Mainwaring, Peter Pastuszko, and John J. Lamberti

Chapter 80: Ventricular Septal Defects 829
Stephanie Fuller

Chapter 81: Aortopulmonary Window 838
Moritz C. Wyler von Ballmoos, Michael E. Barnes, and James S. Tweddell

Chapter 82: Coarctation of the Aorta 849
Karl F. Welke and Ross M. Ungerleider

tahir99-VRG & vip.persianss.ir

Chapter 83: Interrupted Aortic Arch Complex 861
Richard G. Ohye, Takaaki Suzuki, Eric J. Devaney, Jennifer C. Hirsch-Romano, and Edward L. Bove

Chapter 84: Left Ventricular Outflow Tract Obstruction and Valvar Aortic Stenosis 871
Tara Karamlou and Gordon A. Cohen

Chapter 85: Anomalies of the Sinuses of Valsalva and Aortico-Left Ventricular Tunnel 886
Luca A. Vricella and Duke E. Cameron

Chapter 86: Atrioventricular Canal Defects 892
Jeffrey P. Jacobs and Martin J. Elliott

Chapter 87: Truncus Arteriosus 910
Michael O. Murphy and Thomas L. Spray

Chapter 88: Double Outlet Ventricles 924
Kirk R. Kanter

Chapter 89: Transposition of the Great Arteries 937
Aaron W. Eckhauser and Thomas L. Spray

Chapter 90: Congenitally Corrected Transposition of the Great Arteries 954
Ergin Kocyildirim and Victor O. Morell

Chapter 91: Pulmonary Stenosis and Pulmonary Atresia with Intact Septum 963
Deborah J. Kozik and Mark D. Plunkett

Chapter 92: Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collaterals 979
V. Mohan Reddy and Frank L. Hanley

Chapter 93: Contemporary Surgical Therapy for Tetralogy of Fallot 990
Charles D. Fraser, Jr. and Muhammad S. Khan

Chapter 94: Cavopulmonary Shunts and the Hemi-Fontan Operation 1006
Marshall L. Jacobs and Robert D. Stewart

Chapter 95: Tricuspid Atresia/Single Ventricle and the Fontan Operation 1020
Stephanie Fuller and Pradeep Kaushik

Chapter 96: Hypoplastic Left Heart Syndrome 1030
Peter J. Gruber and Thomas L. Spray

Chapter 97: Anomalies of Pulmonary Venous Return 1041
Carlos M. Mery, James J. Gangemi, and Irving L. Kron

Chapter 98: Coronary Artery Anomalies in Children 1055
Julie A. Brothers and J. William Gaynor

Chapter 99: Pediatric Heart Transplantation 1074
Charles B. Huddleston

Chapter 100: Heart and Lung Transplantation in Children 1085
Stephanie Fuller and Thomas L. Spray

Chapter 101: Ebstein's Malformation of the Tricuspid Valve in Children 1098
Sitaram M. Emani and Pedro J. del Nido

Chapter 102: Mitral Valve Repair in Children 1105
Christian Brizard

Chapter 103: Aortic Valve Repair in Children 1118
Chawki el-Zein, Anastasios Polimenakos, and Michel N. Ilbawi

Chapter 104: Aortic Valve Replacement in Children 1129
John W. Brown, Parth Patel, Mark Rodefeld, and Mark W. Turrentine

Chapter 105: Arrhythmia Surgery and Fontan Conversion 1141
Constantine Mavroudis, Barbara J. Deal, and Carl L. Backer

Chapter 106: ECMO and Ventricular Assist Devices 1157
Andrew J. Lodge and Robert D. B. Jaquiss

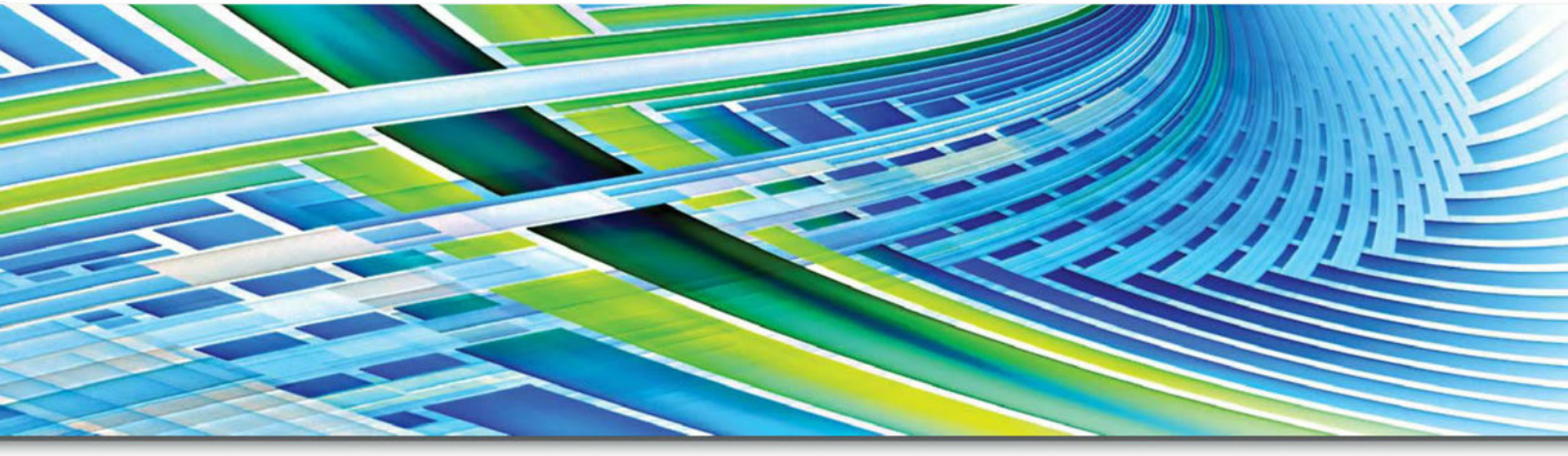
Chapter 107: Introduction and Overview of Pacemaker/ICD Therapy in Children 1168
Peter Aziz, J. William Gaynor, and V. Ramesh Iyer

Chapter 108: Conjoined Twins and Ectopia Cordis 1182
Joseph J. Amato

Index 1193

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General Thoracic Surgery I



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1 Endoscopy: Bronchoscopy and Esophagoscopy

Abbas E. Abbas

INTRODUCTION

Traditional endoscopy allows the evaluation of the endoluminal aspect of the tracheobronchial tree and the foregut. Newer advances such as endosonography and electromagnetic navigation have broadened the scope to include information outside the immediate lumen. The indications have evolved from a simple diagnostic procedure to one that may also be therapeutic or for staging of thoracic malignancies. The impressive technological advances in the field of fiberoptic endoscopy over the last decade now allow us to have access to areas previously considered "blind" by traditional endoscopy. For example, electromagnetic navigation can help to accurately navigate the bronchoscope to a peripheral lung nodule. Endoscopic ultrasound (EUS) allows us to visually assess a mediastinal lymph node in real time while simultaneously performing a transbronchial fine needle aspiration. Rigid endoscopy, whether esophageal or bronchial, remains as important for the thoracic surgeon to master today as it always has been. Although flexible endoscopy has taken away some of the indications for the rigid procedures, such as dilation and stenting, rigid endoscopy still plays a vital role for the thoracic surgeon and at times may be life-saving. The main advantage of rigid endoscopy is the ability to use larger instruments. This allows better biopsy specimens, debridement of tissues, and suctioning of fluids and debris. Rigid endoscopy is also necessary for certain stent placements in the airway. Although it is possible to perform all endoscopic procedures, including even rigid endoscopy, under local anesthesia or with simple sedation, advanced endoscopies are sometimes quite lengthy and safer to perform under general anesthesia. The choice of analgesia and anesthesia should be determined on an individual basis and according to pathology or operator preference.

ANATOMY

It is important for the endoscopist to have a clear understanding of both the endoluminal and the external relationship to overlying mediastinal structures. This is especially important for EUS and this is also where surgeons have a distinct advantage.

Pharynx and Larynx

Aerodigestive anatomy begins with the entry of air into the body via the nasopharynx and the oropharynx. Entry may be gained to the upper aerodigestive system through either the nose or the mouth. At the nares, there is a 3- to 4-mm opening below the lower turbinates, with direct access to the posterior pharynx. This entry is just medial to the pharyngeal tonsil and passes posterior to the soft palate. Entry through the mouth reaches the posterior pharynx beyond the soft palate. As the posterior portion of the tongue is passed, one reaches the vallecula. At this level, the epiglottis can be seen anterior to the aerodigestive tract. Immediately posterior to this structure, the vocal cords can be seen easily. They are attached anteriorly and move apart posteriorly. The vocal cords are bounded by the aryepiglottic fold and posteriorly by the corniculate tubercles. Just posterior to the tubercles is the entrance to the esophagus. Laterally, on both sides of the esophagus, are the pyriform recesses, which are often mistaken for the true esophagus but are false passages with a depth of 2 to 3 cm.

Bronchoscopic Anatomy

A complete bronchoscopic examination will inspect the divisions of the tracheobronchial tree to the level of the segmental and subsegmental orifices.

Trachea

The trachea begins inferior to the glottis, at the level of the sixth cervical vertebra. Distally, it divides into the left and right mainstem bronchi at the carina. The location of the carina is usually at the level of the fifth thoracic vertebra, which is in line with the angle of Louis. This may ascend or descend up to two vertebrae with breathing. The internal diameter is about 16 mm while the length of the trachea varies according to the height of the individual and is about 12 cm. Surgically, the trachea is divided into two parts; the cervical trachea from the glottis to the manubrium and the thoracic trachea from the manubrium to the carina (Fig. 1.1).

The wall of the trachea is formed of fibromuscular tissue which is reinforced anteriorly by 16 to 20 incomplete cartilaginous rings. These rings form the anterior two-thirds of the tracheal circumference. The posterior wall is a flat fibromuscular membrane, which is immediately anterior to the esophagus.

The thoracic trachea is crossed anteriorly by the left innominate vein. Immediately inferior to the vein, it lies behind and to the right of the arch of the aorta and is straddled by the innominate artery on the right and the left common carotid artery on the left. Laterally on the right, the superior vena cava runs longitudinally adjacent to the trachea. Paratracheal and subcarinal lymph nodes lie in direct relationship to the trachea.

Mainstem Bronchi

The right mainstem bronchus (RMSB) arises from the carina at an angle of 30 degrees while the left mainstem bronchus (LMSB) has a sharper turn closer to 45 degrees. The diameter of the RMSB is also larger than the LMSB (16 vs. 13 mm). These are the reasons for the difficulty in intubating the left bronchus compared with the right, and for why inhaled foreign bodies are found more often on the right side.

The RMSB is usually about 1- to 2.5-cm long and ends after giving off the right upper lobe (RUL) branch. Its continuation then becomes the bronchus intermedius

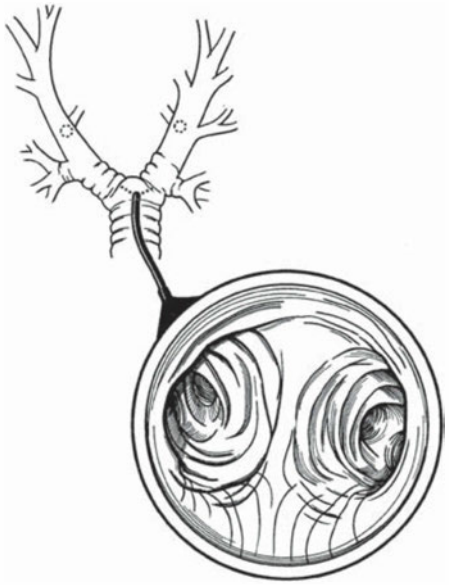


Fig. 1.1. Bronchoscopic view of the trachea and carina.

(BI), which is also about 2-cm long. The RUL bronchus (Fig. 1.2) comes off perpendicularly to the RMSB and gives off three segmental branches. Its location can vary in different individuals and may even arise as a direct branch of the trachea. It is saddled by the azygos vein laterally and lies superior then posterior to the right

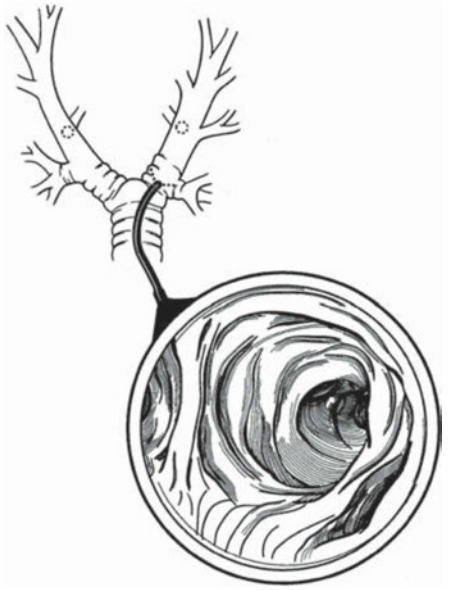


Fig. 1.2. The view down the proximal right mainstem bronchus showing the origin of the right upper lobe bronchus. The close proximity of the upper lobe takeoff to the carina and the angle of its origin provide an anatomic landmark that can be used to determine orientation during bronchoscopy.

pulmonary artery. The BI then terminates as a bifurcation to the middle and lower lobe bronchi (Fig. 1.3). The right middle lobe (RML) bronchus usually arises directly opposite to the superior segmental branch of the right lower lobe (RLL), but may be just higher or lower, a relationship that is important to determine when performing lobectomy.

The LMSB (Fig. 1.4) is longer than the right and averages 5 cm. It passes underneath the aortic arch and crosses anterior to the esophagus, the thoracic duct, and the descending aorta. It lies posterior and then inferior to the left pulmonary artery. The bifurcation of the LMSB into the left upper lobe and the left lower lobe bronchi is often called the secondary carina.

Bronchopulmonary Segmentation

Primary branches of the lobar bronchi are called segmental bronchi and each leads to a structurally separate unit called the bronchopulmonary segment (Table 1.1 and Fig. 1.5). These number 10 in the right lung and 8 in the left. The RUL bronchus divides into three segmental bronchi, namely, the apical, posterior, and anterior segments. The RML bronchus divides into the medial and lateral segments. The RLL bronchus divides into the superior segment and four basal segments, including the anterior, medial, lateral, and posterior basal segments. The left upper lobe bronchus divides into an upper division and a lower division. The upper division, or upper lobe

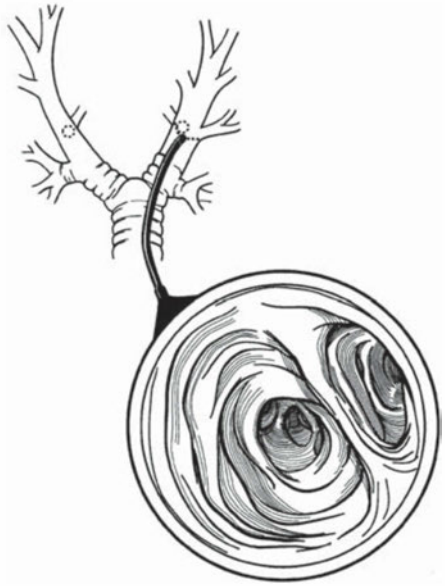


Fig. 1.3. The view down the bronchus intermedius showing the middle lobe and the lower lobe orifice.

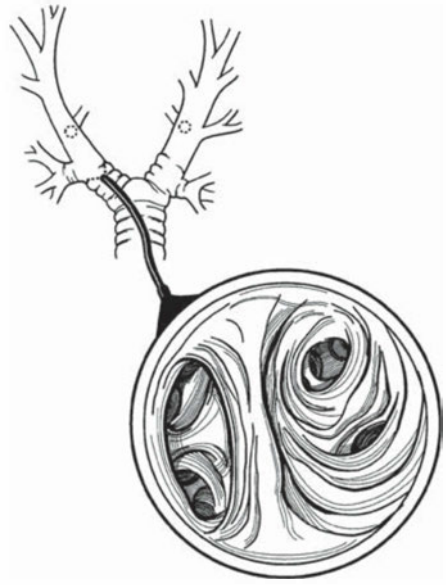


Fig. 1.4. The left mainstem bronchus is significantly longer than the right and terminates at the bifurcation (or minor carina) into the upper lobe bronchus (composed of the upper lobe proper and lingular orifice) and the lower lobe bronchus.

proper, gives rise to the apicoposterior and anterior segments. The lower division, or lingual, gives rise to the superior and inferior segmental bronchi. As on the right side, the first segmental branch of the LLL bronchus is a large superior segmental bronchus. This is followed by the basal segments, which number only 3 on the left side; anterior basal segment, the lateral basal segment, and the posterior basal segment.

The Esophagus

The esophagus, or gullet, is a long muscular tube that connects the pharynx to the stomach. The length of the esophagus in adults is about 25 cm and, like the trachea varies with the length of the individual. It extends from the cricopharyngeus muscle at the level of the cricoid cartilage and the sixth cervical vertebra to the cardiac orifice of the stomach at the level of the 11th thoracic vertebra.

Along its course, the esophagus is in the midline, anterior to the spine except for two leftward curves. At its origin, the esophagus curves gently to the left down to the level of fifth thoracic vertebra where it returns to the midline. The second is a sharper curve to the left and is formed as the esophagus traverses the diaphragm at the hiatus where it turns to cross the descending thoracic aorta and enter into the stomach.

Table 1.1 Segmental Airway Anatomy

Right

Upper lobe: apical (RB1), posterior (RB2), and anterior (RB3)

Middle lobe: lateral (RB4) and medial (RB5)

Lower lobe: superior segment (RB6), medial basal (RB7), anterior basal (RB8), lateral basal (RB9), and posterior basal (RB10)

Left

Upper lobe

The upper lobe proper bronchus: apicoposterior (LB1+2) and the anterior (LB3)

The lingular bronchus: superior (LB4) and inferior (LB5)

Lower lobe: superior segment (LB6), anterior basal (LB8), lateral basal (LB9), and posterior basal (LB10)

There is no medial basal segment (LB7) in the left lung.

The esophagus also follows the convex and then concave curvatures of the cervical and thoracic spine.

In endoscopy, the parts of the esophagus are described according to the distance from the upper incisor teeth. The upper esophageal sphincter (UES), or cricopharyngeus, is at 15 cm whereas the lower esophageal sphincter (LES) is at 40 cm. It is important to remember that these levels can vary depending on the height of the patient and also on the timing when measurements were taken. Measurements made while advancing the endoscope tend to be longer than those made during retraction, a phenomenon commonly seen with a sliding hiatal hernia.

It is also usually divided into cervical, thoracic, and abdominal portions. The cervical portion extends from the cricopharyngeus to the suprasternal notch. The thoracic portion extends from the suprasternal notch to the diaphragm. The abdominal portion extends from the diaphragm to the cardiac portion of the stomach.

The esophagus has three natural constrictions along its course. It is important for the endoscopist to be aware of these locations, especially when searching for impacted foreign bodies or when deciding on the amount of safe dilation for pathologic strictures at those sites. The first constriction is at 15 cm from the incisors, right at the UES and is the narrowest portion of the esophagus. The second is at 23 cm, where the esophagus lies behind the aortic arch and LMSB. The third constriction is at 40 cm, where it traverses the diaphragm at the LES.

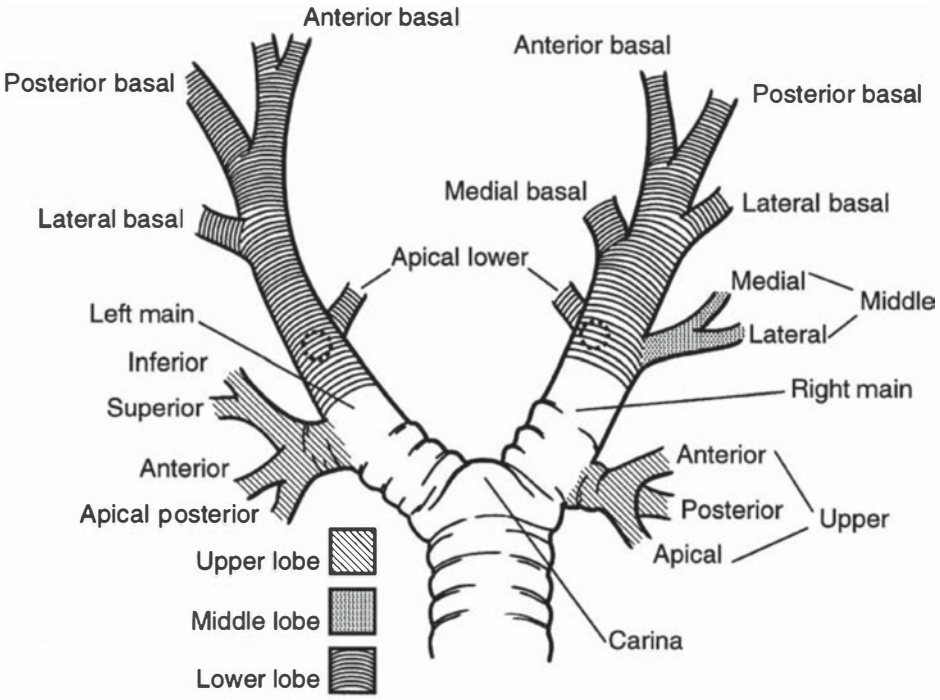


Fig. 1.5. Segmental anatomy of the tracheobronchial tree in an orientation akin to that observed when performing bronchoscopy.

Endoscopic Relationships of the Esophagus

It is important to understand the relationships of the esophagus especially when performing EUS or before contemplating certain maneuvers such as dilation and stenting. The cervical esophagus lies posterior to the membranous trachea. It overlies the bodies of sixth, seventh, and eighth cervical vertebra. The bilateral carotid sheath and lateral lobes of thyroid gland are immediately lateral to the cervical esophagus.

The thoracic esophagus lies in the superior mediastinum between the trachea and vertebral column. It then enters the posterior mediastinum by passing behind the aortic arch. Laterally, it is related to the aorta and left subclavian artery on the left while on the right side, it is related to the azygos vein. Anteriorly, the esophagus is related to the trachea, right pulmonary artery, left bronchus, pericardium with left atrium, and diaphragm. Posteriorly, the esophagus is related to the vertebral column, descending aorta, and diaphragm.

The abdominal esophagus passes through the right crus of the diaphragm to lie posterior to the left lobe of the liver.

EQUIPMENT

A modern fiberoptic endoscope (Fig. 1.6) basically consists of a handle and a fiberoptic bundle that connects to a light source. There are also one or more working channels for suctioning and for the passage of instruments or injection of liquids. The handle contains one or more knobs to control the position of the tip of the endoscope. Most current bronchoscopes also have a camera adaptor allowing connection to a high-definition monitor (Fig. 1.7).

Bronchoscopy

In addition to performing the visual inspection, there are various options for obtaining material from a bronchoscopy. Brushes can be used to obtain cytology material. The brush is retracted into a sheath after

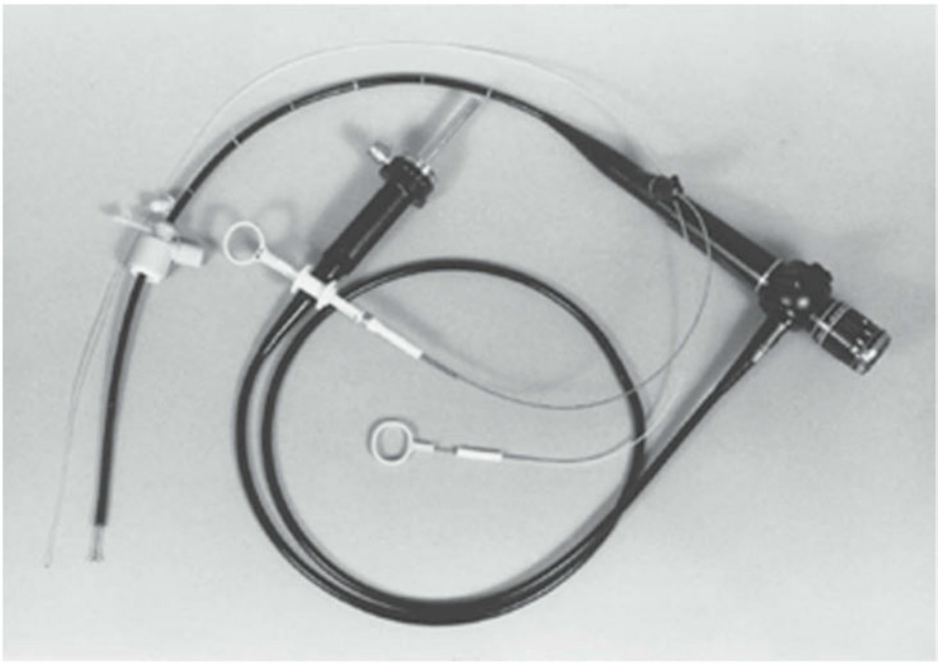


Fig. 1.6. Flexible bronchoscope with standard accessories. The endotracheal tube ventilating adaptor, the biopsy forceps, and a disposable brush are shown.



Fig. 1.7. Flexible fiberoptic bronchoscope showing camera adapter, suction port, and biopsy or irrigation port.

brushing to protect the specimen. A biopsy forceps is used to obtain material either under direct visualization from the proximal airway or from the peripheral lung parenchyma under fluoroscopic guidance. A transbronchial needle may be used to obtain extrabronchial samples with or without ultrasound guidance. A triple lumen catheter with a double-sheathed telescoping brush enables sampling of lower respiratory tract secretions for microbiologic examination without contamination from the inner channel of the bronchoscope. This is especially useful in the setting of the possibility of lobar pneumonia. When performing lavage for either cytology or microbiology, irrigation is administered through the working channel and a trap is connected to the suction port.

The endoscope diameter ranges from 3 to 6 mm, with a working channel ranging from 1.2 to 3 mm. For routine diagnostic purposes, a standard 3.5-mm endoscope is usually sufficient. If one plans more extensive procedures, a larger endoscope may be necessary. It is advisable to have available a range of sizes to accommodate, the planned procedure. Most bronchoscopic procedures require an endotracheal tube of at least 8 mm. A bronchoscopy adapter for the endotracheal tube allows simultaneous ventilation during the procedure.

Rigid Bronchoscopy

Except for minor modifications, today's rigid bronchoscope (Fig. 1.8) is similar to that developed by Chevalier Jackson, the father of American bronchoesophagology. It is a stainless steel straight and hollow tube that is available in a range of sizes. Rigid bronchoscope external diameters can range up to 14 mm, and the length varies according to the age and size of the patient and have a beveled tip used to elevate the epiglottis and thus expose the vocal cords to facilitate passage. The proximal end of the bronchoscope consists of a central opening, which has a removable eyepiece that can also allow placement of a 4- or 5-mm telescope. This telescope usually has a 0-degree viewing angle to enable wide end-on visualization, while a 90-degree viewing angle may be useful to visualize the right upper lobe bronchus or the superior segmental orifices of the lower lobe.

Slit-like openings in the distal end allow ventilation to the contralateral lung.

Most rigid bronchoscopes also contain a light source at the distal end. The side ports may be used for flexible suction and laser fiber or flexible biopsy forceps.

Esophagoscopy

Rigid esophagoscopy has been an integral part of the practice of thoracic surgery since it was first described in 1868 by Kussmaul. Not long after, Jackson developed the ability to end-illuminate the rigid esophagoscope and since then the instrument has undergone very few modifications and has been used extensively for both diagnostic and therapeutic purposes. The rigid esophagoscope (Figs. 1.9 and 1.10) looks much like a rigid bronchoscope but is flatter and has a more bulbous tip. It is also available in adult and pediatric sizes and can vary in length from 20 to 50 cm. The advantages of rigid esophagoscopy include excellent visualization of the upper esophageal and



Fig. 1.8. Rigid bronchoscope set in two different sizes (with permission from Karlstorz, Inc.). There are two side ports, one for ventilation and a second port for placement of flexible tools. The light source is placed within the wall of the scope. The adapter shown at the bottom can be adjusted to provide a seal around different sized inserts. At the top are shown three optical forceps that incorporate the telescope and allow direct visualization while they are being used.

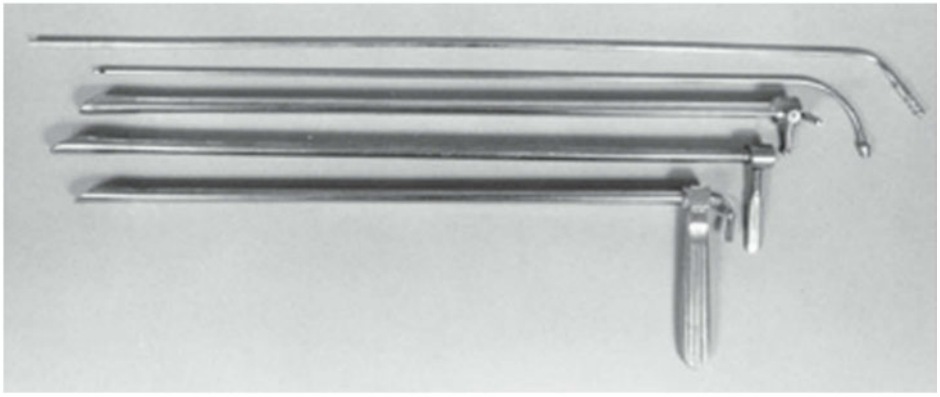


Fig. 1.9. Standard Jackson esophagoscopes. Each varies slightly in the shape at the end, but in general they are flattened and shaped like the esophagus. Suctions have either an end hole or two side holes.



Fig. 1.10. Dilating esophagoscope. This esophagoscope is round and will accept up to a 36F dilator. Several varieties of Jackson rigid dilators are shown coming out of the end of the scope.

hypopharyngeal region in addition to allowing the ability to perform many therapeutic interventions such as foreign body removal and tumor resection. It is especially useful for dilating a stricture with a bougie under direct visualization.

First introduced by Basil Hirschowitz in 1957, flexible esophagogastroscopy, however, quickly became the preferred approach to evaluate the esophagus and stomach. Its flexibility, ease of passage, and superior visualization, in addition to the fact that it is a much easier technique to master, have made it the currently preferred instrument for most diagnostic and therapeutic esophageal procedures.

The fiberoptic esophagoscopy, much like the bronchoscope, also comes in different lengths and diameters. Accessories mirror those that are available for bronchoscopy, but are, of course, longer.

DILATORS AND STENTS

Simple repeated esophageal dilation is the oldest technique for palliation of dysphagia but is rarely associated with long-term relief. Usually, dysphagia recurs after only a few days or weeks. Dilation may be accomplished by using weighted rubber Maloney bougies, wire-guided polyvinyl Savary dilators, or hydrostatic dilating balloons. Maloney dilators (Fig. 1.11) are passed blindly into the esophagus and are quite effective in experienced hands. Savary dilators (Fig. 1.11) are passed over a stiff guide wire, which is inserted under direct vision through the stricture. Successive dilators are passed up to the desired level. Both types of dilators come in diameters ranging from 2 to 20 mm (6F to 60F) and both mm and French size are usually clearly marked on the instrument. They are also marked in centimeters to determine the depth of insertion. These measurements are shown both from the tip (American) and from the widest part (European) of the dilator. Finally, hydrostatic balloon dilators (Fig. 1.12) have been developed that can be passed through the working channel of the endoscope and deployed under vision. They have the same range of diameters available as the rigid dilators. Theoretically, the radial force of a dilating balloon is thought to be safer than the longitudinal force of bougies.

The risk of perforation is higher in very tight lesions and in patients who have had previous radiation therapy. The procedure usually needs to be repeated every few days and is seldom sufficient in producing long-term relief.

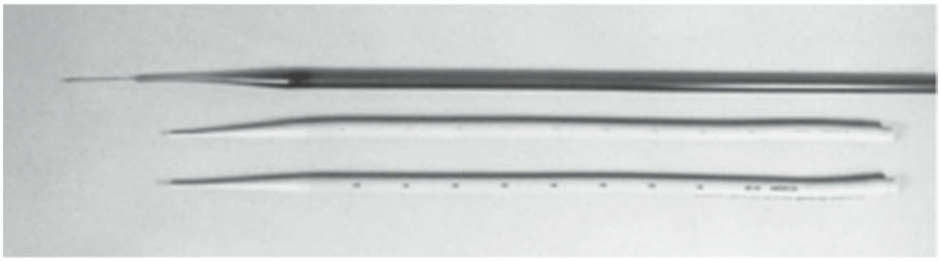


Fig. 1.11. Flexible dilators. The Savary dilator (*top*) is a wire-guided dilator. The Maloney dilators are marked in centimeters to gauge the depth of insertion in both American and European systems (see text).

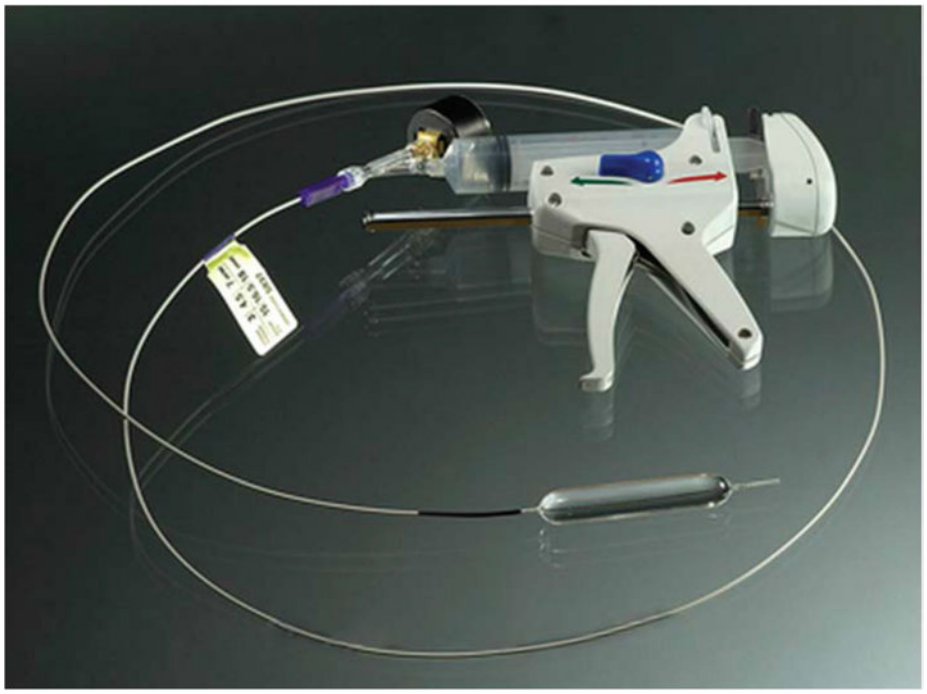


Fig. 1.12. Controlled radial expansion-fixed wire balloon dilator and inflation device. **(b)** Inflation device. (Copyright © 2006 Boston Scientific Corporation or its affiliates. All Rights Reserved.)

We prefer using either wire-guided Savary bougies under fluoroscopic guidance or the hydrostatic balloons under direct endoscopic visualization. However, with the availability of self-expanding metal and plastic stents, dilation is rarely used as the sole treatment for dysphagia.

Prior to the advent of self-expanding metal stents (SEMS), the main methods for maintaining the lumen of the esophagus open was in the form of fixed-diameter plastic stents, for example, the Celestin or Wilson-Cook stents. These were inserted either using rigid esophagoscopy (push technique) or by pulling through a gastrostomy (pull technique). These have largely been abandoned in favor of the easier to place self-expandable stents.

Self-Expandable Metal Stents

SEMS were introduced in 1990 and have now replaced the conventional fixed-diameter plastic prostheses. Numerous studies have shown them to be associated with fewer complications and better efficacy (85% to 100%) than the fixed-diameter plastic stents. In addition, they are much easier to insert, and usually do not require a predilation unless the diameter of the lumen is narrower than that of the delivery system (6 to 11 mm). SEMS (Figs. 1.13A and 1.14A) have a thin wall and a large endoluminal diameter (16 to 23 mm), which ensures effective relief of dysphagia. These stents become embedded into the mucosa and are difficult to remove after several weeks. A silicone or polyurethane coating (Fig. 1.13B) allows stents to resist tumor ingrowth but makes them more prone to migration. They all come preloaded on a small-diameter delivery system (Fig. 1.14B), and when deployed, they exert outward radial force to expand to their full diameter. The radial force of the stent maintains lumen patency, and tumor compression holds the stent in place (Fig. 1.13C). They are all placed in a similar manner, usually over a wire with endoscopic and/or fluoroscopic guidance. They are then deployed by a self-expanding mechanism. They may be placed under either general anesthesia or moderate sedation.

Early complications occur in approximately 10% of patients and include perforation, bleeding, pain, stent migration, and airway obstruction. Perforation is related to the use of excessive force during dilation or passage of the delivery system. Airway obstruction may occur especially in the presence of large cervical or mediastinal adenopathy, which may cause external compression of the trachea after expansion of the stent. This may require emergent stent removal or even placement of a second airway stent.

Late complications include recurrence of dysphagia that may occur due to tumor overgrowth or ingrowth (10% to 15%), granulation tissue hyperplasia or, stent migration (1% to 15%). Food impaction, troublesome gastroesophageal reflux, esophagorespiratory fistula, and bleeding have also been reported with the use of these stents. Tracheoesophageal fistula can result from stent erosion through the esophagus and into the respiratory tree. Massive bleeding may result from a similar process into the aorta.

When tumor ingrowth or overgrowth does occur, it can be managed with several techniques including placement of a second stent within the old one or using a variety of ablation methods including laser, argon plasma coagulation, endoscopic debridement, and photodynamic therapy.

PROCEDURE

Anesthesia

As mentioned above, the choice of whether to use general anesthesia or sedation with local anesthesia depends on the predicted complexity and duration of the procedure. It is safer to perform a lengthy and complex endoscopic procedure under general anesthesia with the control of the airway and the ability to use muscle relaxants.

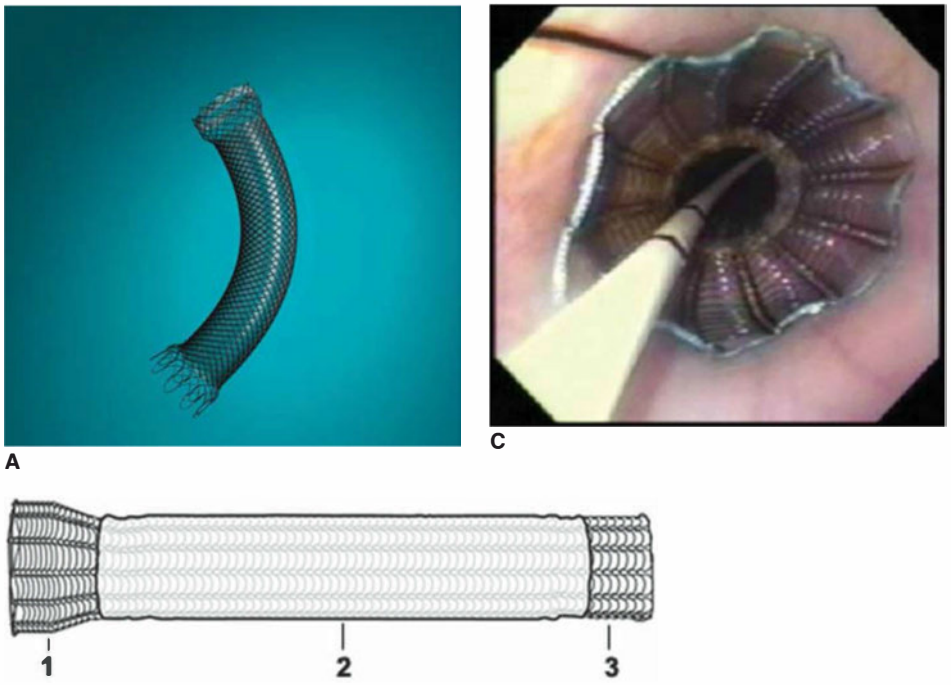


Fig. 1.13. (**A**) Partially covered self-expanding metal stent (Wallflex) (with permission from Boston Scientific, Inc.). (**B**) Diagram of stent construction: (1) large proximal flare, (2) polyurethane covering, (3) flexible knitted-loop design (with permission from Boston Scientific, Inc.). (**C**) Endoscopic view of deployed intraesophageal stent (notice the proximal uncovered flange).

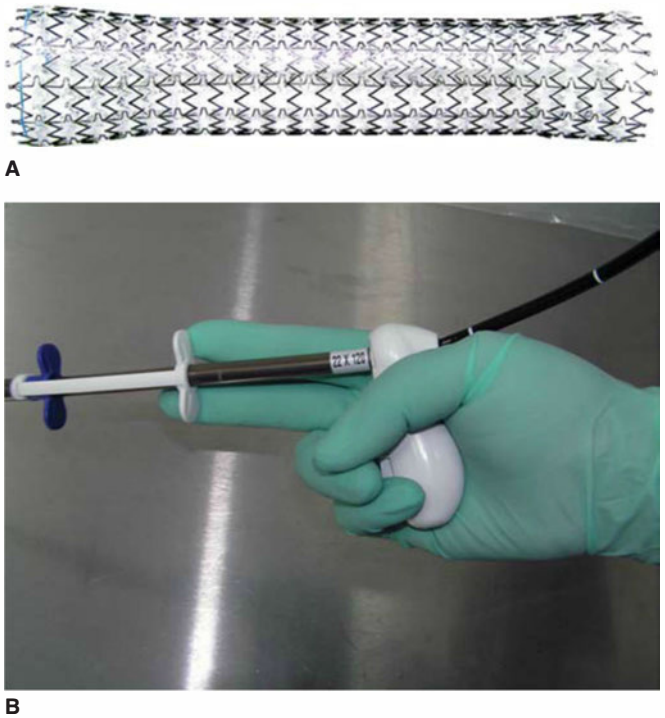


Fig. 1.14. (**A**) Fully covered self-expanding metal stent (Alveolus Alimaxx) with hybrid metal-silicone structure (with permission from Alveolus, Inc.) (**B**) Example of deployment device for a self-expanding metal stent (Alveolus Alimaxx) (with permission from Alveolus, Inc.)

Patients undergoing conscious sedation are usually given a combination of a sedative such as midazolam and an analgesic such as meperidine. These agents are given in increments while monitoring the patient carefully. Monitoring includes an electrocardiogram tracing, blood pressure, heart rate, and a pulse oximeter. Supplemental oxygen before, during, and after the procedure is administered.

The local anesthetic administered depends on the procedure. For esophagoscopy, viscous lidocaine gargle or tetracaine spray may be used. For bronchoscopy, lidocaine is sprayed to the pharynx, the vocal cords, the trachea, and throughout the airway, at the orifice of each bronchus. One must be careful not to exceed the maximum dose of the local anesthetic.

For general anesthesia, it is best to use short-acting agents such as propofol and inhalational agents. Muscular relaxants such as succinylcholine may be used when necessary, such as for rigid bronchoscopy.

BRONCHOSCOPY

Flexible Bronchoscopy

The fiberoptic flexible bronchoscope can be introduced into the airway through different routes. There are distinct advantages and disadvantages to each of these options, and it is important to be both aware and familiar with each. Flexible bronchoscopy may be performed under conscious sedation using either the transnasal or transoral route. If a patient has a tracheostomy stoma, this can also provide a route for bronchoscopy. When general anesthesia is employed, it is possible to perform the procedure through an endotracheal tube, but this allows examination only of the airway distal to the tube, usually just above the carina. An excellent alternative is to perform the procedure through a laryngeal mask airway (LMA), thus allowing full exposure of the glottis and entire airway distally.

When performed under conscious sedation, the operator usually stands behind the patient or on the patient's right side. For the transoral approach, a bite block is placed around the bronchoscope which is then placed between the incisors after the scope is advanced into the trachea. The transnasal approach is a more direct approach to the larynx while the oral route requires navigating the scope around the tongue and the epiglottis to identify the glottis. Both approaches allow complete visualization of the airway.

After instilling 1 to 2 cm^3 of local anesthetic into the larynx, the scope is passed through the vocal cords. Local anesthetic is sprayed at every lobar bronchial orifice before traversing it.

Procedure

Careful inspection of the airway is performed on the first pass before any possible distortion by endoscopic scope trauma. The examination should be done on the contralateral side of the suspected pathology to ensure that every part of the airway was examined in a comprehensive and sequential manner. After the initial inspection, any abnormalities that are noted should be addressed in a manner that will confirm their nature. Secretions should be collected in a trap, taking care to prevent contamination by the oropharynx by only attaching the trap once the bronchoscope is in the airway. The choice of the procedure depends upon the circumstances involved. For an endobronchial lesion, both brushing and biopsy should be performed. For a peripheral lung lesion and for interstitial lung disease, a transbronchial lung biopsy is performed. Fluoroscopy is used to advance the biopsy forceps toward the abnormality and to minimize the risk of pneumothorax. The forceps is introduced into the segment to about 3 cm from the ribs. The forceps is opened and advanced while patient is inhaling until it wedges. The forceps is closed and pulled back at the end of expiration. If the patient develops pleuritic pain during the procedure, the forceps is withdrawn slightly until there is no pain. About six biopsy specimens should be obtained to ensure an adequate sample, which should include respiratory epithelium and alveolar wall. The specimens should be sent for routine histology, microbiology, and immunofluorescent staining. The Wang needle is used for transbronchial biopsy (Fig. 1.15). This is usually done at the carina to evaluate the subcarinal lymph nodes. The needle is contained within a sheath that is passed through the working channel of the bronchoscope. It is advanced out of the sheath and through the wall of the airway into the desired extramural mass or lymph node. After several passes, the material obtained is placed on slides for direct cytologic interpretation and into cell fixative for cell block. Some of the materials may also be sent for culture. Bronchoalveolar lavage is done for diffuse lung disease. Washings or lavage is best performed after brushing. The scope is wedged into a segmental or subsegmental bronchus, and about 100 ml of saline is instilled in 20 ml aliquots. At least 50% must be retrieved in a trap for a successful BAL. The fluid is sent for both cytology and microbiology. After completion of the procedure, the entire airway is reinspected for bleeding, which can be easily controlled by instillation of cold saline or small amounts of a 1:10,000 solution of epinephrine. Blood pressure and pulse should be carefully monitored during this procedure because drugs are rapidly absorbed from the respiratory epithelium. When a transbronchial biopsy is performed, a chest radiograph must be obtained following the procedure to rule out a pneumothorax.

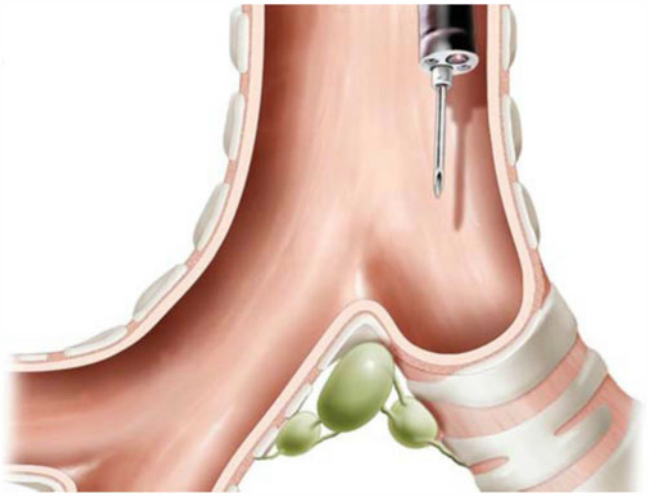


Fig. 1.15. Transbronchial needle for biopsy (with permission Olympus, Inc.).

Rigid Bronchoscopy

Anesthesia and Ventilation

General anesthesia is preferred for both comfort and safety. Because the anesthesiologist gives up control of the airway to the surgeon, it is essential to have clear communication between the two throughout the entire procedure. Induction of anesthesia can be done with inhalational agents but once induced, it is preferable to continue to maintain the depth of anesthesia with a total intravenous technique. A muscle relaxant is usually used for the initial intubation with the rigid bronchoscope. Ventilation during the procedure can be through the side port, using breaths delivered by a combination of spontaneous breathing, hand-assistance, and ventilator delivery. It is important to ensure a good seal at the mouth as evidenced by rise of the chest with breaths and end-tidal carbon dioxide monitoring. Jet ventilation is another option in which oxygen and air mixture is delivered through a small side port using a handheld injector. The advantage of jet ventilation is the ability to provide adequate oxygenation with minimal pressure.

Position

Positioning the patient correctly is critical for successful rigid bronchoscopy. The patient is placed supine with extension of the neck using a rolled sheet or air bag (Fig. 1.16). This allows the trachea to be as anterior as possible and to also be in direct alignment with the larynx (Fig. 1.16).

A guard should be placed over the maxillary front teeth. The videoscope is advanced into the bronchoscope and the bronchoscopist looks either directly down the bronchoscope or at the monitor while the bronchoscope is advanced. The beveled edge should be facing anteriorly toward the tongue. While advancing the scope, the left hand should be used to stabilize it and also to protect the teeth, which should never be used as a fulcrum for the scope. Another technique for insertion is by using a laryngoscope to elevate the tongue.

Once the uvula is in view, the beveled tip is used to lift the epiglottis, thus exposing the vocal cords. The bronchoscope is then rotated 90 degrees to allow its narrowest edge, the bevel, to advance in between the cords. Once in the trachea, it is rotated back so that the beveled tip is once more anterior. Advancing the scope through the trachea should be done with a gentle corkscrew motion of the right hand, while the left hand is used as a fulcrum to stabilize the scope and protect the teeth.

Another method to insert the rigid bronchoscope is by using a Miller laryngoscope to expose the vocal cords. The laryngoscope is pulled back as the rigid bronchoscope is advanced below the epiglottis and into the trachea.

Once the bronchoscope is passed into the trachea, the central airways can be visualized including the trachea, mainstem

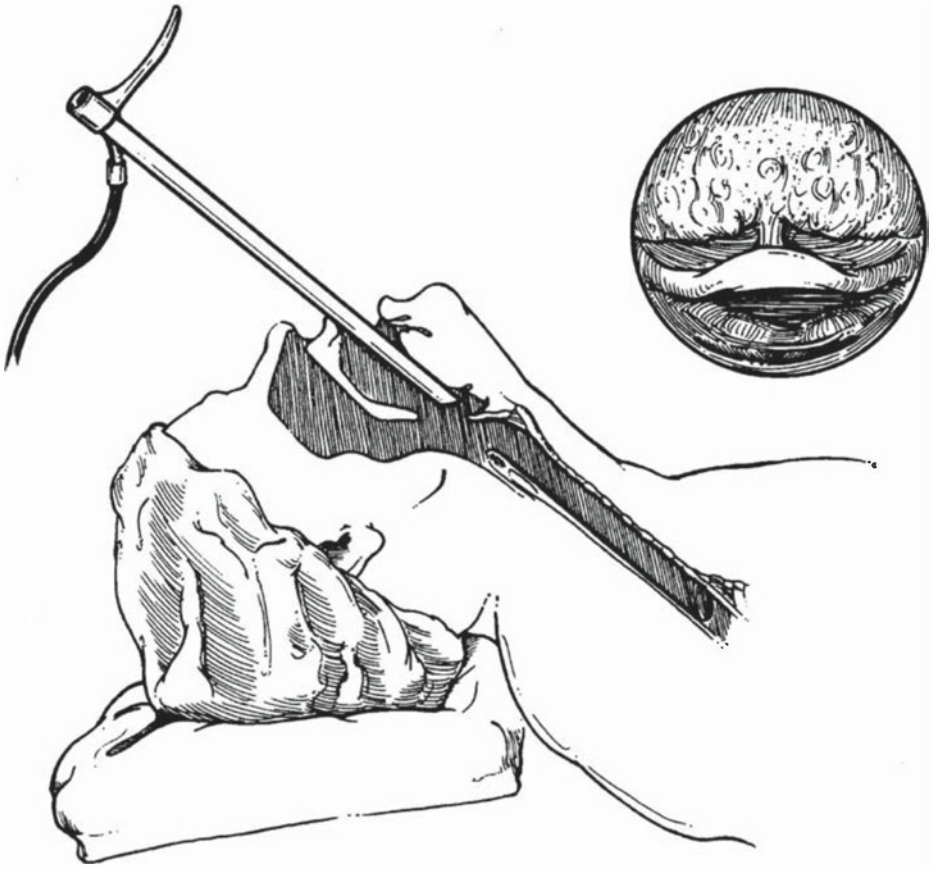


Fig. 1.16. Patient positioning for rigid bronchoscopy. The inset shows the view once the bronchoscope is passed beyond the tongue into the posterior pharynx.

bronchi, and lobar bronchi. At the carina, the patient's head is rotated in the opposite direction to the side of the mainstem bronchus to be intubated, that is, to the left for the RMSB, and vice versa. The segmental bronchi are difficult to evaluate unless an angled telescope or a flexible bronchoscope is used through the rigid bronchoscope.

Esophagoscopy

When flexible esophagoscopy is performed under conscious sedation, the patient is placed in the left lateral decubitus position and the endoscopist stands facing the patient. When done under general anesthesia, the patient is intubated and placed supine while the endoscopist stands to the right side of the head. A gentle C-shaped curve is made at the tip of the scope, which is then passed over the tongue and under direct vision into the esophagus. The scope is inserted into the oropharynx and advanced through the pyriform sinuses into the esophageal lumen.

Using gentle insufflations to distend the esophageal lumen, the scope is advanced through the entire length of the esophagus and into the stomach. Care is taken to avoid passing the scope if any resistance is encountered. Even minimal force can cause disruption of the thin-walled esophagus. The inspection and assessment of the mucosa for pathologist done upon withdrawing the endoscope while the focus should be on the safe passage of the instrument during its insertion.

Once in the stomach, a retroflexion maneuver is performed in order to adequately inspect the cardia and the gastroesophageal junction. This identifies the presence of a sliding hiatal hernia or the extension of a tumor into the stomach. Photo documentation should be performed for any findings.

Rigid Esophagoscopy

Although occasionally performed by some under conscious sedation, the procedure is usually done under general anesthesia generally to provide better relaxation and lower the risk of perforation. The patient is positioned supine with the neck extended. The esophagoscope is introduced into the right side of the mouth and stabilized using the left thumb and forefinger as a fulcrum. It is advanced behind the right arytenoid cartilage into the right pyriform fossa. The head of the bed is lowered as the scope is advanced past the cricopharyngeus. Lowering the head further allows the scope to pass through the gastroesophageal junction by directing to the right side at the distal esophagus. Full examination is done on withdrawal to avoid missing any pathology under folded mucosa during advancement of the scope.

Any post-procedural chest pain should be considered related to an esophageal perforation until proven otherwise. This should be evaluated with an immediate esophagogram first with water-soluble contrast followed by dilute barium if no extravasation is seen with the water-soluble material.

TECHNOLOGICAL ADVANCES IN ENDOSCOPY

Since the introduction of fiberoptic endoscopy, several other enhancements have been developed to improve the yield of standard endoscopy including EUS, electromagnetic navigation bronchoscopy (ENB) and the use of different wavelengths for light.

Endosonography is the usage of special flexible endoscopes fitted with an ultrasound scanner at the tip that provide both an endoscopic and sonographic image. The images obtained are fed to a processor that can control settings such as gain, Doppler, and so on. Both endoesophageal and endobronchial endoscopes (Figs. 1.17 and 1.18) have been developed for this purpose. The endobronchial ultrasound (EBUS) scope has a convex probe while for esophageal EUS, scopes are available with both radial and linear probes. Both scopes utilize a saline-inflatable balloon at the tip to be inflated when direct probe to wall cannot be accomplished. For EUS, the radial probe provides axial images while the linear probe provides longitudinal ones. The endoscopic view from these scopes is at an angle. It is therefore important for the endoscopist to know that to obtain normal endoscopic straight view, the scope must be flexed toward the wall. Inexperience with this technique may result in accidental injury.

Once the scope is at the location of interest, the balloon may be inflated to maximize acoustic coupling to the wall. Lymph nodes are visualized and nodal size, station, and sonographic characteristics are recorded prior to biopsy. It is helpful to have any prior computer tomography (CT) scan or positron emission tomography scan imaging available



A



B

Fig. 1.17. (**A**) Endobronchial linear ultrasound endoscope tip. (**B**) Ultrasound processor.



Fig. 1.18. Endoscopic ultrasound esophagogastroscope.

in order to correlate imaging findings with the procedural findings. The tip of the scope is gently flexed and pushed against the wall, aiming to maximize the observed diameter of the lymph node on the monitor. A 21- or 22-gauge needle is advanced under direct vision into the node. The stylet is then removed and the needle passed several times into the substance of the node. This material is then processed onto slides and into cell preservant for rapid onsite cytologic exam and cell block studies. During this procedure, a thorough knowledge of the surrounding vascular landmarks is essential. The Doppler function can confirm these structures.

The EUS scope can readily access mediastinal nodes at stations 1, 2L, 4L, 7, 8L, 8R, 9L, and 9R in addition to the left adrenal gland. EBUS can access 1, 2R, 2L, 4R, 4L, 7, 10R, 10L, and 11. It is obvious that EUS and EBUS should, therefore, be viewed as complimentary as the ability to combine both techniques in one setting can provide access to all the mediastinal stations except for the paraaortic and aortopulmonary window (stations 5 and 6).

The diagnostic power of flexible bronchoscopy is limited by its inability to guide biopsy instruments directly to certain lesions that are either extramural (e.g., mediastinal lymph nodes) or are too peripheral in the lung. In fact, the diagnostic yield of flexible bronchoscopy ranges from 36% to 84%, depending on the size and location of the lesion.

ENB (Fig. 1.19) provides a system to guide the bronchoscope to a specific point within the airway. The first part of this system is the three-dimensional reconstruction of axial CT scan images using specific software that also reconstructs the bronchial anatomy (virtual bronchoscopy). The second part of the system is a steerable probe that has a position sensor and is navigated through the airway that feeds its location to an electromagnetic field generator located on a board beneath the patient. The exact position of the probe is then correlated to the reconstructed images, allowing the ability to guide the bronchoscope and its instruments to the desired location. Possible applications for this system include needle biopsy of a peripheral lung lesion, the ability to place fiducials for stereotactic radiation, and the ability to inject a dye marker at the pleural surface of a lung nodule to facilitate finding it at the time of surgery.

Another technological addition to endoscopy has been the introduction of light with different wavelengths. This helps differentiate normal from pathological mucosa, which may both appear similar under standard white light endoscopy. A biopsy forceps can then be better directed at the tissue most likely to yield the abnormality. One example is autofluorescence bronchoscopy that utilizes ultraviolet light at 440 nm to inspect the mucosa. Normal mucosa fluoresces green, while mucosa from dysplasia or carcinoma appears pink.

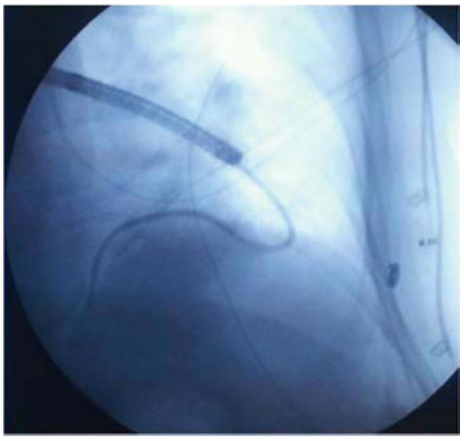
Another modality is narrow band imaging in which the light of specific wavelengths (blue at 440 to 460 nm and green at 540 to 560 nm) is also used to inspect the surface of the mucosa. A special filter is electronically activated by a switch in the endoscope. Blood vessels appear dark and distinct, resulting in increased contrast between the epithelial surface and the underlying vascular network. This can help differentiate abnormal, hypervascular epithelium from the surrounding normal lining.

CONCLUSION

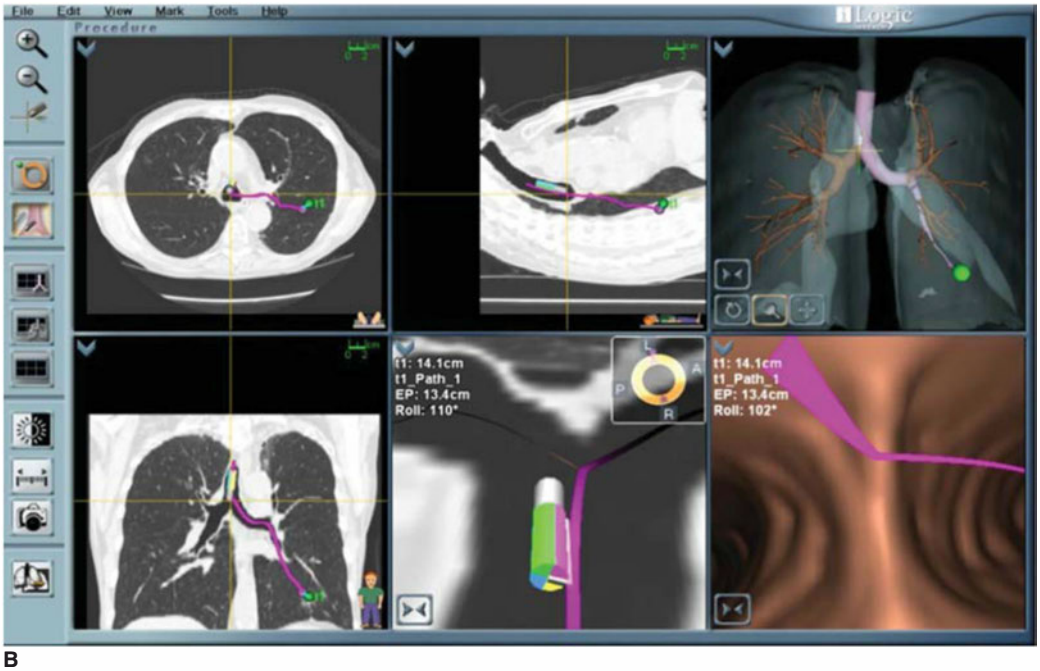
The ability to perform accurate thoracic endoscopy is essential for appropriate evaluation of thoracic disease. This is an area where formal training is provided by different specialties and to varying degrees depending on the specific interests of the center. The vast technological advances in this field have made it more challenging to acquire the necessary skills to become proficient at these modalities. Despite this, it is still important for the thoracic surgeon to become both familiar and facile at all aspects of thoracic endoscopy. Even when a patient has had a previous endoscopy, it is invaluable for the surgeon to visualize the pathology to allow for better planning of the approach for the operative procedure. Furthermore, many of these endoscopic procedures such as endoscopic stenting may be necessary adjuncts to patient care.



A



C



B

Fig. 1.19. (**A**) Electromagnetic navigation bronchoscopy (ENB) unit (with permission from iLogic). (**B**) ENB Navigation Screen. After loading the reconstructed CT scan data, a virtual bronchoscopy is obtained. The computer-generated pathway to reach the target lesion in the left lower lobe is shown. The endoscopist follows this pathway that can be seen in different views including virtual bronchoscopy, three-dimensional view, and CT views. (**C**) Fluoroscopy aids in performing the biopsy safely and accurately.

SUGGESTED READINGS

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EDITOR'S COMMENTS

This is a superb chapter that nicely summarizes both the old and the new of thoracic endoscopy. As the author mentions, endoscopy is now shared among a number of specialties but it remains incumbent on the thoracic surgeon to not only be familiar with the techniques but be expert at performing them. The threat of losing this expertise looms if thoracic surgeons become too comfortable at letting others perform the endoscopic procedure and simply depend on a report. This is not to say that our medical colleagues are not expert at performing endoscopic procedures, it is simply to say that optimal planning of a surgical procedure often requires the surgeon to visualize the anatomy and the pathology. The perspective of the surgeon differs from that of the medical endoscopist though both should work closely for the benefit of the patient.

I feel it is necessary to comment on the status of rigid endoscopy, procedures that may become a lost art. To me it is absolutely necessary for a thoracic surgeon to be facile with rigid bronchoscopy as the need surely will arise for the emergency establishment of an airway whether obstructed by tumor or foreign body. There are just situations where flexible bronchoscopy is not the procedure of choice. The ability to work through a large working channel while being able to ventilate the patient through the same instrument provides significant advantages at times. The thoracic surgeon should be able to place a rigid bronchoscope using only the scope itself to visualize and lift the epiglottis to expose the vocal cords and safely pass the instrument into the trachea. This can be a life-saving procedure. I am afraid that we are not doing a great job of exposing current thoracic surgical residents to enough rigid bronchoscopies for them to become expert and there is a potential for patients to suffer. The ease with which flexible bronchoscopy may be done has, in many situations but not all, obviated the need for rigid instrumentation. But in certain situations, there is no substitute for the rigid endoscope.

Rigid esophagoscopy, I fear, is a dying art that is not being taught. Passing the rigid esophagoscope can be fraught with danger specifically of perforation if not done with perfect technique. The area of the cervical prominence at C7 is the area most commonly perforated if the instrument is not lifted up and away from that area as it is passed. The lumen must be visualized at all times. Choosing the appropriate length of instrument based on the procedure to be accomplished is also critical. If only the upper esophagus needs to be evaluated with the rigid instrument, a shorter scope that is easier to manage may be utilized. It goes without saying that managing esophageal strictures is greatly facilitated at times with the use of the rigid instrumentation but our gastroenterology colleagues, who are not trained in the use of these, use the flexible endoscopes and seemingly dominate these procedures in the current era. In a way, it is sad that current thoracic surgical trainees may have to forego the management of patients with esophageal strictures or the palliative management of esophageal tumors as the gastroenterologists have claimed much of this work. Hopefully, thoracic surgical education will continue to include EUS and EBUS so we do not cede these procedures completely to our medical colleagues but work collaboratively with them.

LRK

2 Mediastinoscopy and Other Thoracic Staging Procedures

Michael Lanuti

INTRODUCTION

Lung cancer staging is continuously evolving as technological advances improve the imaging and endoscopic modalities available to better stratify patients into treatment and prognostic categories. The benefits of enhanced staging modalities, both local and metastatic, are to enrich the population of patients who can benefit from surgery and to avoid surgery in those patients who are not likely to benefit. The tumor-node-metastasis (TNM) staging system for lung cancer has undergone multiple revisions since 1986 when it was first published by Clifton Mountain based on 2155 lung cancer patients derived from a single institution, University of Texas, MD Anderson Cancer Center. The current seventh edition staging system for lung cancer (revised in 2009, and summarized in Tables 2.1 and 2.2) has been validated in a data set of 81,015 lung cancer patients from 19 countries, but excludes some patient populations in Africa, South America, the Indian subcontinent, and parts of China. It is the basis on which treatment strategies are constructed and is highly dependent upon accurate staging.

Operative intervention is routinely offered for stage I (lymph node negative, $T_{1a-b}N_0$ and $T_{2a}N_0$) and stage II ($T_{1a-b}N_1$, $T_{2a}N_1$, $T_{2b}N_{0-1}$, T_3N_0) patients. Complete resection in stage I yields a cure rate of ~58% to 73% and is not enhanced by adjuvant regimens except in a subset of stage IB tumors >4 cm. Recent evidence seems to support a survival advantage for adjuvant therapy in resected stage II and III disease. Node-negative superior sulcus tumors stage IIB ($T_3N_0M_0$) tumors that can be resected en bloc are best treated surgically, with selective use of concurrent chemotherapy and external beam radiation given preoperatively. Stage IIIA disease ($T_{1-2}N_2$, T_3N_{1-2}, T_4N_{0-1}) is a heterogeneous group of locally advanced tumors where multimodality therapy including surgery should be carefully considered. At the other end of the spectrum, stage IIIB and IV patients, with rare exceptions, do not benefit from surgical resection. A subset of stage IV patients with oligometastatic disease of solid organs (such as brain and adrenal gland) and no N2 disease may benefit from resection of the metastatic site and lung to achieve a 5-year survival approaching 20%.

Techniques for the identification and subsequent staging of lung cancer have improved; however, no single modality has been able to noninvasively confirm definitively the presence of lymph node metastases. Computed tomography (CT), positron emission tomography (PET), and less frequently routine magnetic resonance imaging (MRI) of the chest have been employed as clinical staging modalities for lung cancer. Lung cancer staging should include a diagnostic quality CT, a PET to evaluate the mediastinal lymph nodes and exclude extrathoracic or bone metastases, and liberal use of more invasive types of mediastinal staging such as, mediastinoscopy, endobronchial ultrasound (EBUS), and/or esophageal ultrasound-guided fine needle aspiration (EUS-FNA) of lymph nodes, and in selected patients video-assisted thoracoscopy (VATS). All of these modalities have different sensitivity and specificity for lymph node staging. Lymph node metastasis portends a worse prognosis, and surgical resectability for cure is greatly influenced by mediastinal lymph node involvement.

Evaluation of mediastinal lymph node metastases by CT alone has been extensively reviewed in the literature and has historically yielded an overall sensitivity of 59% (range 20% to 81%) and an overall specificity of 78% (range 44% to 100%). PET evaluation of the mediastinum for stage I and II nonsmall cell lung cancer (NSCLC) has yielded a sensitivity of 85% and specificity of 88%. Its negative predictive value for the mediastinum is 87%, whereas the positive predictive value is 56%. Specificity of PET is limited as both neoplastic and inflammatory nodes can have increased fluorine-18–labeled deoxyglucose (FDG) uptake. Meta-analysis of 44 relevant studies that explored the diagnostic accuracy of PET compared with CT imaging in detecting mediastinal lymph node metastases demonstrated that PET was more accurate. Accurate detection of hilar nodal involvement in lung cancer has historically been difficult, but with CT-PET and more widespread use of EBUS transbronchial needle aspiration biopsy, pathologic correlation has proven that PET is very good at detecting N1 disease.

The widespread use and availability of PET-CT fusion imaging has increased the proportion of patients found to have additional lesions that contraindicate surgical treatment but require tissue confirmation to exclude a false-positive examination. For optimal results from PET-CT, patients fast for 4 to 6 hours and then receive 15 mcuries of 18-FDG (550 MBq) as an intravenous injection 1 hour before study. 18-FDG has a 110-minute half-life. Transmission CT (5 mm cuts) is acquired during quiet breathing and then the PET evaluation is acquired. CT-PET acquisition takes about 30 to 35 minutes. Importantly, the transmission CT scan that is generated by this modality is not as accurate as a diagnostic quality CT chest. This technology has been evaluated in the literature with increasing frequency. In a sentinel study from Switzerland in 2003, the authors studied PET-CT in 50 patients with NSCLC. Integrated PET-CT provided additional information in 41% of patients and was significantly more accurate in precise staging compared with CT alone, PET alone, or visual correlation of PET and CT.

MRI of the lung has historically been challenged by poor spatial resolution and high noise-to-contrast ratio and has been reserved for characterizing mediastinal masses or detecting tumor invasion into chest wall, mediastinum, vasculature structures, diaphragm, pericardium, or bone. MRI has greatly contributed to the clinical staging of superior sulcus tumors. A recent report suggests that new MRI techniques such as diffusion-weighted imaging (DWI)

Table 2.1 TNM 7th Edition International Lung Cancer Staging System

Tis	Carcinoma in situ
T1	Tumor <3 cm surrounded by lung, no bronchoscopic invasion
T1a	Tumor <2 cm
T1b	Tumor >2 cm but <3 cm
T2	Tumor >3 cm but <7 cm, or tumor with any of the following features: involves main bronchus <2 cm from carina Invades visceral pleura, or associated with obstructive pneumonitis extending to hilum
T2a	Tumor >3 cm but <5 cm
T2b	Tumor >5 cm but <7 cm
T3	Tumor >7 cm or one that directly invades any of the following: chest wall (including superior sulcus), diaphragm, phrenic nerve, mediastinal pleura, parietal pericardium or tumour <2 cm from carina without involvement of carina, or **Separate tumor nodules in same lobe**
T4	Tumor of any size that invades any of the following: mediastinum, heart, great vessels, trachea, RLN, esophagus, vertebral body, carina or **Separate tumor nodules in different ipsilateral lobe**
N1	Mets to ipsilateral peribronchial or hilar LNs and intrapulmonary LNs, including involvement by direct extension
N2	Mets to ipsilateral paratracheal or subcarinal LNs
N3	Mets to contralateral paratracheal, contralateral hilar, ipsilateral or contralateral scalene, or supraclavicular LNs
M1a	Separate tumor nodules in a contralateral lobe, tumor with pleural nodules, or malignant pleural or pericardial effusion
M1b	Distant metastases

LNs, lymph nodes; RLN, recurrent laryngeal nerve.
(American Joint Committee on Cancer, 7th Edition Lung Cancer Staging Manual, 2010, Chicago, IL.)

Table 2.2 7th Edition International Lung Cancer Staging System with 5-Year Survival Based on Pathologic Staging

Stage	T	N	M	5-y survival
0	Tis	N0	M0	
IA	T1a,b	N0	M0	73%
IB	T2a	N0	M0	58%
IIA	T1a,b	N1	M0	46%
	T2a	N1	M0	
	T2b	N0	M0	
IIB	T2b	N1	M0	36%
	T3	N0	M0	
IIIA	T1,T2	N2	M0	24%
	T3	N1,N2	M0	
	T4	N0,N1	M0	
IIIB	T4	N2	M0	9%
	Any T	N3	M0	
IV	Any T	Any N	M1a,b	

and short tau inversion recovery turbo spin echo (STIR) sequences can improve upon the accuracy of PET/CT for distinguishing benign from malignant lymph nodes. Furthermore, PET-MRI is the newest modality that is being evaluated for lung cancer staging.

MEDIASTINOSCOPY

The gold standard for staging the mediastinum is cervical mediastinoscopy, which was developed by Harken and associates in 1954 and then promulgated by Carlens in 1959 and later Pearson in 1965. Carlens and Pearson recognized that mediastinoscopy potentially was useful not only for lung cancer staging but also for the diagnosis of lymphoma, metastatic disease from an extrathoracic origin, infectious etiologies, and sarcoidosis. It is the benchmark against which all comparisons of lymph node accuracy are currently analyzed. The mediastinoscopes used today are modifications of the original instruments, with distal illumination, a beveled end, and a lateral slit for instrumentation (see Fig. 2.1). The efficacy of mediastinoscopy has been well established in the assessment of enlarged mediastinal lymph nodes with 100% specificity and ~90% sensitivity. In patients with known or suspected lung cancer, the routine use of mediastinoscopy can change the plan of care in up to 25% of patients. Large studies confirm false-negative rates from 5% to 8%, as demonstrated in Table 2.3. The false-negative rate of mediastinoscopy may be attributed to the diligence of the surgeon dissecting and sampling the nodes. Ideally, five nodal stations (stations 2R, 4R, 7, 2L, and 4L) should be routinely examined (see Fig. 2.2), with at least one



Fig. 2.1. The Carlens mediastinoscope. (image © 2014 Teleflex Incorporated)

Table 2.3 Morbidity and Mortality Reported in Large Studies of Patients Undergoing Mediastinoscopy for Lung Cancer

Study	*N*	No. with lung cancer	No. (%) with false-negative results	No. (%) of complications	No. (%) of deaths
Lemaire (2006)	2,145	1019	56 (5.5)	23 (1.07)	1 (0.05)
Park (2003)	3,391	NA	NA	14 (0.04)	0
Hammoud (1999)	2,137	947	76 (8.0)	12 (0.06)	4 (0.2)

NA, not applicable.

node sampled from each station unless none are present after dissection in the region of a particular nodal station. Compared with conventional mediastinoscopy, video mediastinoscopy appears to yield some improvement in sensitivity (92%) and false-negative rates (7%).

Routine mediastinoscopy remains somewhat controversial in that many lung cancer treatment centers use the modality selectively. A national survey of 729 hospitals (31% teaching or university hospitals, 38% community cancer centers, 46% comprehensive community cancer centers) sponsored by the American College of Surgeons identified more than 11,668 patients whose initial management included surgical therapy for lung cancer.

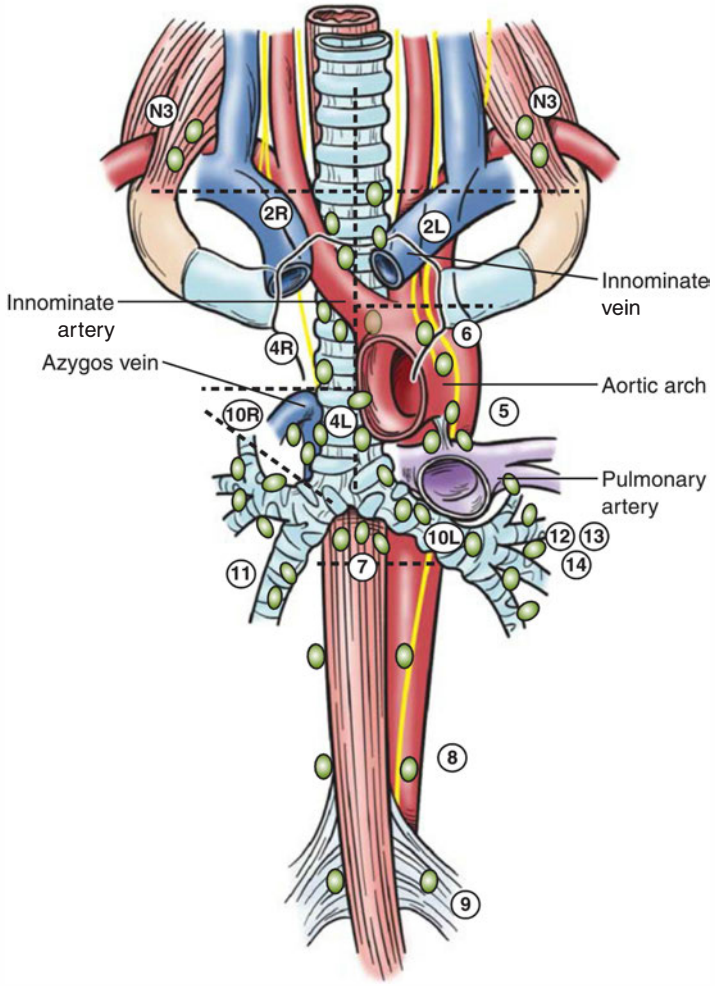


Fig. 2.2. The Mountain-Dresler modification of the regional lymph node map originally proposed by the American Thoracic Society. (Redrawn after Mountain CF, Dresler CM. Regional lymph node classification for lung cancer staging. Chest 1997;111:1718-1723c.)

The mediastinum was evaluated preoperatively with mediastinoscopy in only 27% of these surgical patients and only 26% underwent a staging PET. The underuse of aggressive mediastinal staging in both academic and community lung cancer care is sobering. Additionally troublesome is that only 42% had lymph nodes sampled at any mediastinal level during the definitive surgical procedure.

Indications and Contraindications

Mediastinoscopy should be performed in any patient harboring a suspicious lung nodule with enlarged (>1 cm in short axis measured on CT) or FDG-avid mediastinal lymph nodes (N2 or N3), those with central tumors, and those with peripheral tumors >2 cm. T1 tumors with an aggressive histology (i.e., large cell neuroendocrine carcinosarcoma, small cell or pleomorphic carcinomas) should also undergo mediastinoscopy. Peripheral T1a lesions (tumors ≤2 cm) with PET-negative mediastinal lymph nodes can be regarded as the one exception to the routine use of mediastinoscopy. Mediastinoscopy is the procedure of choice for lung cancer staging (*endorsed by the American College of Chest Physicians, and European Society of Thoracic Surgeons*) but endoscopic techniques such as EBUS or EUS-FNA are recognized as modalities where a cytologic diagnosis can be achieved to initiate a treatment strategy. If EBUS or EUS-directed biopsies are negative in a pathologically enlarged or FDG-avid mediastinal lymph node, mediastinoscopy is still recommended.

Mediastinoscopy does not have many strict contraindications except for perhaps inability to extend the neck because of severe kyphosis or fused cervical vertebra. Relative contraindications to mediastinoscopy include large thyroid goiter, severe atherosclerotic disease in the aortic arch, vertebral arteries (vulnerability to ischemic events with neck extension), or the innominate artery which can contribute to embolic stroke. End tracheal stoma after laryngectomy in association with neck radiation can present anatomic difficulty with increased risk of cervical wound infection. Previous mediastinoscopy is associated with fibrosis of the pretracheal tissue plane and makes redo mediastinoscopy challenging. Despite this phenomenon, repeat mediastinoscopy is feasible and safe in most cases and can be carefully used to restage the mediastinum after induction therapy. Particular care must be exercised in separating the

innominate artery from the trachea. Sharp dissection under direct vision is safer than the blunt approach used in first-time mediastinoscopy. Many surgeons reserve EBUS-directed biopsy for preinduction mediastinal lymph node sampling in patients suspected of harboring locally advanced lung cancer (stage IIIA) with positive N2 nodes.

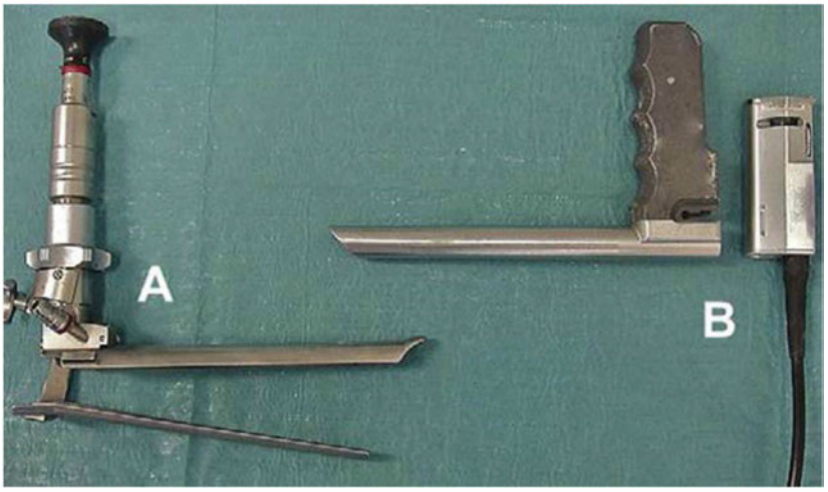


Fig. 2.4. Video mediastinoscopes (**A**) Linder-Dahan scope with dilating and parallel adjustment blades. (**B**) Lerut scope where camera module is fully integrated in the hand piece. (Obtained from Thorac Surg Clin. May 2012;22(2):180. WB Saunders, Elsevier, Inc.)

Technique

Mediastinoscopy is an outpatient procedure that needs to be performed in a hospital setting to efficiently manage the potential for hemorrhage. The patient is placed supine on the operating room table where the head is placed at the top of the bed (Fig. 2.3). After establishing general endotracheal anesthesia, the neck is extended with either a thyroid bag or a roll under the shoulders. A pulse oximeter or radial artery cannula can be used to monitor compression of the innominate artery (observation of a dampened wave form) so as not to compromise blood flow to the right carotid artery for an extended period. Most surgeons include the entire sternum in the operative field in the event that serious hemorrhage is encountered and median sternotomy is necessary. In the event that a patient already has a previous median sternotomy, a right or left anterior thoracotomy can be considered for exploration in the setting of acute hemorrhage. Mediastinoscopy can be achieved with direct visualization through a standard scope (Fig. 2.1) or via a magnified view with a video mediastinoscope (Fig. 2.4). Video mediastinoscopy permits both direct and monitor viewing and facilitates teaching since all participants in the operating theater can observe. As previously mentioned, video mediastinoscopy has been shown to improve lymph node sampling over conventional mediastinoscopy.

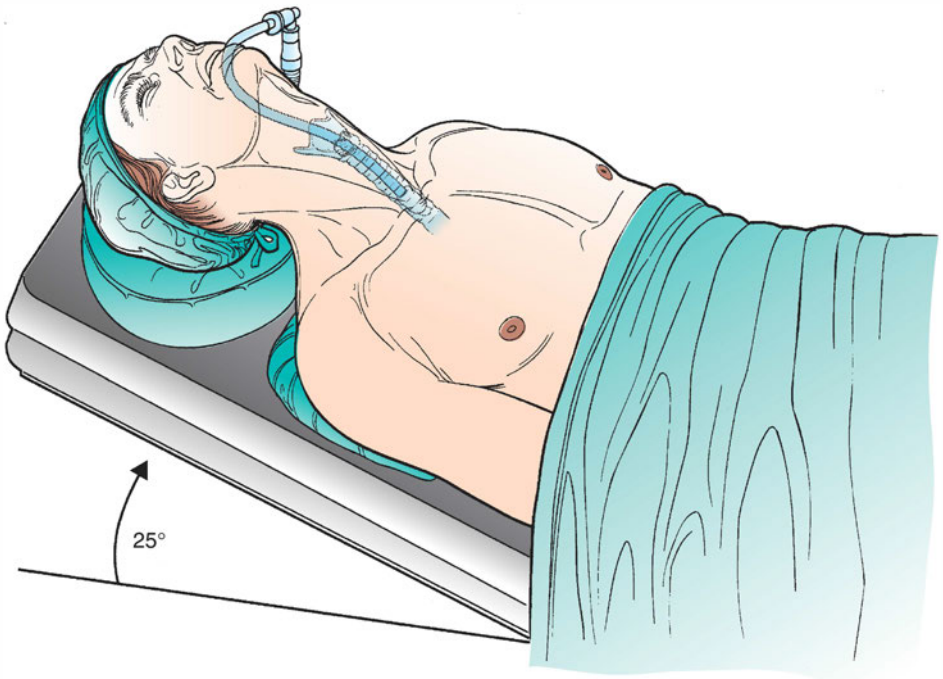


Fig. 2.3. Position for mediastinoscopy with general anesthesia and endotracheal intubation. Shoulders are elevated and the patient's head at top of the table.

Dissection is carried down through the incised platysma muscle to the strap muscles (sternohyoid and sternothyroid). The thyroid isthmus is rarely divided. Palpation of the trachea helps identify the soft tissue midline where the strap muscles (see Fig. 2.5) can be vertically separated to allow sharp entry into the pretracheal alveolar plane. There is often an adipose layer in the midline. The anterior tracheal wall is visualized (Fig. 2.5C) and a tunnel can be created by bluntly spreading with a Metzenbaum scissors or by insertion of the index finger. Blunt finger dissection caudally (Fig. 2.6) can often permit palpation of the innominate artery pulse which later can be vulnerable to compression when using the mediastinoscope. Finger dissection is continued down the trachea where the surgeon can appreciate tissue resistance and texture of mediastinal lymph nodes (Fig. 2.7). It is essential to assess the area just above the sternal notch early in the dissection because neck extension may elevate the innominate artery up into the base of the neck where it can be injured by sharp dissection or cautery.

The mediastinoscope can be inserted into the pretracheal tunnel where a suction cautery dissector is used to bluntly dissect this alveolar plane with occasional fibrous bands (Figs. 2.8 and 2.9). The instrument is advanced only if it passes easily and there is a visible tunnel ahead. The surgeon can explore the entire length of the trachea and both main stem bronchi. The landscape is carefully surveyed to identify lymph nodes in the various stations depicted in Figure 2.10. Nodes are identified by color and consistency. Particular care is taken when

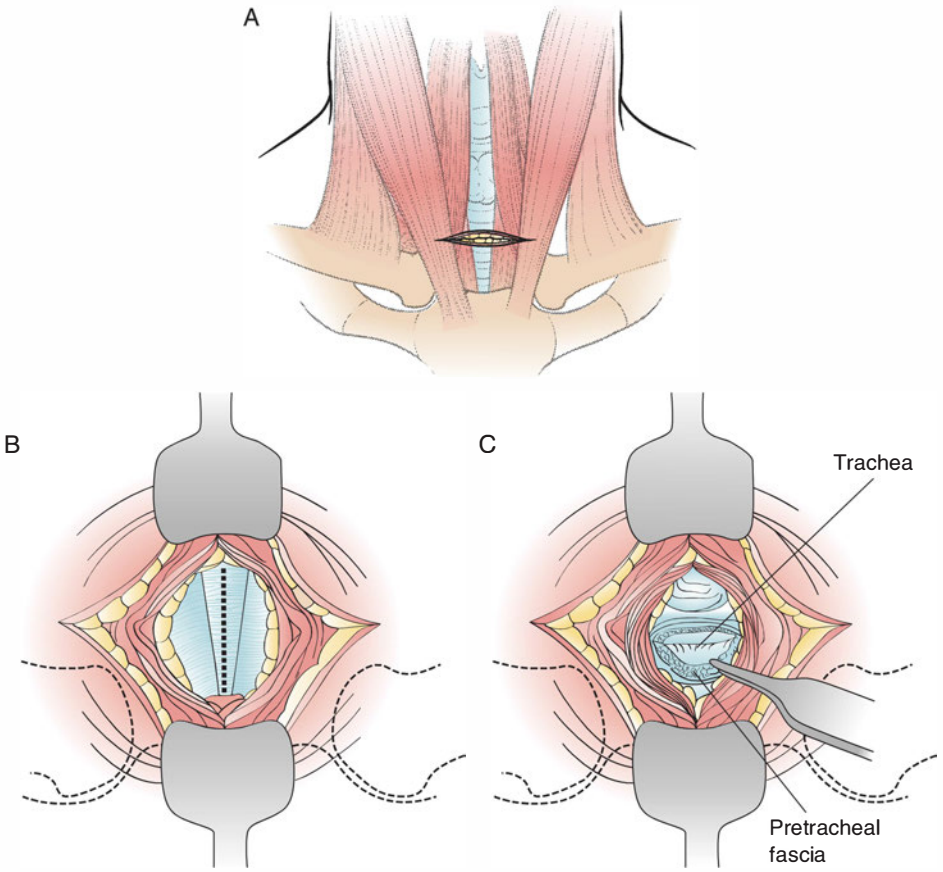


Fig. 2.5. Mediastinoscopy: small cervical neck incision (**A**) just about the sternal notch. After division of platysma muscle, strap muscles are exposed to identify the midline (**B**). Pretracheal fascia is sharply divided and raised to expose trachea (**C**).

Complications

The incidence of complications for mediastinoscopy across large series is extremely low (summarized in Table 2.4). Although catastrophic hemorrhage can occur during mediastinoscopy, most bleeding is minor and can be controlled with application of Surgicel (oxidized cellulose), transient packing, or partial withdrawal of the scope to tamponade the mediastinum. Occasionally hemoclips can be used in the face of a visible small vessel particularly in the subcarinal area where bronchial arteries are prevalent. Cautery should not be used anywhere along the left paratracheal space to avoid thermal injury to the recurrent laryngeal nerve. If significant bleeding occurs, the mediastinum is packed and left in place for at least 10 minutes and then gently removed. Blood for transfusion should be made available in the operating theater along with additional instruments. If hemorrhage continues, packing is reinstituted, and median sternotomy or thoracotomy is emergently performed. Median sternotomy is the most versatile incision and preferred for hemodynamic instability, or injury to innominate artery, aortic arch, or right pulmonary artery. It also allows better access for cardiopulmonary bypass if necessary to control hemorrhage from a major vessel. Performing pulmonary resection after control of major bleeding is

retrieving specimens near the tracheobronchial angles because of the proximity of the azygos vein and apical branch of the truncus anterior pulmonary artery on the right and the recurrent laryngeal nerve on the left. The right main pulmonary artery is observed superior to the carina and can be injured with cautery while achieving hemostasis from biopsies of subcarinal lymph nodes. All dissection and lymph node biopsies should be in close proximity to the airway. The blue-gray hue of venous structures can sometimes be mistaken for an anthracotic node. If there is any doubt about the solid or vascular nature of the tissue in question, aspiration is performed using a 20- or 22-gauge spinal needle and 10 to 20 cc syringe (Fig. 2.11). Target lymph nodes should be partially dissected before any biopsy specimens are taken to reduce bleeding. Once sufficiently freed, the node is grasped with a cupped biopsy forceps, and traction is applied under direct vision (Fig. 2.11C). If the node cannot be extracted by gentle pulling and twisting, further dissection is carried out or an additional instrument can be introduced down the scope to divide tethering attachments or stabilize the origin of the lymph node to avoid traction injury to adjacent vasculature. The goal of standard mediastinoscopy is to sample lymph nodes and not perform lymphadenectomy. When sampling lymph nodes for lung cancer, it is important to sample multiple lymph node stations. Samples are routinely obtained from stations 2, 4, and 7 (see Fig. 2.10). Of note, station 10 is a hilar node (N1) and by definition resides inside the pleural envelope and cannot routinely be accessed with standard mediastinoscopy techniques. EBUS is the modality of choice for accessing this nodal station. Sampling nodes contralateral to the primary tumor is paramount to identify patients who harbor N3 disease (stage IIIB) where definitive surgical resection would be contraindicated most of the time. Labeling all specimens by numerical station is less subject to error and removes ambiguity from pathology reports. This behavior should be aggressively embraced when training since it improves communication (common language among all treating physicians) and provides more accurate data when performing clinical research.

Table 2.4 Complications of Mediastinoscopy

Death <0.2%
Major complications ≤1%
Major hemorrhage intraoperatively
Recurrent laryngeal nerve paralysis
Cerebrovascular accident
Esophageal perforation
Mediastinitis
Mediastinal hemorrhage postoperatively
Tracheobronchial injury
Phrenic nerve paralysis
Thoracic duct injury
Venous air embolism
Minor complications 2.5%
Pneumothorax
Recurrent laryngeal nerve palsy
Wound infection
Minor bleeding
Autonomic reflex bradycardia

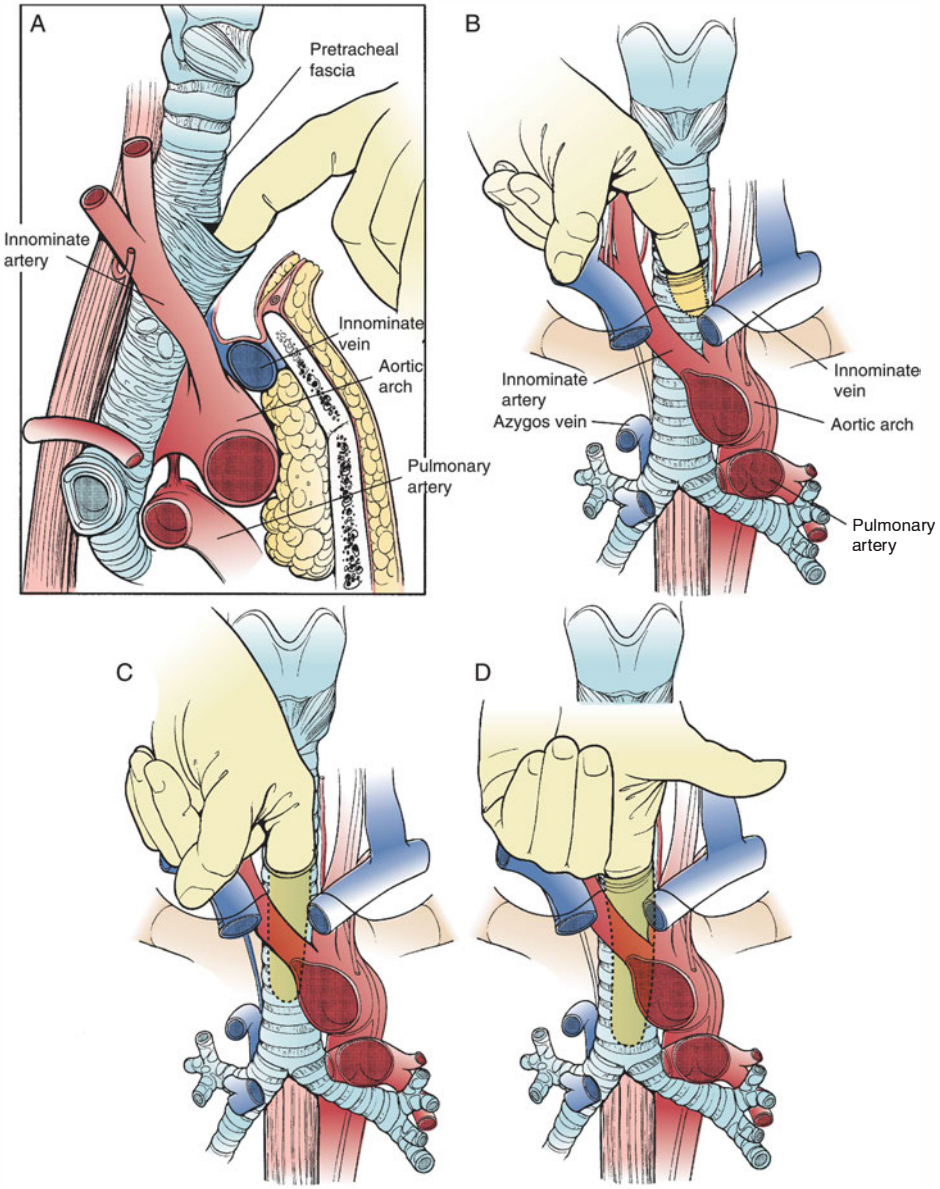


Fig. 2.6. Sagittal view (**A**) and coronal view (**B**) of finger dissection to develop the pretracheal plane. Digital palpation of the innominate pulse (**C**) with additional caudal dissection toward carina (**D**).

a disadvantage of median sternotomy. Right thoracotomy is often employed for the more common azygos vein injury and for the less common superior vena caval injury. Large studies confirm low morbidity and mortality as described in Table 2.3. The most underreported complication from mediastinoscopy is left vocal cord paralysis from recurrent nerve injury. This can be observed with traction injury from the scope as well as thermal injury from cautery. Esophageal injury is extremely uncommon and can be encountered in association with aggressive biopsy at the low left paratracheal or subcarinal space. This injury may not be immediately recognized and the patient may present postoperatively with mediastinal air, mediastinitis, or pleural effusion. Esophagram should be obtained and the injury managed with techniques similar to those for managing other traumatic injuries to the esophagus. Pneumothorax can be observed when violating the parietal pleura and is not often associated with a parenchymal injury. This can be managed intraoperatively using a flexible rubber catheter that is brought out through the cervical wound. After the mediastinoscope is removed, pleural air is evacuated during a Valsalva maneuver as the tube is removed. If there is a recognized lung parenchymal injury, tube thoracostomy must be performed.

Extended Cervical Mediastinoscopy

Extended cervical mediastinoscopy is a modality that is performed only by few thoracic surgeons and therefore is not routinely part of thoracic surgery education. It was designed to stage patients with left upper lobe lung cancers in whom standard cervical mediastinoscopy results are negative. Ginsberg and colleagues devised a method for access to the paraaortic (station 6) and aortopulmonary (station 5) lymph nodes. The approach originates through a standard cervical incision where a tunnel is created over the aortic arch, between the innominate artery and the left carotid artery (see Fig. 2.12). The mediastinoscope is inserted obliquely to reach the subaortic space. The distinction between the subaortic and para-aortic space is difficult and frequently limited by the bony chest wall. In addition to potential injury to the innominate and carotid artery, injury can be observed to the left main pulmonary artery, left recurrent laryngeal nerve, and left phrenic nerve. Alternatively, these nodal stations can be accessed by anterior mediastinotomy or VATS with improved visualization.

Anterior Mediastinotomy/ Mediastinoscopy

This procedure was described in 1966 by McNeill and Chamberlain and is often employed in lieu of extended cervical mediastinoscopy for lung cancer staging. It is an outpatient procedure that can target enlarged prevascular (station 6) or aortopulmonary (station 5) lymph nodes in the setting of lung cancer or anterior mediastinal masses that require a diagnosis. When mediastinotomy is indicated by CT findings, it is usually preceded by mediastinoscopy in the setting of lung cancer staging. The procedure requires general anesthesia and a single-lumen endotracheal tube. It can be performed through a 5-cm transverse incision at the left sternal border over the second or third interspace. The fibers of the pectoralis muscle are split to expose the cartilage. Although the original description recommends excision of the second costal cartilage (Fig. 2.13), this can be obviated with the use of a mediastinoscope (Fig. 2.14). The mediastinum is entered after incising the endothoracic fascia. The internal mammary artery and vein may be ligated if necessary but can often be retracted and preserved (Fig. 2.13C). Although an extrapleural plane can be developed to access station 5 or 6, this is not always feasible. When sampling a mediastinal mass suspicious for a thymic neoplasm, entering the pleural space can be a risk for pleural dissemination of disease and should be avoided if possible. The mediastinotomy site is often excised (full thickness) during

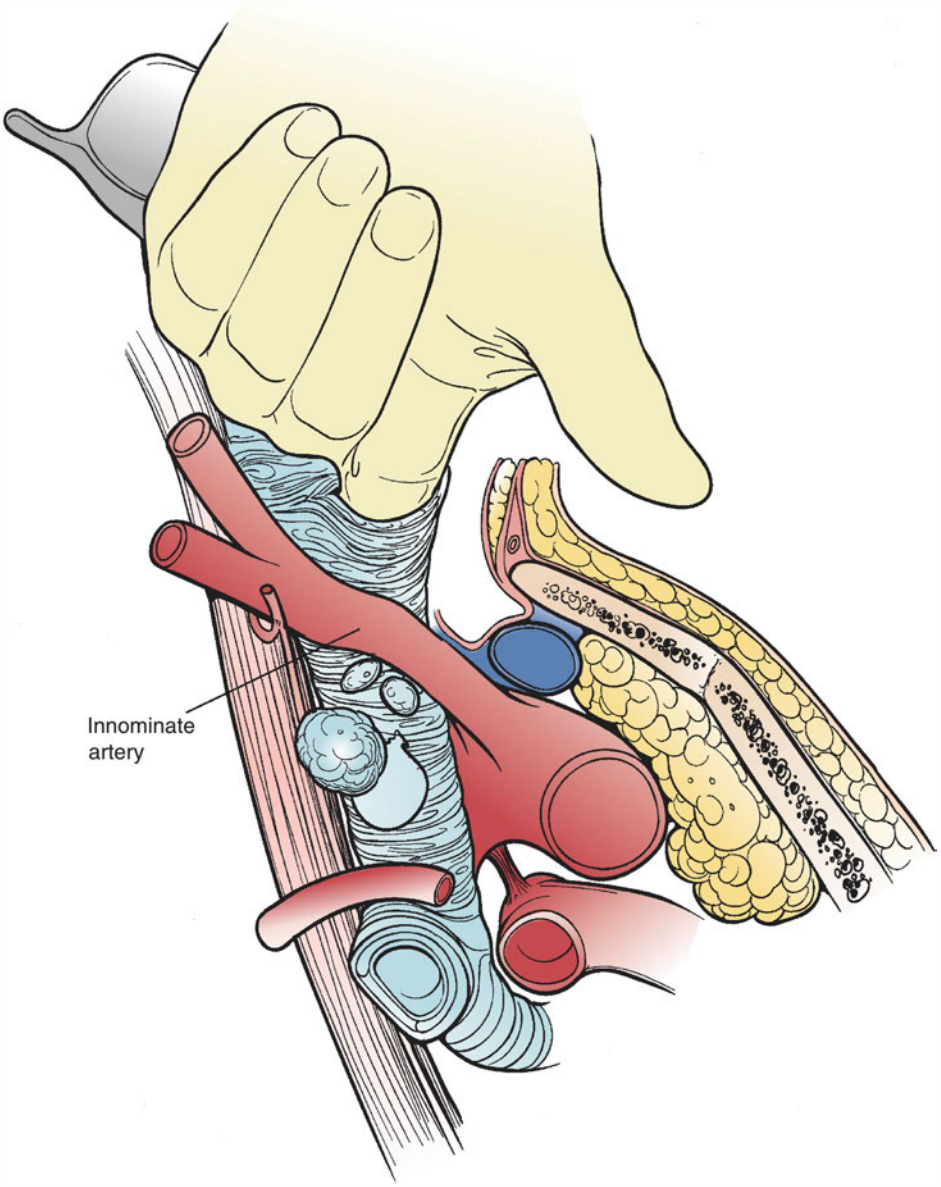


Fig. 2.7. Oblique view of finger bluntly dissecting the pretracheal fascia to enter the node-containing space.

definitive surgical management of thymic neoplasms to reduce the incidence of local tumor recurrence by virtue of tumor cell implantation at the time of mediastinotomy. When mediastinotomy is done in conjunction with cervical mediastinoscopy, the neck incision is left open to allow bidirectional palpation of the aortopulmonary region (Fig. 2.15). A mediastinoscope is useful for improved lighting when performing biopsy with a cup forceps. If the pleural cavity is entered, one can aspirate the pneumothorax using a flexible rubber catheter that is brought out through the wound edge. After wound closure, pleural air is evacuated during a Valsalva maneuver as the tube is removed. Potential pitfalls here are inadvertent biopsy of the left main pulmonary artery or aortic arch or injury to phrenic or recurrent laryngeal nerve. Chylothorax after mediastinotomy is rare, but has been reported. Adequate visualization is mandatory and if it cannot be achieved, thoracoscopy can be added through additional port incisions while intermittent ventilation is employed.

OTHER STAGING MODALITIES

Supraclavicular (Scalene) Lymph Node Biopsy

Supraclavicular lymph node biopsy is currently reserved for palpable disease in the setting of lung cancer or other disease processes such as lymphoma, metastatic disease from an extrathoracic site, sarcoidosis, or infectious disease. Enlarged lymph nodes in this region may be amenable to needle aspiration, but a negative aspirate in an enlarged lymph node does not rule out disease. Excisional biopsy is preferred for enlarged lymph nodes where a formal excision of the scalene fat pad is less utilized. Historically, scalene fat pad excision was the mainstay of nodal staging in lung cancer before the advent of mediastinoscopy. The positivity rate in the absence of palpable adenopathy varied from 3% to 20%, with central adenocarcinomas having the highest incidence. Excisional or incisional biopsy of a supraclavicular lymph node can be the first diagnostic procedure in patients who present with suspicious lung mass and enlarged neck nodes. Confirmation of node-positive disease with this technique invokes at least pathologic TxN3 lung carcinoma (stage IIIB). Pitfalls in executing this procedure are injury to the phrenic nerve, chyle leak when dissecting in the left supraclavicular fossa, and bleeding.

Esophageal Ultrasound

EUS-FNA is a minimally invasive alternative technique for mediastinal staging of NSCLC. The concept was introduced in 1996. Access to all mediastinal nodes is limited to left paratracheal (stations 2L and 4L), subcarinal (station 7), paraesophageal (station 8), and inferior pulmonary ligament (station 9) lymph nodes (depicted in Fig. 2.10). EUS can visualize stations 5 and 6 lymph nodes, but the feasibility of needle biopsy is limited to the inferior aspect of station 5 with favorable anatomy. Advanced users have described a method to sample the para-aortic (station 6) lymph node without traversing the aorta. EUS can also detect and sample metastatic (M1) disease in the adrenal glands and liver. Aside from thoracoscopy or thoracotomy, EUS is the only modality that can access N2 lymph nodes along the esophagus and inferior pulmonary ligament (stations 8 and 9). EUS-FNA has been shown to be highly accurate in detecting mediastinal lymph node metastases with an overall diagnostic yield of 90% and a false-negative rate of 22%. Because of the insufficient negative predictive value, negative results with EUS-FNA in an enlarged lymph node should be verified by surgical staging. A meta-analysis was performed to estimate the diagnostic accuracy of EUS-FNA for staging mediastinal lymph nodes (N2/N3

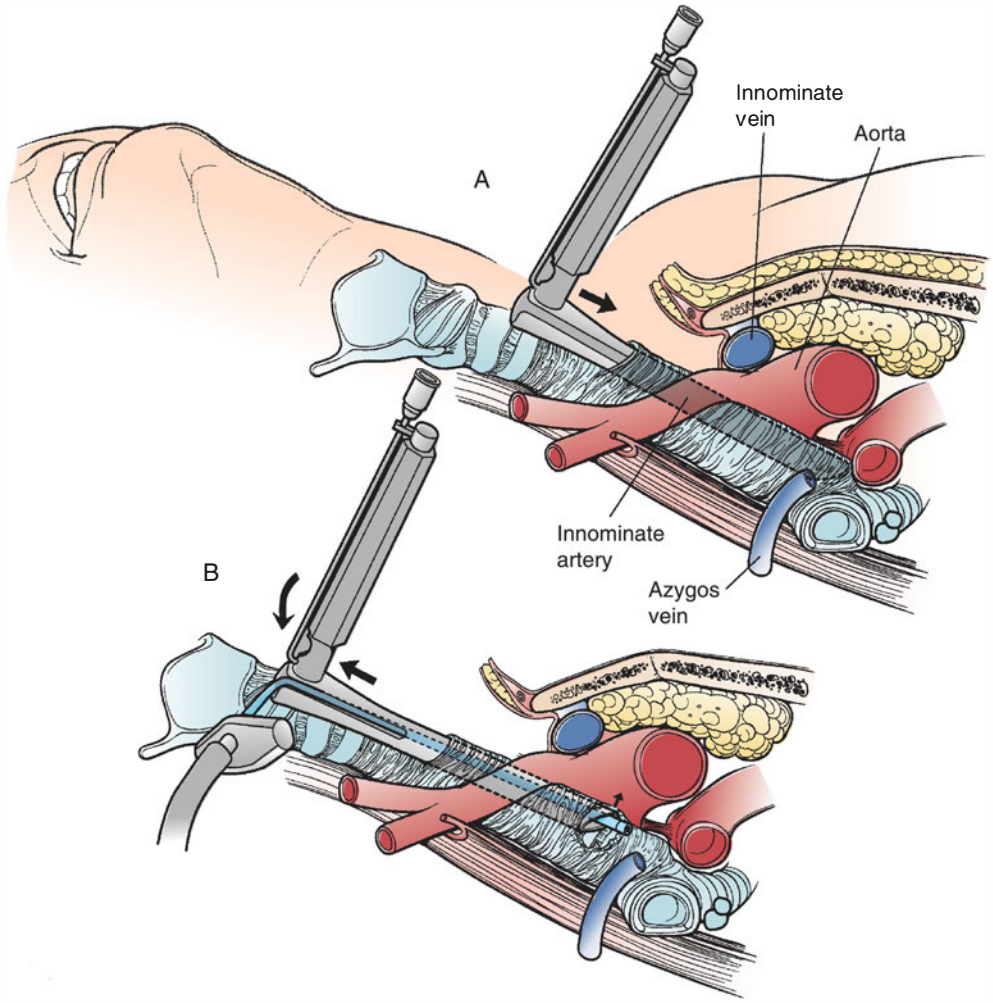


Fig. 2.8. The mediastinoscope is passed into the pretracheal tunnel (**A**). Note the adjacent structures. Suction cautery (**B**) is used to further develop the pretracheal plane to expose carina and bilateral main stem bronchi.

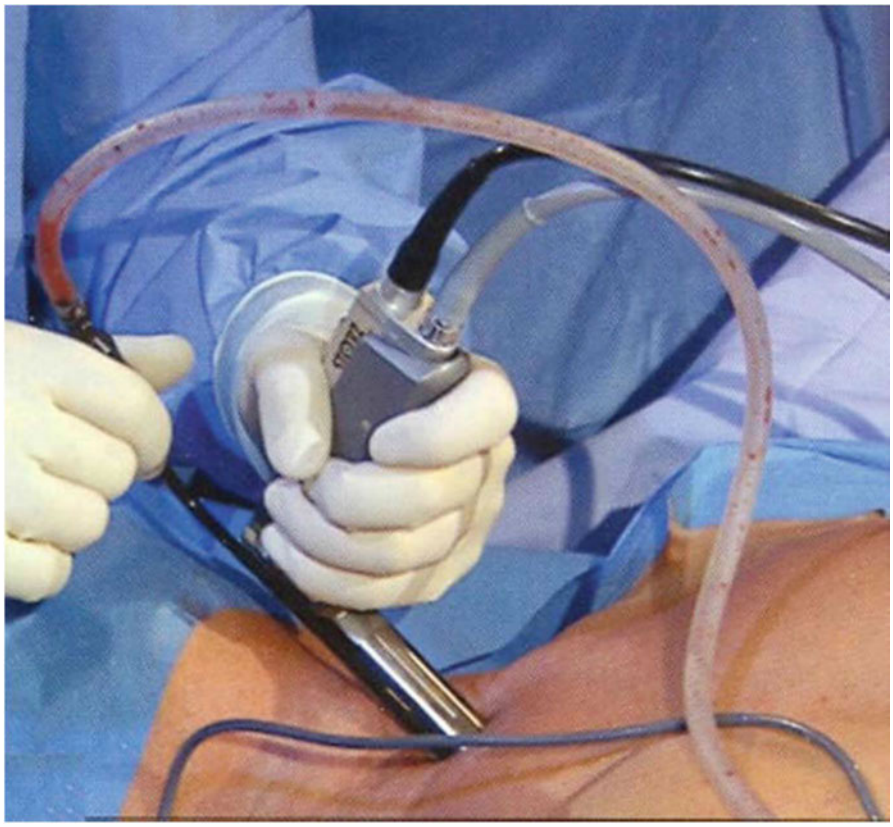


Fig. 2.9. Video mediastinoscope with suction cautery. (Obtained from Video mediastinoscopy copyright 2011 Endo Press Tuttlingen, Germany.)

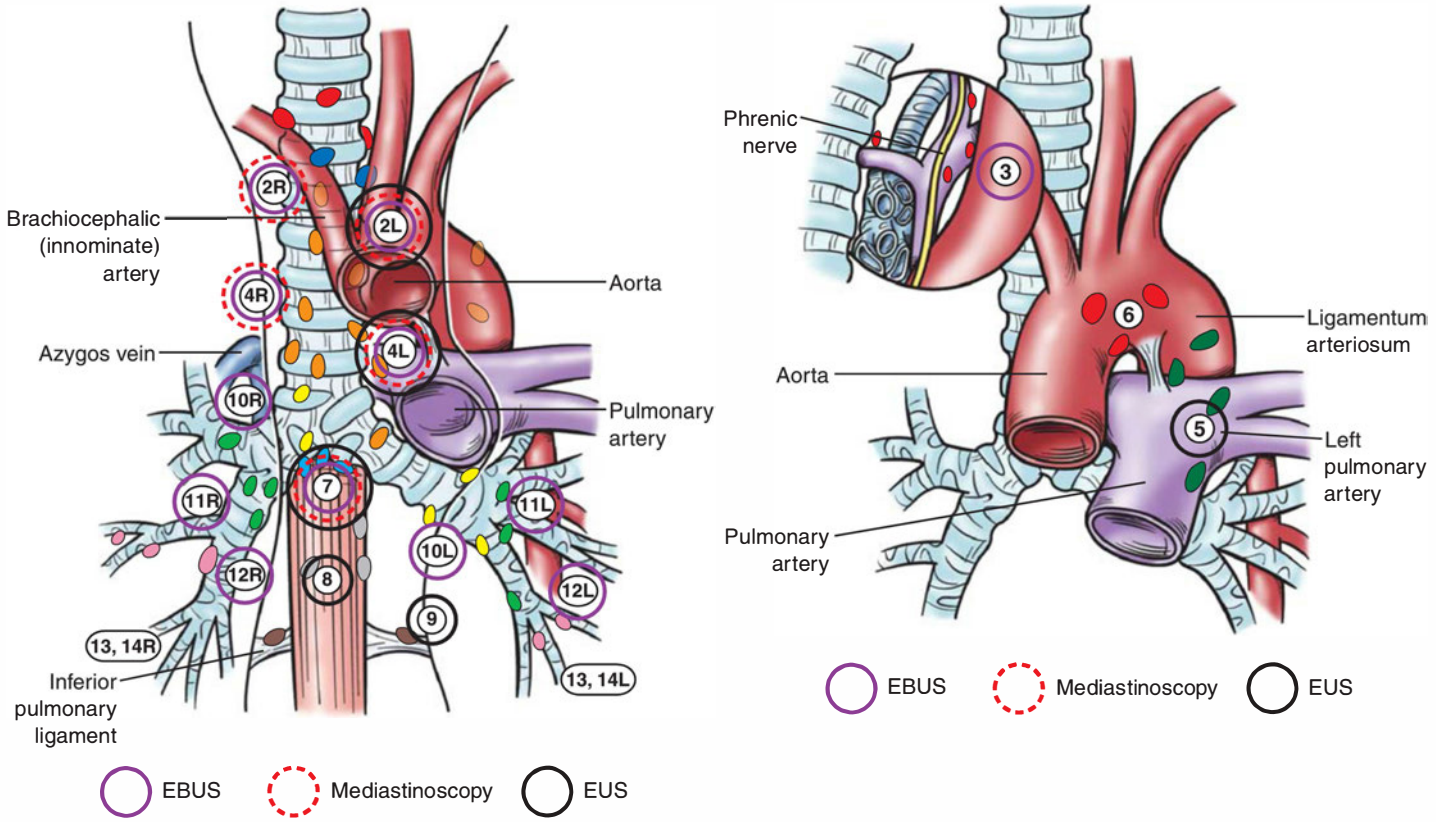


Fig. 2.10. Map depiction of regional lymph nodes according to accessibility by staging modality including mediastinoscopy, endobronchial ultrasound (EBUS) and esophageal ultrasound (EUS). (Redrawn after Mountain CF, Dresler CM. Regional lymph node classification for lung cancer staging. Chest 1997;111:1718-1723c.)

disease) in patients with lung cancer. Pooled meta-analysis of EUS staging of lung cancer demonstrated an 83% sensitivity (78% to 87%) and 97% specificity (96% to 98%). When analyzing EUS for radiographic abnormal lymph nodes seen on CT, the sensitivity was 90% and specificity was 97%.

Endobronchial Ultrasound

There is mounting data regarding the use of EBUS and staging of lung cancer. The modality was originally applied to lung cancer staging by Yasufuku and colleagues in 2004. Real-time EBUS transbronchial needle aspiration (TBNA) is highly accurate and safe for sampling enlarged mediastinal lymph nodes. Integrated color power Doppler ultrasound is used to avoid intervening vessels immediately before needle puncture. EBUS can be performed in an outpatient setting with either conscious sedation or general anesthesia. Biopsies can be performed using a 21-or 22-gauge needle with a maximal extension of 40 mm (Fig. 2.15). Fine needle aspiration is performed by passing a needle through the tracheal or bronchial wall into adjacent lymph nodes or central or perihilar parenchymal masses under real-time ultrasound control (Fig. 2.16). Specimens can be smeared on glass and fixed with ethanol or sprayed with Cytofix. Additional specimens can be placed in 10% formalin or saline in preparation of a cell block to perform immunohistochemistry. Of great importance in the contemporary treatment of lung cancer, this small amount of tissue can be used for molecular genotyping (i.e., EGFR, ALK, ROS1, BRAF, HER2, and others) to identify patients who will benefit from targeted therapy, or be eligible for clinical trials, particularly in the setting of metastatic disease.

Compared with mediastinoscopy, EBUS-TBNA has the advantage that it is also able to routinely access posterior mediastinal (station 7) and hilar lymph nodes (stations 10, 11, and 12) in addition to bilateral paratracheal nodes (stations 2 and 4). Figure 2.10 shows the extent of accessible lymph nodes when using EBUS. A recent study by Herth and colleagues suggests that patients with a normal CT and PET of the mediastinum can be primarily evaluated and staged during a diagnostic bronchoscopy with EBUS-TBNA of all nodes larger than 5 mm, especially if the primary tumor is known to be an adenocarcinoma. Unfortunately, champions of the procedure tend to have better results than those obtained by all other users. The false-negative rate of EBUS has been reported to be 15% to 28%. A meta-analysis of EBUS in lung cancer staging reviewed 365 publications where only 10 were suitable for analysis. EBUS-TBNA biopsy had a pooled sensitivity of 88% (range 80% to 94%) and a specificity of nearly 100% (range 92% to 100%).

To date there have been two randomized controlled clinical trials comparing mediastinal nodal surgical staging (mediastinoscopy) of lung cancer to combined EBUS-TBNA and EUS-FNA. The trials enrolled over 100 patients per study arm and were designed to implement mediastinoscopy as a follow-up modality to the EBUS/EUS arm if no nodal metastases were detected on needle aspiration biopsy.

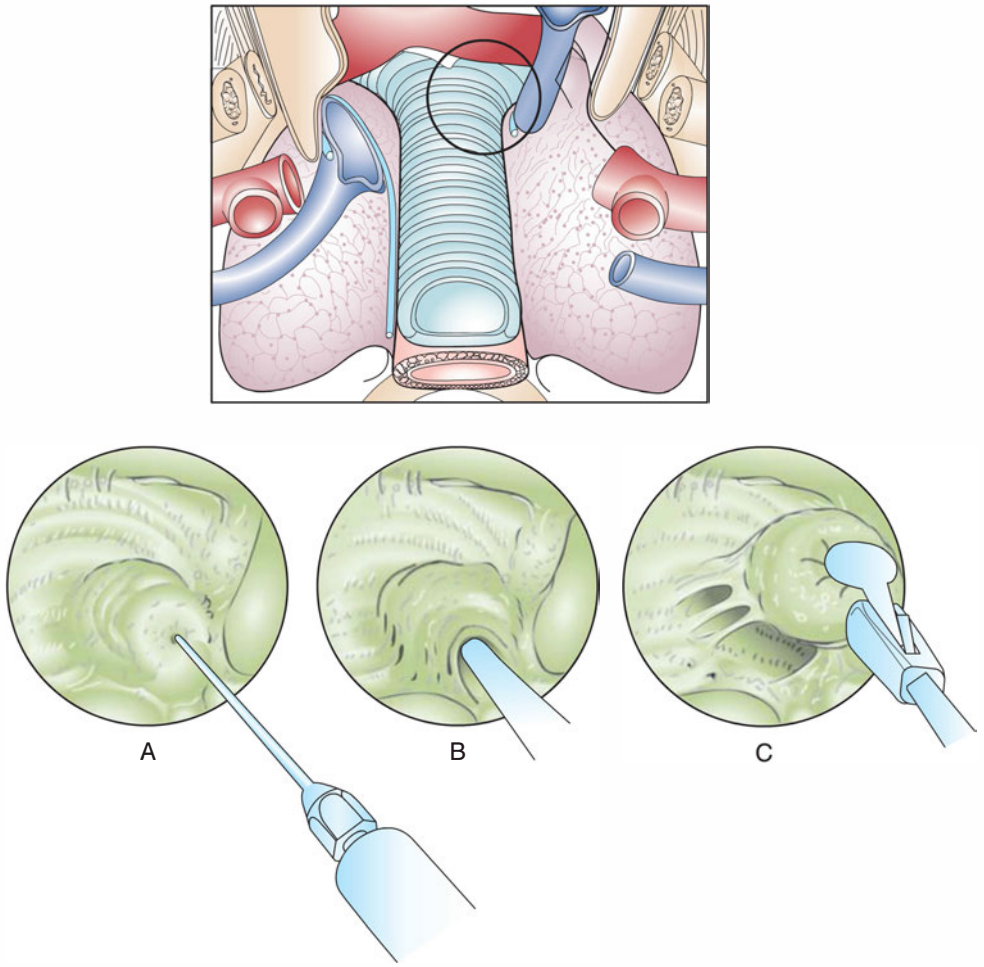


Fig. 2.11. The circle depicts the right tracheobronchial angle (station 4R). Tissue is identified and aspirated with a needle to ensure the absence of a vascular structure (**A**). An edge of lymph node is dissected free (**B**) and biopsied with a cup forceps (**C**).

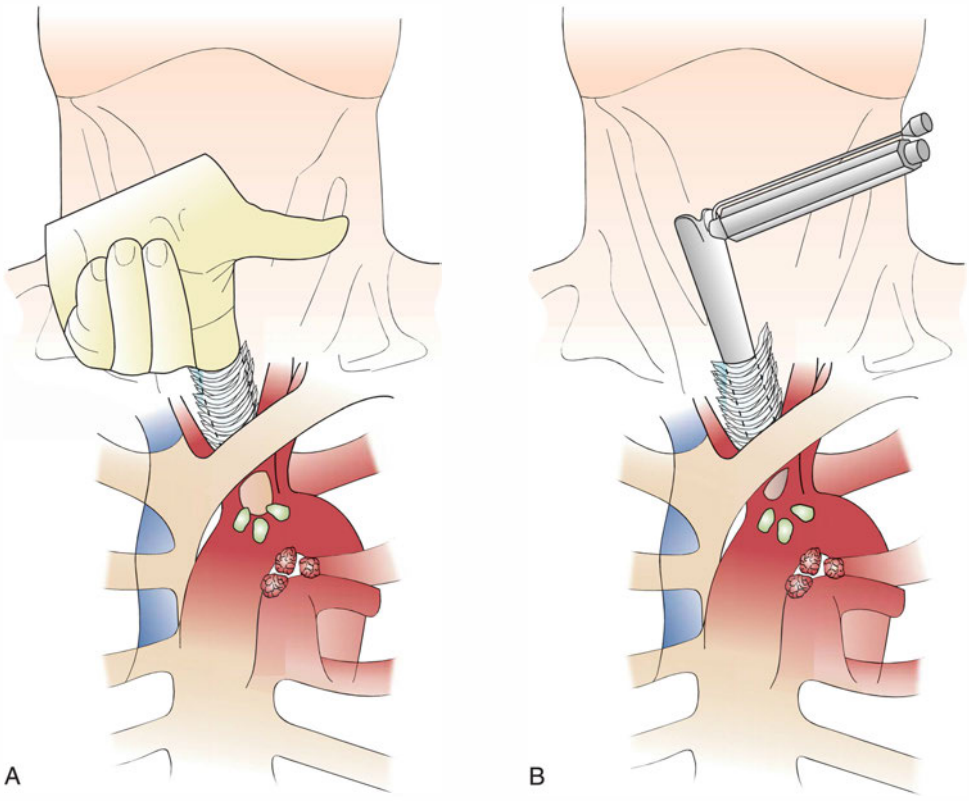


Fig. 2.12. Extended cervical mediastinoscopy: tunnel over the aortic arch is created (**A**) to reach the aortopulmonary window with the standard mediastinoscope (**B**).

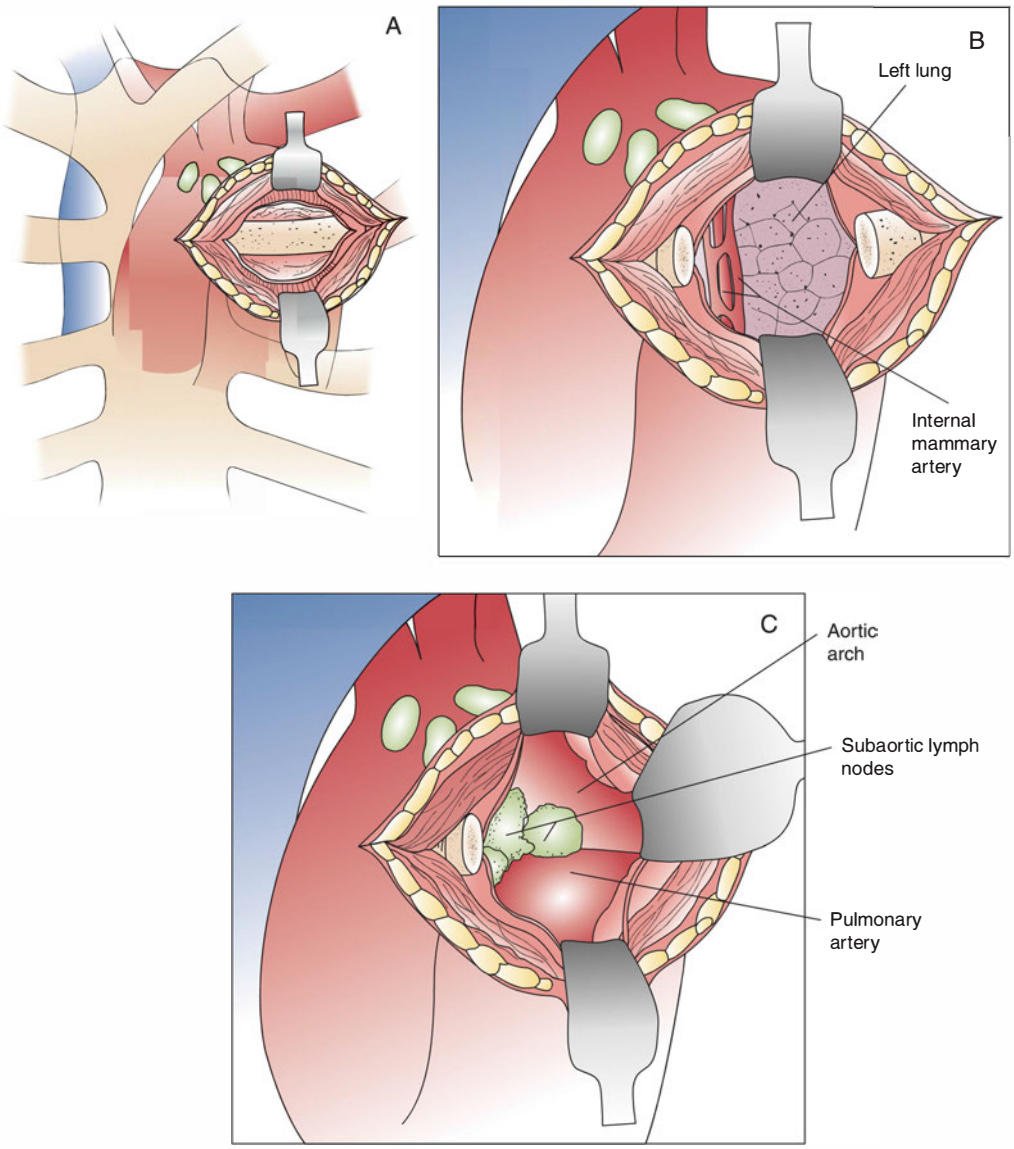


Fig. 2.13. Anterior mediastinotomy: transverse incision over the second costal cartilage (**A**). Cartilage is removed (**B**) exposing endothoracic fascia, then mediastinal pleura and internal mammary artery and vein. View of enlarged subaortic lymph nodes (station 5) (**C**).

In both of these randomized studies thoracotomy with lymph node dissection was performed when there was no evidence of mediastinal tumor spread. Both studies demonstrated that the endosonographic modalities (EBUS/EUS) and mediastinoscopy were complementary and not mutually exclusive in the comprehensive evaluation of mediastinal lymph nodes. Endosonography (followed by surgical staging if negative) resulted in greater sensitivity and improved negative predictive values for mediastinal nodal metastases and fewer unnecessary thoracotomies. EBUS can be used for nodal staging of lung cancer, restaging the mediastinum after neoadjuvant therapy, assessment of isolated hilar or mediastinal lymphadenopathy, and sampling of perihilar parenchymal masses. The American College of Chest Physicians recommends 50 supervised procedures to achieve proficiency; however, there is evidence to suggest that thoracic surgeons can achieve proficiency after 10 supervised procedures.

EBUS is also highly sensitive and cost effective for isolated mediastinal lymph node enlargement in diseases other than lung cancer including both benign and malignant disease (sarcoidosis, metastatic disease from extrathoracic sites, and even lymphoma). The sensitivity and negative predictive value of EBUS-TBNA in patients with isolated mediastinal lymph node enlargement is 92% and 40%, respectively.

Video-Assisted Thoracoscopy

VATS is another powerful modality in the armamentarium of thoracic surgeons for improving pathologic staging of locally

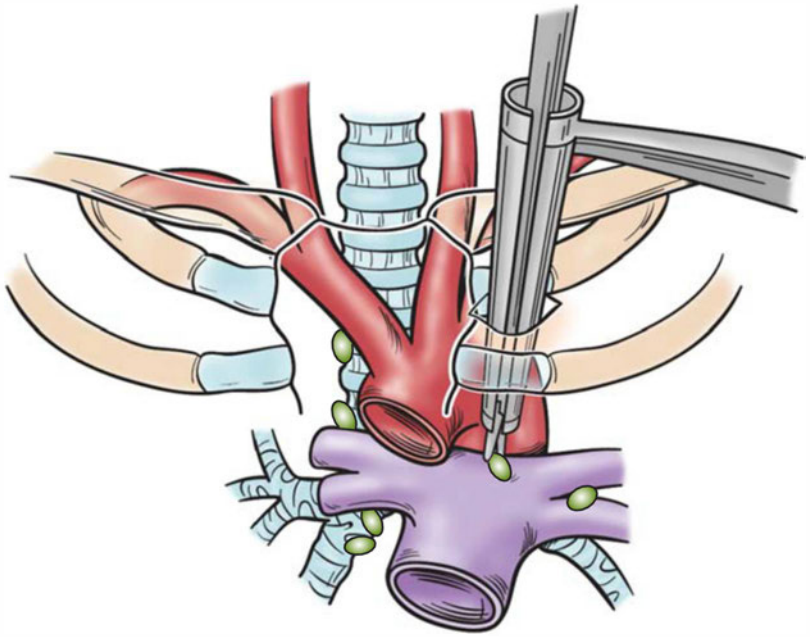


Fig. 2.14. Anterior mediastinoscopy: mediastinoscope is placed through a parasternal incision in the second intercostal space allowing access to aortopulmonary lymph nodes.

advanced lung cancer. It is frequently used at the time of pulmonary resection to rule out pleural metastases and more accurately assess T stage, particularly when the tumor abuts the chest wall, vascular, or mediastinal structures. VATS can also be used to assess enlarged prevascular (station 6) and aortopulmonary (station 5) lymph nodes in the setting of a left upper lobe NSCLC. More is discussed about this technique in another chapter.

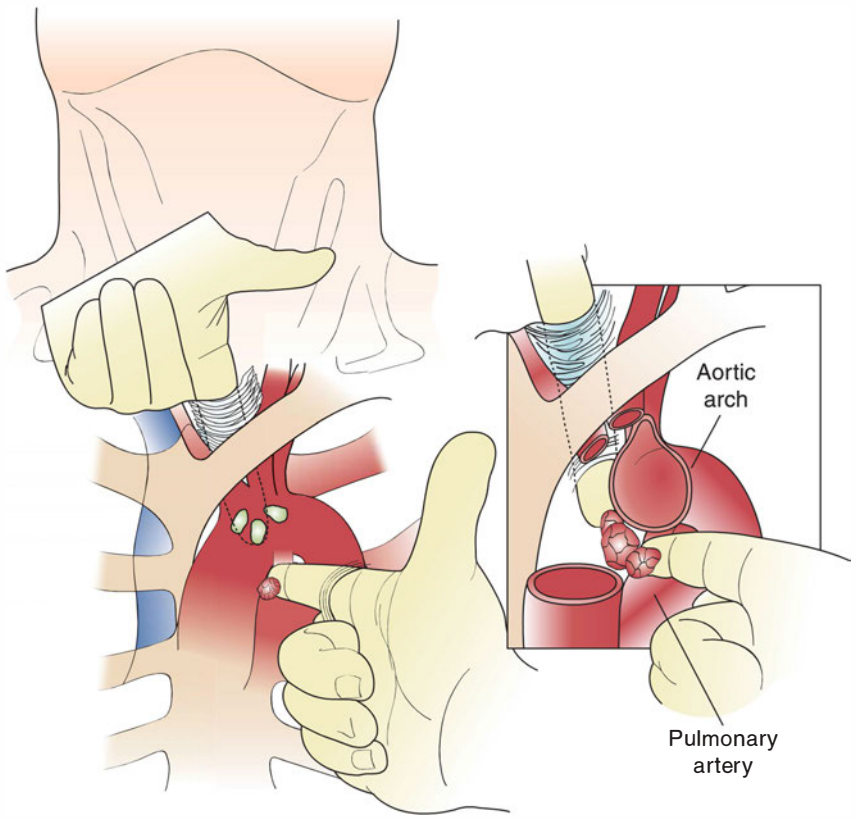


Fig. 2.15. Bidirectional digital palpation of the aortopulmonary region at the time of combined cervical and left anterior mediastinotomy.

CONCLUSION

All of these modalities must be included in the modern education of a thoracic surgeon including the endoscopic techniques of EBUS and EUS for mediastinal lymph node sampling. Each of these modalities is complementary and can be performed in an orderly sequence during a single general anesthesia.

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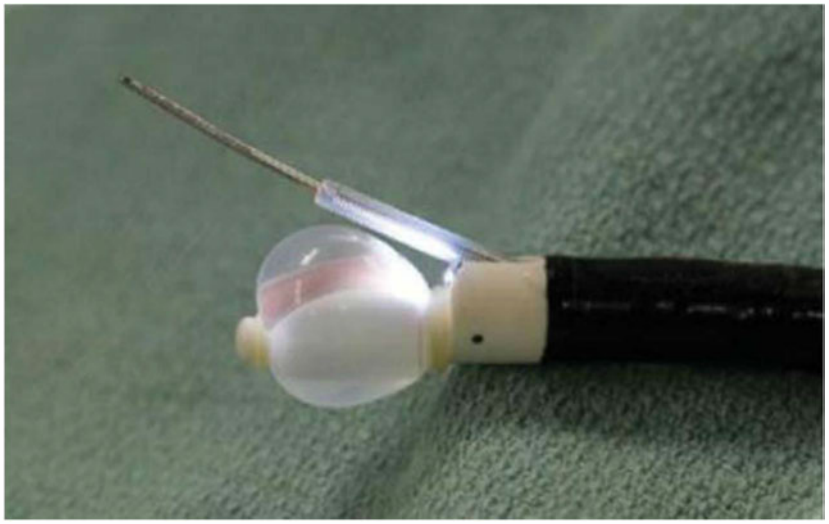


Fig. 2.16. Linear endobronchial ultrasound with balloon inflated and a 22-guage needle passed through the biopsy channel.

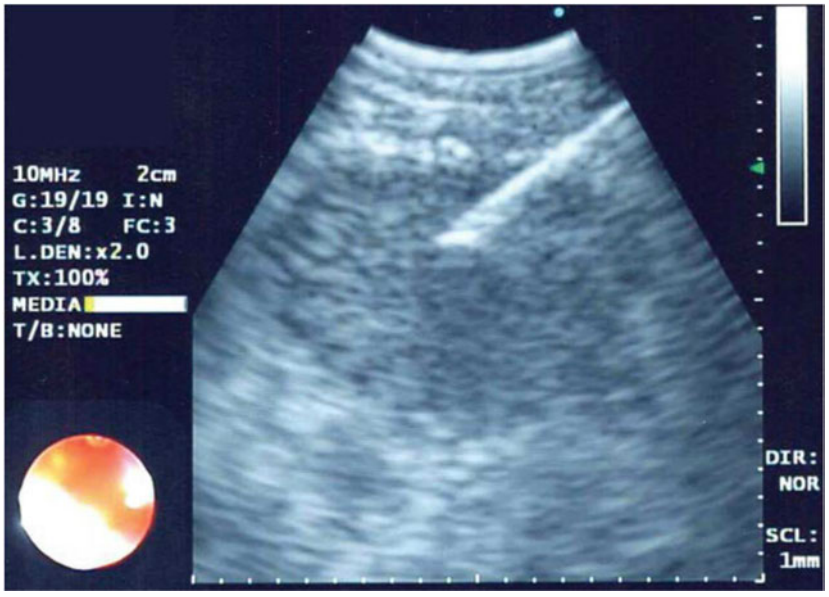


Fig. 2.17. Endobronchial ultrasound transbronchial needle aspiration (EBUS-TBNA) biopsy of subcarinal (level 7) mediastinal lymph node. (Origin Lanuti.)

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EDITOR'S COMMENTS

As Dr. Lanuti points out, mediastinoscopy remains the definitive procedure for staging the mediastinum in patients with nonsmall cell lung cancer. In addition, it remains the benchmark against which all other mediastinal staging procedures are compared. Endoscopic bronchial ultrasound-guided biopsy clearly has added to the armamentarium of the thoracic surgeon and provides visual confirmation of the lymph node being sampled. However, a negative needle aspirate of an enlarged lymph node even when the node is sampled under ultrasound guidance should not be accepted truly as negative until a sample obtained at mediastinoscopy confirms the finding. Not every patient requires separate mediastinal staging prior to resection, specifically if the fused CT/PET scan shows no enlarged lymph nodes and no increased uptake of radioisotope.

Mediastinoscopy is difficult to teach because it essentially is a single-operator procedure though the advent of the video mediastinoscope has made it somewhat easier. Part of the difficulty in teaching the procedure resides in the fact that the tactile sense is particularly important for optimal performance and that is difficult to convey even if the trainee can see what is being done on the video screen. The feel conveyed through the suction tip as it is used to dissect is critical to being able to distinguish lymph node from vessel. The azygous vein, for example, looks very similar to a lymph node, yet clearly there is a distinct difference in feel when the suction tip is pushed against it as compared to when touching the lymph node. The possibility of major vascular injury makes mediastinoscopy a formidable procedure that causes the inexperienced operator to be somewhat tentative in the exploration of the mediastinum. This tentativeness also results in many mediastinoscopies done where no nodal tissue is obtained, and this not only subjects a patient to needless risk but also does nothing to gain additional information that may be critical to establish the optimal treatment regimen.

As Lanuti points out, nodes at multiple levels should be sampled in order to obtain the maximum amount of information. However, one should avoid getting greedy during the procedure and recognize that the aim is not to perform a lymphadenectomy but to just obtain representative samples. Nodes from both the right and left side should be obtained specifically at levels 2 and 4 as well as a sample of the subcarinal (level 7) lymph node. When dissecting in the left paratracheal region, the left recurrent laryngeal nerve is at risk of injury and care must be taken when using the electrocautery. The left paratracheal nodes occur somewhat more cephalad than those on the right and pushing posteriorly with the tip of the mediastinoscope aids in identifying the left-sided nodes.

It should be kept in mind that it is okay to think when performing mediastinoscopy and not simply sample everything in sight. For instance, if a contralateral lymph node is suspected of being involved, tumor sampling of that node first may obviate the need for any further sampling if on frozen section the node proves to be positive. The disease at that point has been staged with the demonstration of N3 disease and further sampling to prove N2 disease is not only pointless but also subjects the patient to additional needless risk. If one is performing mediastinoscopy to assess enlarged lymph nodes in the absence of a primary lung cancer, the first demonstration of a pathologic lymph node on frozen section terminates the procedure.

The most common cause of bleeding during the procedure is not a major vessel injury but simply bleeding from a lymph node with the subcarinal space being the most common site. Use of electrocautery usually controls this but packing with a hemostatic agent such as oxidized cellulose may also be used. Intimate familiarity with the anatomy of the superior mediastinum gained by experience with the procedure usually serves to avoid major vessel injury but the azygous vein and the apical anterior branch of the right pulmonary artery are the vessels most at risk. Injury to the right main pulmonary artery or to the innominate artery is also possible though these structures are readily visible. The first indication of a major vessel injury occurs when the field of vision suddenly goes dark. The mediastinum should immediately be packed with gauze (vaginal packing serves this purpose well) and preparations made for a median sternotomy. After a waiting period of 10 minutes or so, the packing is moistened with saline and slowly removed. If bleeding has stopped, a further period of observation is warranted but if there is any doubt a median sternotomy should be performed and the vessel repaired. Continued bleeding after the pack is removed calls for repacking and immediate median sternotomy. Additional help should be summoned as well. Extended cervical mediastinoscopy is mentioned here only to be condemned, in my opinion. If access to the aortopulmonary window is desired either anterior mediastinotomy or video thoracoscopy should be the procedure of choice.

Anterior mediastinotomy provides access to the aortopulmonary window and may be accomplished in an extrapleural plane especially if a piece of the second costal cartilage is removed. One should keep in mind that level 5 (aortopulmonary lymph node) disease in the presence of node negativity in the rest of the mediastinum essentially behaves like N1 disease and whether it needs to be documented prior to resection is debatable. In my own practice, isolated level 5 disease is a straightforward surgical issue.

LRK

3 Thoracic Approaches

M. Blair Marshall

The optimal approach to the thorax depends on a number of variables including bony and hilar anatomy, location and extent of pathology, the objectives of the procedure, and the experience of the surgeon. Historically, the posterolateral thoracotomy was the workhorse for the majority of major thoracic procedures, the majority of approaches to thoracic pathology were performed through this, and there was significant short-term and long-term morbidity associated with the operations we performed. Today, with advances in surgical techniques, emphasis on less invasive procedures, optimization of perioperative care, and approaches to thoracic pathology have become more diverse and specialized. In addition to the prerequisite factors required for planning an approach, one may also consider alternative strategies for the management of lung isolation, positioning, and the inherent limitations of the instrumentation or the additional time required to complete a minimally invasive or multi phased procedure.

Given the short-term morbidity as well as chronic pain associated with traditional approaches, alternative approaches are becoming routine. These approaches may favorably affect morbidity, operative time, postoperative pulmonary function, muscle strength, cost, and postoperative pain. Although the specific costs associated with a minimally invasive approach appear to be increased, these are offset by shortened time in intensive care units and decreased length of stay. In the future, less invasive, tissue-sparing approaches may also demonstrate additional benefits as it relates to oncologic outcomes. It is not clear if this is a benefit of the approach in relation to decreased tissue damage and decreased cytokine response versus improved recovery and a thus earlier initiation of adjuvant therapy. Given the limited access provided through these minimally invasive approaches, careful preoperative planning and operative steps and strategy should be routinely visualized prior to performing the operation. Anticipation of intraoperative steps and exposure will go a long way toward preventing misadventures during the surgical procedure.

LUNG ISOLATION

For traditional and minimally invasive approaches, lung isolation with a double-lumen endotracheal tube remains the key maneuver to facilitate working within the pleural cavity. Given the length of the left main stem bronchus, left-sided tubes are the preferred choice, except in the setting of a left-sided sleeve resection.

At times, lung isolation through a double-lumen tube may be unnecessary or problematic, as in patients with an end tracheal stoma. Alternative lung isolation strategies include placement of a bronchial blocker or direct compression of the lung with CO_2 insufflation. When using a bronchial blocker, it is important to instruct the anesthesiologist to deflate the bronchial balloon and disconnect the ventilator. This will allow the ipsilateral lung to collapse more readily releasing any air that might be trapped behind the balloon.

Visualization within the pleural space may be facilitated with CO_2 insufflation if a single-lumen endotracheal tube must be used. In this setting, unlike that of laparoscopic surgery, a pressure of 10 mmHg is sufficient to collapse the lung and obtain a working space. Pressures >10 mmHg are often poorly tolerated due to the creation of tension physiology. Lesions accessible with this type of approach are simple wedge resections, as well as anterior and posterior mediastinal lesions. However, this technique is not suitable for complex pulmonary resections or any other circumstance where the complexity would necessitate absolute control of ipsilateral ventilation.

When working within the diaphragmatic hiatus via a laparoscopic approach, inadvertent entry into the pleural space is not uncommon potentially creating tension physiology. If hypotension ensues, it can be resolved by decreasing the insufflation pressure to 10 mmHg.

POSITIONING

The lateral decubitus continues to be the mainstay for the management of the majority of pulmonary pathology (Fig. 3.1), but with increasing frequency alternative positions are used to manage lesions in the region of the posterior or anterior mediastinum. With axillary approaches and also our preference for video-assisted thoracoscopic surgery (VATS) lobectomy, the opposite shoulder is elevated with a roll to expose the anterior axillary line. This allows better direct exposure through the intercostal space and also allows one to move the edge of the latissimus to a more posterior position minimizing the need to divide even a portion of this muscle (Fig. 3.2).

With either of these positions, the hip is placed below the area of flexion in the bed so that this allows for a slight widening of the intercostal space. It is critical when performing a chest wall reconstruction with prosthesis that the table should be taken out of flexion prior to securing the mesh in place. If this step is omitted, the mesh will not be caught and may result in a flail segment.

The semi-Fowler's position with the arms supported at right angles allows one to access both sides of the chest without a need to reposition the patient, as for a bilateral thoracoscopic sympathectomy (Fig. 3.3A and 3.3B). Apical bullectomy and pleurodesis may also be done through this approach. For additional approaches to the posterior mediastinum, as in the thoracoscopic portion of a minimally invasive esophagectomy, the prone position allows for excellent visualization of the posterior mediastinum while allowing the airway and lungs to fall away from the esophagus. For this position, the patient's arms are placed above his head so that the scapula is brought up as high as possible, minimizing interference with access to the chest. (Fig. 3.4A– 3.4C)

In contrast, access to the anterior mediastinum can be performed in the lateral decubitus as with traditional operations



Fig. 3.1. The patient is positioned on his side with the hip below the point of flexion in the bed. The lateral and posterior chest wall is optimally exposed for this approach. A limited muscle sparing thoracotomy is delineated by the *solid line* and a standard posterolateral approach by the *dashed line.*

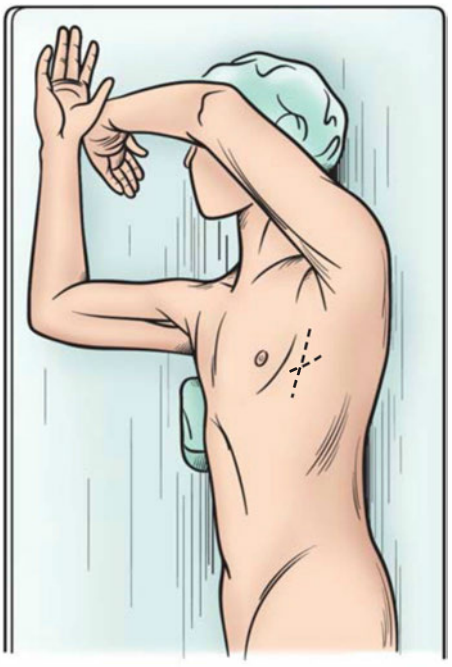


Fig. 3.2. The patient is again positioned in the lateral decubitus; however, the axilla is opened up and the anterior axillary line and ipsilateral breast are predominantly exposed. The axillary vertical muscle sparing incision oblique incisions are demonstrated.

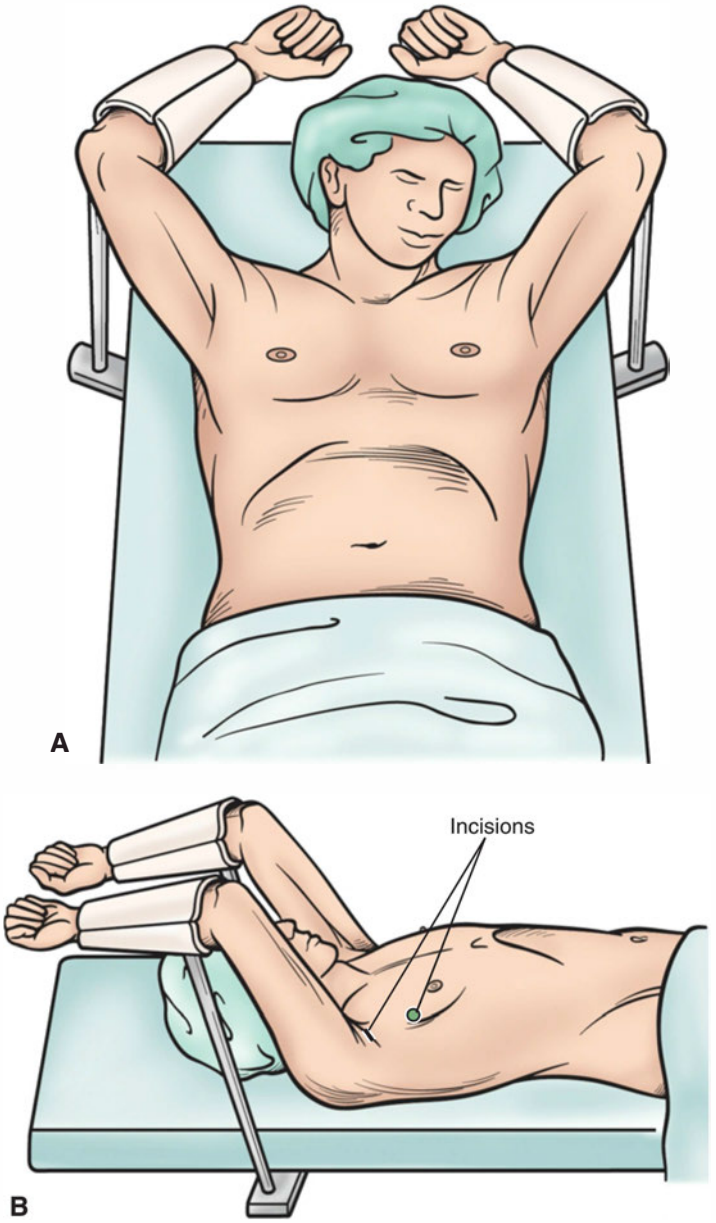


Fig. 3.3. (**A**) The patient is positioned with both arms elevated to expose the bilateral axilla, taking care not to extend the brachial plexus. (**B**) The location of the port sites for a video-assisted thoracoscopic surgery sympathectomy.

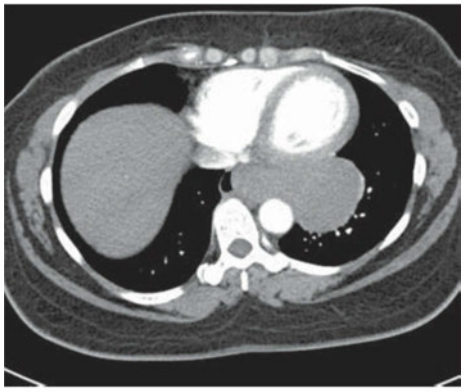
or in the supine or semi-supine position. The latter positioning allows for visualization of the contralateral side of the thymus when performing VATS resections. When utilizing such positioning, access to the lateral chest wall should be planned for in advance if a combined approach will be used. In this setting, we will usually place a roll under the side of the chest we wish to expose, and pull the arm and shoulder posterior so that the cervical as well as thoracoscopic approaches can be simultaneously used without the need to reposition (Fig. 3.5).

VIDEO-ASSISTED THORACOSCOPIC SURGERY APPROACHES

Historically, video thoracoscopic procedures started with simple thoracic procedures, and the ports were placed along the line of an anticipated thoracotomy incision. Currently, port positioning is more commonly chosen based on the objectives of the operation, and the limitations of the instrumentation as well as experience of the surgeon. The potential need to convert to an open procedure no longer is a major



A



B



C

Fig. 3.4. (**A**) The patient is positioned prone with exposure of the lateral and posterior chest wall. A vacuum bean bag may be used to rotate the bed with stabilization of the patient as well as to provide increased access to the lateral chest wall. (**B**) Posterior esophageal duplication cyst resected with the patient in the prone position. (**C**) Location of port sites with patient in the prone position are demonstrated.

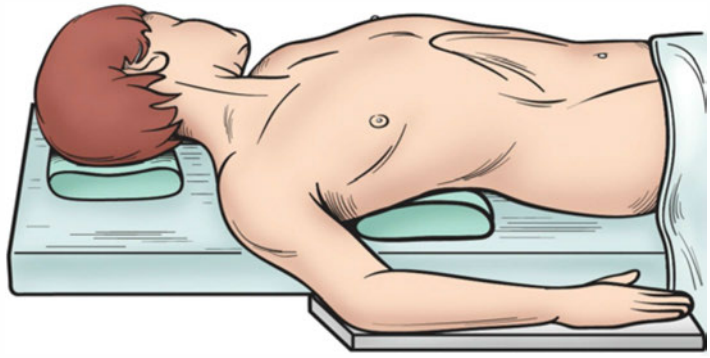


Fig. 3.5. The patient is positioned semi-supine with a support under the chest. This allows the ipsilateral arm to fall posterior to minimize interference with instrumentation.

consideration as ports have become much smaller and the need to convert to a standard posterolateral thoracotomy much less likely.

Today, the complexity of cases performed with this approach has increased significantly. Re-operations after previous thoracotomy, lobectomy with mediastinal lymph node dissection, thymectomy, and resection of mediastinal lesions are all performed under thoracoscopic guidance. In these settings, port sites are strategically located in order to minimize the challenges associated with instrumentation, the bony and hilar anatomy, as well as the pathology. Given this, one must alter port location, as well as patient positioning to facilitate not only exposure but also freedom of instrumentation. Although we typically use three-port sites, we will add a fourth when needed for more complex procedures. Upper extremity positioning or other hindrances may interfere with the full mobility of instruments, and these limitations should be anticipated when planning the operative procedure. In particular, when dealing with benign disease, where margins are less of a concern, a utility port is unnecessary as tumors may be morcellated prior to extraction (Fig. 3.6A and 3.6B).

Because the nuances associated with port positioning are critical to successful



A



B

Fig. 3.6. (**A**) Large benign solitary fibrous tumor. (**B**) Postoperative image of the port sites after resection of the large solitary fibrous tumor. The location of port sites was altered to facilitate dissection of the lesion at the posterior and inferior chest.

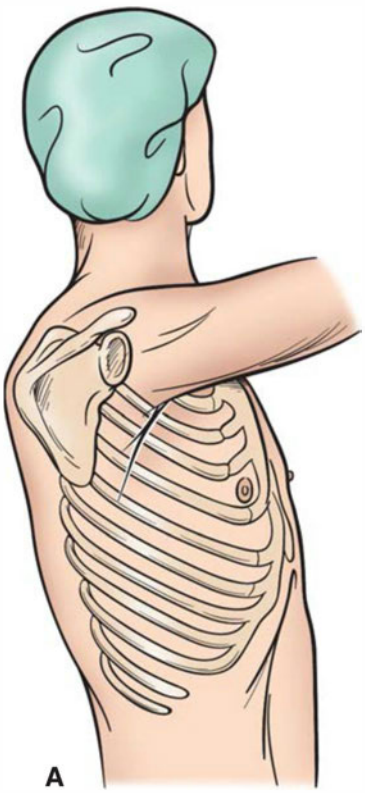
performance of the procedure, we will briefly discuss the minimally invasive approaches to specific pathology.

Video-Assisted Thoracoscopic Surgery Wedge Resection

For any video-guided surgery, triangulation of port sites is essential to minimize instrument interference and improve visualization. Port placement is fairly standard with two usually placed along the anterior axillary line and one posterior below the tip of the scapula. We try to minimize the posterior location as the intercostal spaces are narrower and trauma to the intercostal nerve may be more likely to occur. However, if one is planning for a VATS wedge resection with subsequent lobectomy with lymph node dissection, it is important to locate the superior port along the planned incision for the utility port. In our experience,

we typically use two 5-mm ports for the superior ports and a 10-mm port for the inferior port along the anterior axillary line. The intercostal spaces are larger here to allow for placement of a stapler and this site functions as the subsequent chest tube site (Fig. 3.7A and 3.7B).

For complex procedures, port site location is modified based on anatomic and clinical factors. One must identify where the target lesion is to be found. If direct palpation is necessary to identify the lesion, we typically place the larger port site specifically within "finger reach" of the target lesion. We tend to keep two-third of the port sites in the 5 mm range when possible. In this setting, it is only possible to palpate through one port or pass the stapler for a wedge excision through one port and the angle and trajectory that this port takes must be taken into account. Another critical factor is the width of the patient's intercostal spaces, as the more posterior aspect of the space is significantly narrower. We do not hesitate to enlarge an incision when necessary or resect a small segment of rib to accommodate direct palpation when one cannot identify the lesion. For any thoracoscopic procedure, the surgeon should be positioned on the side of the camera to facilitate orientation with the projected image.




B

Fig. 3.7. (**A**) The patient is in the lateral decubitus position with the typical planning for video-assisted thoracoscopic surgery port sites demonstrated. (**B**) Operative photograph demonstrating operative planning for a typical video-assisted thoracoscopic surgery procedure.

Video-Assisted Thoracoscopic Surgery Lobectomy

For a VATS lobectomy, when placing the port sites for the diagnostic portion and then proceeding with a lobectomy, the anterior–superior port site is placed overlying the superior pulmonary vein for an upper lobectomy and above the pulmonary artery within the fissure for a lower lobectomy, as this will become the location of the utility port. There is great variation among surgeons as to the optimal location of ports for VATS lobectomy. We typically use two incisions along the anterior axillary line, one for the utility port as described above and the inferior one at the seventh intercostal space for the camera. We use a second 5-mm port posteriorly approximate one inner space below the tip of the scapula. We tend to again manipulate the patient's ipsilateral elbow to open up the axilla, as in an axillary thoracotomy, as this allows the utility port to be muscle sparing as the edge of the latissimus dorsi muscle is reflected posteriorly and the serratus anterior muscle is split along the intercostal space. We use a soft tissue retractor as it minimizes the potential trauma from metal retractors and the exposure it provides, especially in the larger patient, is unparalleled (Fig. 3.8A and 3.8B).

Although some thoracic surgeons perform the entire procedure without moving the camera position, we typically will place the camera through the posterior port site in order to perform the posterior dissection of the airway as well as to dissect the level 7 lymph nodes.


A

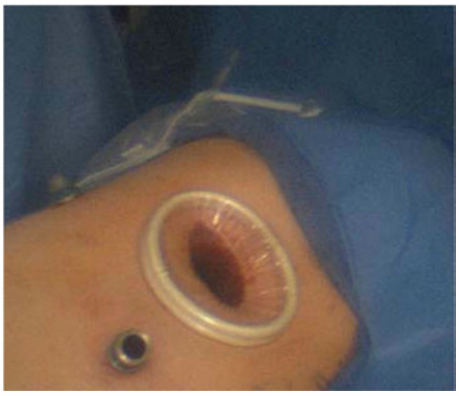

B

Fig. 3.8. (**A**) An operative photograph demonstrating the video-assisted thoracoscopic surgery lobectomy port sites. (**B**) A view of the soft tissue wound retractor in place.

VIDEO-ASSISTED THORACOSCOPIC SURGERY APPROACH TO THE ANTERIOR MEDIASTINUM

For the VATS approach to anterior mediastinal lesions, location of port placement is dependent upon the overall strategy and position of the patient. We tend to use a semi-supine position and combine a transcervical thymectomy with a left-sided VATS for thymectomy as the aorto-pulmonary window can be particularly challenging through a limited transcervical approach (Fig. 3.9). As well, for complex lesions or in the setting of mediastinal resection in patients who have undergone previous sternotomy, direct exposure is critical and we have found placement of a port in the infraclavicular fossa to be a useful position for access to the mediastinum. In this setting, the patient is either positioned supine or if in the lateral decubitus, the ipsilateral upper extremity is positioned posterior

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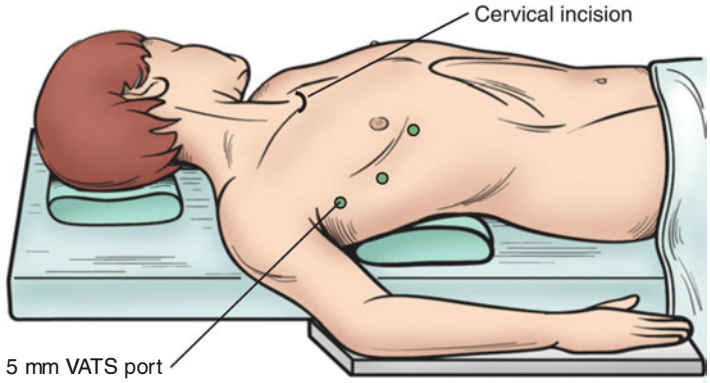


Fig. 3.9. Illustration of positioning and incision location for the combined transcervical and left video-assisted thoracoscopic surgery approach to thymectomy.

rather than anterior to open the access to that space. When working on lesions where margins are not an issue, there is no need for a utility port as the lesion can be morcellated and brought out through a 10-mm site (Fig. 3.10A and 3.10B).

Video-Assisted Thoracoscopic Surgery Approach to Posterior Mediastinum

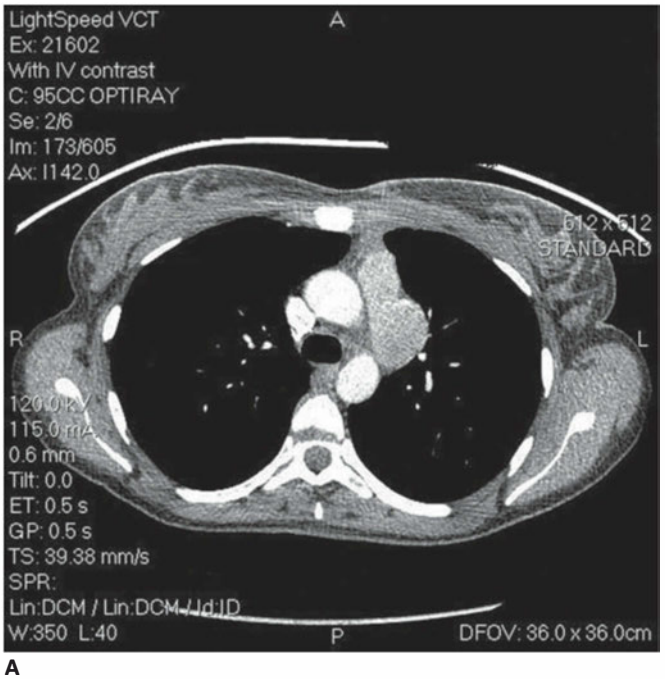
Positioning for posterior mediastinal approaches is dependent upon the pathology and its location, as port placement for an esophagectomy, sympathectomy, or repair of a Bochdalek hernia will vary greatly. For a sympathectomy or other lesions at the apex of the chest, we use a modified semi-Fowler's position. This allows for bilateral exposure with a single position, eliminating the additional operative time required to change the patient's position (see Fig. 3.3A and 3.3B).

The lateral decubitus or prone position for thoracic esophageal dissection or other lesions provides excellent exposure of the esophagus. Again, benign lesions may be approached through well-placed minimally invasive ports with morcellation used for extraction (Fig. 3.11A– 3.11D).

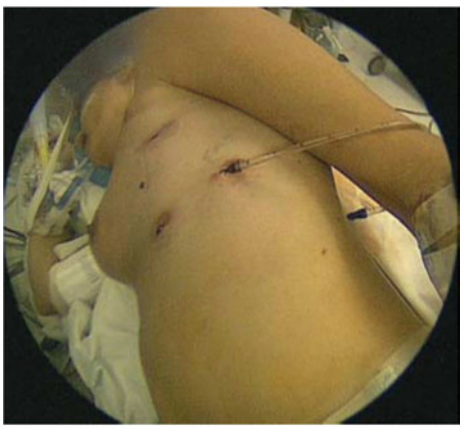
Although for some there is a concern that one should be able to "bail out" with a posterolateral thoracotomy if necessary, we have not found this to be the case. One can convert to a lateral thoracotomy if necessary to manage bleeding or if more extensive disease than expected is found. Since in this position the lateral and posterior chest is exposed, an incision can be made, the intercostal muscles are divided, and ample open access to the chest is created. The more of these alternative approaches one uses, the more comfortable one becomes with handling these issues as they arise.

ROBOTIC APPROACHES

Although it may not have yet been reported, essentially any operation that can be performed with VATS can be performed with the robot. The challenges of nonrobotic thoracoscopic approaches lie in the lack of articulation of the instruments as well as the limitation of two-dimensional imaging. Robotic surgery mitigates these challenges given the 360 degrees of freedom offered by the instruments and the binocular camera that produces 3D visualization. However, repositioning of the camera or instruments through different port sites, when this is needed can be a challenge. This has been improved with the DaVinci SI, as the technology allows a fourth arm so that the surgeon can alternate between instruments thus facilitating the technical performance. The ideal pathology for a robotic approach allows for the entire dissection to be performed from one strategic positioning of the camera and robotic arms, thus negating the challenges of this approach. In this setting, port placement is typically performed in a gentle arc so that the instruments focus on the area of dissection.



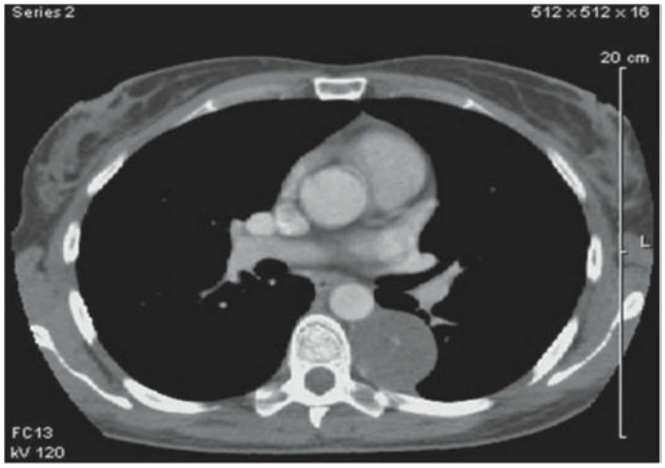
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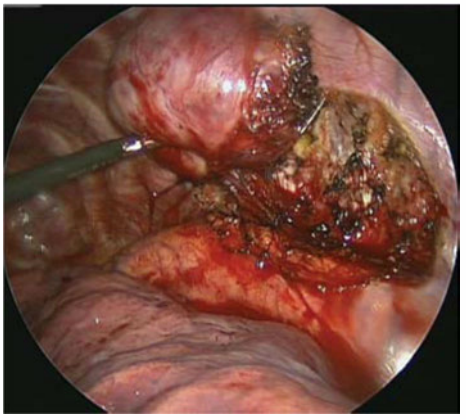
B

Fig. 3.10. (**A**) Computed tomography demonstrating a benign central anterior mediastinal lesion (Castlman's). (**B**) Photograph demonstrating posterior positioning of the ipsilateral arm providing access to the axillary space and room for laparoscopic instruments. The superior infraclavicular port site can be seen just below the clavicle.

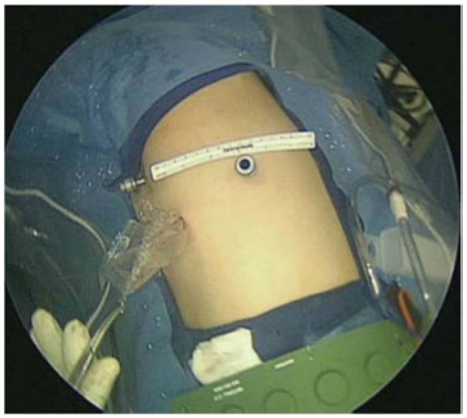
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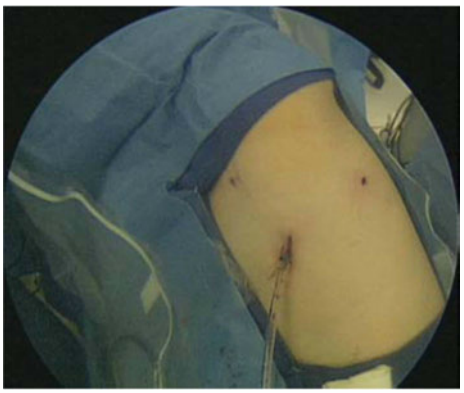
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D

Fig. 3.11. (**A**) Computed tomography demonstrating a posterior mediastinal schwannoma. (**B**) Intraoperative view through a 5-mm camera of the resected schwannoma video-assisted thoracoscopic surgery resection of a large schwannoma. (**C**) The lesion is placed in a sac and the edges of the sac are retrieved through the 10-mm port site. The mass is morcellated prior to extraction. (**D**) The 10-mm port site is used to place the chest tube.

TRANSCERVICAL APPROACH

The transcervical approach is used in the management of a number of thoracic pathologies. Access to the neck, as well as posterior and anterior mediastinum can be achieved through a cervical incision. For thymectomy, the patient is positioned supine for the transcervical approach or with a roll under the left chest for a combined approach with VATS. An inflatable pillow is placed behind the shoulders to maximally extend the neck and allow the surgeon to work sitting at the head of the table. One must make sure that in elderly patients, they can sufficiently extend their neck to allow for adequate exposure.

A curvilinear incision is made approximately one finger breadth above the clavicular heads. The platysma is divided and sub platysmal flaps are elevated from sternal notch to the thyroid cartilage. Once this has been done, the strap muscles are separated in the midline exposing the thymus.

When exposing the esophagus for esophagectomy or a Zenker's diverticulum, the cervical incision is made higher usually within a skin crease for improved cosmesis. Although the traditional incision for this approach has been an oblique incision along the border of the sternocleidomastoid muscle, the transverse incision allows for improved cosmesis in a highly visible area and does not limit the exposure (Fig. 3.12A and 3.12B).

POSTEROLATERAL THORACOTOMY

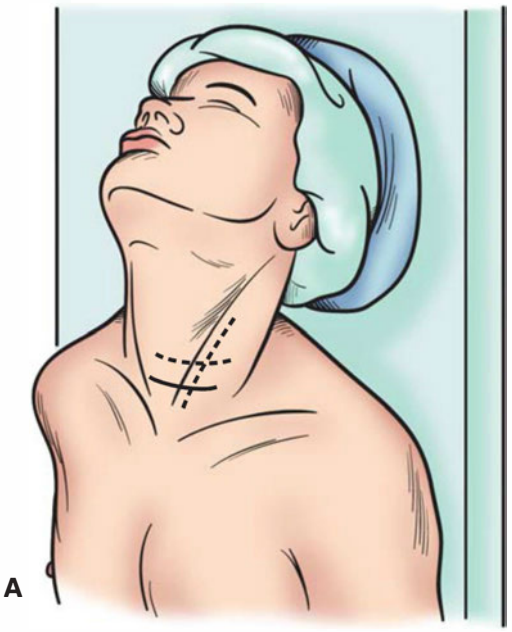
The posterolateral thoracotomy historically has been the most widely used approach for thoracic pathology. Although any pulmonary resection may be performed through this incision, which has led to its particular versatility, muscle sparing options and less invasive techniques have, in many centers, begun to replace this as the routine approach.

In positioning, it is important to prepare the skin over a broad area and place the drapes widely, particularly along the posterior spine. One must appropriately pad and support the upper and lower extremities so as to avoid inadvertent neurologic or pressure injury. Care must be taken with the elbows and wrists.

The landmarks for the posterolateral thoracotomy include the spine and the scapula. A gentle curve is drawn at the midpoint between the posterior border of the scapula and spine to approximately one fingerbreadth below the tip of the scapula over to the anterior axillary line (see Fig. 3.1). We tend not to use the full extent of this incision for most procedures so as to minimize tissue trauma and improve healing. Once the skin and subcutaneous tissues are divided, the latissimus dorsi is divided as well. Anteriorly, we routinely spare the serratus anterior muscle by freeing the fascial attachments to the chest wall and between the muscle and the latissimus dorsi muscle; this is done obliquely along the chest wall to avoid division of the serratus fibers. Depending on the posterior extent of the incision, additional thoracic muscles such as the trapezius or rhomboids may need to be divided. This is particularly important when elevating the scapula off of the chest wall as in a chest wall resection or posterior approach to an apical lung tumor.

The scapula is elevated with a right-angled retractor, and the ribs are counted by manual palpation. Relying on the first rib can be misleading because the posterior first rib may be obscured. One can avoid errors by using the insertion of the

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A



B

Fig. 3.12. (**A**) Cervical approaches to the neck and superior mediastinum. A support is placed behind the shoulders to extend the neck. The *solid line* represents the incision for a transcervical thymectomy or tracheal resection and the *dotted lines* represent approaches to the esophagus. (**B**) Intraoperative photograph demonstrating the superior horns of the thymus through the cervical incision.

posterior scalene muscle onto the second rib as a reference to begin the rib count.

A fifth intercostal space thoracotomy incision is made by dividing the intercostal muscles directly above the superior margin of the sixth rib. This avoids injury to the neurovascular bundle that lies just inferior to the fifth rib in a slightly recessed groove. The division along the intercostal muscles extends fairly far beyond the limits of the skin incision to maximize rib separation. In patients with brittle ribs, excising a subperiosteal segment of the sixth rib at the costovertebral angle, the so-called shingling," can help to avoid rib fracture. A 1-cm segment is removed to minimize discomfort from periosteal friction following reapproximation. Resection of larger segments of rib is reserved for repeat thoracotomies. In this situation, entering the pleural space through the bed of the resected rib improves exposure for adhesiolysis of the pleural space. In approaching diaphragmatic pathology, the seventh or eighth intercostal space is suitable for optimal exposure.

Once the chest is entered, a Finochietto-type retractor is used to separate the ribs. For smaller incisions, smaller retractors provide adequate exposure. On completion of the procedure, a single modified thoracostomy tube usually provides adequate drainage. Additional drainage holes are cut in the tube, and it is tunneled transversely so as to lie in the diaphragmatic sulcus and course posterior to the apex of the chest; thus, it functions as both a basilar and an apical tube. Large absorbable sutures are used to reapproximate the ribs. One must be careful not to over approximate the ribs because this may contribute to postoperative pain. The remainder of the incision is closed meticulously in layers with absorbable sutures.

This incision is suitable for pulmonary resections and extrapleural pneumonectomies. For esophageal disease, depending on the location of the pathology, the incision may be made an innerspace or two lower in order to get access to the hiatus. In this setting, it is useful to place a traction suture on the diaphragm in order to retract it out of the way. In addition, when on the left side, one may place a suture in the pericardium to pull the heart up and out of the way improving exposure in a challenging area. When performing an extrapleural pneumonectomy, often an additional thoracotomy, two intercostal spaces below can be divided to improve access for the diaphragmatic resection as well as the reconstruction.

AXILLARY THORACOTOMY (MUSCLE SPARING)

The axillary thoracotomy has replaced the posterolateral thoracotomy as my incision of choice for open resections. Because of the inherent limitations of exposure, it did not initially gain widespread use. Improved stapling devices and increased surgical experience have demonstrated its superiority. The advantages of this incision are several: (1) the major thoracic muscles are left intact, (2) there is increased ease and speed of both opening and closing the chest, and (3) cosmesis is improved. Because the incision is small it may be difficult for two surgeons to have adequate simultaneous visibility. In addition, most posterior chest wall resections are more optimally suited for a posterolateral thoracotomy because elevating the muscles and scapula off of the chest wall facilitates the dissection. We have found that having a thoracoscope within a future chest tube site allows the scrub nurse, student, and others to follow the operations. In addition, the camera facilitates division of the inferior pulmonary ligament which on occasion may be a challenge with the incision being kept particularly small.

Some authors have suggested that complex resections, such as bronchial sleeve resections or pulmonary arterial reconstructions, should not be performed through this approach, but we have not found this to be the case. We routinely perform complex procedures through this incision.

The patient is positioned in the lateral decubitus position with a few adjustments. The elbow is rotated cephalad to open up the axilla (see Fig. 3.2). The body is rotated posterior so that more of the anterior and lateral chest is exposed. The skin incision for this approach may be oriented vertically or obliquely. A 5- to 7-cm incision is made along the anterior axillary line aligned with the anterior–superior iliac spine centered usually on the nipple in male patients or the fourth intercostal space. One should avoid placing this incision too far posterior because injury to the long thoracic nerve may occur. The intercostal brachial nerve runs in the superior half of the incision and should be preserved if possible. Patients should be counseled preoperatively that

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they may have numbness in the area of distribution of this nerve. In addition, numbness along the lateral breast is common with this incision, though this eventually resolves.

After the subcutaneous tissues are divided, the pectoralis major muscle is undermined and the anterior border of the serratus anterior muscle is visualized. Often the pectoralis minor muscle must be reflected in order to expose the anterior insertions of the serratus muscle. The serratus insertions are mobilized off the ribs by reflecting the attachments or split along the fifth interspace. Thus, the insertions of the serratus anterior muscle are mobilized and reflected off of the fourth and fifth ribs. Because this is essentially an anterior incision for pulmonary resections, we enter the chest through the fourth intercostal space, whereas for thoracic tracheal resections we enter through the third. Because this incision is anterior, one cannot rely on the posterior scalene muscle for counting the ribs. The first rib is identified by palpating the tubercle. If one is unsure of the first rib, one can reach over the superior border and identify the thoracic outlet because there are no intercostal muscles above the first rib.

The intercostal muscles along the appropriate space are divided as in a posterolateral thoracotomy. The intercostal incision is carried well beyond the limits of the skin incision, almost to the level of the vertebral bodies. The incision in the intercostal space is kept away from the overlying chest wall muscles and is facilitated by progressive opening of the rib spreader, which provides countertraction. A pediatric Finochietto-type rib spreader is used to separate the ribs, and a Balfour retractor is placed perpendicular to the rib spreader to retract the skin and subcutaneous tissues. The skin is quite pliable, and thus excellent exposure can be achieved through this relatively small skin incision because the intercostal incision essentially rivals or even exceeds that made with a posterolateral incision. At the completion of the procedure, a single chest tube is placed, pericostal sutures are placed to reapproximate the ribs, the insertions of the serratus anterior muscle are sewn to the pectoralis minor muscle anteriorly, and the rest of the closure proceeds as usual.

ANTERIOR THORACOTOMY

With recent trends toward minimally invasive cardiac surgery, the anterior thoracotomy has regained an element of popularity. Otherwise, it remains the incision of choice for open-lung biopsy and emergent thoracotomy. Open-lung biopsy, in contrast to a video procedure, may be the approach of choice in hypoxic, ventilated patients who require lung biopsy. These patients may not tolerate single-lung ventilation, and this procedure can be performed while both lungs are being ventilated by a single-lumen endotracheal tube.

The patient is positioned with a roll under the operative side. The ipsilateral arm is placed over the patient on an arm rest or alongside on a support. An incision is made from the anterior axillary line curving under the breast toward the sternum. The fourth or fifth intercostal space is entered. For an open-lung biopsy, we make a fairly limited incision, approximately 3 cm in size. For an emergency thoracotomy, the incision is obviously much larger and can be extended across the sternum to increase exposure as needed (Fig. 3.13).

MEDIAN STERNOTOMY

The median sternotomy is most commonly used for cardiac surgical procedures but can be a useful tool in general thoracic surgery operations requiring access to both pleural spaces or for resection of mediastinal disease.

The patient is placed supine with one or both arms tucked at the side. A roll is placed behind the shoulders to extend the neck and expose the sternal notch. The midline of the sternum is marked, and a knife divides the skin (Fig. 3.14). The subcutaneous layers are divided though the decussating fibers of the pectoralis major muscle, and the midline of the sternum is carefully marked with cautery. Palpating the intercostal spaces along the sternum allows one to accurately identify the midline. The interclavicular ligament is divided, and the retrosternal space is bluntly dissected. The xiphoid tip is dissected, and again the space posterior to this is developed. A reciprocating saw is used to divide the sternum. The bleeding periosteal edges are carefully and judiciously cauterized. An appropriate sternal retractor is used to gradually spread the two sternal edges.

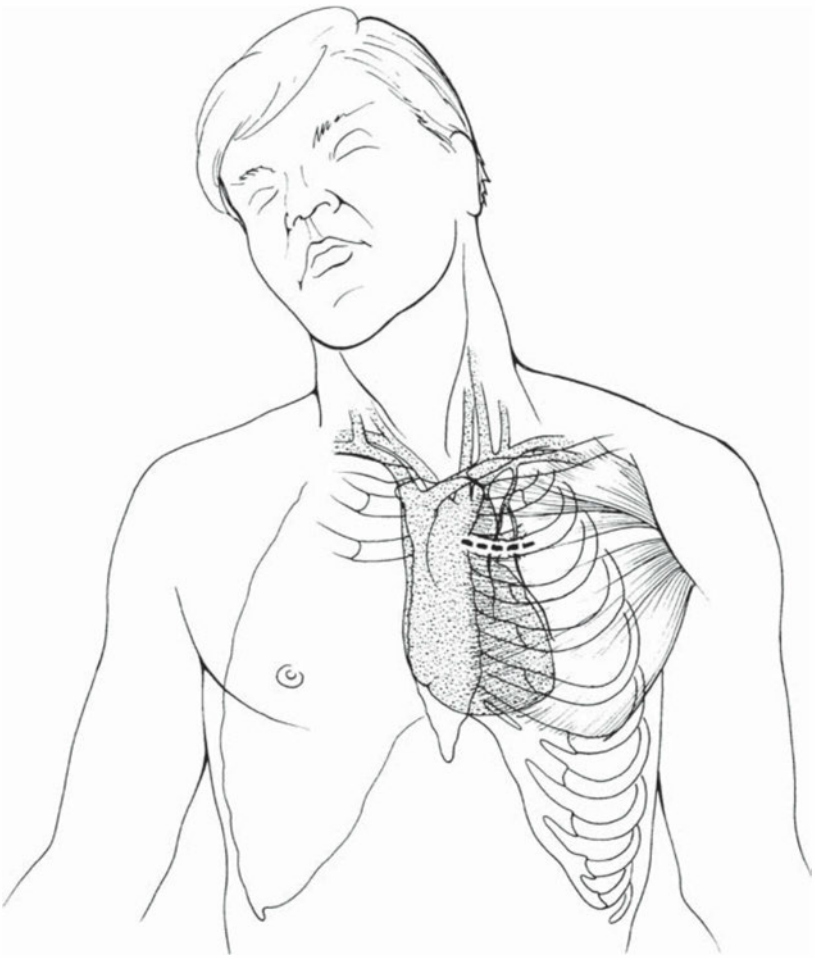


Fig. 3.13. The patient is positioned supine, with a support for the ipsilateral side. The incision for an anterior thoracotomy is illustrated. Depending on the purpose for this incision, the patient's position and the length of the incision can be modified.

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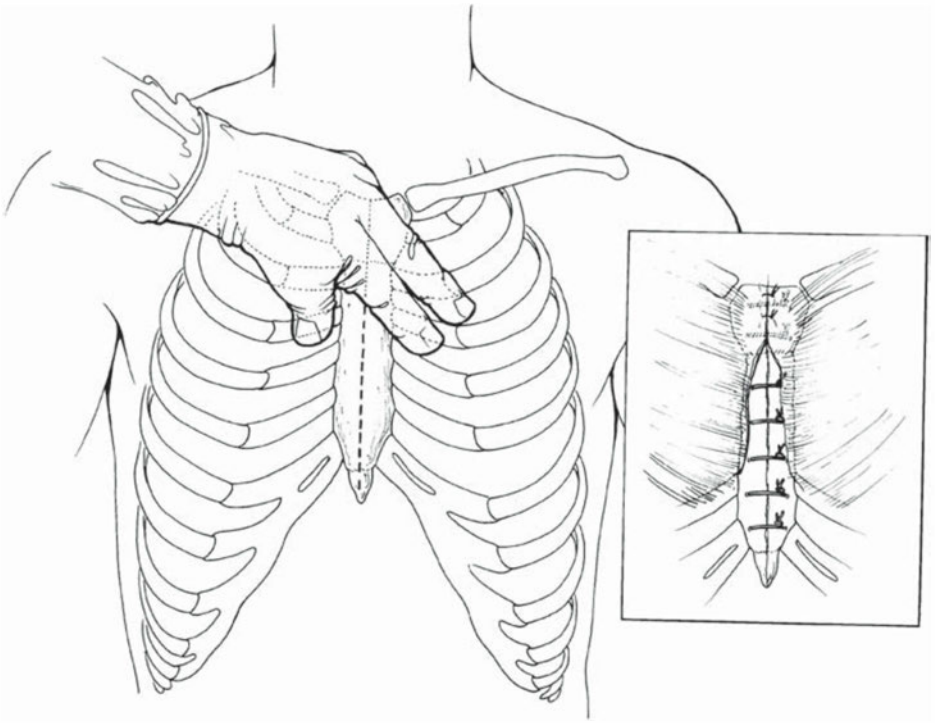


Fig. 3.14. For the median sternotomy, the patient is positioned supine and a roll is placed under the shoulders to optimize exposure of the sternal notch.

large tumors of the anterior mediastinum, although admittedly it is used sparingly.

A transverse skin incision is made extending from one anterior axillary line along the inframammary crease to the other anterior axillary line (Fig. 3.15). The subcutaneous tissues are divided, and the fourth or fifth intercostal space is identified. The intercostal muscles along this space are divided, and the internal mammary vessels are identified and ligated prior to dividing the sternum. The Lebsche knife is used to divide the sternum transversely. Rib spreaders are usually placed on both sides to allow for maximal exposure. After the procedure, paracostal sutures are placed and the sternum is reapproximated with wire.

THORACOSTERNOTOMY (HEMICLAMSHELL)

The thoracosternotomy is a useful approach for large central pulmonary lesions and other special situations. Such tumors can interfere with one's access to the hilum, making the dissection and proximal control

Brachial plexus injuries may occur when the retractor is opened too quickly or too widely.

At the conclusion of the procedure, one or two chest tubes are placed. Wires are used to reapproximate the sternum. We place two in the manubrium and four around the sternal edges. The pectoralis fascia is closed over the sternum, and the remainder of the wound is closed in layers. One should thoroughly irrigate the wound and carefully obliterate any potential spaces at the time of closure to attenuate the possibility of infection. Newer sternal closure systems that employ plates and screws may also be used for sternal closure and may be especially useful for the more fragile sternums.

BILATERAL THORACOSTERNOTOMY (CLAMSHELL)

The bilateral thoracosternotomy had previously been the incision of choice for bilateral lung transplantation. Due to complications relating to the transverse sternal division, most centers have abandoned this technique in transplant patients. However, it remains a very useful approach when wide access to both pleural spaces is required, as in selected patients with bilateral pulmonary metastases or those with

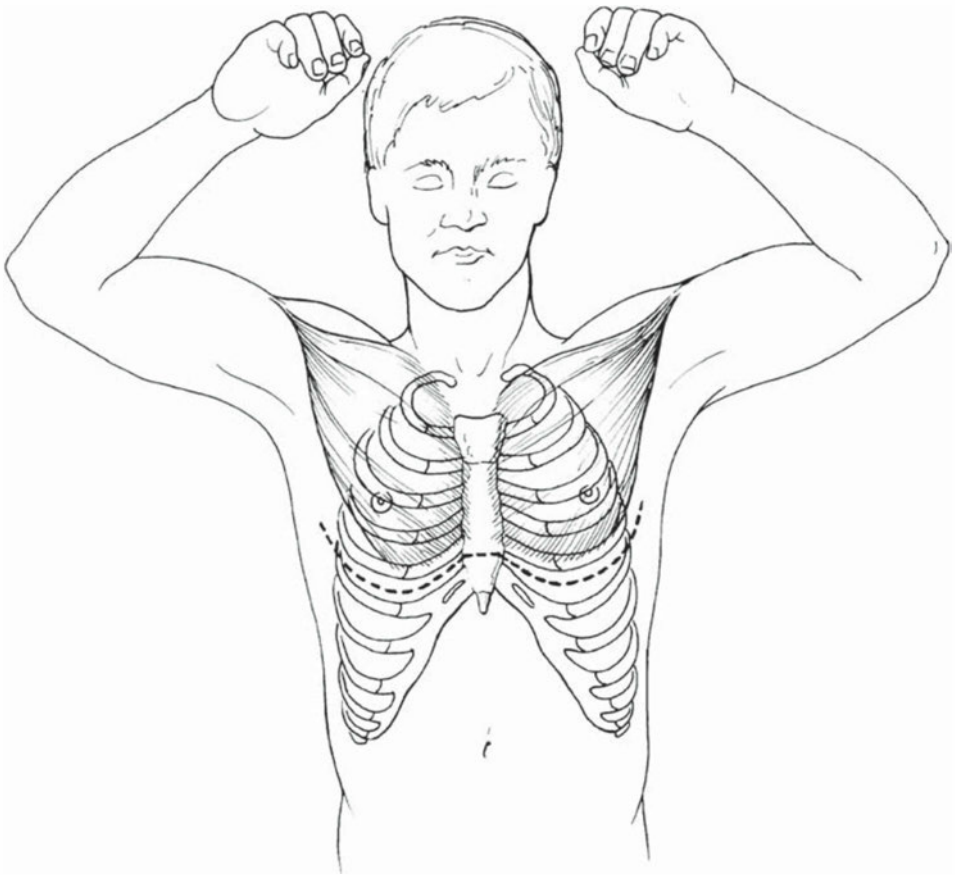


Fig. 3.15. The clamshell incision is illustrated coursing along the inframammary creases and across the sternum.

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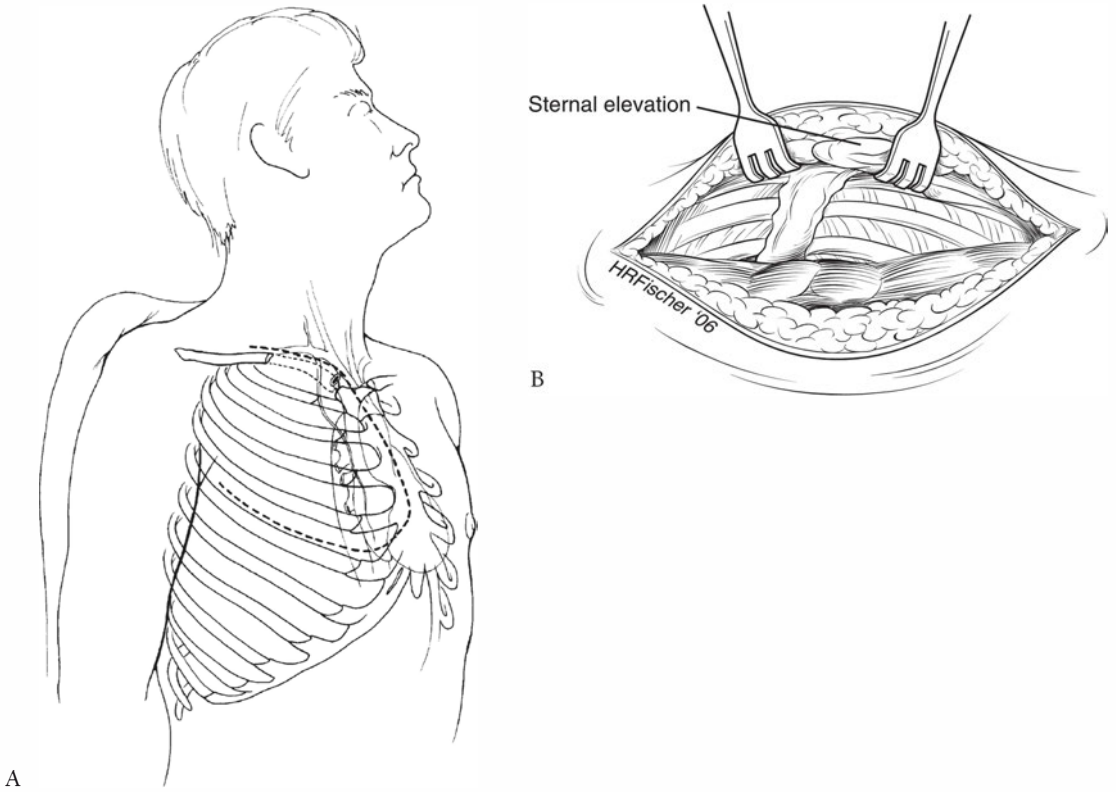


Fig. 3.16. (**A**) The hemi clamshell incision incorporates the sternotomy with extension to a thoracotomy to increase the intrathoracic exposure. This incision can also be extended along the sternocleidomastoid when exposure of the brachiocephalic vessels is required. (**B**) A hemisternal retractor is used to elevate the sternum and increase the intrathoracic exposure.

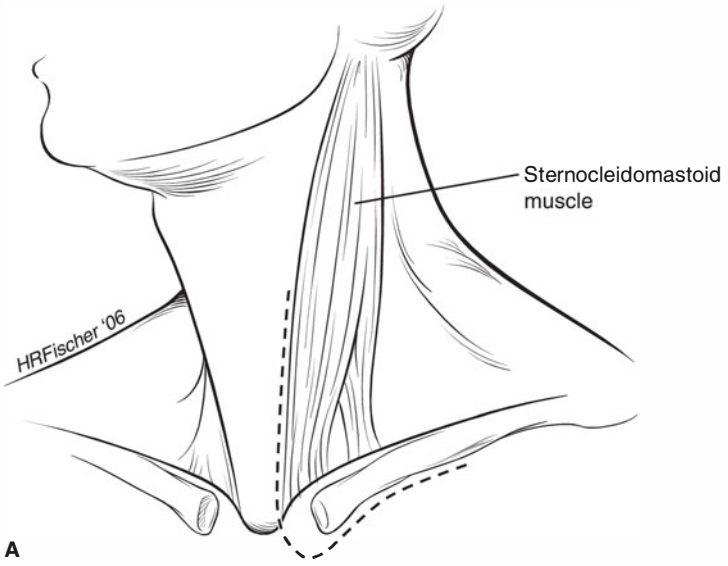
of the pulmonary artery difficult (Fig. 3.16). With the thoracosternotomy, one can approach the dissection anteriorly and dissect the hilar vessels from within the pericardium without being impeded by a large tumor as occurs when trying to approach the hilum from a posterior approach. In addition, the brachiocephalic vessels are much more readily seen, exposed, and controlled through this anterior approach. This incision is extremely versatile, and we do not hesitate to use it for these special situations.

The patient is positioned supine or with a small support elevating the ipsilateral chest. Both arms may be tucked, or the ipsilateral arm may be extended if additional exposure of the lateral chest is needed. The initial skin incision is limited to the inframammary portion and the fourth intercostal space is entered to assess resectability and ensure that there is no diffuse pleural spread or other reason why the entire incision should not be made. The skin incision is then extended over to the sternum and up toward the sternal notch and then for a short distance along the anterior border of the ipsilateral sternocleidomastoid muscle. The subcutaneous tissues are divided through the decussating fibers of the pectoral muscles and then along the fourth intercostal space. Before dividing the sternum, the intercostal muscles in the fourth space are divided up to the mammary vessels. These are dissected, doubly ligated, and divided in advance. The sternal saw is used to perform a partial sternotomy curved out to the fourth interspace. A hemisternal retractor of the type used for harvesting the internal mammary artery is then placed and used to elevate the chest wall. Although the skin incision will usually be limited to the anterior or posterior axillary line, the division of the intercostal muscles should extend back further posteriorly to allow for full elevation of the chest wall. This will assist in avoiding inadvertent fractures.

ANTERIOR CERVICOTHORACIC APPROACH (MODIFIED DARTEVELLE APPROACH)

For pathology at the level of the thoracic outlet or apex of the chest, a modified thoracosternotomy is used. This particular incision is used most commonly for apical intrathoracic pathology that may or may not involve resection of the apical chest wall.

A double-lumen tube is used. The patient is placed supine, with a soft support under the shoulders. The head is turned away from the side of pathology. An L-shaped skin incision is made along the anterior border of the ipsilateral sternocleidomastoid muscle to the sternal notch and then curving out below the clavicle going toward the deltopectoral groove (Fig. 3.17). The retrosternal space is freed with blunt dissection. The intercostal muscles are divided into either the first or second intercostal space, whichever is more suitable for the pathology. We enter the intercostal space laterally and divide the muscular fibers medially until meeting the manubrium. During this, we use blunt dissection to sweep the mammary vessels away from the chest wall. The Lebsche knife is used to divide the manubrium in an L-shaped fashion from the midline down to the appropriate intercostal space (Fig. 3.17). If the chest wall is to be resected, the cut with the Lebsche knife is extended to divide the first rib just lateral to the sternum. The internal mammary vessels are divided after the manubrium and clavicle have been separated. This improves exposure of the vessels as they come through the outlet, avoiding inadvertent



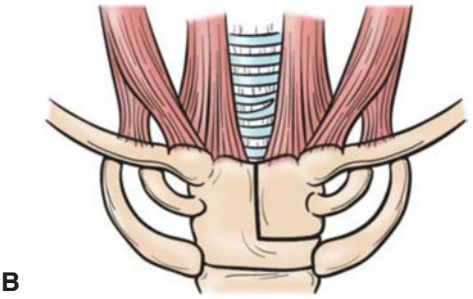


Fig. 3.17. (**A**) The L-shaped incision for a cervicothoracic approach. The incision begins along the border of the sternocleidomastoid muscle, proceeds to the sternal notch, and then curves out along the infraclavicular groove. (**B**) The modified Dartevelle approach is performed with an "L"-shaped manubrotomy allowing elevation of the clavicle and exposure of the thoracic outlet.

injury to the phrenic nerve. A hemisternal retractor is used to elevate the chest wall and provide exposure.

Given the angle the ribs take from anterior to posterior, exposure of the posterior third intercostal space posteriorly can be viewed through the first intercostal space anteriorly. The extent of disease is evaluated, and the chest wall may be divided in the appropriate locations as identified by palpating the lesion. After all of the chest wall and intercostal attachments have been completely divided, the chest wall bloc with the attached underlying pulmonary parenchyma is dropped into the thorax through the defect. The pulmonary parenchymal resection may be completed without difficulty through this defect.

On completion of the procedure, a chest tube is placed, and the chest wall is reconstructed when necessary. Anterior defects should routinely be reconstructed with a rigid prosthesis to prevent a "flail-like" physiology especially in the patient with borderline pulmonary function. To close this incision, the divided interspace is reapproximated with paracostal sutures and the manubrium is reapproximated with No. 5 stainless steel wires. The remainder of the wound is closed by reapproximating the muscle and fascia in the usual fashion.

THORACOABDOMINAL INCISION

The thoracoabdominal incision provides wide exposure of the lower chest, retroperitoneum, and upper abdomen, especially the diaphragmatic hiatus. It is a common approach for the management of abdominal aortic aneurysms when done as an open procedure, occasional distal esophageal tumors, and anterior approach to the lower thoracic and lumbar spine, and in those patients who may have undergone previous distal esophageal or upper gastric procedures. As well, a right-sided thoracoabdominal incision may be used for an Ivor Lewis type of approach to esophagectomy. In this setting, for performing the traditional open approach, we use a vertical muscle sparing incision for the thoracic portion. Positioning in this fashion obviates the need for repositioning during the procedure.

The exact location of the skin incision varies in relation to the pathology. The chest portion of the incision may be over the sixth, seventh, or eighth intercostal space, and the abdominal portion may be either midline or paramedian in location (Fig. 3.18, previous edition Fig. 3.10). The patient is positioned supine, usually with a small support extending from the hip to chest of the ipsilateral side. The arm can be supported at the patient's side, extended out at 90 degrees or suspended across the chest. The skin incision is made extending from the anterior axillary line, across the costal arch onto the abdomen. The costal arch and the intercostal muscles are divided. The diaphragm can be divided radially from the chest wall to the hiatus or circumferentially leaving 3 to 4 cm along the chest wall for later reapproximation. One must be careful to avoid injury to the phrenic nerve. A rib spreader is placed to separate the ribs, or a self-retaining type of retractor that attaches to the operating table may be used to provide exposure in both the chest and abdomen.

On completion of the procedure, a chest tube is placed. The diaphragm is closed with large nonabsorbable, interrupted sutures. We routinely excise the edges of the costal arch to prevent postoperative pain and discomfort that can result from overlapping of the costal edges. Pericostal sutures are used to reapproximate the ribs, and the abdomen is closed in layers, again with large nonabsorbable sutures for the fascia.



Fig. 3.18. The thoracoabdominal incision courses over the costal margin down onto the abdomen in a paramedian or median location depending on the objectives of the procedure.

SUMMARY

Given the increased focus on minimally invasive surgery, alternative strategies have been developed to deal with almost every type of thoracic pathology. Time will tell which ones ultimately prove to be most useful to the surgeon and most beneficial to the patient. Unlike open approaches, which are somewhat forgiving given the wide access provided, for less invasive options additional factors must be considered. The ergonomics of the instruments, angle of approach, and the surgeons' ability to handle the pathology

all contribute to the preoperative planning strategy. Careful consideration of all the potential challenges associated with positioning, the anatomy and pathology, as well as instrumentation are critical to the successful performance of the procedure.

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EDITOR'S COMMENTS

Dr. Marshall has provided a detailed and complete summary of the incisions available to the thoracic surgeon. As the current generation of thoracic surgical residents is likely being exposed mostly to minimally invasive approaches, it still remains important to have the full range of incisions in one's armamentarium since situations clearly will arise where a minimally invasive approach is not either optimal or feasible. Patient positioning is important for each particular incision both to maximize exposure and to protect against injury either from pressure or stretch. Working together with the anesthesiologist will maximize the chance of avoiding injury.

When working via a minimally invasive approach, the surgeon should be intimately familiar with the procedure to be followed should the need arise to rapidly open the chest. I particularly worry about a pulmonary artery injury when utilizing either a VATS or robotic approach. The initial maneuver, of course, should be to control hemorrhage simply with pressure applied over the bleeding point while help is mobilized and blood for transfusion is made available. Having appropriate help in an emergency situation cannot be overemphasized especially if there is difficulty controlling the bleeding. It should never be considered a sign of weakness to call for the help of a colleague in such a situation.

I have found the thoracosternotomy (hemiclamshell) incision particularly useful when dealing with large pulmonary lesions since it provides ideal exposure of the pulmonary hilum that otherwise would be obscured if approached via a posterolateral approach. From this approach, it is usually best to divide the appropriate pulmonary vein as the first step in a resection.

The anterior cervicothoracic incision also has proven to be extremely versatile for lesions occurring at the thoracic inlet. The approach provides excellent exposure of the lower cervical and upper thoracic spine, especially at the T1 level, a region that is difficult to expose either from the neck or from the chest. Leaving the sternoclavicular junction intact by dividing and elevating the manubrium avoids problems with postoperative upper extremity dysfunction. This approach essentially has replaced the posterior approach when dealing with an apical lung tumor or so-called Pancoast's tumor. Even when the lesion appears to be posterior, this anterior exposure is the procedure of choice since the anterior–posterior distance at the apex is extremely short, so there is no issue in dealing with the posterior chest wall at this level. When chest wall is to be resected, it is important to divide the first rib at the sternum as one competes the division of the manubrium. Otherwise, if the first rib is left intact, it is elevated with the manubrium making its excision particularly difficult. The exposure of the brachial plexus and brachiocephalic vessels is optimal from this approach and is much more readily available than when approached through the extended posterior thoracotomy. The incision is also considerably less morbid than the extended Paulson-type posterior incision that mandates division of the latissimus dorsi, serratus anterior, rhomboids, and trapezius muscles in order to elevate the scapula off the chest wall to gain exposure to the first rib. Utilizing the anterior cervicothoracic approach does require the surgeon to be intimately familiar with the anatomy of the thoracic inlet.

LRK

4 Right-Sided Pulmonary Resections

Larry R. Kaiser

GENERAL INTRODUCTION TO PULMONARY RESECTIONS

Pulmonary resection is the operation that defines the thoracic surgeon. The specialty of thoracic surgery is relatively new dating back to less than 50 years but really being defined during the past 30 years. Because of problems relating to positive pressure ventilation in the patient with an open chest, the development of anatomic pulmonary resection moved slowly. The initial procedure performed for a carcinoma of the lung was a pneumonectomy carried out by mass ligation of the pulmonary hilum with subsequent suturing of individual hilar structures. During the first four decades of the 20th century carcinoma of the lung was an uncommon disease and most pulmonary resections were performed for inflammatory conditions or tuberculosis. Most lung cancers were treated by total removal of the lung when they were deemed operable and this clearly was the operation thought to be required. Lesser resections were reserved for benign disease, mostly infectious problems. It took a number of years before surgeons recognized that an anatomic resection of a lobe, though a more difficult operation, provided an acceptable alternative for the treatment of lung cancer, not unlike the recognition that a more conservative resection than a radical mastectomy could be done for breast cancer with comparable survival rates.

Recognizing that surgical excision is the optimal treatment for otherwise operable lung cancer, it is important that the appropriate procedure be performed. Lobectomy remains the definitive resection since it is an anatomic resection that assures removal of the regional lymph nodes that course along the lobar bronchus and thus provides the best staging information and local control. Doing less than a lobectomy must be considered a compromise though often it is tempting to consider a nonanatomic wedge excision for small primary tumors. Not only does a wedge excision not include the lobar bronchus, precluding evaluation of lobar lymph nodes, but usually also provides only a minimal parenchymal margin and thus is accompanied by a significant incidence of local recurrence. The Lung Cancer Study Group (LCSG) addressed the question of lobectomy versus limited resection for T1N0 lesions (tumor <3 cm, negative lymph nodes) in a prospective randomized trial. The initial analysis of the data demonstrated an increased incidence of local recurrence in the limited resection group (>30% incidence) but failed to demonstrate a decrease in survival. The final analysis, however, revealed superior survival for patients in the lobectomy group. A number of other studies have looked retrospectively at patients undergoing limited resection, including anatomic segmental resection, and have demonstrated long-term survivors but the LCSG study stands as the only large randomized trial. Limited resection is associated with lower morbidity and decreased hospital stays but the best evidence supports a higher incidence of loco regional recurrence when compared to lobectomy. That said, limited resection may be comparable to lobectomy in the elderly (>70 year old) and in those with small peripheral tumors.

There are patients in whom lobectomy is not feasible and a lesser resection offers the best alternative, though admittedly a compromise. Patients in this category are those with borderline pulmonary function or those who have had previous pulmonary resections. Whenever possible the lesser resection should be an anatomic segmental resection, which by definition involves taking the appropriate segmental artery and vein as well as the segmental bronchus with its accompanying lymph nodes. Wedge resection, a nonanatomic form of resection where the bronchovascular structures are not isolated and taken separately along with regional lymph nodes, is another alternative though not ideal for patients with primary lung cancers. With the advent of videothoracoscopic techniques and the simplicity of wedge resection via this approach for a time there was renewed interest in utilizing this technique for T1N0 lung cancers. The American College of Surgeons Oncology Group (ACOSOG) has an ongoing study looking at limited resection combined with local implantation of radioactive seeds to assess whether there is a decreased incidence of local recurrence and whether this translates into a survival benefit. Preliminary results at 60 and 90 days show no increase in morbidity with the addition of brachytherapy when compared with sublobar resection alone. As it currently stands based on the LCSG data, wedge excision mostly should be avoided and patients who are found to have a primary lung cancer should be offered the best possible procedure which is, to the best of present knowledge, a lobectomy. Wedge resection, at best, is a compromise and patients who otherwise can tolerate an anatomic resection are not well served by having a lesser procedure, at least based on the best evidence to date. This has become even more important as we are identifying more early stage, small lung cancers picked up in patients who present themselves for screening spiral computed tomography (CT) scans. As noted above sublobar resection of small peripheral tumors, especially some of those identified on screening spiral CT scans, may have a survival comparable to lobectomy.

Prior to operation a decision must be made regarding which, if any, other studies should be carried out. The type and extent of the staging evaluation depends on a number of clinical factors. At a minimum patients should have a recent chest radiograph and CT scan of the chest. Most, if not all, should have a recent set of pulmonary function studies including diffusion capacity. Positron emission tomography (PET) scanning has become standard for the evaluation of a solitary pulmonary nodule and there is convincing evidence of its usefulness in staging the mediastinum as well. The fluorinated glucose used for the PET scan is trapped preferentially in malignant cells as opposed to normal cells and thus shows up as hot. Numerous studies have looked at the sensitivity and

specificity of PET for both pulmonary nodules and mediastinal lymph nodes. A nodule that shows up positive on PET has a high (approaching 90%) likelihood of being malignant. A negative PET scan does not rule out malignancy as bronchioloalveolar carcinoma and carcinoid tumors do not readily take up the fluorinated glucose and thus have a significantly higher incidence of false-negative results. An elevated blood glucose level also could be the source of a false-negative scan. A solitary nodule that is negative on PET needs to be followed with serial CT scans and resected if growth is detected. In assessing mediastinal lymph nodes, PET has a specificity of greater than 90% but the sensitivity is somewhat lower and approximates to 80%. A CT scan without mediastinal lymph nodes greater than 1.5 cm in size and a negative PET scan obviates the need for further mediastinal staging while a PET-positive lymph node mandates the need for invasive mediastinal staging with either mediastinoscopy or endoscopic bronchial ultrasound (EBUS)-guided needle aspiration.

The decision to search for disseminated disease is a difficult one and precise criteria to define if and when it should be done remain controversial. Our bias is to err on the side of performing a complete extent of disease evaluation if there is any reason at all to do so. Reasons would include any organ-specific or nonspecific signs or symptoms. Nonspecific signs include weight loss, easy fatigability, or anemia while organ-specific symptoms include bone pain, elevated liver enzymes, or localized neurologic findings. If any of these findings is present a complete staging evaluation should be obtained, not just the study pointed to by the organ-specific complaint. A PET scan obviates the need for a separate radionuclide bone scan but an MRI (or less likely) a CT scan of the brain is required for a complete extent of disease evaluation. Any patient with a past history of malignancy should have a search for distant disease as should the patient who is at a higher risk for operation, such as an individual with multiple medical problems or borderline pulmonary function. Also, any patient with locally advanced disease in whom the indications for operation are being extended (i.e., N2 disease, T4 disease) or the nonsmoker with a lung mass should have distant disease ruled out. The aim is to avoid operating on a patient who proceeds to manifest disseminated disease within 1 year of operation, a finding which ideally should have been identified preoperatively.

An important aspect of the preoperative evaluation of a patient with lung cancer is the assessment of pulmonary function. Not all patients undergoing thoracotomy require pulmonary function testing but the majority of patients with lung cancer also have some element of underlying lung disease as a result of the same risk factor that is associated with their cancer, for example, cigarette smoking. Assessment of pulmonary function serves both to identify those patients at a significantly increased likelihood of postoperative morbidity as well as those patients who stand to benefit from preoperative manipulations designed to attenuate those risks. There is no single best test to evaluate pulmonary function in a patient who is slated to undergo pulmonary resection. Also there are no absolute values that contraindicate resection though using a combination of studies it is at least possible to make a judgment as to which patients are at an increased risk for postoperative morbidity or mortality. Preoperative spirometry to measure flows and volumes should be performed. Important measurements include forced expiratory volume in 1 second (FEV_1), maximal voluntary ventilation (MVV), diffusing capacity, FEV_1/forced vital capacity (FVC) ratio, and the ratio of the residual volume (RV) to total lung capacity (TLC). An FEV_1 of less than 40% of predicted has been associated with increased postoperative morbidity and mortality. A reduced diffusing capacity also has been associated with postoperative morbidity. Any assessment of postoperative morbidity and mortality has to take into account the extent of the proposed resection. Often this is difficult to determine and the experience of the surgeon is closely related to the likelihood of pneumonectomy. The surgeon operating on patients with compromised lung function should be experienced in the performance of segmental resections, sleeve resections, and nonanatomic resections, all techniques of lung preservation. Techniques of video-assisted thoracic surgery (VATS) may also be of use in patients with limited pulmonary reserve who require pulmonary resection. There are a number of measures that can be instituted that are designed to attenuate the postoperative risk in patients with compromised lung function. That said, most often it comes down to a judgment on the part of the surgeon based on both objective and subjective factors that is the deciding factor. There are some patients who just are not candidates for pulmonary resection for a variety of reasons that may sometimes be difficult to articulate.

Experience with lung volume reduction surgery in patients with severe end-stage emphysema has changed our approach regarding operation for lung cancer in patients with compromised lung function. Nutritional assessment and therapy and a supervised program of pulmonary rehabilitation may further optimize borderline patients for pulmonary resection. Currently our approach is to be quite aggressive in considering patients with pulmonary malignancies for resection. Rarely is a patient turned down for resection solely on the basis of his/her pulmonary function. At times, the resections have to be somewhat creative and there are a number of intraoperative factors that may contribute to a reduction in postoperative problems. Patients with borderline lung function have to be strongly motivated toward resection. These patients are not the ones to be talked into an operation even if their other treatment options are limited; they really have to be motivated. The major morbidity and potential mortality in these patients occur in the early postoperative period but one must also keep in mind the long-term sequelae of the resection of lung parenchyma in these individuals. Paradoxically there may be some improvement in lung function following pulmonary resection in these patients especially if the lung parenchyma removed receives only a minimal amount of the pulmonary perfusion. Most commonly this occurs with a heterogeneous pattern of emphysema with the upper lobe being the most diseased portion of parenchyma. A preoperative quantitative ventilation–perfusion lung scan is useful in assessing the significance of the loss of lung parenchyma. An estimate of the predicted postoperative FEV_1 may be obtained by subtracting the percentage removed by the proposed resection based on the percent of perfusion received by that area of lung parenchyma. A residual FEV_1 of less than 800 ml has been associated with an increased risk of postoperative morbidity and mortality but this is entirely dependent on what percent of the predicted FEV_1 the 800 ml represents. For instance, in a 50 kg female 800 ml may represent a predicted postresection FEV_1 of 60% or more. An algorithm for the preoperative assessment of risk in patients with lung cancer is presented in Figure 4.1.

In addition to pulmonary function studies an assessment of the exercise capability of a patient with compromised lung function may be appropriate. This assessment may range from something as simple as having the patient climb one or two flights of stairs while monitoring oxygen

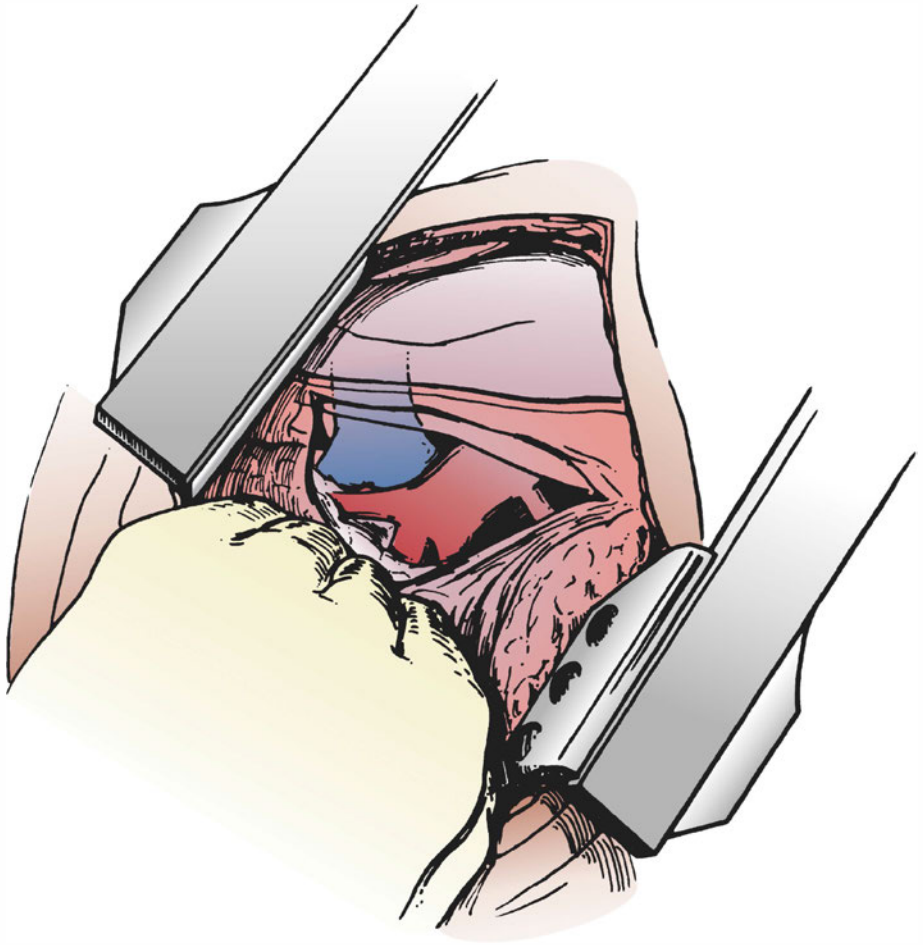


Fig. 4.1. View of the right hilum as seen from the *right side* of the table. The hilar pleura has been incised and the right main pulmonary artery and superior pulmonary vein are well seen. Note the middle lobe vein draining into the superior pulmonary vein. This vein must be preserved when performing a right upper lobectomy. The continuation of the artery distal to the apical-anterior trunk passes posterior to the superior pulmonary vein.

saturation and pulse rate to formal exercise testing and calculation of maximal oxygen consumption (VO_2 max). One can be reasonably certain that a patient who can walk up two flights of stairs can tolerate a lobectomy, a crude assessment to be sure but one that intuitively makes sense. For truly borderline patients, measurement of VO_2 max may be the deciding test. A value of less than 15 ml/kg/min has been associated with significantly increased postoperative morbidity and mortality. Patients in this category should be scrutinized closely before deciding to proceed with resection. It is this type of patient who may benefit from a limited resection, albeit a compromise, such as a video-assisted wedge excision if the lesion is small and peripheral. It is highly unlikely that a patient with this severe degree of pulmonary compromise could tolerate a pneumonectomy. Other parameters suggesting high risk include PCO_2 >45 torr and elevated pulmonary artery pressures.

Some patients may undergo invasive staging prior to pulmonary resection. The decision to perform mediastinoscopy may be based on CT scan findings of enlarged mediastinal lymph nodes and the results of the PET scan. The criteria for defining "enlarged" vary and the sensitivity and specificity of the technique vary depending on the size that is set. We perform mediastinoscopy when lymph nodes greater than 1.5 cm in size are seen on the CT scan. Others perform mediastinoscopy on all patients prior to pulmonary resection recognizing that the majority of procedures will reveal only nodes without evidence of metastatic disease. Mediastinal nodes that are positive on PET scan mandate the need for mediastinoscopy unless there is such bulky adenopathy seen on CT scan that tissue confirmation would be redundant. EBUS assessment of the mediastinum with selective needle aspiration biopsy also is being used more frequently to interrogate the mediastinum.

Whether mediastinoscopy is used selectively or routinely, the key point is accurate staging of the mediastinum in the patient with lung cancer. If mediastinoscopy is performed, it is important that lymph node material be obtained for pathologic examination. Accurate mediastinal staging mandates at the least mediastinal lymph node sampling at thoracotomy or, preferably, complete systematic lymph node dissection. The problem with lymph node staging alone is the issue of how lymph nodes are chosen to be sampled, a problem not present when a complete lymph node dissection is carried out. Mere palpation of a node or an assessment of nodal size will miss nodes harboring intranodal or microscopic disease. It is not clear what percentage of nodal disease is missed with a staging procedure because of the variability in the selection of nodes to be sampled and the lack of a study where nodes are first sampled followed by a complete lymph node dissection. From our own experience 10% to 20% of resections where mediastinal lymph node disease is not suspected result in positive lymph nodes being identified by the pathologist. An operation without lymph node staging information must be considered incomplete. Accurate staging allows the surgeon to discuss prognosis realistically with the patient and allows the patient the opportunity to either participate in a trial of postoperative adjuvant therapy or be evaluated for treatment outside of a protocol setting. With prospective randomized clinical trials demonstrating improved survival of patients with completely resected N1 or N2 disease treated with postoperative adjuvant chemotherapy, the quality and completeness of the intraoperative staging has become even more important.

Despite the bias on the part of most physicians that postoperative radiation therapy is of value in resected patients who are found to have either N1 or N2 disease there are no prospective data that demonstrate a survival advantage in patients so treated. One prospective randomized trial of postoperative radiation therapy versus no further treatment for resected patients with squamous cell carcinoma demonstrated a significant reduction in local recurrence but absolutely no difference in survival. A randomized trial conducted by the collaborative efforts of the national cooperative cancer groups compared postoperative chemotherapy (cisplatin, VP-16) and radiation therapy with radiation therapy alone. This trial required that patients either have mediastinoscopy performed or have a completely negative CT scan in addition

to mandating what lymph nodes had to be sampled at the time of thoracotomy. The results of this trial showed no advantage for the combined radiation therapy and chemotherapy arm over radiation therapy alone and no significant difference when compared to historic controls. Interestingly, there did appear to be a survival advantage in the subgroup of patients who had mediastinal lymph node dissection as opposed to lymph node sampling only.

Long-term survival following pulmonary resection depends both on characteristics of the primary tumor (T stage) and presence or absence of lymph node disease (N stage). Any analysis of survival is greatly dependent on how thoroughly the lymph nodes are staged, as discussed above. These data, accumulated from studies performed by the LCSG, are particularly enlightening because of the stringent requirement for nodal staging that was mandated for entering patients into these studies. Thus, when a patient was staged as N1 we can be assured that was an accurate staging evaluation since the mediastinal lymph nodes would have been sampled and found to be free of tumor.

INTRODUCTION TO RIGHT-SIDED RESECTIONS

There are a number of significant anatomic features specific to right-sided pulmonary resections. The right main pulmonary artery is relatively long and courses posterior to the superior vena cava and traverses the carina. This extra length of the artery, at times, is an advantage for some proximal lesions that in a similar location on the left side would not be resectable because of the short length of the left main pulmonary artery relative to the bifurcation. The distance between the carina and the origin of the right upper lobe bronchus usually is less than 2 cm and the carina is readily mobilized from the right side. Access to the proximal left main stem bronchus is significantly easier from the right side compared to the left side where the aortic arch limits access both to the origin of the left main bronchus and the carina. Mobilization of the carina is not possible from the left chest and even visualization of the carina from the left is difficult. Carinal resections are preferentially performed through a right thoracotomy or, at times, through a median sternotomy.

The superior mediastinum, the space accessed by the mediastinoscope, is well visualized from the right side. The area bounded by the azygous vein (inferior), the trachea (posterior), the subclavian vein (superior), and the superior vena cava (anterior) delineates this compartment whose lymph node-bearing contents may be removed en bloc from the right side. No such access exists on the left side where the left paratracheal nodes are relatively inaccessible because of the location of the aortic arch. As mentioned above, with the access afforded to the carina from the right side it follows that the subcarinal space is readily dissected for lymph node removal. The subcarinal space also is easily accessed on the left side as well.

On the right side the azygous vein is an important anatomic landmark. The vein courses from posterior to anterior across the main stem bronchus to drain into the superior vena cava. Just inferior to where it crosses the main bronchus is the origin of the upper lobe bronchus, a key anatomic feature. Rarely is it necessary to divide the azygous vein but it may be taken with impunity if involved by tumor or limits access to the lymph node-bearing area.

In assessing resections from the right side the upper lobectomy probably is the most straightforward resection though the location of the posterior segmental arterial branch may, at times, be problematic. Right lower lobectomy is complicated by the location of the middle lobe artery and bronchus and the middle lobectomy is considered difficult by some because of the minor fissure.



SURGICAL TECHNIQUE

Right Upper Lobectomy

Right upper lobectomy is the prototypical pulmonary resection and is a good starting point for the trainee just starting out in pulmonary surgery. The long right main pulmonary artery with the apical-anterior branch and the discrete takeoff of the upper lobe bronchus makes this an ideal resection. The so-called truncus anterior, the apical-anterior branch of the pulmonary artery, also facilitates segmental resections of the right upper lobe. Once this branch is divided the segmental bronchi are easily visualized.

A left endobronchial double-lumen tube is placed to allow for single-lung ventilation so that the right lung is collapsed for the lobar resection.

With the patient positioned on the left side (left lateral decubitus position), the chest is entered through either a standard posterolateral thoracotomy incision or a vertical axillary muscle-sparing incision. The chest is entered through the fifth intercostal space for the posterolateral incision or the fourth intercostal space for the more anterior muscle-sparing incision. The hilum and mediastinum are palpated to assess the extent of involvement and determine resectability. Access incisions for VATS lobectomy are described in detail in the chapter dealing with VATS pulmonary resection but a small utility incision usually is made anteriorly not only to facilitate the dissection but also to allow for removal of the lobe from the chest. Whether done by a VATS approach or open thoracotomy, the dissection of the pulmonary hilum essentially is the same in that the superior pulmonary vein, the pulmonary arterial branches, and the lobar bronchus must all be taken individually.

The lung is retracted posteriorly and the hilar pleura anteriorly and superiorly is incised. The superior pulmonary vein is identified and dissected distally toward the lung parenchyma and proximally to the pericardial reflection. The vein is encircled once the appropriate plane of dissection is entered. Care must be taken to avoid injury to the pulmonary artery which lies directly posterior to the superior pulmonary vein (Fig. 4.1). The middle lobe venous branch, which usually drains to the superior pulmonary vein, is identified and preserved. This vein almost always is a tributary of the superior pulmonary vein but occasionally may be found entering the inferior pulmonary vein. Infrequently aberrant venous drainage on the right side, usually a small venous branch emptying directly into the vena cava, may be encountered. Lying just superior and posterior to the vein is the right main pulmonary artery. The artery is dissected circumferentially and followed proximally where it is observed as it courses posterior to the superior vena cava. The artery is easily encircled with a finger once the appropriate plane of dissection is entered. For an open thoracotomy, proximal control of the artery is usually established in this fashion if there is even a suggestion that the dissection may be difficult. It is not necessary to encircle the right main pulmonary artery in every case but one cannot be too safe in taking the extra precaution of having proximal control. After encircling the artery an umbilical tape is passed and a Rumel tourniquet is placed. Should the artery be inadvertently entered it is a simple matter to snug down on the tourniquet and control the bleeding. The artery is dissected distally and the apical-anterior arterial branch is identified (Fig. 4.2). This usually occurs as a common trunk but the individual segmental branches may arise separately from the main artery with the

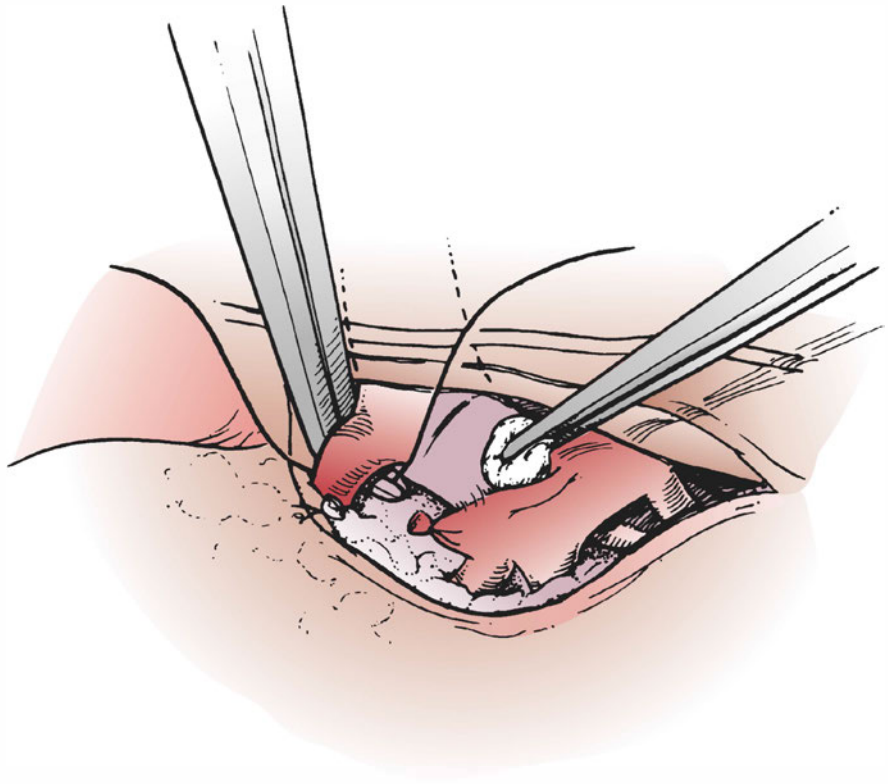


Fig. 4.2. At times the apical venous branch obscures the origin of the apical-anterior arterial trunk and must be divided prior to division of the arterial branch. Here the venous branch has been divided in preparation for division of the apical-anterior trunk. The right angle is around the arterial branch. The artery is divided between carefully placed 0 silk ligatures.

branch to the anterior segment coming off the artery as the most proximal branch.

Dissecting out the pulmonary artery and its branches differs significantly from the manner in which other arteries are handled. Lacking a muscular coat the main strength of the artery comes from the intima. Thus the artery is extremely fragile and must be handled with the utmost care. When dissecting the artery great care must be taken in using any "spreading" maneuver. Whereas in peripheral vascular surgery it is perfectly acceptable to dissect out an arterial branch by using the scissors to spread adjacent tissue, this move, if applied to a pulmonary arterial branch, can easily result in avulsion of the branch from the artery trunk. Likewise, when attempting to use a right angle to encircle a branch of the pulmonary artery little or no force should be applied since one gets no tactile cues from this artery. An arterial branch should be completely dissected free circumferentially prior to trying to pass a right-angled clamp. To do this the thin connective tissue overlying the artery is grasped with a forceps and lifted to allow a cut by the scissors. Once the correct plane on the artery is reached, a closed, blunt-tipped scissors is used to gently push the artery away while continuing to hold the incised tissue. This rapidly creates the plane of dissection and frees up the adjacent arterial branch. If an attempt to pass a right angle is met with any resistance, further dissection should be performed rather than persisting with the right angle and potentially perforating the back wall of the arterial branch. A peanut dissector may also be used to dissect the pulmonary artery once the correct plane of dissection on the artery is encountered but I personally do not favor use of this instrument.

Once the artery has been identified, the dissection proceeds superiorly along the hilum to enter the plane of the bronchus. The azygous vein is a significant landmark. This vein crosses the right main stem bronchus just superior to the origin of the right upper lobe bronchus. The vein courses from posterior to anterior and the bronchus lies medial to the vein. Once the artery is dissected away from the bronchus, it is possible to encircle the upper lobe bronchus at this point if deemed necessary. At times it is advantageous to divide the bronchus first, especially if the primary tumor or lymph nodes involve the branches of the pulmonary artery.

The lung is retracted anteriorly by the assistant to reveal the posterior aspect of the hilum. Unlike operating in the abdomen, it quickly becomes clear when doing pulmonary surgery that the surgeon must be familiar with the anatomy and especially the relationships between structures from both an anterior and posterior orientation. In the chest the surgeon needs to think in three dimensions. To facilitate the performance of an upper lobectomy dissection is begun by incising the overlying pleura in the bifurcation formed by the upper lobe bronchus and the bronchus intermedius (Fig. 4.3). This is one of the most significant moves of the entire procedure and if understood it greatly simplifies and speeds up the operation. Taking care to coagulate small bronchial vessels that are present in this "crotch" the pleura is incised. A lymph node is a constant finding in this location and the dissection frees this node anteriorly away from the bifurcation. The upper lobe bronchus may be encircled at this point and taken, if desired (inset, Fig. 4.3). Just anterior to this lymph node, however, lies the branch of the pulmonary artery to the superior segment of the lower lobe, which is easily visualized from this posterior approach. Once this arterial branch is identified, the posterior portion of the major fissure may be completed with a firing of the linear stapler (Fig. 4.4). The lower lobe superior segment arterial branch is the most posterior branch of the artery within the fissure and the stapler may be safely passed just posterior to the branch. The pleura within the fissure needs to be incised and the appropriate location for placement of the stapler may be found by placing the forefinger in the crotch just dissected posteriorly and the thumb in the fissure. The fissure at the appropriate spot is quite thin and may be further thinned out by finger dissection of the parenchyma held between the thumb and forefinger. This move is safe because the location of the artery is known and it has been dissected free. It avoids extensive dissection in the fissure in a search for the pulmonary artery. Until the location of the artery is known, firing a stapler across the fissure, as is commonly done, adds little; it really brings you no closer to the artery. Taking advantage of the anatomy posteriorly allows easy identification of the artery in the fissure without actually dissecting the fissure and thus avoids air leaks. It is important to remember that the fissure, for that matter any fissure, is defined by the artery. Once the artery is identified and dissected in the correct plane, the posterior portion of the major fissure is very simple to complete.

Alternatively, the upper lobe bronchus may be encircled and divided at this point,

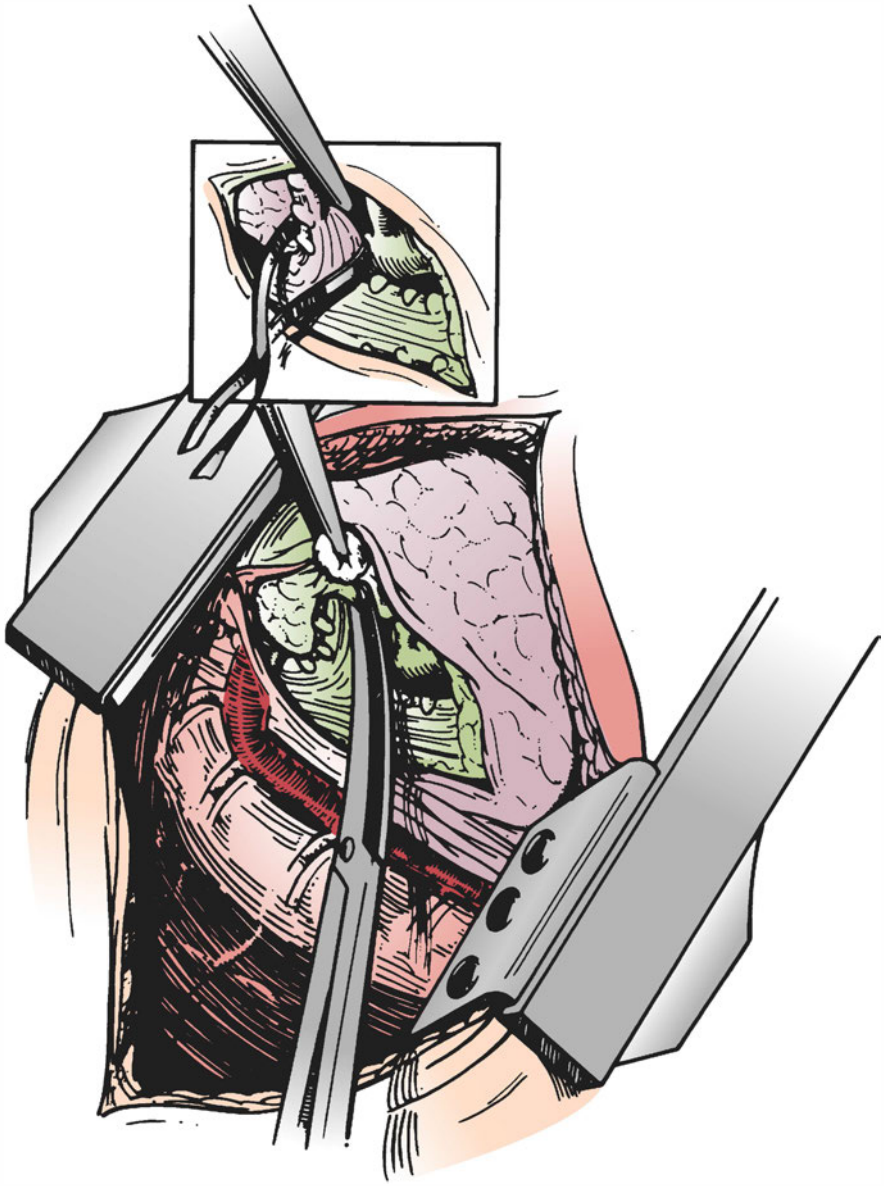


Fig. 4.3. One of the most important concepts in pulmonary surgery is visualization of the anatomy in three dimensions. Much of the dissection may be done from behind or the posterior aspect of the hilum. Here the bifurcation between the bronchus intermedius and upper lobe bronchus is being dissected and the upper lobe bronchus skeletonized. There is the constant finding of a lymph node at this "crotch." Incision at the bifurcation and elevating the lymph node out of the area reveals the pulmonary artery within the fissure. **Inset**. Once the bifurcation is dissected the bronchus may be encircled. Superiorly, if the artery has been dissected the bronchus may be taken at this point.

which allows complete visualization of the artery from "behind" the fissure. This is the preferable move when there is nodal involvement in the fissure that makes dissection on the artery difficult. The arterial branch to the posterior segment of the upper lobe is adjacent (superior) to the superior segmental branch and occasionally may arise from this branch (Fig. 4.5). The arterial supply to the middle lobe usually arises just opposite the takeoff of the superior segmental branch. There is usually one middle lobe arterial branch, but two branches are not uncommon.

Resectability must be assured prior to dividing any vascular structures. The ability to remove all disease, including mediastinal lymph node disease, if encountered, must be clear. It offers the patient no benefit to proceed with resection if gross disease is left behind. There is no advantage to a palliative resection, only the disadvantage of the risk of the procedure. At times the surgeon must be prepared to be somewhat creative in completing a resection especially if the tumor involves the hilum of the lung and obscures the origin of the lobar vessels making individual dissection of these vessels impossible. Clamping the artery proximally allows the portion of the arterial wall containing the origin of the branches to be excised and the arterial wall either sutured primarily or patched with a piece of pericardium. Otherwise, proximal involvement of the pulmonary artery may mandate pneumonectomy but parenchymal conservation should always be considered as long as oncologic principles are not compromised. Rarely should proximal tumor involvement or encasement of the right upper lobe bronchus in and of itself mandate pneumonectomy as long as the surgeon is familiar with the technique of sleeve resection. The arterial trunk to the anterior and apical segments is divided between ligatures, preferably silk because of the easy handling and tying features of this material. The ties must be placed so as to leave enough distance between them to allow the branch to be divided and leave a cuff of artery on each side. No upward traction should be exerted when tying since the artery may be easily disrupted at the origin of the segmental branch. Usually, a single tie is placed on an arterial branch but a suture ligature may also be placed adjacent to the proximal tie for added safety. To avoid any accidental pulling on a tie, each suture is cut after the knot is complete and prior to dividing the ligated arterial branch. How much force one exerts when securing a knot on a pulmonary arterial branch is critical to avoid injury to the vessel. Only enough force to feel the intima "crunch" should be applied and no more. The pulmonary artery is a low pressure system which does not mandate a huge amount of force to ligate a vessel. More force than is necessary may result in avulsion of the arterial branch. Alternatively, a vascular stapler may be used to divide the apical-anterior arterial branch. Once this branch is divided the origin of the upper lobe bronchus is readily apparent. Lymph nodes lying along the bronchus are dissected upward to be removed with the specimen, a maneuver which completes the dissection around the bronchus.

The superior pulmonary vein is divided either with a vascular stapler or tied and suture ligated (Fig. 4.6). Alternatively, a vascular clamp may be placed and the divided vein closed with a horizontal mattress stitch, the clamp removed, and the suture run as a simple stitch anterior to the mattress stitch back to where it was begun. Once the vein is divided the continuation of the pulmonary artery is identified as it lies posterior to the vein. The middle lobe arterial branch is readily visualized from the anterior aspect of the hilum and, to facilitate division of the fissure and separation of the middle lobe from the upper lobe, the branch should be mobilized

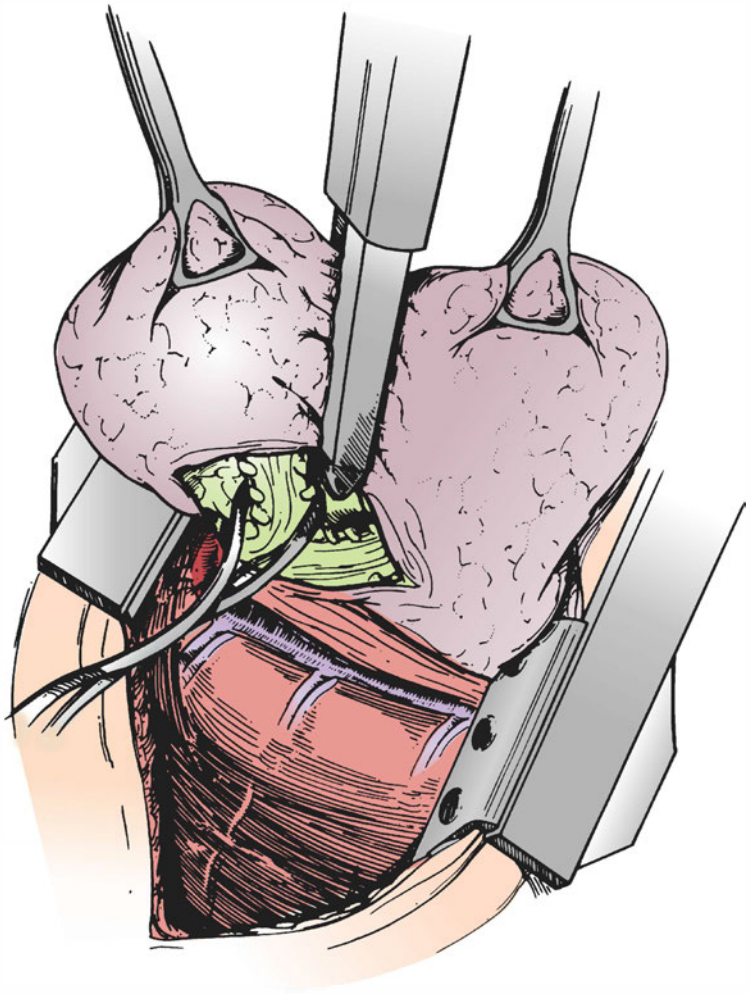


Fig. 4.4. Once the artery is identified and the branch to the superior segment of the lower lobe defined, the posterior aspect of the major fissure may be divided with a linear stapler. There are no arterial branches posterior to the superior segmental branch, thus allowing the fissure to be defined and divided. Here the stapler has been placed facilitated by the upper and lower lobes grasped with Duval lung clamps.

for a short distance. The arterial branch to the posterior segment of the upper lobe at times may be seen through this anterior exposure or an additional anterior segmental branch may be identified. Once the artery is identified from this anterior approach and the middle lobe artery is seen, the minor fissure, which is usually incomplete, may be divided with an application of a linear stapler.

The posterior segmental branch of the artery is ligated and divided within the fissure. With the lung again retracted anteriorly the origin of the upper lobe bronchus is well seen and a stapler is used to close the bronchus (Fig. 4.7). The bronchus is taken as close as possible to its origin without compromising the lumen of the right main stem bronchus. The bronchus is divided and the upper lobe removed if the minor fissure has been divided. If the minor fissure remains it is completed with a firing of the linear stapler. In order to obtain definitive staging information, a complete mediastinal lymph node dissection should be performed.

Following division of the minor fissure and removal of the right upper lobe the middle lobe is left without tether since the oblique fissure usually is relatively complete. Postoperatively this situation may predispose to torsion and infarction of the middle lobe. In order to prevent this very significant complication the middle lobe is "reattached" to the lower lobe either by placement of several absorbable sutures placed in a figure-of-eight fashion or by placing a row of staples between the two lobes. The middle lobe must be properly oriented prior to attaching it to the lower lobe.

There are several potential pitfalls to avoid when performing a right upper lobectomy. For the most part it is one of the most straightforward of the pulmonary resections but problems may occur. The middle lobe vein must be identified and preserved when dividing the superior pulmonary vein. Once the superior pulmonary vein has been divided, great care must be taken to avoid injury to the middle lobe artery especially when dividing the minor fissure. Traction exerted on the upper lobe while it is still attached to the middle lobe by the intact minor fissure may result in an avulsion injury to the middle lobe artery. As already mentioned, the right main bronchus may be narrowed if the upper lobe bronchus is closed and subsequently divided with a stapler placed too close to its origin. This usually occurs when the lobe is retracted upward tethering the main stem bronchus prior to placing the stapler. If the main stem bronchus has been narrowed it is best to proceed with a sleeve resection and reanastomosis instead of trying to "repair" the damage. Waiting for a stricture to become symptomatic creates far more problems in the long run.

Right Middle Lobectomy

Middle lobectomy is thought by many to be the most difficult lobectomy because of the problems presented by the fissures. This is an erroneous concept since it is possible to accomplish the bronchovascular dissection and division from an anterior approach if the fissures are problematic. The middle lobe is a common site for inflammatory disease and bronchiectasis. It is a common location for mycobacterial infections other than tuberculosis (MOTT).

The illustrations (Figs. 4.8-4.10) depict the view from the left side of the table, the ideal position from which to operate. If the major fissure is well developed and the pulmonary artery is visualized easily within the fissure, the overlying pleura is incised. If the artery is not visible, the fissure overlying the artery must be divided in order to identify the artery. This creates air leaks and is generally messy and time consuming. To proceed with this "standard" approach, significant dissection in the fissure is required and it is safest to gain proximal control of the pulmonary artery by incising the hilar pleura overlying the main pulmonary artery with the lung retracted posteriorly. Within the fissure the middle lobe arterial branch is identified as it originates from the main pulmonary artery usually just opposite the branch to the superior segment of the lower lobe (Fig. 4.5). Most commonly, there is a single-arterial branch to the middle lobe but occasionally two branches are identified. The arterial supply to the middle lobe is ligated

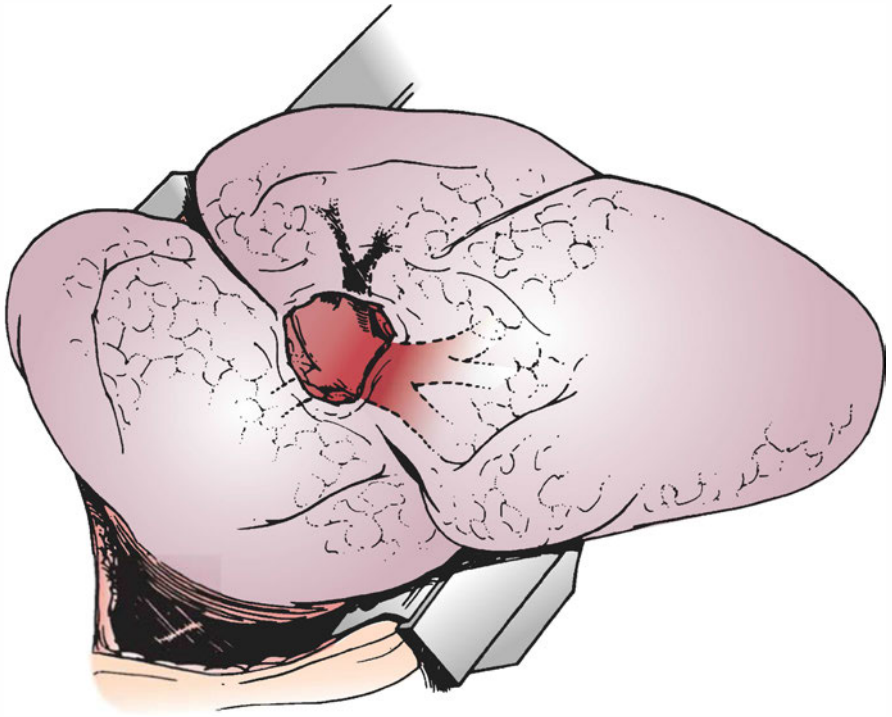


Fig. 4.5. View of the pulmonary artery from within the fissure showing the position of the arterial branch to the posterior segment of the upper lobe in relation to the middle lobe branch and the superior segmental branch. If the fissure is well developed, the overlying pleura may be incised and dissection carried down directly on the vessel. If the fissure is poorly developed, it is easier to divide the bronchus, identify and divide the posterior segmental branch, and then complete the fissure with several firings of the linear stapler.

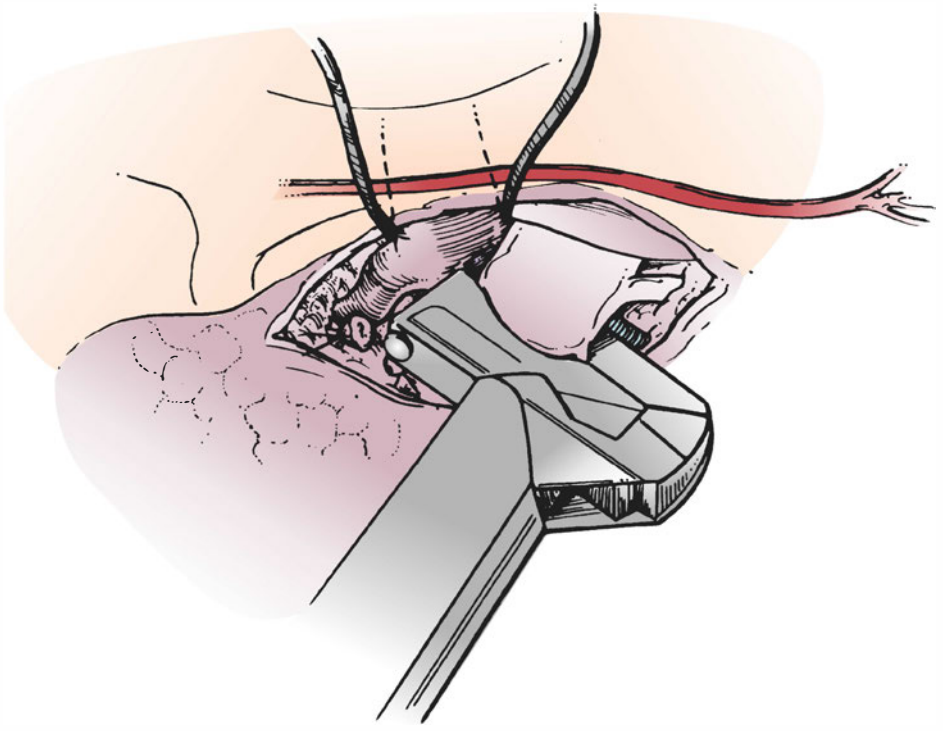


Fig. 4.6. Proximal control of the artery has been obtained by placing an umbilical tape around the artery. The superior pulmonary vein, with the middle lobe vein left intact, is divided between rows of vascular staples. Here the stapler is being applied to the vein.

with silk ligatures and divided. Once the arterial supply is divided, the middle lobe bronchus may be seen lying deep to the artery and slightly inferior, as viewed from within the fissure. The bronchus is dissected back to its origin from the bronchus intermedius, stapled, and divided. Alternatively, the bronchus may be divided and closed with interrupted sutures of braided or monofilament absorbable material of size 3-0 or 4-0.

With the lung retracted posteriorly, the anterior hilar pleura over the superior pulmonary vein is incised. The middle lobe venous tributary or tributaries most commonly drain into the superior pulmonary vein but can drain into the inferior vein on rare occasions (Fig. 4.1). To assure that a tributary is coming from the middle lobe, the lobe may be grasped with a lung clamp and retracted laterally (upward toward the incision). This will avoid division of small branches coming from the upper lobe. The venous branch is then divided after ligating with silk ligatures and securing with a suture ligature.

Once the bronchovascular structures have been divided the minor fissure is completed with a firing of the linear stapler. The anterior portion of the major fissure is likely well developed and is easily completed and the lobe removed.

An alternative technique for middle lobectomy is also illustrated and is likely to be more useful and versatile. This technique does not rely on the pulmonary artery being visible within the fissure and does not require extensive dissection in the fissure to identify the artery. The lung is retracted posteriorly. The hilar pleura overlying the superior pulmonary vein is incised and the middle lobe vein identified. The vein is ligated and divided and immediately posterior and slightly superior to the vein lies the middle lobe bronchus (Fig. 4.8). The bronchus is surrounded by connective tissue that is divided taking care to coagulate any bronchial arterial branches that are encountered. The bronchus should be followed back to its origin at the bronchus intermedius. The middle lobe arterial branch lies just posterior and slightly superior to the middle lobe bronchus but may not be visible prior to dividing the bronchus. The bronchus is encircled with a right-angled clamp staying close to the bronchial wall to avoid damage to the adjacent arterial branch (Fig. 4.9). The bronchus is divided with a scalpel using a right-angled clamp as a guide. The bronchial stump is closed with interrupted sutures. Alternatively a stapler may be applied to close the middle lobe bronchus.

Following division of the bronchus, the middle lobe arterial branch is easily seen and is circumferentially mobilized (Fig. 4.10).

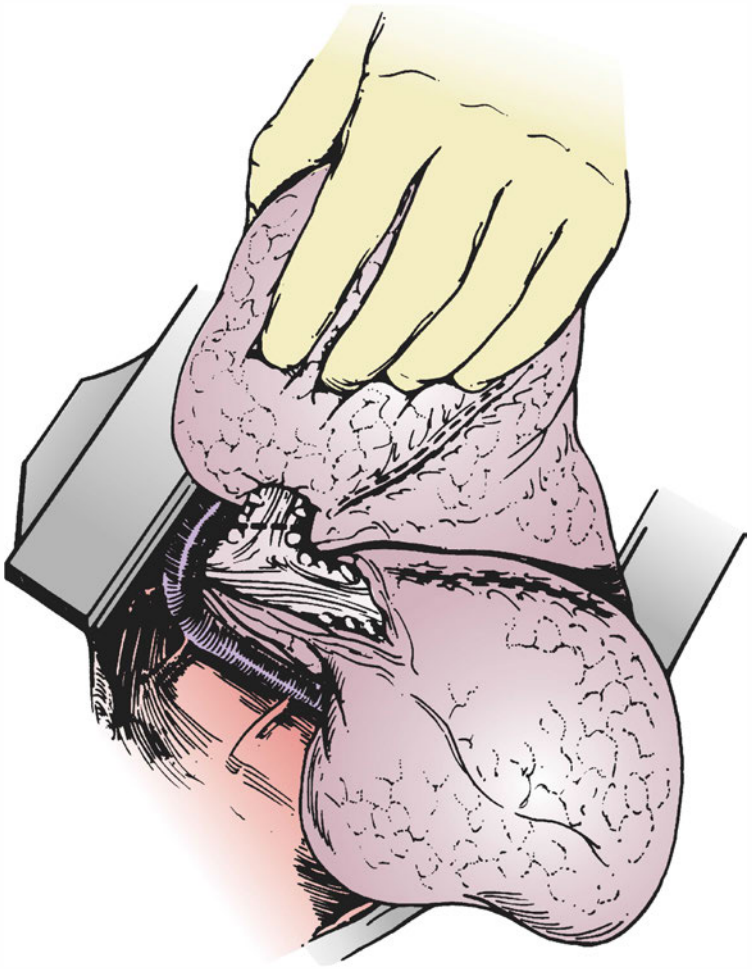


Fig. 4.7. The right upper lobe bronchus is divided as close to its origin as possible (*dotted line*). The point of division should not be so close that it compromises the lumen of the main bronchus or bronchus intermedius. It is safest to take the bronchus from this posterior approach since its origin is well seen.

The arterial branch or branches are ligated and divided. Once the bronchus, artery, and vein are divided, the minor fissure as well as the portion of the oblique fissure in contact with the middle lobe is divided with several firings of the linear stapler and the lobe is removed. A mediastinal lymph node dissection is then completed in order to obtain the most accurate and complete staging information.

Despite the apparent simplicity of middle lobectomy as described, there are several potential trouble spots. The middle lobe bronchus originates from the bronchus intermedius at essentially a right angle and is quite fragile and susceptible to injury. The origin of the middle lobe bronchus is not easily seen from the anterior approach and the bronchus intermedius may be damaged when the middle lobe bronchus is taken with a stapler. The middle lobe arterial supply may also present problems if there is more than one branch. Sometimes, the additional branch may be obscured and injured if its presence is not recognized. Care must also be taken when encircling the middle lobe arterial branch from the anterior approach in order to avoid injuring the main pulmonary artery within the fissure.

Right Lower Lobectomy

Because of the close proximity of the bronchovascular structures of the middle lobe, resection of the right lower lobe provides several unique challenges and is one of the more difficult lobectomies. Similar to the middle lobectomy, the pulmonary artery must be identified within the fissure in order to complete the resection and in those cases where the fissure is poorly developed, a direct attack through parenchyma often proves to be quite challenging. The chest is entered through either a standard posterolateral thoracotomy incision (5th intercostal space) or a vertical axillary muscle-sparing incision (4th intercostal space). If disease is noted within the fissure or if the hilum is involved it is safest, in my opinion, to obtain control of the proximal right main pulmonary artery. The hilar pleura is incised anteriorly and superiorly with the lung retracted posteriorly and the proximal pulmonary artery is encircled just lateral to the superior vena cava.

If the fissure is reasonably well developed, the pleura overlying the pulmonary artery is incised and the dissection is carried down onto the plane of the artery (Fig. 4.11). The branch to the superior segment of the lower lobe is first identified and the middle lobe arterial branch is most commonly found arising from the opposite aspect of the artery just across from the superior segmental origin. The dissection may be extended posteriorly along the superior aspect of the branch to the superior segment, which leads to the bifurcation of the upper lobe bronchus and bronchus intermedius. With the lung retracted anteriorly, the pleura overlying this bifurcation posteriorly is incised and a linear stapler encompassing the parenchyma within the fissure may be inserted from just above the superior segmental arterial branch through the area of the bifurcation. This move is possible since there are no vascular structures present posterior to the origin of the superior segmental arterial branch. On the superior aspect of the artery just opposite the superior segment the posterior segmental branch, the so-called recurrent branch (posterior segmental), to the upper lobe arises and is easily visualized. Rarely, this branch to the upper lobe may arise from the superior segment branch to the lower lobe but this possibility should be kept in mind. The posterior aspect of the major fissure is then divided and completed.

The relationship of the superior segmental branch to the middle lobe arterial branch determines whether the lower lobe artery may be divided as a complete trunk or whether the superior segmental branch and basal arterial trunk need to be taken separately. As illustrated in Figure 4.11 the superior segmental branch must be taken separately to avoid damage to the middle lobe arterial supply. The dotted line indicates the position for division of the basal segmental trunk. This is usually a 1- to 2-cm trunk and should be double ligated with a suture ligature or divided with a linear vascular stapler. The simplest stapling maneuver utilizes the endoscopic linear stapler with a vascular cartridge for closure and division (inset, Fig. 4.11). The stapler may be placed obliquely to include the superior segment branch while avoiding the middle lobe artery.

Dividing the pulmonary artery reveals the bronchus that lies just deep (medial) to the artery. With the artery retracted

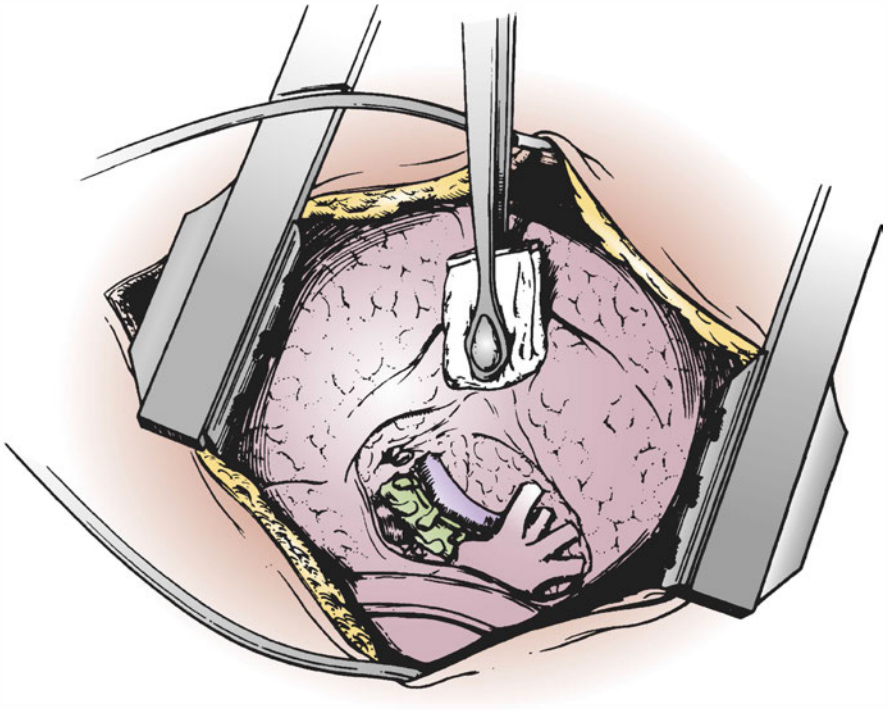


Fig. 4.8. View of the right hilum from the *left side* of the table. The superior pulmonary vein has been dissected out and the middle lobe vein identified, ligated, and divided. The ligated stump of the middle lobe vein is seen. Immediately posterior to the vein lies the middle lobe bronchus. Usually, the bronchus has to be divided to have optimal visualization of the middle lobe artery, but at times the artery is seen slightly superior and posterior to the bronchus. Division of the bronchus facilitates division of the artery and provides the exposure to assess if there are other middle lobe arterial branches.

superiorly, the origin of the middle lobe bronchus may be visualized and the location for division of the lower lobe bronchus established (Fig. 4.12). The middle lobe artery lies superficial and superior to the middle lobe bronchus. Care must be taken to avoid compromising the origin of the middle lobe bronchus when stapling or dividing the lower lobe bronchus (inset, Fig. 4.12). The bronchus may be closed either with a stapler or divided with a scalpel and closed with interrupted absorbable sutures.

With the lung retracted toward the apex of the chest, the inferior pulmonary ligament is divided up to the level of the inferior pulmonary vein (Fig. 4.13). An inferior pulmonary ligament lymph node (level 9) should be excised for staging purposes. The inferior pulmonary vein is dissected and encircled in preparation for division. A finger is passed around the vein after entering the appropriate dissection plane and the vein is divided with a vascular stapler (inset, Fig. 4.13). Alternatively, the vein may be clamped, divided, and sutured with a running monofilament thread, or doubly ligated prior to division. At the minimum a tie and a suture ligature are placed to secure the vein. The anterior aspect of the major fissure is now easily completed with a firing of the linear stapler that allows the lobe to be removed.

It is a common misconception that right lower lobectomy is difficult because of the necessity to identify the pulmonary artery within the fissure. If a "difficult" fissure is encountered, it is always best to obtain proximal control of the right main pulmonary artery as the initial maneuver. The artery may then be followed distally beyond the middle lobe branch that leads up to the fissure and facilitates dissection of the fissure minimizing air leaks. Alternatively, the artery may be identified posteriorly from within the crotch formed by the bronchus intermedius and the upper lobe bronchus and the posterior aspect of the fissure completed. Once the artery is identified further dissection within the fissure proceeds expeditiously. Rarely, it is necessary to dissect through the depths of the fissure to identify the artery. In my opinion there is no such thing as a "difficult fissure."

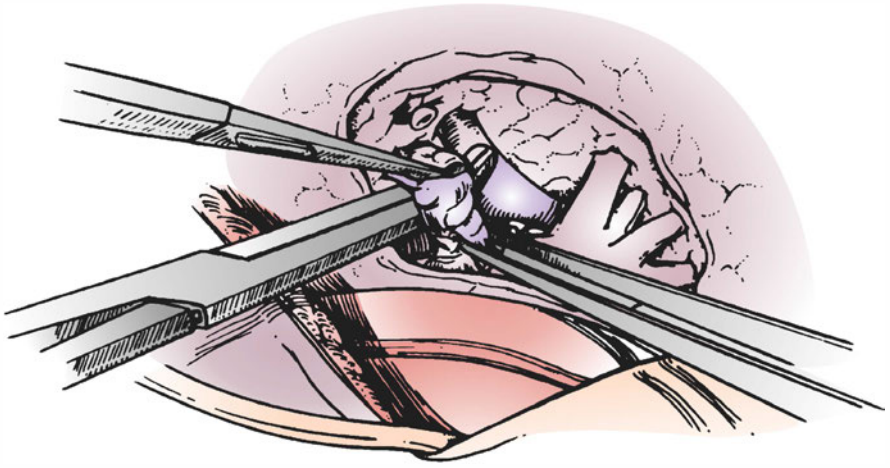


Fig. 4.9. The middle lobe bronchus is encircled with a right-angled clamp and divided with a scalpel. It is important to identify the origin of the middle lobe bronchus to avoid damage to the bronchus intermedius. The bronchial stump is closed with interrupted absorbable sutures or alternatively a stapler may be placed and the bronchus closed. Note the relationship of the middle lobe artery to the bronchus.

BILOBECTOMY

Occasionally, the location of a lesion will mandate removal of the middle and lower lobes, a procedure which can be accomplished en bloc because of the common origin of these lobes from the bronchus intermedius. A tumor originating in the bronchus intermedius usually requires removal of both lobes but a lower lobe lesion that involves the external aspect of the lobar bronchus may also mandate taking the middle lobe. Where an indication exists for bilobectomy, the vascular supply for each lobe is isolated and divided as described for each individual lobectomy. Once the pulmonary arterial branches have been divided the point of division of the bronchus becomes obvious; the bronchus should be divided above the origin of the middle lobe bronchus just distal to the origin of the upper lobe bronchus (Fig. 4.14). Morbidity and mortality for bilobectomy exceed that for lobectomy alone; so this resection should not be performed solely for ease or convenience. The middle lobe should never just be assumed to be

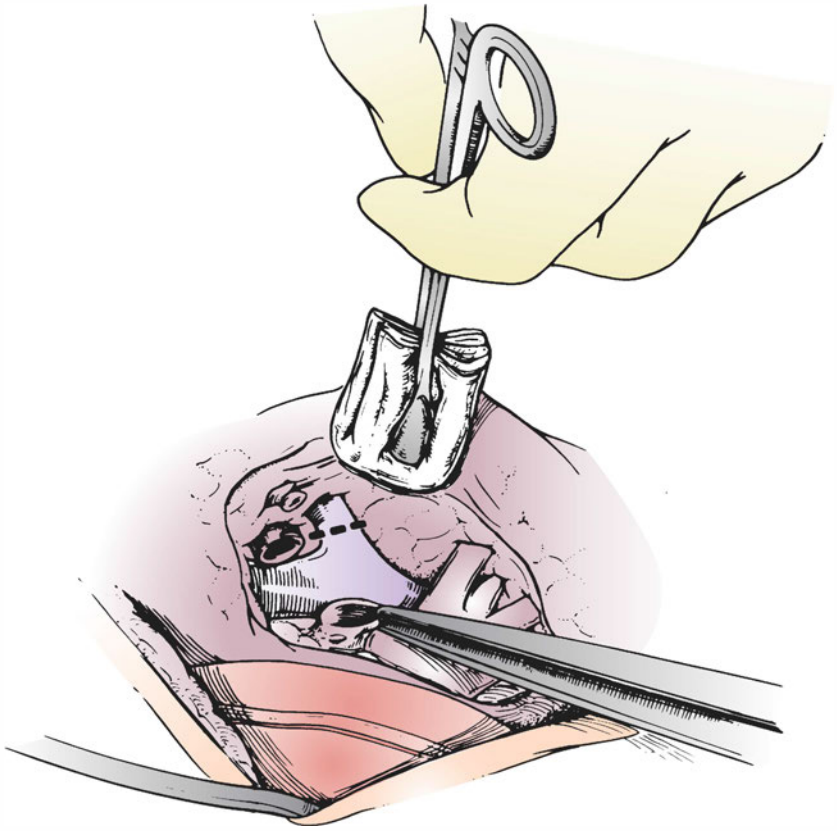


Fig. 4.10. Following division of the middle lobe bronchus, the arterial branch (or branches) is well seen and can be mobilized and divided (*hatched line*). Once the artery is taken the minor fissure is divided with a linear stapler, as is the anterior portion of the major fissure.

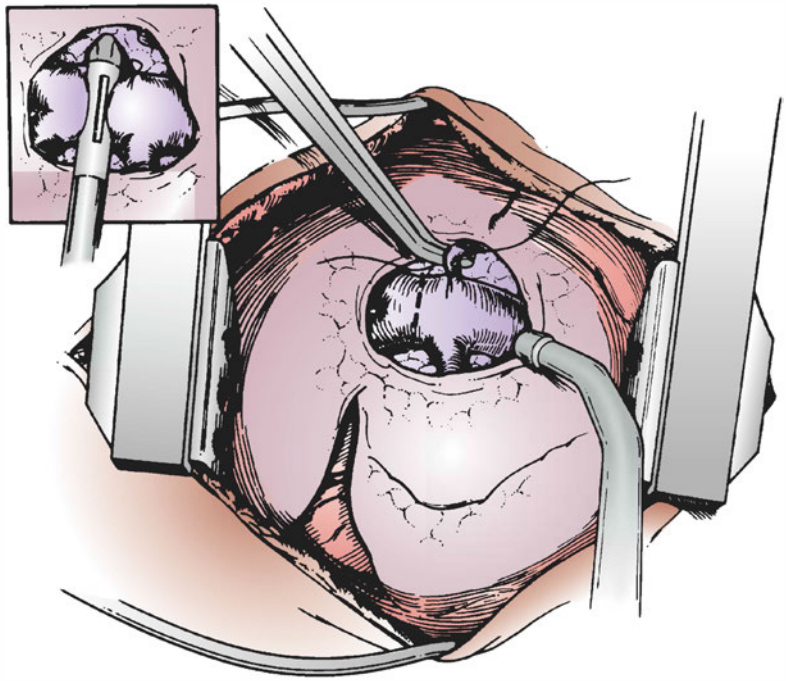


Fig. 4.11. The pulmonary artery within the fissure has been identified and dissected. The position of the branch to the superior segment is such that it must be ligated and divided separately from the rest of the arterial supply to the lower lobe. The superior segmental branch is shown here being encircled by a right-angled clamp and the point of division of the basal arterial trunk is marked (*hatched line*). Note the position of the superior segmental branch relative to the middle lobe artery. **Inset**. The basal arterial trunk is shown being divided by the endoscopic vascular stapler that both ligates and divides. The angle is usually ideal for placement of this stapler, which provides an extremely secure closure of three parallel rows of staples.

expendable. The bronchial stump placed so close to the upper lobe bronchus may be at somewhat increased risk for breakdown compared to other bronchial closures.

Lesions within the bronchus intermedius often present additional problems for the surgeon. These tumors may invade the inferior pulmonary vein occasionally with proximal extension into the left atrium. Careful exploration is mandated to assess resectability prior to dividing any structures. Proximal involvement of the inferior pulmonary vein need not preclude resection if the vessel is able to be encircled or if the extent of atrial involvement is not excessive. Some of these lesions may demand intrapericardial pneumonectomy because of the proximal involvement of the atrium. A sleeve resection of the main bronchus may also be performed if the proximal extent of the tumor involves the bronchus at the level of the upper lobe bronchus. The right main bronchus may be divided proximal to the upper lobe origin and the upper lobe bronchus severed at its origin. The middle and lower lobes together with a portion of main bronchus are removed and the upper lobe anastomosed to the open end of the main bronchus.

Postoperative Mortality

Thirty-day mortality from pulmonary resections is approximately 4%. Lobectomies and lesser resections have mortality between 1% and 2% while pneumonectomies still carry a mortality of 6% to 7%. The mortality rate is directly proportional to increased age, associated diseases, and the extent of resection. Respiratory complications, not surprisingly, are the most common cause of postoperative mortality in patients undergoing pulmonary resection. Cardiac complications also account for a significant percentage of mortality while technical problems such as hemorrhage, bronchopleural fistula, and empyema account for a small but significant percentage of complications leading to death.

Postoperative Morbidity

Approximately 30% of patients undergoing pulmonary resection will sustain a postoperative complication of which approximately two-thirds are minor and the other one-third nonfatal major complications. The most common complication is supraventricular arrhythmia that occurs in up to 20% of patients, depending upon how closely patients are monitored. Most of these respond to simple pharmacologic manipulation and rarely are hemodynamically

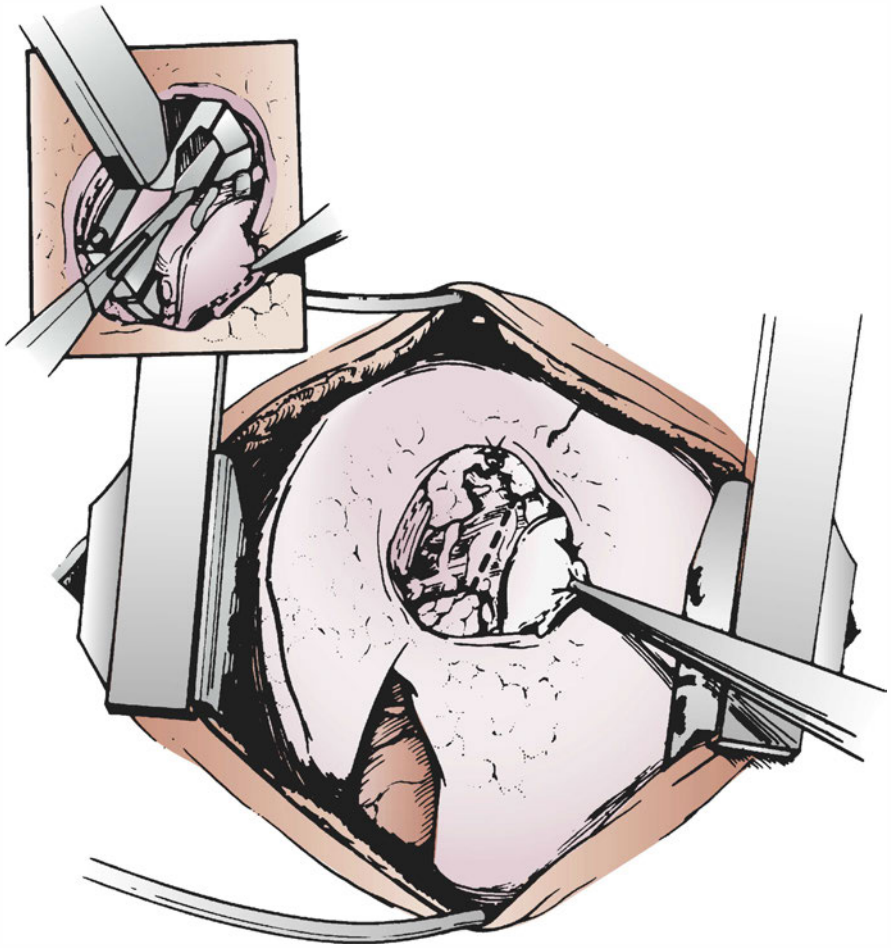


Fig. 4.12. The stump of the lower lobe pulmonary artery is retracted superiorly to expose the lower lobe bronchus. The middle lobe bronchus that comes off the bronchus intermedius at a 90-degree angle must be identified and preserved. If a stapler is to be used, it must be placed in such an orientation as to avoid compromising the orifice of the middle lobe bronchus. The site of bronchial division is shown (*hatched line*). The bronchial division includes the bronchus to the superior segment as shown. Occasionally, it is necessary to close and divide the superior segment bronchus separately. **Inset**. A stapler is placed across the lower lobe bronchus distal to the origin of the middle lobe bronchus. Often the stapler has to be oriented in an oblique fashion to include the superior segment bronchus and avoid the middle lobe.

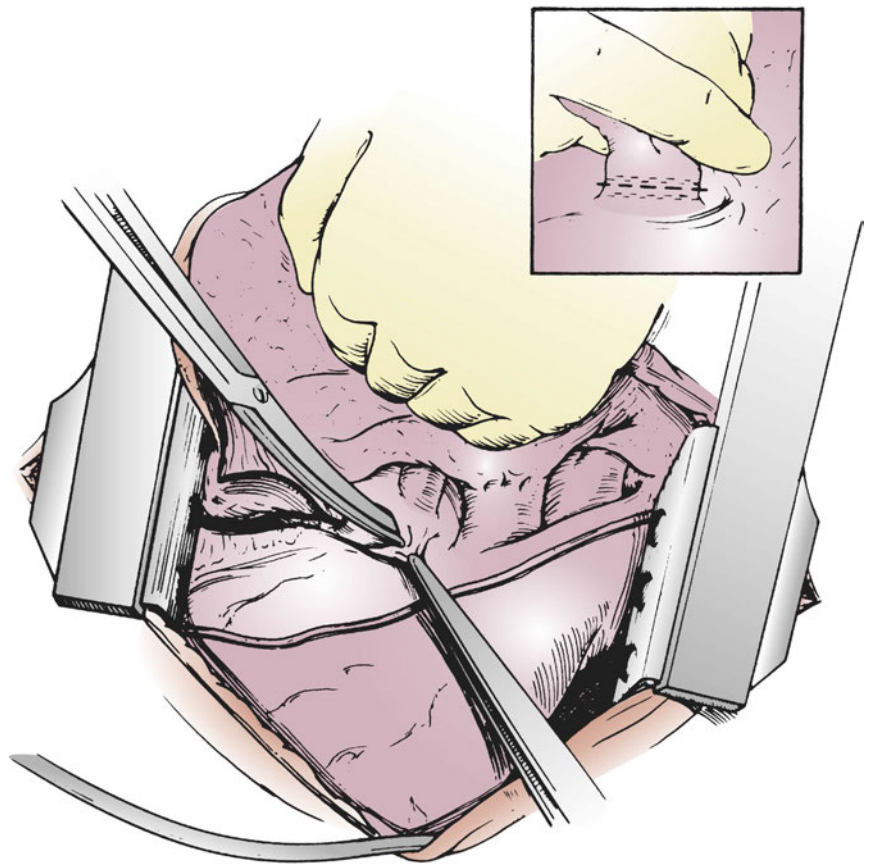


Fig. 4.13. With the lung retracted superiorly the inferior pulmonary ligament is incised up to the level of the inferior pulmonary vein. The vein is shown being dissected by incising the overlying pleura. The vein may then be encircled with a finger and divided between rows of vascular staples or clamped, divided, and sutured closed. **Inset**. The vein has been encircled and two rows of staples placed. The line of division is marked (*hatched line*). Often division of the vein precedes bronchial division, but there is no set order in which structures must be taken.

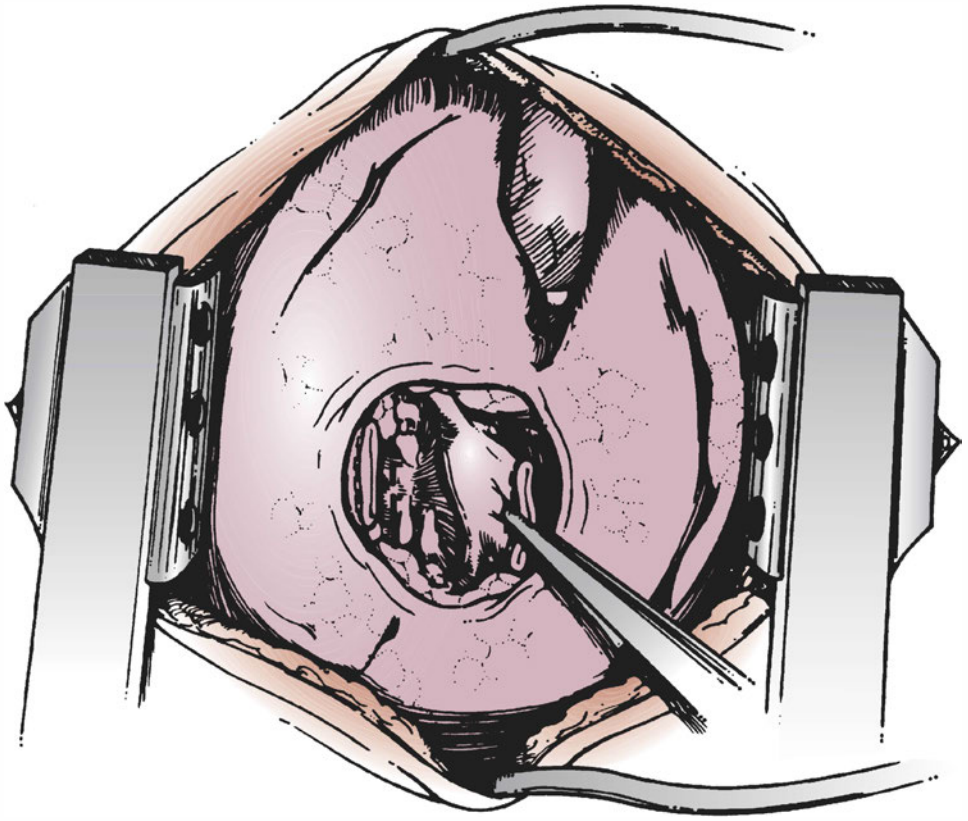


Fig. 4.14. To complete a bilobectomy the pulmonary artery within the fissure is divided proximal to the middle lobe arterial origin, if possible. Often the middle lobe artery must be taken as a separate branch. The bronchus intermedius is then exposed by retracting the artery. The bronchus intermedius is divided proximal to the middle lobe origin and just distal to the takeoff of the right upper lobe (*hatched line*). Surprisingly, the right upper lobe usually is adequate to fill the residual space and space infection problems are unusual.

significant at onset. With appropriate treatment the rhythm reverts to sinus quickly and patients may be taken off the antiarrhythmic drugs usually after 1 month. Other minor complications include postoperative air leaks lasting greater than 7 days and atelectasis. Major nonfatal events most commonly are respiration related with patients developing significant infiltrates and pneumonitis. A small percentage of patients require reintubation in the postoperative period for respiratory failure usually related to the development of an infiltrate. There are no definitive predictors for postoperative pulmonary complications though significant risk factors for major complications include age >60 years, FEV_1< 2 L, weight loss >10%, associated systemic disease, and extent of disease. Pulmonary complications can be minimized with meticulous attention to postoperative respiratory maneuvers including chest physiotherapy and preoperative teaching.

Other complications of pulmonary resection include wound infections, and disturbances in mental status especially in older patients. Notwithstanding our best efforts to avoid them, complications do occur. If recognized early many can be treated without sequelae. Meticulous attention to detail in all phases of management, preoperative, intraoperative, and postoperative goes a long way toward keeping problems to a minimum.

SUGGESTED READINGS

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EDITOR'S COMMENTS

It is a pleasure to be able to comment on my co editor Larry Kaiser's approach to right-sided pulmonary resections. It's been a long time since I have done lung surgery but I certainly have had more recent involvement in lung transplantation. Dr. Kaiser gives an approach that demonstrates his experience at not only performing the operations safely but also treating complications. The technical focus of this chapter is clear and important to every level of reader.

I think the most important aspect from my standpoint is the approach to the dissection of the pulmonary artery. As Dr. Kaiser mentions there is no muscular wall, so the strength is all in the intima. The vessels don't take a lot of traction or inappropriate dissection. He discusses the importance of complete dissection before trying to encircle the vessel. Certainly avoidance of complications is the key here for doing these operations with mortality as low as 1%.

ILK

5 Left-Sided Pulmonary Resections

Larry R. Kaiser

INTRODUCTION TO LEFT-SIDED PULMONARY RESECTIONS

Left-sided resections have a number of unique features distinct from those carried out on the right. The aortic arch is a left-sided structure and its position relative to the pulmonary artery and the left main bronchus is the major defining feature of resections on this side. Access to the proximal left main bronchus and carina is limited because of the aortic arch. Thus the left paratracheal region, a lymph node-bearing area, is difficult to access at thoracotomy. There is no well-defined area where a lymph node dissection is carried out as on the right side. Lymph nodes are dissected from the aortopulmonary window and the subcarinal space and, at times, from the paratracheal area.

Access to the most proximal aspect of the left main pulmonary artery, literally at the level of the pulmonary artery bifurcation, may be gained by dividing the ligamentum arteriosum. The left recurrent laryngeal nerve is highly vulnerable to injury because of its position in relation to the inferior aspect of the aortic arch. This nerve originates from the vagus nerve as it traverses the aortic arch and then "recurs" around the ligamentum arteriosum. Any dissection in the aortopulmonary window places the left recurrent laryngeal nerve at risk of injury.

The left main bronchus also varies significantly from the right. On the left there is a long segment of main stem bronchus prior to the bifurcation of the lobar bronchi as opposed to the right where the right upper lobe bronchus originates within 2 cm of the carina. Sleeve resection of either the upper or lower lobe is certainly feasible though left-sided sleeve resections account for only a minority of these resections in any series.

The lingular segment is analogous to the middle lobe in that it has separate pulmonary arterial supply and venous drainage as well as a distinct bronchus. Lingular segmentectomy was one of the first segmental resections described likely because of its well-defined, discrete bronchovascular anatomy.

Contralateral mediastinal lymph node involvement is much more common with left-sided lesions particularly lesions of the left lower lobe. For that reason, mediastinoscopy or other pathologic staging of the mediastinum is particularly important when assessing lesions of the left lower lobe.

SURGICAL TECHNIQUE

Left Upper Lobectomy

In the realm of pulmonary resections the left upper lobectomy is, perhaps, the most technically challenging. The location of the pulmonary artery in relation to the aorta and the branching pattern of the left pulmonary artery contribute to the technical difficulties. There are a number of potential pitfalls that must be avoided in order to safely complete a left upper lobectomy. Lymphatics from the left upper lobe commonly drain to lymph nodes in the aortopulmonary window (level 5) or paraaortic location (level 6), and these lymph nodes must be removed in order to obtain complete staging information. Despite the classification of these nodal locations as mediastinal (N2), involvement of these lymph nodes with tumor in the absence of other nodal disease is associated with a better prognosis than N2 disease in any other location. Survival with isolated involved level 5 or 6 lymph nodes approximates that of patients who have only N1 lymph node involvement (approximately 40% at 5 years) as long as a complete resection is able to be performed. Access to the superior mediastinum is difficult from the left side because of the location of the aortic arch in relation to the left main bronchus and tracheobronchial angle. For this reason, mediastinoscopy is extremely useful for left-sided lesions even without enlarged lymph nodes present on the CT scan since it allows accurate sampling of the paratracheal area in a manner that is significantly simpler than trying to access this area during thoracotomy. Needle aspiration of left paratracheal and tracheobronchial angle lymph nodes with the aid of endoscopic bronchial ultrasound (EBUS) guidance in some hands may provide equivalent staging information.

Left upper lobectomy is begun by incising the hilar pleura anteriorly and superiorly with the lobe retracted posteriorly. The pulmonary artery emerges from beneath the aortic arch and is located superior and posterolateral to the superior pulmonary vein. The apical segmental branch of the superior pulmonary vein may cross the artery partly obscuring the apical-posterior segmental trunk of the pulmonary artery necessitating division of the venous branch first (Fig. 5.1). The appropriate plane of dissection on the pulmonary artery is entered proximal to the takeoff of the first branch and careful circumferential dissection is carried out. The left main pulmonary artery is safely encircled with the index finger and a blunt-tipped C-clamp is directed toward the encircled finger to allow an umbilical tape to be passed around the vessel. A Rumel tourniquet is placed but not snared to allow the main pulmonary artery to be occluded should it prove necessary. The superior pulmonary vein is dissected and encircled taking care to include the lingular branch. The vein may be doubly ligated or stapled with a vascular stapler and divided. It is both safe and expeditious to employ vascular staplers to ligate and divide both pulmonary veins and arteries. Parallel rows of staples are placed and the vessel is divided. Alternatively, the endoscopic vascular stapler that cuts between parallel rows each with three layers of staples may also be used.

The apical-posterior segmental branch of the pulmonary artery is a short, broad vessel that may be easily avulsed or torn if too much traction is applied when

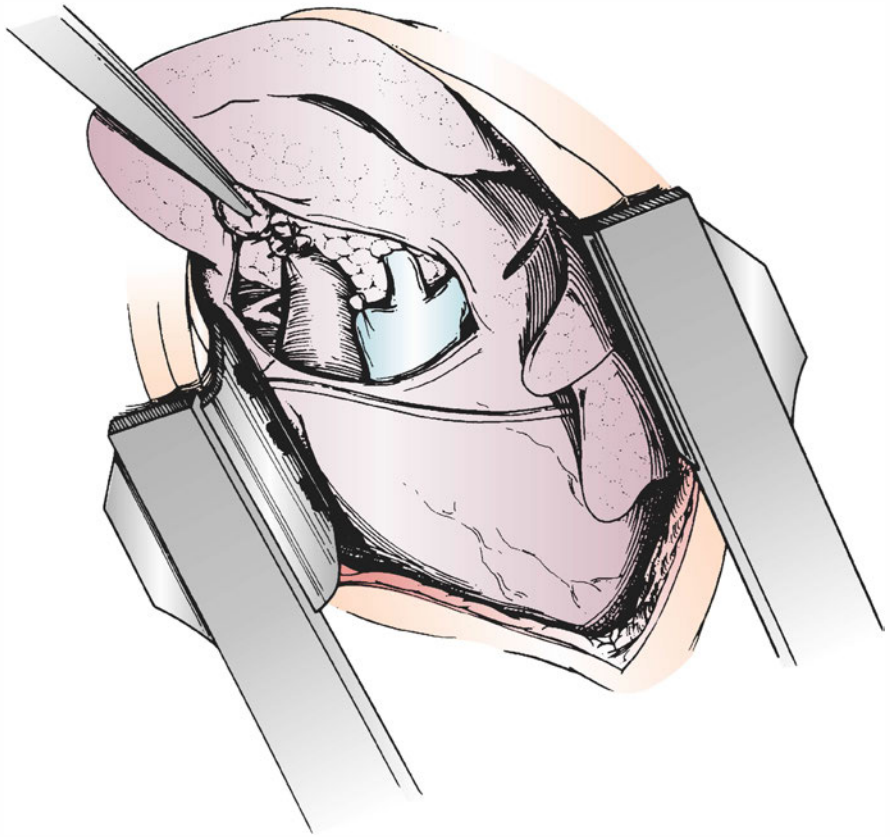


Fig. 5.1. View of the left pulmonary hilum from the right side of the table showing the pulmonary artery and vein. The line of division of the apical-posterior segmental branch of the artery is shown (*hatched line*). A venous branch partially obscuring the artery has been divided and the ligated stump is shown.

retracting the lobe (Fig. 5.1). This is a dreaded complication of left upper lobectomy, which may force a pneumonectomy depending on the extent of the injury. If proximal control of the artery has not been secured, as discussed above, there exists a possibility for a particularly disastrous complication. Trying to get around the left main pulmonary artery in order to place a clamp on the vessel is extremely difficult when at the same time it is necessary to staunch the ongoing hemorrhage from the injury to the artery. Often a significant amount of blood is lost during this maneuver. One should avoid the temptation to wildly try to place a clamp on the vessel. Without encircling the vessel this results in, at best, only partial occlusion and, at worst, further injury to the vessel. Proximal extension of a tear in this portion of the pulmonary artery may result in an irreversible situation with the patient dying from exsanguination. If an injury to the artery occurs and proximal control has not been obtained, one should gently occlude the rent with a finger and assess the situation. It is difficult at best to attempt to suture the pulmonary artery without first gaining proximal control. Attempts to place sutures in the artery under these conditions may result in further injury to the artery as the torrential bleeding does not allow enough visualization to accurately place the sutures. It may be best to open the pericardium to secure additional length of the artery and encircle the artery within the pericardium for proximal control while maintaining the digital pressure on the arterial rent. Once proximal control is achieved, the branch is completely divided and the artery repaired with 5-0 or 6-0 monofilament, nonabsorbable sutures. Rather than avulsion of the apical-posterior arterial branch, a more common traction injury is a hematoma in the vessel from an intimal tear. The intima is the structure responsible for the integrity of the pulmonary artery; so this has the potential to be a disastrous situation. Proximal control should be obtained and the branch vessel ligated preferably proximal to the intimal tear. Placing a ligature at the area of the tear may result in complete avulsion of the branch when the knot is placed. As noted, it is best to recognize that this arterial branch presents special problems and avoiding problems with the pulmonary artery is far better and simpler than having to repair the artery no matter how good you may be at getting out of trouble.

Dissection on the artery continues distally following the artery toward and into the fissure. The left main pulmonary artery resides in an epibronchial location relative to the left main bronchus (Fig. 5.2). As the fissure is entered, the anterior segmental arterial branch to the upper lobe is encountered just proximal and opposite to the origin of the superior segmental branch to the lower lobe. Once the superior segmental branch is identified, the posterior portion of the fissure may be completed with a stapler. The anterior segmental branch is divided between silk ligatures (Fig. 5.3) and the artery is followed further distally until the lingular branches are encountered. There may be a single lingular trunk or two separate branches. The lingular branches are ligated and divided and the anterior portion of the fissure is completed with a firing of the linear stapler (Fig. 5.4). Alternatively, the fissure may be taken following division of the bronchus when it is all that remains holding the lobe in place.

Within the fissure the artery is bluntly reflected inferiorly away from the underlying bronchus, which is located medial (or deep) (Fig. 5.5). The bifurcation of the left main bronchus is visible at this point and care should be taken to avoid damage to the lower lobe bronchus when the upper lobe bronchus is divided. The bronchus needs to be skeletonized and encircled prior to its division. Bronchial vessels should be either electrocoagulated or occluded with metal clips and divided, depending on their size. It should not be assumed that the stapler used to close the bronchus will occlude these vessels. Staplers used on the bronchus are not particularly hemostatic because of the size (3.5 or 4.8 mm) of the staples. To avoid postoperative bleeding from a bronchial artery these vessels should be identified and ligated prior to dividing (or stapling) the bronchus.

Exposure of the left upper lobe bronchus is achieved either from both the anterior aspect of the hilum or from within the fissure. Division of the superior pulmonary vein exposes the bronchus as seen when viewing the hilum from the anterior aspect. After dividing the superior pulmonary vein, there is adequate exposure to work on cleaning off the bronchus and sweeping any lymph nodes upward with the specimen.

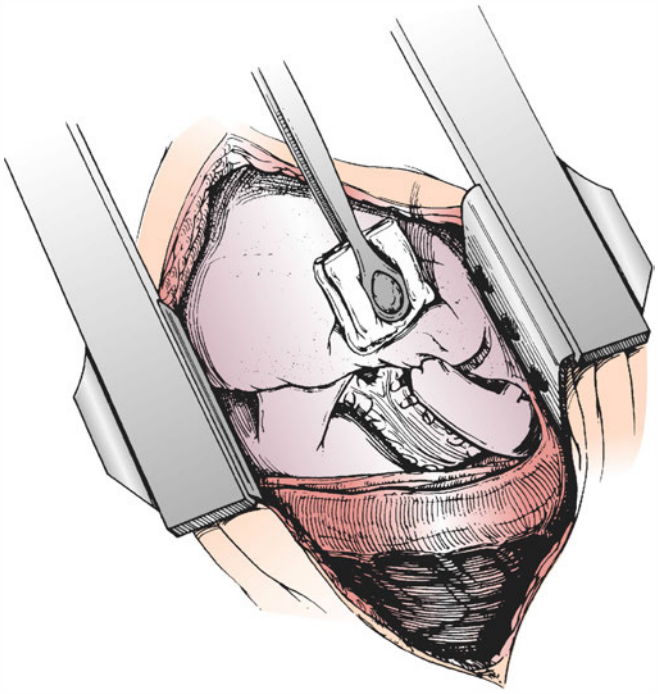


Fig. 5.2. Relationship of the left main pulmonary artery to the left main bronchus seen from the posterior aspect. The main pulmonary artery lies in an epibronchial location. The inferior pulmonary vein is also seen from this posterior view.

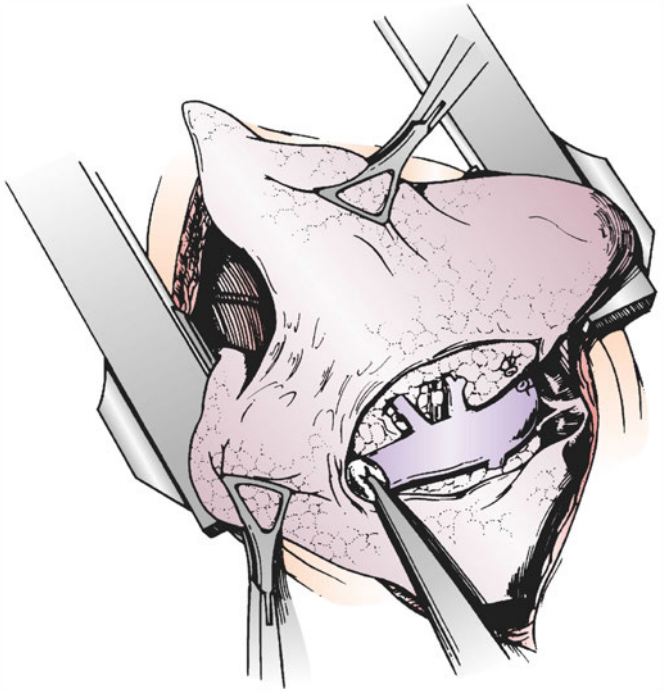


Fig. 5.3. The anterior segmental branch of the pulmonary artery has been divided and the artery exposed in the fissure. Two lingular arterial branches are seen as is the branch to the superior segment of the lower lobe.

Incising the fibrous tissue on the plane of the bronchus at the level of the bifurcation also facilitates division of the anterior portion of the fissure. The thumb and first finger placed at the bifurcation may be used to thin out the parenchyma in this location so as to be able to divide the fissure by firing the linear stapler after placing it through the hole formed by the opposed thumb and forefinger. The upper lobe bronchus is stapled and divided as close as possible to the bifurcation (Inset, Fig. 5.5). Alternatively, the bronchus may be divided with a scalpel (open technique) and closed with individual sutures of either a 3-0 or 4-0 monofilament or braided nonabsorbable material (Fig. 5.6). The bronchus is divided in an open fashion, that is, incised with a scalpel in the presence of endobronchial disease that may be close to the bronchial margin since the stapler, by virtue of its width, obscures what otherwise might be an adequate margin. The importance of a negative margin is obvious and a frozen section of the bronchial margin should be obtained whenever an endobronchial lesion is present if not routinely for all but small peripheral lung cancers. The bronchial stump is checked under saline to assure that the closure is airtight. The anesthesiologist is asked to inflate the lung and hold a pressure of between 25 and 30 cm H_2O.

The inferior pulmonary ligament is incised freeing up the lower lobe, which it tethers though the value of this maneuver is questionable. The intent of incising the so-called inferior pulmonary ligament is to allow the lower lobe to more adequately fill the residual space following removal of the upper lobe. Lymph nodes in the paraaortic (level 6) and aortopulmonary window (level 5) locations are taken. The subcarinal space is opened by incising the mediastinal pleura posteriorly and just inferior to the main bronchus. Small vagal branches going to the lung are divided between metal clips. The contents of the subcarinal space (level 7) are removed using blunt and sharp dissection along with the liberal use of metal clips. The left paratracheal and tracheobronchial angle lymph nodes are most easily sampled at mediastinoscopy but if exposure of these nodal locations is required, it is obtained by dissecting inferior to the aortic arch heading medially. The pulmonary artery must be retracted inferiorly to permit this dissection, which is facilitated by dividing the ligamentum arteriosum (Fig. 5.7). On the left side there is not as well-defined a packet of superior mediastinal lymph nodes that yields a nice distinct packet of lymph nodes as is found on the right side. The nodes must be removed individually. Great care must be taken to avoid the left recurrent laryngeal nerve that "recurs" around the ligamentum arteriosum. If the patient is hoarse in the postoperative period, the vocal cords should be examined with a laryngoscope to assure that the left vocal cord is moving. If the

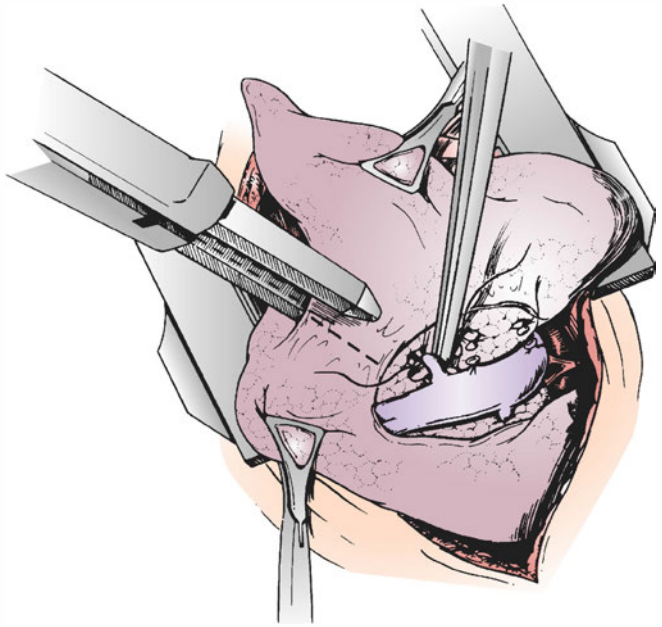


Fig. 5.4. The anterior aspect of the fissure is taken with the linear stapler after the artery has been identified and dissected. A right angle clamp is around a lingular arterial branch in preparation for ligation and division of the branch.

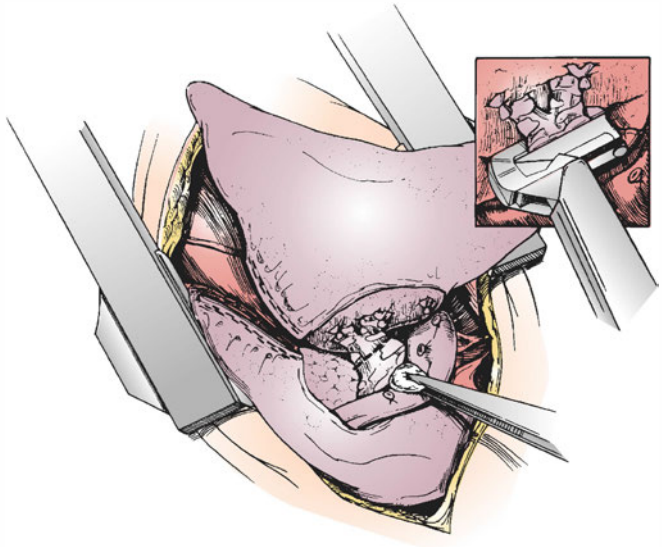


Fig. 5.5. The pulmonary artery is retracted inferiorly to expose the origin of the left upper lobe bronchus. The point of division of the bronchus is shown (*hatched line*) just proximal to the bifurcation. The lingular bronchus and upper lobe proper are easily seen. **Inset**. The stapler is applied to the bronchus proximal to the lingula and upper lobe proper bifurcation. This is done from within the fissure.

left vocal cord is paralyzed, the patient's ability to cough and clear secretions in the postoperative period is markedly impaired and aspiration with subsequent pneumonia becomes a significant risk. Paralysis of the left vocal cord may not become apparent until several days after the operation because of edema of the vocal cord allowing for adequate glottic closure.

A variation of the standard left upper lobectomy is a lingular-sparing upper lobectomy that is useful in patients with borderline lung function where the primary tumor is in the apical or posterior segment of the upper lobe. Lesions in the anterior segment sometimes are amenable to this approach as well. Basically the dissection proceeds as for a standard left upper lobectomy but the lingular branch of the superior pulmonary vein is preserved as are the lingular arterial branches as they are identified in the fissure. The dissection on the bronchus is carried further distal than for standard upper lobectomy to identify the bifurcation between the upper lobe proper and the lingular bronchus. The lingular bronchus is spared as the stapler is applied to the upper lobe proper bronchus. The parenchyma is either "stripped," as described classically for segmentectomy, or divided with a stapler guided by the location of the remnants of the distal ends of the bronchovascular structures and the anterior segmental arterial branch.

Left Lower Lobectomy

In contradistinction to the left upper lobectomy, left lower lobectomy can be one of the most straightforward pulmonary resections and is an excellent resection for those just beginning their experience in pulmonary surgery. The anatomy is quite well-defined and there are only a few traps into which one may be led. Again the role that mediastinoscopy plays in lesions of the left side, particularly left lower-lobe lesions, must be emphasized. Carcinomas of the left lower lobe involve contralateral mediastinal lymph nodes more commonly than lesions in any other lobe because of the almost constant occurrence of lymphatics that cross the midline. For left lung lesions, mediastinoscopy is not only the definitive invasive procedure for sampling right paratracheal lymph nodes but also provides access to the left paratracheal (level 2) and tracheobronchial angle (level 4) lymph nodes. The left-sided lymph nodes are actually easier to access with the mediastinoscope than at thoracotomy because of the location of the aortic arch relative to the left main bronchus. In experienced hands EBUS-guided needle aspiration biopsy of the left-sided lymph nodes may substitute for mediastinoscopy.

The chest is entered through either a standard posterolateral thoracotomy incision (5th intercostal space) or a muscle-sparing vertical axillary incision (4th intercostal space). Even for lower lobectomy there is no advantage to being in a lower intercostal (6th) space since the

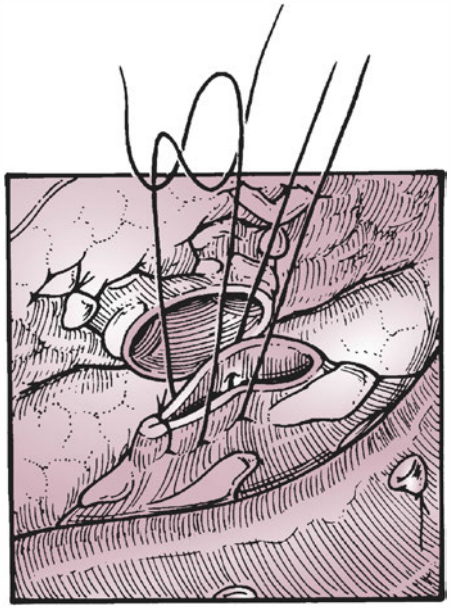


Fig. 5.6. The bronchus has been divided with a scalpel and is being closed with sutures. The first suture should be placed at the midpoint of the closure and all sutures should be placed prior to tying. The sutures should be evenly spaced and should be tied snugly but not so tight that they pull through the membranous bronchus which is quite fragile. The bronchus is closed so that the membranous portion is apposed to the cartilaginous bronchus.

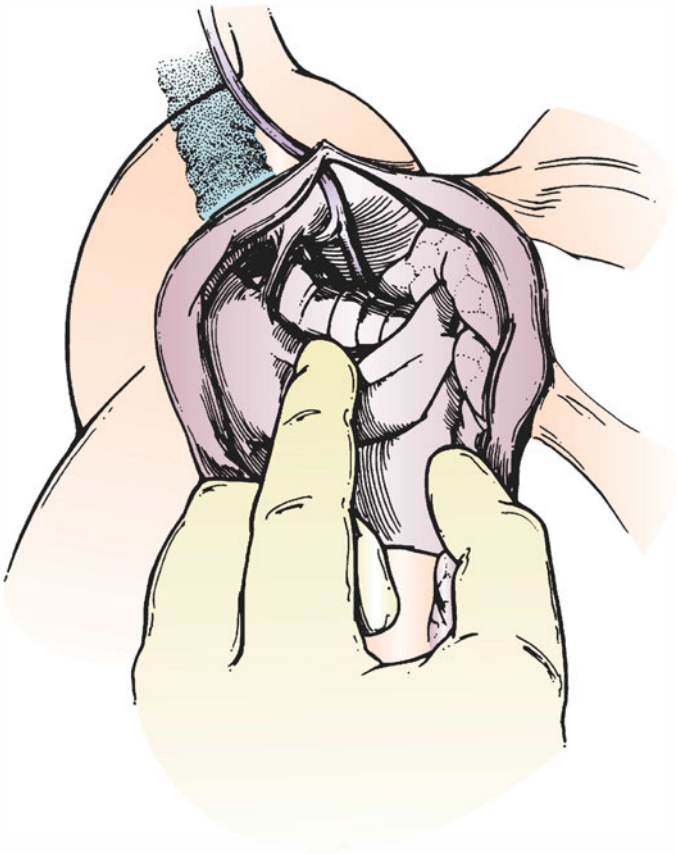


Fig. 5.7. Exposure of the tracheobronchial angle is difficult from the left side because of the relationship of the left main bronchus to the aortic arch. In order to gain access to the tracheobronchial angle the pulmonary artery must be retracted inferiorly. The ligamentum arteriosum is shown intact but access to the left tracheobronchial angle is facilitated by dividing the ligamentum. The vagus nerve and the left recurrent laryngeal nerve are nicely demonstrated showing how vulnerable the recurrent nerve is to injury during dissection in the aortopulmonary window.

position of the hilar structures remains constant though intuitively one would have a tendency to think that if the fifth space is used for upper lobectomy the sixth must be appropriate for lower lobectomy. The pleural space is thoroughly explored to rule out visceral or parietal pleural spread of tumor, to assess lymph node involvement, to assess nodal disease within the fissure, and to establish whether left lower lobectomy is the procedure of choice. It is safest, especially for the less-experienced operator, to begin the dissection with the upper lobe retracted posteriorly to allow access to the proximal pulmonary artery, which is exposed by incising the pleura anteriorly and superiorly and dissecting down onto the artery. The appropriate plane is entered and the artery is encircled with a finger, an umbilical tape is passed, and a Rumel tourniquet is placed but not cinched down. This establishes proximal control if needed, a maneuver which is much easier to carry out at this point than at a time of sudden hemorrhage if the pulmonary artery or one of its branches is entered inadvertently.

If the fissure is near complete, that is, if the pulmonary artery is visible from within the fissure, dissection may begin in the fissure by incising the pleura overlying the pulmonary artery to enter the appropriate plane on the vessel (Fig. 5.8). Once this plane is reached, dissection may proceed in both an anterior and posterior direction along the artery. The superior segmental arterial branch is identified usually just opposite the lingular branch. This branch often needs to be divided separately depending on its location, relative to the lingular branch (Inset, Fig. 5.8). At times the basal segmental trunk of the artery may be divided along with the superior segmental branch but this is dependent on the superior segment branch coming off distal (i.e., inferior) to the takeoff of the lingular branch. The artery to the lower lobe may be taken with a vascular stapler or doubly ligated. The linear endoscopic vascular stapler is ideally suited for ligation and division of the basilar arterial trunk. Once the superior segmental branch of the artery is identified, the posterior portion of the fissure may be completed by incising the pleura overlying the pulmonary artery posteriorly just before the artery enters the lung parenchyma. There are no branches coming off the inferior aspect of the artery posterior to the superior segment branch and a finger may be inserted along the artery between this branch and the posterior aspect of the lung. This allows for placement of a linear stapler and completion of the fissure. Rarely should it be necessary to directly cut into lung parenchyma overlying the artery in order to complete a fissure. Likewise, the anterior aspect of the fissure may be completed with the linear stapler once the artery has been identified.

The inferior pulmonary ligament, a fibrofatty band tethering the lower lobe medially, is incised using electrocautery and divided up to the level of the inferior pulmonary vein (Fig. 5.9). The inferior pulmonary vein may be visualized from either the anterior or posterior aspect of

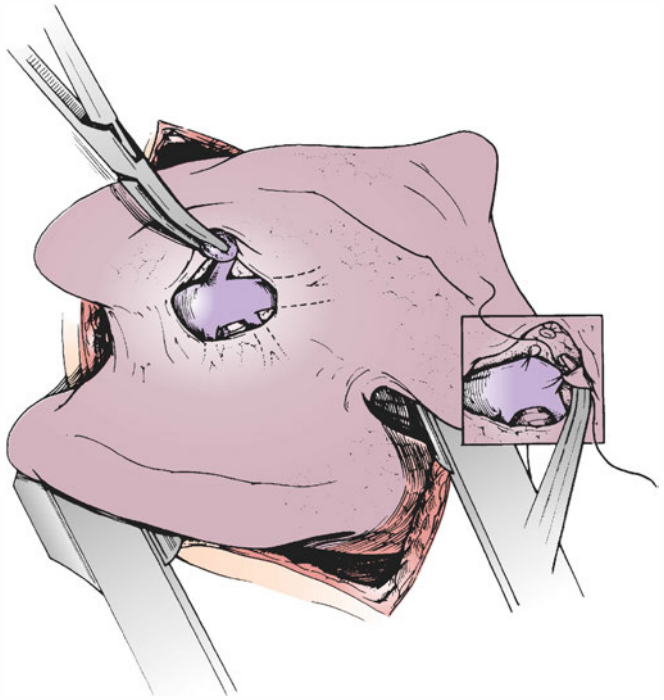


Fig. 5.8. View of the left pulmonary artery in the fissure showing the branch to the superior segment to the lower lobe being exposed. Note the position of this branch relative to the anterior segmental branch (*upper lobe*) and lingular branch. Often the superior segment branch needs to be taken separately but may, at times, be able to be taken in combination with the basilar trunk. **Inset**. The superior segment branch has been ligated and divided, and the basilar trunk is now being ligated.

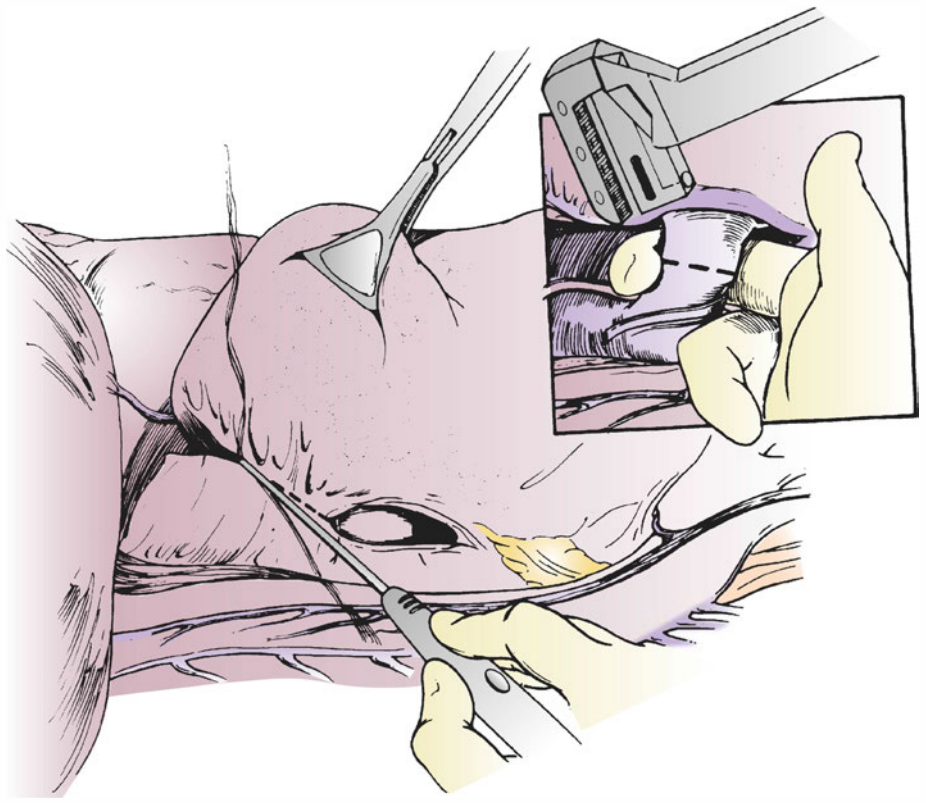


Fig. 5.9. The inferior pulmonary ligament, a fibrofatty band tethering the lower lobe, is being incised with electrocautery as the lower lobe is elevated upward. The ligament is incised up to the level of the inferior pulmonary vein. **Inset**. Once the inferior vein is dissected free it is encircled with a finger or clamp. A vascular stapler may be used to ligate the vein or alternatively the vein may be doubly ligated or clamped, divided, and sutured closed.

the hilum and is encircled with a finger or right angle clamp once the plane of dissection is entered. The vein is then ligated and divided with a vascular stapler (Inset, Fig. 5.9), or clamped and sutured.

By retracting the proximal pulmonary artery stump superiorly the bronchus to the lower lobe is identified (Fig. 5.10). The bifurcation of the left main bronchus will be seen with this maneuver confirming its identity. Care must be taken to include the superior segmental bronchus with the division of the lower lobe bronchus. This may require division of the bronchus in a slightly oblique orientation and this should be done as close as possible to the bifurcation. Identification of the lower lobe bronchus is also facilitated following division of the inferior pulmonary vein as the bronchus lies just posterior and superior to the vein. This identification may be helpful when there is lymph node involvement or tumor within the fissure, making it difficult to approach the bronchus from that aspect.

A lymph node dissection is performed by taking the contents of the aortopulmonary window (level 5), the para-aortic location (level 6), and the subcarinal space (level 7). The subcarinal space is entered by retracting the lung anteriorly and incising the pleura just inferior to the left main bronchus posteriorly. Several vagal branches are encountered going to the lung, which need to be clipped and divided. The contents of the subcarinal space are removed with the aid of metal clips placed on the small bronchial vessels. An inferior pulmonary ligament lymph node (level 9) is also taken. This is most often encountered as the inferior pulmonary ligament is incised and is usually found near the inferior pulmonary vein.

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Patterson GA, Piazza D, Pearson FG, et al. Significance of metastatic disease in subaortic lymph nodes. Ann Thorac Surg 1987;43:155-159.

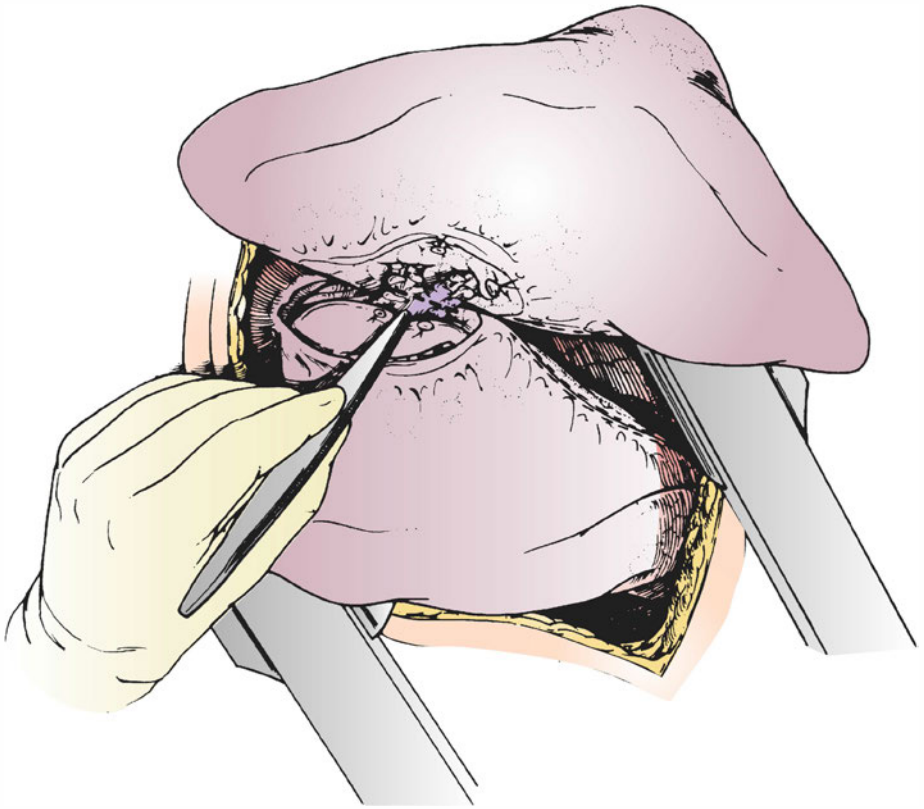


Fig. 5.10. The pulmonary artery is retracted superiorly to reveal the lower lobe bronchus. The line of division (*hatched line*) is just proximal to the bifurcation between the superior segment bronchus and the basilar segmental bronchus. The line of division needs to be slightly oblique to encompass the origin of both of these. Most commonly the bronchus is closed with a stapler, but it may also be cut and sutured closed.

EDITOR'S COMMENTS

Clearly, there are major differences in the pulmonary anatomy on the left side. The techniques are well described and the figures are clear.

I think the most important issue that Dr. Kaiser has raised here is what happens if the left pulmonary artery is injured. It is clear that sewing the vessel in a bloody field is nearly impossible. What is extremely important is the ability to open the pericardium and gain proximal control. This obviously is a technique well used in vascular surgery and it can't be overemphasized in this situation. This is the difference between a surgical mortality and a benign postoperative course. He also emphasizes the importance of cutting the inferior pulmonary ligament and filling the entire chest cavity. These are basic principles in lung surgery but can't be emphasized enough.

ILK

6 Pneumonectomy

Robert J. Cerfolio and Ayesha S. Bryant

Pneumonectomy, defined as the removal of an entire lung, is technically one of the easiest operations and yet one of the riskiest operations performed in the chest. This risk is inherent to the final result of the procedure—having only one lung. Elective pneumonectomy when performed for nonsmall-cell lung cancer (NSCLC) has an operative mortality that ranges from 3% to 12%. This is significantly higher than that for an elective coronary artery bypass grafting. Unlike other paired organs that are removed for malignancy, the lungs are unusual in the fact that a right-sided pneumonectomy has a significantly higher operative risk than a left. The indication for pneumonectomy affects the operative risk as well. When a pneumonectomy is performed for a destroyed lung from an inflammatory process such as tuberculosis, the reported operative mortality is higher and ranges from 3% to 30%. The morbidity from this has been quoted as high as 44%. In this chapter, we focus on pneumonectomy for NSCLC and do not discuss the specific issues and considerations of pneumonectomy for mesothelioma, for destroyed lung, or other less common clinical scenarios. Finally, since many regard a pneumonectomy as a disease in and of itself, a surgeon rarely should go into an operation planning to do a pneumonectomy. Parenchymal preservation should always be front and center in the mind of the surgeon and a pneumonectomy should be done only if there exists no other way to completely remove all of the cancer and achieve negative margins. There are several different types of pneumonectomies. These various types are only mentioned for reasons of completeness but the description of most is out of the scope of this chapter.

TYPES OF PNEUMONECTOMIES

There are several different types of pneumonectomies with the most radical being extrapleural pneumonectomy, defined as the removal of the entire lung along with the ipsilateral pleura, hemidiaphragm, and hemipericardium. This type of operation is most commonly done on patients with mesothelioma. Other types of pneumonectomy include completion pneumonectomy, which is removal of the entire remaining lung after a patient has had some other portion of that lung removed at a previous operation. There are also intrapericardial and extrapericardial pneumonectomies. The former is often performed for anatomic reasons such as for large central tumors. A carinal pneumonectomy refers to the removal of an entire lung in addition to the carina. This requires an anastomosis between the remaining main stem bronchus and the distal trachea. This chapter focuses only on the surgical techniques of a standard pneumonectomy.

INDICATIONS FOR PNEUMONECTOMY

Prior to taking any patient to the operating room for a possible sleeve resection and/or what might turn out to be a pneumonectomy, the patient's pulmonary function tests and cardiovascular status must be carefully assessed and a decision rendered regarding suitability. In addition, a pneumonectomy for a presumptive malignancy should never be performed until tissue confirmation has been obtained. Often the inability to perform a parenchymal-sparing resection is discovered intraoperatively; so the preoperative evaluation should always include an assessment of the patient's ability to tolerate a pneumonectomy. This possibility should be discussed with the patient preoperatively. The thoracic surgeon and the patient must weigh the risks and benefits of pneumonectomy, including increased morbidity and mortality and decreased exercise tolerance against the benefit of potential cure and increased survival.

We perform a cardiac stress test and an echocardiogram on all of these patients. Reversible cardiac ischemia is a contraindication to elective pneumonectomy; therefore, a thorough evaluation of the patient's cardiac status must be undertaken. Patients should have areas of reversible myocardial ischemia revascularized and/or cardiologic clearance prior to elective pulmonary resection. On echocardiogram, the presence of a patent foramen ovale should be known preoperatively and significant valvular disease assessed and corrected if severe.

The baseline pulmonary function and the extent of the planned resection help guide the surgeon. Historically, there have been multiple articles proposing lower limits of various pulmonary function study variables below which operative risks become prohibitive. These limits should be used only as guidelines. However, a decision must be made for each patient on an individual basis. The old teaching that the FEV_1 must be 800cc or greater at the completion of a pulmonary resection is obsolete. This old surgical dictum obviously does not take into account the size of a patient. An FEV_1 of 800cc for a four-and-three-quarter-foot tall, thin female is more than adequate. Thus, the percent predicted FEV_1 (FEV_1%), which takes into account the various shapes and sizes that patients come in, is a much better representation. The percent postoperative predicted ($popFEV_1$%) and percent postoperative predicted diffusion capacity of the lung for carbon monoxide (popDLCO%) have been shown to be reliable predictors of postoperative morbidity and mortality. These values should be calculated with the aid of a quantitative perfusion lung scan when patients have radiologic evidence of segmental, lobar, or greater areas of atelectasis. Important studies have shown that when the $popFEV_1$% is less than 40, and the popDLCO% is less than 40, the operative risks are significantly increased. These values are guidelines since each patient and clinical situation is different.

Another important assessment besides the $popFEV_1$% and the popDLCO% is the arterial blood gas measurement. The presence of significant hypercapnia raises an important red flag. When the preoperative arterial blood gas, a mandatory test prior to possible pneumonectomy, shows a $PaCO_2$

of 48 mmHg or greater the operative risk is significantly increased. We have even lowered this in our practice to 45 mmHg. However, the surgeon and patient again must decide together before operation as to what will be done if a sleeve resection with parenchymal sparing is not technically feasible or if after the completion of a sleeve resection a positive bronchial margin is identified.

Finally, since a pneumonectomy is associated with an increased risk compared to a lobectomy, certain oncologic principles need to be considered. The presence of residual or recalcitrant N2 disease (after neoadjuvant therapy for biopsy-proven N2 disease) is, in our opinion, an absolute contraindication for pneumonectomy (except in certain young patients with high popFEV$_1$% and popDLCO% or those who are having significant hemoptysis, etc.). Once the decision has been made to perform a sleeve lobectomy or pneumonectomy, the operation should follow certain principles and be carried out in a certain sequence.

THE SURGICAL PROCEDURE: RIGHT-SIDED PNEUMONECTOMY

Despite the fact that there have been vast improvements in the field of general thoracic surgery and anesthesia, a pneumonectomy done today is not much different from the one done 30 years ago, except perhaps for the advent of stapling devices. After preoperative staging with the use of integrated positron emission tomography (PET)/ computed tomography (CT) scan and CT scan, along with a careful assessment of the cardiopulmonary reserve, and the establishment of the absence of N2 disease via mediastinoscopy, transesophageal ultrasound with fine needle aspirate, or VATS, the patient is finally prepared for pneumonectomy. Preoperative bronchoscopy is performed and occasionally that alone can tell a surgeon that a pneumonectomy is required to achieve an R0 (negative margin) resection. If a lesion is attached (not invading, but attached) to the right proximal bronchial wall and one is able to get distal to it with the bronchoscope and it continues as one contiguous lesion down into the bronchus intermedius and into the right lower lobe, then right pneumonectomy is the only appropriate procedure. One must be sure that it is not simply tumor thrombus coming out of the right lower lobe bronchus, but rather is tumor adherent to the bronchial main stem wall. Similarly, if a lesion is quite large and involves the main pulmonary artery on the right and courses distally involving branches of the basilar pulmonary artery, a right pneumonectomy is required. Otherwise, in the vast majority of patients a sleeve resection should be the desired approach and the decision to do a pneumonectomy is not made on preoperative bronchoscopy, but rather on surgical exploration.

After the preoperative bronchoscopy the appropriate devices are placed—epidural catheter for postoperative analgesia (place prior to anesthesia), a double-lumen endotracheal tube (placed by the surgeon if there is a large and/or bloody lesion in the proximal airway to avoid any blood from spilling into the soon-to-be-only lung), an arterial line, central line, warming blanket, and serial compression devices on the legs. The patient is turned onto the left lateral decubitus position, carefully padded, and secured. There are several types of possible surgical approaches including a vertical axillary thoracotomy, and so on, but we focus on the most common approach for a pneumonectomy, a posterolateral thoracotomy. This can be performed in several ways as well, but we prefer cutting about half of the posterior aspect of the latissimus dorsi muscle, sparing all of the serratus anterior muscle and entering the chest over the uncut, unshingled, and unbroken sixth rib. We also strongly believe that an intercostal muscle flap should be harvested in any patient who is going to undergo a sleeve lobectomy or possible pneumonectomy (as shown in Fig. 6.1) for use either in covering a bronchial stump or wrapping around a bronchial anastomosis. We have demonstrated the usefulness of this pedicled graft in over 400 patients. It should be harvested and mobilized prior to the placement of a chest retractor. This provides a well-vascularized, soft, pliable muscle that is free of periosteum that does not calcify over time when harvested in this manner. It can reach to buttress any bronchus and it provides a barrier between sleeved pulmonary artery and/or bronchus though it should not be circumferentially wrapped around any anastomosis. It only takes a few minutes to mobilize and we have shown in a prospective randomized trial that it also is associated with a decrease in the pain of thoracotomy. If a sleeve resection is not performed, the muscle can then be tacked to the bronchus with the use of interrupted sutures as shown in Figure 6.2.

Once the rib spreader is placed, the chest is explored and carefully inspected to rule out pleural effusion that could be consistent with T4 disease, metastatic nodules on the pleura or diaphragm that could represent M1 disease, or previously nonimaged pulmonary nodules. We prefer to remove all the mediastinal (N2) lymph nodes and perform a complete systematic thoracic lymphadenectomy as opposed to simple nodal sampling. If any of the N2 nodes look or feel suspicious or were imaged by CT and/or integrated

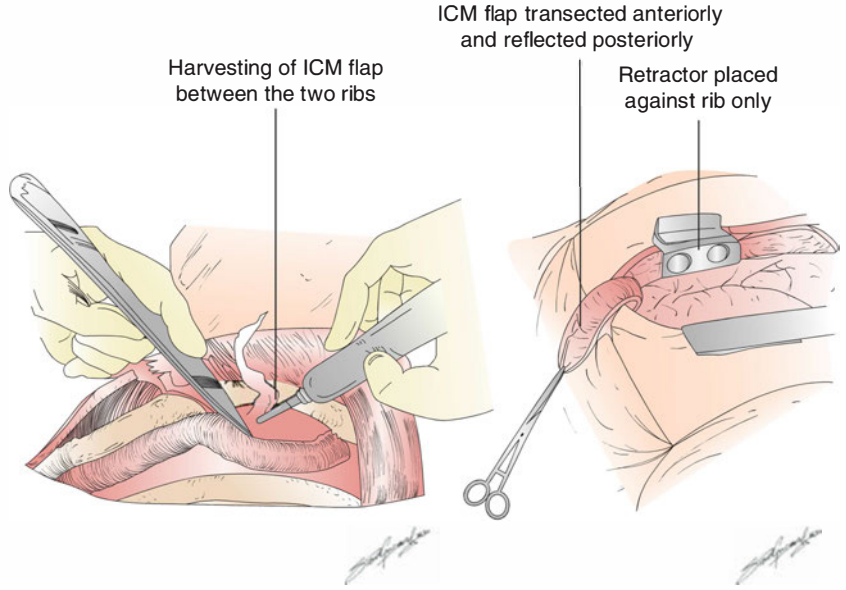


Fig. 6.1. Harvesting of an intercostal muscle off of the inferior aspect of the sixth rib and carefully off of the top bottom part of the fifth rib using a cautery so it is devoid of periosteum.

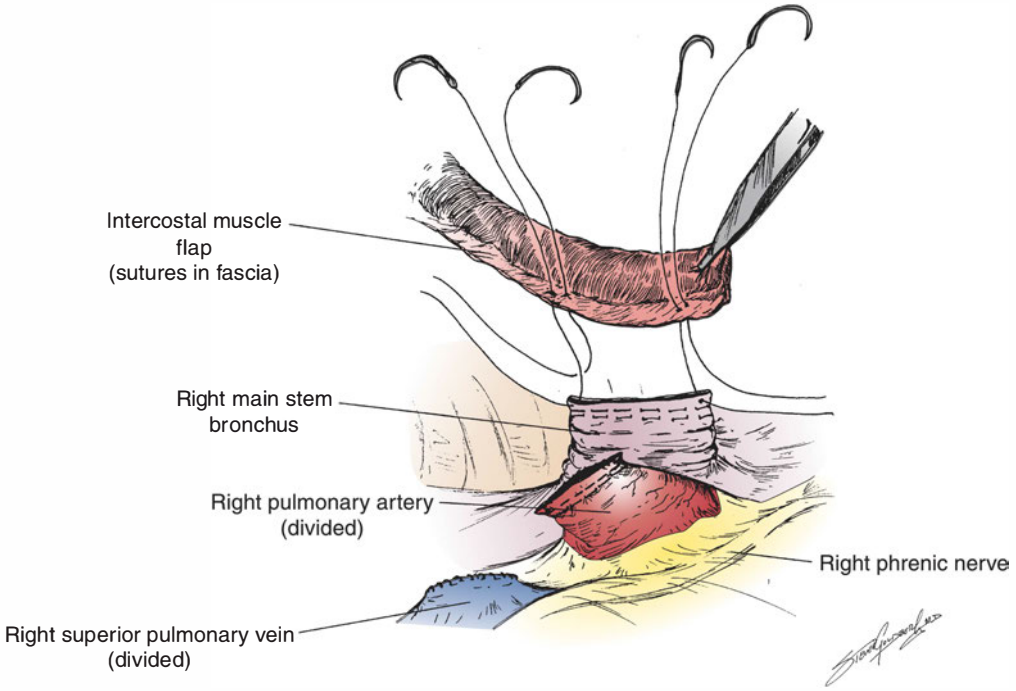


Fig. 6.2. The intercostal muscle is parachuted down onto the right main stem bronchi for coverage. Note the small bites on the bronchus that are taken distal to the staple line so as not to injure its blood supply. Note that the muscular aspect of the flap is brought down onto the right main stem bronchus.

fluorine-18 fluorodeoxyglucose (FDG)-PET-CT as suspicious and were not ruled out as harboring cancer prior to thoracotomy, frozen section analysis should be performed. Once the decision has been made that resection is the appropriate option, the lung is retracted posteriorly and the anterior hilum is exposed. The hilar pleura is incised posterior to the phrenic nerve. The superior pulmonary and the inferior pulmonary vein are dissected free. Lymph nodes from this area are removed and the small veins that lead to the phrenic nerve are carefully coagulated so as not to injure the nerve since a functional phrenic nerve is important even following pneumonectomy. In rare situations, a single right pulmonary vein is found that makes a sleeve resection more difficult because the blood has to be baffled back into the left atrium. The inferior pulmonary ligament should have already been released during the inspection phase to allow removal of the level 9 and 8 lymph nodes. The inferior pulmonary vein is then easily encircled with an index finger as shown in Figure 6.3. If the subcarinal lymph nodes have been completely removed, this move is facilitated since the finger is placed in between the superior pulmonary vein and middle lobe vein and then comes around anterior to the right main stem bronchus. Dissection is then carried out anteriorly and more superiorly. Since a sleeve resection is usually attempted first, we often isolate the superior pulmonary vein next,

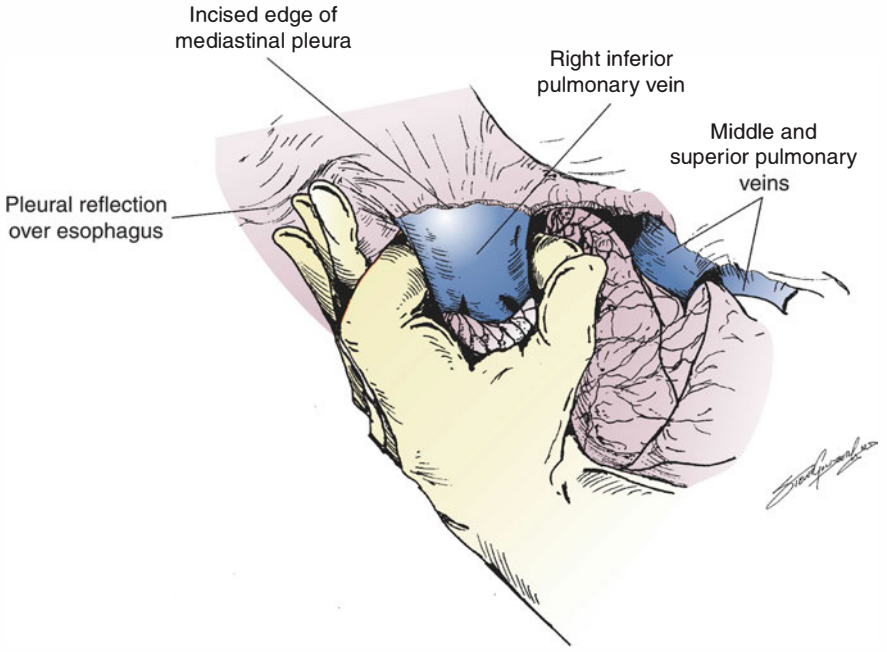


Fig. 6.3. A finger is placed around the right inferior pulmonary vein and it is encircled.

depending on the size and location of the tumor and remove the N1, level 10, and level 11 lymph nodes. Very commonly, there is a large hilar lymph node that interdigitates between the superior aspect of the superior pulmonary vein and the main pulmonary artery just as it courses distally to the superior vena cava. If one is able to completely remove this large N1 node, the course of the distal main pulmonary artery becomes visible. This makes encircling the superior pulmonary vein safer and easier. The vein is carefully encircled around its posterior aspect so as not to injure the main pulmonary artery that runs just beneath it. If the tumor is large and proximal, the pulmonary artery may have to be encircled as the initial maneuver. At times it may be necessary to obtain control of the intrapericardial portion of the right main pulmonary artery. The pericardium should be opened away from the phrenic nerve,

which must be visualized and spared. Opening the pericardium allows central inspection to assess resectability and allows for proximal control of the vessels. However, the intrapericardial mobilization may increase the incidence of postoperative atrial fibrillation (AF). A finger is then used to encircle the main pulmonary artery and the superior pulmonary vein.

The sequence of ligation of the hilar structures is highly dependent on the position of the lesion and the surgeon's preferences. Prior to taking any vessels, we have shown in a nonrandomized but prospective trial that giving 250 mg of Solu-Medrol may help prevent the complication of postpneumonectomy pulmonary edema (PPE). Thus, we recommend giving this 5 to 10 minutes prior to ligating the vessels. One should also test clamp the pulmonary artery for a minute or two and ensure that the patient's hemodynamics tolerate the shunting of all of the pulmonary blood supply into the left lung. If the patient's blood pressure drops quickly and if the clamp or the fingers/hand used to clamp the pulmonary artery is not compressing the heart, this suggests that the patient will not tolerate a pneumonectomy. This move should be repeated several times, but if it continues to occur the vessels should not be taken and the operation should be aborted. This is a very rare occurrence but is a final check of the patient's suitability for pneumonectomy. In this situation, despite the careful preoperative evaluation that suggested that the patient was a candidate for pneumonectomy, the procedure cannot be carried out.

There is little data that documents the advantage or disadvantage of taking the vein prior to the artery. Kurusu has reported an oncologic advantage in taking the vein prior to the artery because it may prevent the accidental dislodgement of cancer cells into the systemic circulation during the manipulation and dissection of the tumor. Others believe there is less blood lost in the removed lung when the vein is taken after the artery. There is no data to support these latter claims. However, if a pneumonectomy (or lobectomy) is performed robotically, there is distinct advantage to taking the vein last. This prevents engorgement of the specimen. If the lung to be removed becomes swollen from early vein ligation, it leads to more bleeding from the specimen, less working room in the chest during minimally invasive surgery, and more difficulty in manipulating the specimen from one side to the other.

If a thoracotomy is the chosen surgical approach then the vessels can also be ligated in several different ways. We prefer to take the vein with a vascular stapler that staples and cuts simultaneously, but prefers to take the artery with a vascular stapler that only staples. The staplers disperse the closure of the vessel over its entire length as opposed to bunching it up into one suture ligature. Another method involves clamping the artery, cutting, and then oversewing the end with a running Prolene suture (Ethicon, Cincinnati, Ohio) along its length. This obviously takes longer to do.

Once the vessels have been cut, the bronchus is the only remaining structure that is keeping the lung in vivo. We prefer not to take the bronchus first on a right or left pneumonectomy as we often do on a right upper lobectomy because it is easier to achieve a short stump if the bronchus is taken after the vessels are divided. However, on the right side the bronchus can be taken prior to the vessels if needed to obtain better access to the pulmonary artery. It can make the dissection easier for large anterior tumors that are too big to allow for anterior dissection of the hilar structures. It can be taken with a knife or a stapler (we use 4.8-cm-long staples) before the artery or veins or divided. If taken with a knife, one can visualize the trachea and ensure the position of the double-lumen tube. If taken with a stapler, one should ask the anesthesiologist to inflate and then deflate the balloon on the tracheal cuff side after closing the stapler but before firing the instrument to ensure that the tracheal balloon of the double-lumen tube has not been entrapped. Either way a short stump is a crucial and mandatory part of a successful right pneumonectomy.

We divide the azygous vein first with a vascular stapler prior to stapling the right main stem bronchus. One must ensure the central venous line is not in the azygous vein. It is usually very easy to see the blue tip of the central line in the superior vena cava or even in the azygous vein. In rare situations when an intercostal muscle is not available and if there is no pericardial fat pad, the azygous vein can be used as a form of bronchial coverage. This is accomplished by dividing the vein as far posteriorly as possible and then ligating the other end tied flush against the superior vena cava. The vein is cut as distally as possible and this provides a long flap of a defunctionalized vein that can be split and used to cover the right main stem bronchus. We frequently use this technique for a radical extrapleural pneumonectomy for mesothelioma because the intercostal muscle is not useable as it may be infiltrated with tumor cells. It is important to note that a pleural flap does not provide adequate coverage since it is paper thin and is often nonviable within 48 hours after surgery.

Once the pulmonary vessels have been ligated and divided, the bronchus is taken last. The lung is retracted so the right main stem is exposed. If all of the subcarinal lymph nodes have been dissected and the large bronchial artery that comes from the undersurface of the carina to the subcarinal nodes has been clipped, the left main stem bronchus is already exposed. A stapler is placed (more easily on the right than on the left because of the absence of the aortic arch on the right in most patients) and fired flush with the trachea. As described above, one must ensure that there are no parts of the double-lumen tube in the staple line and that no suction catheters are within the bronchus before stapling and cutting. Although there is some literature about handsewing the bronchus, there is no real data that suggests that handsewing is better than stapling or vice versa. We recommend the stapling technique.

The bronchial margin should be sent for frozen section analysis as should the artery or vein margin if the tumor is close. The chest is then filled with warm water, not saline. We prefer the former because of the oncologic possibility that it may lyse tumor cells that are floating in the chest. Also it is easier to see through it than saline to check the stump for a leak. The double-lumen tube should now be opened to both chests and the stump is tested. If a bronchial leak is noted, nonabsorbable monofilament sutures are placed under the staple line in a vertical mattress fashion with knots opposite the pulmonary artery. The bronchial closure should be interrogated again to assure that there is no stump leak. The intercostal muscle flap is then set in place to cover the entire bronchus (as shown in Fig. 6.2). The sutures that are placed in the bronchus should be small superficial bites so as not to injure the stump or its blood supply. The entire stump is covered. We have used this method to cover the bronchus after lobectomy and main stem bronchus after pneumonectomy in over 500 patients, including over 100 who have had preoperative radiation and have had only two BPFs.

We prefer leaving a chest tube in the postpneumonectomy space mainly to monitor postoperative bleeding but also to allow for setting the mediastinum. The best way to handle the postpneumonectomized space has never been described. Some surgeons recommend aspirating the air out with a syringe after the chest has been closed, just prior to leaving

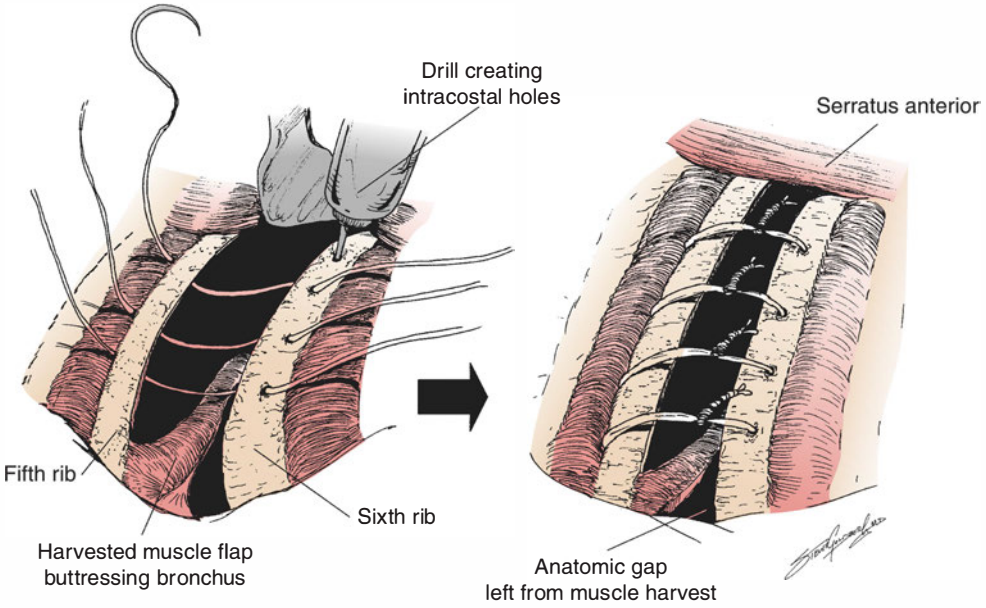


Fig. 6.4. Closure of the chest after pneumonectomy using intracostal sutures.

the operating room. Others recommend placing a chest tube and rolling the patient on his or her back after the operation is completed, and then removing it prior to going to the recovery room. We prefer leaving it in overnight on water-seal only to assess for postoperative bleeding. We attach the tube to a conventional pleural drainage system and place a sign on it that reads "NO SUCTION." This avoids errors postoperatively. A special "pneumonectomy"- balanced drainage system is also available and we have come to prefer it, but first aspirate the air out of the space just prior to connecting to the chest tube.

After a pneumonectomy, the mediastinum shifts over time toward the operated side and this shift can cause serious problems such as postpneumonectomy syndrome, which is described later in this chapter. We believe it is best to have the trachea just to the left of the midline after a right pneumonectomy, since the remaining left lung will slowly displace the trachea toward the pneumonectomy space.

The chest is closed after the placement of a chest tube using intracostal sutures to decrease pain as shown in Figure 6.4.

THE SURGICAL PROCEDURE: LEFT-SIDED PNEUMONECTOMY

The oncologic and physiologic principles for a left pneumonectomy are the same as for a right. However, because the anatomy is different, some special considerations must be mentioned. The left pulmonary artery is shorter than the right. The ligamentum arteriosum tethers the left main pulmonary artery. This structure can be divided to access the most proximal portion of the left main pulmonary artery if necessary but it is not necessary for a standard left pneumonectomy. Routinely this should be avoided since the left recurrent laryngeal nerve is wrapped around this structure and may be injured when the ligamentum is divided. Because of the short left main pulmonary artery, intrapericardial dissection is more commonly needed on the left than on the right. If a patient has had a previous coronary artery bypass procedure, great care must be taken to avoid injuring the left internal mammary artery and the vein grafts during the pneumolysis phase of a left pulmonary resection. Once the decision has been made that a left pneumonectomy is indicated (the patients is N2 negative, a sleeve is not possible, etc.,) the dissection starts, as on the right, by incising the hilar pleura reflection. The avascular plane between the pleural reflection and the pericardium is dissected and the inferior and superior pulmonary veins are identified. Again, the phrenic nerve must be avoided. As described on the right side, the inferior pulmonary ligament is released and the level 9 inferior pulmonary ligament lymph nodes are removed in addition to the level 8 periesophageal and the level 7 subcarinal lymph nodes. If this is done it, becomes easy to get a finger between the inferior pulmonary vein and the left main stem bronchus and encircle the inferior pulmonary vein with a vessel loop (as shown in Fig. 6.5). A finger can be used to get around the superior pulmonary vein as well, but prior to doing this, the edges of the vein must first be dissected free. Care must be taken when dissecting between the superior edge of the pulmonary vein and the inferior border of the left main pulmonary artery being certain to avoid the anterior-apical trunk of the left pulmonary artery. Additionally, the superior pulmonary vein lies anterior to the left main stem bronchus. Large proximal tumors with extraluminal disease can present difficulty when attempting to encircle the vein. Occasionally, the bronchus can be taken first; however, on the left side especially, we prefer taking the bronchus last since it is much more difficult to achieve a short bronchial stump on the left than on the right because of the position of the aortic arch relative to the trachea. Once the superior pulmonary vein is encircled (as shown in Fig. 6.6), a loop is placed around it and the pulmonary artery is mobilized just distal to the ligamentum. The index or middle finger can be placed around it to fully mobilize the vessel. It should be test clamped and the patient's hemodynamics observed as described on the right side. Once all three vessels are encircled, we favor giving 250 mg of intravenous Solu-Medrol and test clamping the artery as described above. The veins and artery are taken as described on the right side leaving the left main stem bronchus.

The biggest difference between a right and left pneumonectomy probably is the position and length of the bronchus. The left main stem bronchus is the longer of the two main stem bronchi and its location with respect to the aortic arch makes it more difficult to achieve a short bronchial stump. A short bronchial stump is

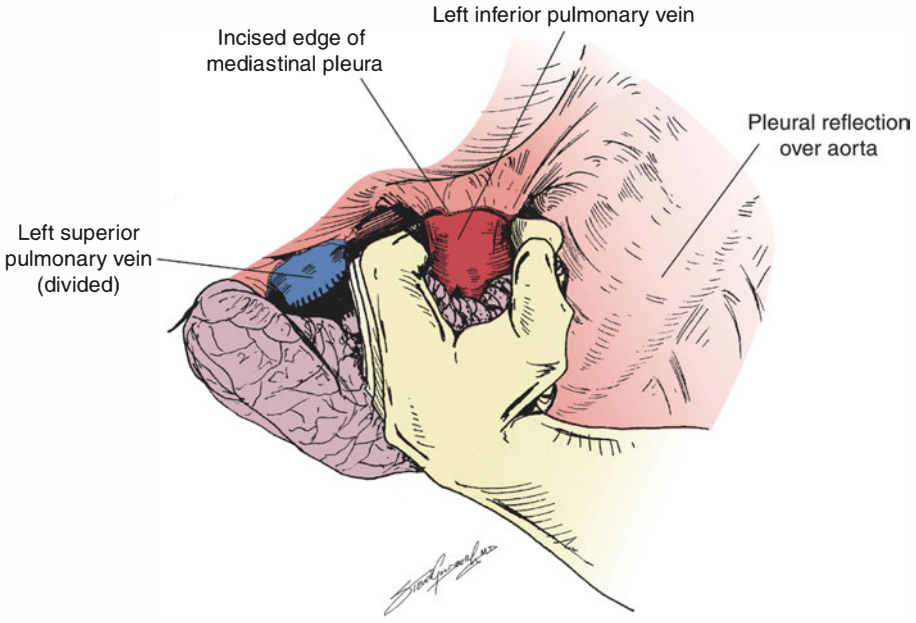


Fig. 6.5. A finger is placed around the left inferior pulmonary vein and it is encircled.

a mandatory part of a pneumonectomy as it keeps the incidence of a BPF low by preventing the pooling of secretions and likely remains better vascularized. The best technique involves pulling the lung up and aggressively retracting it out of the chest. In order to do this, the pericardial reflection must be taken off of the left main stem bronchus. Often there may be a piece of a subcarinal lymph node left that was not completely removed in the lymphadenectomy portion of the case. The remainder of that node should be removed to allow for optimal visualization of the left main stem bronchus. Care must be taken on the left side, as on the right, not to impinge on the opposite side main stem. The tracheal balloon of the double-lumen tube can distend the membranous part of the opposite main stem bronchus, which is paper thin. This can be injured as the subcarinal lymph node is swept off of the opposite chest's main stem. Once the entire left main stem is seen and its junction with the trachea visible, a stapler is carefully slid under the arch. The stapler is fired, the bronchus is cut, and the specimen is removed. As described on the right side, the bronchial margin should be examined on frozen section and the stump checked for a leak with a water submersion test. When taken properly, the left main stem bronchus will immediately retract well under the aortic arch. It should be difficult to see and it should be covered by the surrounding mediastinal structures. This reduced exposure is probably a factor for the lower incidence of a BPF on the left compared to the right, though we still recommend coverage with an intercostal muscle flap. To place these sutures in this short retracted stump one must retract the arch back carefully and avoid the pulmonary artery as well. The chest is closed after the placement of a chest tube using intracostal sutures to decrease pain. Although the operation is often straightforward, the postoperative course can be challenging. Keys to successful outcomes include fluid restriction, strict aspiration precautions, and aggressive pulmonary toilet and cardiophysiotherapy.

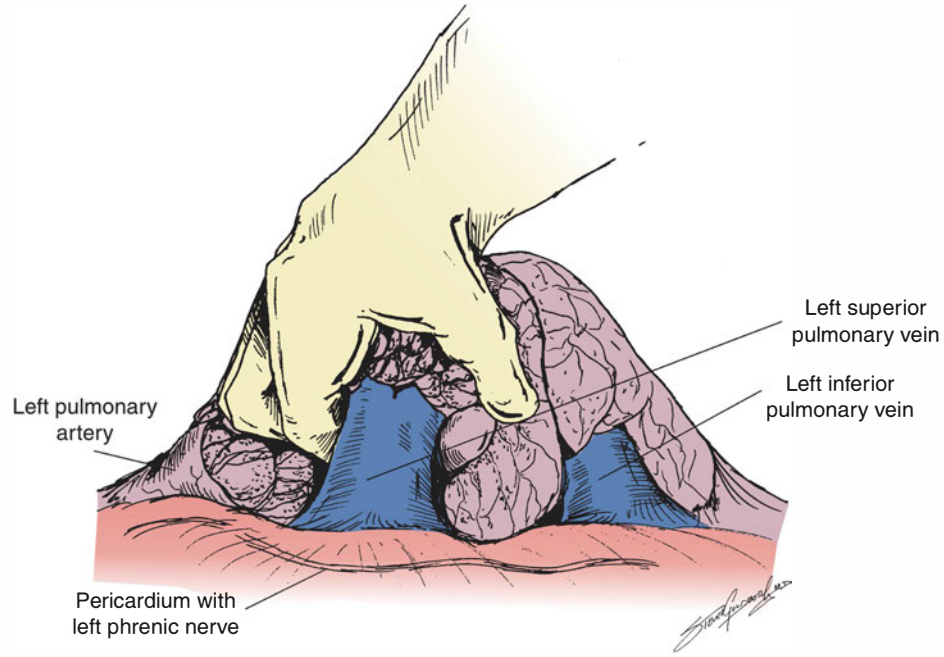


Fig. 6.6. A finger is placed around the left superior pulmonary vein and it is encircled.

MANAGEMENT OF COMMON COMPLICATIONS AFTER PNEUMONECTOMY

Vocal Cord Dysfunction

Because of the location where the left vagus nerve traverses, the aortic arch with the recurrent laryngeal "recurring" around the ligamentum arteriosum, one must be vigilant during the dissection in this area to avoid injury to the nerve. This is especially true when dissecting in the hilum for removing of the aortopulmonary lymph nodes. The right recurrent laryngeal nerve is at risk at the apex of the right chest when the mediastinal lymph node dissection extends up to the subclavian artery, around which the nerve "recurs."

The reported incidence rate of vocal cord paralysis after thoracic interventions is relatively high, ranging from 4% to 31% for postoperative recurrent laryngeal nerve paralysis after primary thoracic cancer surgery. There is also an increased morbidity, as recurrent laryngeal nerve and vagus nerve injuries are associated with increased rate of reintubation, arrhythmias, and aspiration. Risk factors for recurrent nerve injury and vocal cord dysfunction include preoperative radiotherapy, left pneumonectomy, and pericardiotomy.

Mechanism of injury varies and can be caused by heat, stretch devascularization,

or direct transection. Heat injuries usually result from using the electrocautery for hemostasis in the vicinity of the nerve. Stretching of the nerve causing neuropraxia occurs while dissecting out the hilum, especially when associated with a hilar tumor. The perineural vascular supply is delicate and can be injured easily during dissection. The anterior terminal branch of the recurrent nerve, designated as the motor branch, is located laterally, a position which makes it particularly vulnerable to surgical wound or trauma.

Symptoms of recurrent laryngeal nerve injury on either side include hoarseness, aspiration, poor cough, and dysphagia. Diagnosis is made based on symptoms. When symptoms become apparent, a laryngeal examination should be done to confirm the diagnosis and be vigilant of the consequences such as aspiration, bronchial obstruction, pneumonia, and reintubation. On examination, recurrent laryngeal nerve paralysis results in fixation of the vocal fold in a nonmedian position and causes incomplete glottal closure. Treatment varies depending on the extent of injury. Patience is usually best since most injuries are not secondary to severing of the nerve. The surgeon is usually aware if the nerve had to be cut to remove the tumor. If the nerve is still intact, the edema will usually resolve in several weeks and the voice will return to normal. If the nerve has been severed then medialization thyroplasty is indicated. If the nerve is preserved then most clinicians would recommend waiting 6 months before undertaking medialization thyroplasty. During that time functional voice and swallowing therapy is indicated. Often these patients will regain function of their recurrent laryngeal nerve with no need for further therapy.

Arrhythmias

Arrhythmias after pulmonary resection continue to be a frustrating and common problem. An incidence ranging from 5% to as high as 40% following pneumonectomy has been reported depending on how closely the patients are monitored and how it is defined. AF is by far the most common arrhythmia associated with pulmonary resection. Supraventricular tachycardia (SVT) and ventricular tachycardia (VT) are much less frequent than AF. The incidence of SVT following pneumonectomy is reported to be between 13% and 26%. Most arrhythmias occur within the first 2 to 3 days after operation and rarely later than postoperative day 5. The incidence is higher if an intrapericardial dissection has been performed.

The physiology and cause underlying these arrhythmias is poorly defined. Some studies have shown that an increase in right heart pressure and an increase in pulmonary vascular resistance predisposes a patient to clinically significant arrhythmias. Factors that have been associated with postoperative arrhythmias following pulmonary resection include hypoxia, intrapericardial dissection, pericardial manipulation, vagal irritation, pulmonary hypertension, older age, and preexisting cardiac or pulmonary disease. The prognostic significance of arrhythmias, especially AF, is difficult to determine because they are often associated with other more serious cardiac or pulmonary complications. There is no difference in mortality between patients who develop AF, not associated with other complications, who are appropriately treated following pulmonary resection than in those patients who maintain normal sinus rhythm. However postoperative arrhythmias increase the length of stay and thus the cost of the hospital stay.

The data suggests that prophylactic use of an antiarrhythmic drug perioperatively can decrease the incidence of AF in patients undergoing pneumonectomy. Historically, digoxin has been used to prevent and treat arrhythmias following pulmonary resection; however, there have been multiple randomized controlled trials demonstrating minimal to no effect. Prophylactic beta blocker administration has been shown in some studies to be effective in both prevention and treatment of postoperative AF, but its use after a pneumonectomy should be judicious. Amiodarone has been proposed for prophylaxis and is currently used by many to treat AF and SVT. A prospective trial using intravenous amiodarone following pulmonary resection was stopped due to the increased incidence of adult respiratory distress syndrome (ARDS) following pneumonectomy. There have, however, been other studies demonstrating that prophylactic use of oral amiodarone resulted in a decrease in the incidence of SVT when compared to placebo with no reported increase in ARDS. Magnesium sulfate infusion postoperatively has been shown in one randomized trial to be an effective agent in reducing the incidence of AF. The most commonly used drugs at this time for both prophylaxis and treatment of AF/SVT are calcium channel blockers. We prefer a calcium channel blocker, specifically diltiazem that has been shown in multiple studies to decrease the incidence of postoperative AF/SVT when given both orally and intravenously. In a randomized controlled trial by Amar it was shown that postoperative SVT occurred significantly less in the diltiazem-treated group versus placebo. Specifically, a standard intravenous dose is given postoperatively on the day of surgery and subsequently converted to oral dosing on postoperative day 1. When a patient has any sign of an arrhythmia, underlying causes such as hypoxia, electrolyte abnormalities, and even silent myocardial ischemia must be ruled out. Symptomatic arrhythmias must be taken seriously and cardioverted when necessary. If the arrhythmia continues for more than 48 hours, the risks and benefits of anticoagulation and/or chemical or electrical conversion should be considered.

Postpneumonectomy Pulmonary Edema

Postpneumonectomy pulmonary edema (PPE) was first described by Gibbon in 1942 but not extensively covered in the literature until almost 40 years later. Currently no consensus exists with regard to the definition of the syndrome or ways to prevent its occurrence. It most commonly presents 12 to 96 hours postoperatively and can be confused with, or occur in combination with, any of the following: congestive heart failure, pulmonary thromboemboli, or ARDS due to sepsis, pneumonia, and aspiration, all of which make PPE a diagnosis of exclusion. Most authors agree that in order to make the diagnosis there must be progressive and refractory hypoxemia, pulmonary edema on chest imaging, and widened alveolar to arterial oxygen gradient following lung resection.

The incidence of this complication ranges widely due to a lack of a consensus definition. Reported incidences of PPE range from 4% to 7% but may be as high as 12% to 15% if less severe cases are included. The reported mortality also varies but is uniformly high, ranging from 50% to 75% and even 100% in a few studies. Risk factors are variable but generally include perioperative transfusion (result of an immunologic reaction similar to transfusion-related lung injury (TRALI)), higher ventilation pressures intraoperatively, right pneumonectomy (reportedly a result of poor lymphatic drainage), perioperative fluid overload, older age, poor perioperative lung function, preexisting cardiac disease, extensive lymphadenectomy, neoadjuvant therapy, and the hyperoxia, volutrauma, and hyperinflation associated with one-lung ventilation.

Histologic changes in PPE are almost identical to those that are well described in ARDS prompting some authors to suggest that they are one and the same. Initially

the endothelial integrity is lost with resulting extravasation and hemorrhage. Type I pneumocytes undergo necrosis and platelet/fibrin microthrombi form. After about 5 days organization and repair begins with the proliferation of fibroblasts and type II pneumocytes. Also during this time squamous metaplasia of the epithelium begins, as well as hyaline membrane formation. Interstitial and alveolar fibrosis occurs at 10 days with widespread thrombotic, fibroproliferative, and obliterative changes occurring, along with extensive remodeling of the pulmonary vascular bed. As the process continues past 14 days, collagen deposition dominates the remainder of the disease process.

Often despite careful preoperative planning, intraoperative technique, and postoperative management, postpneumonectomy pulmonary edema occurs. Once it does occur, treatment is at best difficult and resource consuming. The successful use of nitric oxide has been reported. A precise mechanism, even with the knowledge of the histology, has never been clearly delineated, making prevention impossible. Treatment traditionally consists of fluid restriction, diuretics, and ventilator support. Similar to ARDS, PPE patients benefit from sedation and paralysis to improve oxygenation. Attempts should be made to keep peak airway pressures as low as possible using pressure-controlled ventilation and permissive hypercapnia. Positive end-expiratory pressure should also be used to keep the inspired oxygen concentration as low as possible.

We have described above the use of Solu-Medrol prior to pulmonary artery clamping. This assumes the steroid bolus helps reduce the ongoing and subsequent inflammatory cascade caused by the manipulation of the lung and sudden shift in pulmonary blood flow. Although these data are interesting there have been no large prospective randomized trials to date that either prove or disprove the utility of this intervention or elucidate potential complications.

Chylothorax

Chylothorax fortunately is a very uncommon complication following pneumonectomy. It should be completely avoidable with careful operative planning and intraoperative techniques. It is an extremely difficult diagnosis to make after a pneumonectomy since one expects the pneumonectomized space to fill with fluid and one is reluctant to perform a thoracentesis for fear of infecting that space. The diagnosis should be considered when a patient returns with a significant mediastinal shift toward the nonoperated side instead of toward the pneumonectomized side. A thoracentesis yields chylous (milky) fluid and the diagnosis is confirmed by the presence of chylomicrons and a triglyceride content of 110 mg/dl or greater, a lymphocyte count more than 90% of the total white blood cell count, and a total protein concentration approaching that of plasma. There are reports suggesting that a history of cirrhosis does increase the risk secondary to the increased rate of flow in the lymphatics of the liver and the thoracic duct. Other risk factors have been identified including gross N2 disease, radical mediastinal lymphadenectomy, and pleural flap elevation along the thoracic aorta.

Chylothorax is often caused by leaking lymphatic ducts from dissection of malignant mediastinal lymph nodes. However, even with complete thoracic lymphadenectomy the incidence of chylothorax is very low. As opposed to esophageal resections, chylothorax following pulmonary resections, including pneumonectomy, is mainly due to injury to small branches of the thoracic duct and is rarely due to injury to the main duct itself. Those patients with N2 disease, especially with extracapsular N2 disease, carry an increased risk of developing a chylothorax, likely due to the obstruction of lymphatic channels by metastatic disease. The mortality rate in the literature after iatrogenic/traumatic chylothoraces ranges from 4.5% to 10%.

Chylothorax following pulmonary resection occurs more often on the right side. This is explained both pathologically and anatomically. Bronchogenic carcinomas occur more often on the right side (ratio of 6 to 4) thus leading to more frequent mediastinal lymph node dissections on the right. Anatomically, the lymphatic drainage of the lungs predominates in the right chest for two reasons: (1) the right lung is larger; (2) the lymph flow to the right paratracheal nodes from the left lung is greater than the lymph flow from the right lung to the left paratracheal nodes.

Initial management of iatrogenic chylothorax following pulmonary resection is predominantly conservative, using medium-chain triglyceride diets or occasionally total parenteral nutrition, as well as drainage via tube thoracostomy. There are a few nonsurgical treatments that have been reported for chylothorax. Lymphangiography can be used both for diagnostic and anatomic detail. There are some studies that report spontaneous closure of the defect following the procedure due to the beneficial effects of the iodinated contrast on the laceration (if small). Percutaneous catheterization of the thoracic duct with embolization of the defect has also been reported. Pleuroperitoneal shunting has been used permitting reabsorption of the chyle by the peritoneum; however, this procedure makes documenting closure of the defect difficult and is mainly reserved for postoperative pediatric patients and cases of nontraumatic malignant chylothoraces.

Surgical management is reserved for those patients that fail conservative measures. Prolonged drainage of a chylothorax leads to neutropenia and carries significant morbidity. Reexploration via thoracotomy and sutured ligation of the main duct with fibrin glue placement is best. The main duct can also be ligated via a laparotomy or laparoscopic approach. No matter what treatment is used, the thoracic surgeon must be aware of the potential for severe protein malnutrition, dehydration, and immune deficiency due to large losses of lymphatic constituents, as the daily volume of the thoracic duct can approach 3 L. Prompt treatment is necessary after diagnosis and aggressive fluid and nutritional monitoring must be instituted. If conservative measures do not stop the leak within 5 to 7 days and if the output remains high (>800cc/day), surgical ligation of the duct is best.

Bronchopleural Fistula and Empyema

Postpneumonectomy BPF is defined as a communication between the main stem bronchial stump and the ipsilateral pleural space. It is a life-threatening problem because infected material can be aspirated into the only remaining lung. It is by definition associated with a postpneumonectomy empyema defined as the presence of purulent material in the postpneumonectomy space and is usually but not always associated with a BPF. When it occurs within the first 6 months after a pneumonectomy there commonly is a BPF associated with it. Early recognition and aggressive treatment with reoperation are the keys to survival.

The overall incidence of BPF and empyema is greater following pneumonectomy than after lesser resections. Following pneumonectomy, for any reason, the incidence of BPF/empyema ranges from 2% to 16%. When stratified by disease state, the incidence for empyema and BPF following pneumonectomy for primary lung cancer are 5.8% and 4.1%, for metastatic disease 3.1% and<1%, and for benign disease 24% and 9.9%, respectively. When it occurs, BPF

following pneumonectomy is associated with a mortality rate that ranges from 30% to 50%. Also noted is an increased incidence of BPF following pneumonectomy if operation has been preceded by neoadjuvant treatment. There is also an increase in both cardiac and pulmonary complications and a concomitant increased hospital stay and associated costs with the occurrence of a BPF. Multiple studies have identified both local and systemic factors that are associated with the development of empyemas and/or BPF following pulmonary resections. Local risk factors include the following:

1. Presence of carcinoma at the bronchial margin
2. Long bronchial stump
3. Disrupted bronchial blood supply
4. Technique of stump closure
5. Preexisting empyema
6. Extended resections
7. Preoperative radiation
8. Postoperative need for mechanical ventilation
9. Right versus left pneumonectomy
10. Right pneumonectomy following neoadjuvant therapy

Systemic factors include the following:

1. Poor nutritional status
2. Diabetes
3. Sepsis
4. Preoperative chemotherapy
5. Underlying lung disease (including chronic infection and COPD predicted by decreased ppoFEV$_1$ and ppoDLCO)
6. Preoperative immunosuppression (steroid therapy)
7. Older age (>70 years)
8. Postoperative sputum positive for AFB

Prevention of postpneumonectomy BPF and empyema centers on attention to careful intraoperative technique. One must limit the length of the bronchial stump, which will help to prevent pooling of secretions leading to increased risk of infection at the site of bronchial closure. Perioperative prophylactic antibiotics have a well-known benefit following general thoracic procedures. For patients with preexisting infection/empyemas or the potential for significant pleural space contamination (destroyed lung or preoperative BPF) specific culture-directed antibiotic regimens may be indicated. There have been reports of intraoperative factors such as blood transfusion that are an independent risk factor for postpneumonectomy respiratory complications. Method of closure of the bronchial stump remains controversial in the current literature. There are a variety of studies that report the superiority of both handsewn and stapled closures. No matter what closure is chosen most authors suggest bronchial stump reinforcement following pneumonectomy, especially in cases where breakdown is more likely (neoadjuvant therapy, presence of infection, and right-sided procedures) is mandatory. As described above, we prefer an intercostal muscle flap. Other options are a pericardial fat pad, defunctionalized azygous vein flap, or a pleural flap.

In cases where postpneumonectomy BPF or empyema is suspected, early diagnosis is critical because the earlier it is diagnosed and treated the better the prognosis. Early empyemas without BPF, though uncommon, are best managed by debridement of the space. When the space has been sterilized, it should then be filled with antibiotic solution (a Clagett procedure) and the tubes removed. Although management of postoperative BPF remains a problem, immediate drainage of the pleural space can be a lifesaving procedure because it may prevent aspiration of accumulated fluid into the contralateral lung. A variety of treatment strategies have been proposed for subsequent management. For small fistulas (<3 mm in size) with a short stump occasionally tube drainage alone is sufficient along with the injection of fibrin glue into the fistula. Sometimes these small fistulas close spontaneously, but that is very rare. In those cases where tube drainage does not result in closure of the small BPF, some authors advocate the use of videoscopic drainage and debridement combined with bronchoscopic closure using cauterization or metallic coils with the application of fibrin glue. For larger BPFs attempts at endoscopic closure are not recommended. Definitive surgery to close a postpneumonectomy BPF is quite involved and thus the patient's medical and nutritional status must be optimized, the postpneumonectomy space must be clean and healthy, the need for postoperative mechanical ventilation minimized, and there must be no evidence of recurrent carcinoma. A variety of procedures have been used for bronchial stump closure following development of a large (3 mm or greater) BPF. The traditional, well-described procedure initially developed by Pairolero involved multiple debridements, the use of muscle flaps or omentum to plug the fistula, and finally closure via a Clagett. Others have suggested a transsternal, transpericardial approach to the bronchial stump via the posterior pericardium and subsequent mobilization of the affected stump, reamputation, and a reinforced closure.

Postpneumonectomy Syndrome

Postpneumonectomy syndrome is an unusual complication occurring most often after right pneumonectomy. Resulting from an extreme shift and rotation of the mediastinum and contralateral lung into the empty pleural space, it produces symptomatic proximal airway obstruction and causes air trapping. The main stem bronchus is stretched and compressed against the vertebral bodies, descending aorta, and/or remaining pulmonary artery branches. Patients present months to years following pneumonectomy with dyspnea, stridor, and recurrent pneumonias. Chest radiographs and CT show an extreme shift of the mediastinal structures and bronchoscopy reveals severe proximal airway extrinsic obstruction and sometimes bronchomalacia. Other causes must be ruled out including recurrent malignancy, pulmonary hypertension, progression of underlying lung disease, pulmonary thromboembolism, and congestive heart failure.

Although it is impossible to predict, the syndrome is described mostly in children and young adults; however, there are occasional reports in the adult literature as well. Most surgeons attempt to prevent the acute shift of the mediastinum by avoidance of chest tubes following pneumonectomy, or if they are used, tubes are either clamped or left to water-seal drainage only. To correct the shift when it does occur, the mediastinal structures must be shifted back to a more anatomic location. This can be done via anterior pericardiorrhaphy, which anchors the pericardium to the parasternal chest wall and by placing expandable saline-filled breast implants into the postpneumonectomy space, which is usually spared of adhesions, stabilizing the mediastinum. However, these techniques have fallen out of favor for fear of infecting the pneumonectomy space. Tracheomalacia after long-standing obstruction is an indicator of poor outcome. Treatment is often the placement of silicone removable stent that helps keep the airway open.

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EDITOR'S COMMENTS

The authors have provided a very complete review of pneumonectomy, a procedure that many say results in a disease in and of itself since it leaves one with a single lung that is the recipient of the entire cardiac output. If patients make it through the postoperative period without a significant complication, many will lead a reasonably normal life though with some limitation. Despite the fact that individuals can live a normal life with one lung, it remains our view that every effort possible should be made to conserve pulmonary parenchyma even if that means performing a sleeve resection of the main stem bronchus and resecting and reconstructing a portion of the pulmonary artery. Rarely if ever do I enter the operating room knowing that the operation is going to result in pneumonectomy, I am always thinking of what can be done to preserve pulmonary parenchyma. But there are situations where pneumonectomy is the only choice if a complete resection is to be accomplished. If the main pulmonary artery is involved so far proximal that it is not possible to place a clamp for control and leave enough cuff for a reconstruction, then pneumonectomy is the only choice. If the bronchus is involved by tumor at the bifurcation on the left and sleeve resection is not feasible, then the entire lung must be removed. On the right side there are more options in terms of bronchial reconstruction and I have sleeved the main stem bronchus and reimplanted only the basal segments.

Several points made by the authors raise some questions and areas of disagreement. The authors state that mediastinal lymph node involvement is an absolute contraindication for pneumonectomy while many of us would argue that that is not the case especially if there is a response to neoadjuvant therapy. That said, the incidence of complications in patients undergoing pneumonectomy who have received neoadjuvant therapy

(*continued*)

EDITOR'S COMMENTS (*continued*)

clearly is higher than in those who have not received any preoperative therapy. It certainly is our belief that patients who are candidates should be offered the therapy that offers the best opportunity at long-term survival and currently that is complete resection even in the face of N2 disease. Often the wisdom as to whether or not a given therapeutic intervention should have been carried out must be determined in retrospect. Subjecting a patient to a pneumonectomy that, even in the most experienced hands is associated with a mortality in the range of 5% to 10%, not to mention the potential for postoperative morbidity, may be said to have been ill-advised if the patient lives for less than 1 year following the procedure. Yet, currently there is no way to predict which patients will live for less than 1 year so that operation in that group could be avoided. Thus if a patient with documented N2 disease is otherwise a reasonable candidate for operation they should at least be considered for an aggressive treatment regimen that likely will include resection.

If a patient is being considered for pneumonectomy, it is probably reasonable to carry out a complete extent of disease evaluation to include brain imaging as well as combined CT/PET imaging. It is a travesty to subject a patient to the risks inherent in pneumonectomy only to find evidence of disseminated disease early in the postoperative period that could have been identified prior to operation saving the patient from an invasive procedure from which he/she will derive no survival benefit.

The authors have recommended the use of Solu-Medrol prior to dividing the pulmonary artery based on the thought that this steroid might prevent PPE. Currently no evidence exists to support this contention though admittedly it would be difficult, if not impossible, to mount a study to prove efficacy since the incidence of PPE, fortunately, is very low.

Despite the author's recommendation to use distilled water, as opposed to normal saline, to irrigate the chest there exists no evidence that such a regimen results in any tumor cell lysis. If one wants to irrigate the pneumonectomy space prior to closure normal saline works just fine and is certainly more physiologic.

As the authors point out there are a number of ways to manage the pneumonectomy space in the immediate postoperative period. We prefer to place a chest tube in the space connected to a balanced drainage system (Pneumonectomy PleurEvac) that allows for the mediastinum to be "set" as well as monitors the amount of bleeding. The tube is removed on the morning of the first postoperative day. Connecting a tube that is within a pneumonectomy space to a standard drainage system that could inadvertently be connected to suction not only is dangerous but also could prove deadly. Simply placing a sign warning to not connect to suction is not enough precaution as far as I'm concerned. Aspirating the space with syringe and needle following chest closure also is acceptable as a maneuver to "set" the mediastinum but does not offer the opportunity to monitor postoperative drainage.

The authors discuss a number of other technical issues that bear some comment. The bronchial stump should be left as short as possible to promote healing that is not likely influenced adversely by "secretions collecting" but more by compromised vascularity if the stump is left too long. The incidence of recurrent laryngeal nerve injury cited by the authors seems high. Injury to the nerve should be uncommon since its anatomic location is well known. A patient presenting with paralysis of the left vocal cord caused by involvement of the recurrent nerve in the aortopulmonary window usually proves to have unresectable disease. Occasionally, tumor involves the nerve in a location where it can be resected thus sacrificing the nerve. Many patients have no difficulty with a paralyzed vocal cord even early following the injury but the major issue remains aspiration and inability to mount a productive cough to clear secretions. Vocal cord injection for augmentation is a reasonable temporizing procedure but there should be at least 6 months of observation before attempting a cord medialization procedure since most patients accommodate very well to unilateral vocal cord paralysis. Contrary to the author's contention, rarely does nerve function return but the contralateral vocal cord allows for glottic closure by extending across the midline to effect cord apposition.

Postpneumonectomy empyema with or without bronchopleural fistula is a devastating complication though patients can be managed. An infected space without a fistula may only present with the patient losing weight and failing to thrive. There should be a low threshold for interrogating the space with thoracentesis to assess whether infection is present. An air fluid level associated with coughing up watery sputum should also prompt thoracentesis and drainage of the space initially with a chest tube. Definitive management requires an open window thoracostomy placed in a dependent location. Usually, adequate drainage allows for the bronchial fistula to close, but it may take months. Once the space is clean, transposition of a muscle flap and closure may be carried out. In our experience the Clagett procedure, where the space is filled with antibiotic solution and closed, works only rarely but in certain situations may be attempted. Transsternal division and closure of the bronchus should be reserved for those cases with a very large leak that is unlikely to close on its own.

LRK

7 Bronchoplastic Procedures

Anna Maria Ciccone, Federico Venuta, Camilla Vanni, and Erino A. Rendina

HISTORY

The first sleeve lobectomy was performed by Price-Thomas in 1947 for an endobronchial adenoma. Five years later, Allison reported a sleeve lobectomy for carcinoma. It was Paulson and Shaw, however, who popularized sleeve resection with their 1955 paper, entitled "Bronchial anastomosis and bronchoplastic procedures in the interest of preservation of lung tissue." The use of their techniques allows the modern thoracic surgeon to perform parenchymal-sparing procedures, even in association with reconstruction of the PA (described for the first time by Allison and Thomas in the 1950s), on patients who would not tolerate a pneumonectomy. In the case of malignancy, sleeve resections may be performed without compromise of oncologic principles.

PRINCIPLES AND JUSTIFICATION

Pneumonectomy is associated with an increased morbidity and mortality when compared with lobectomy and sleeve lobectomy. Thus, in our practice, we make every effort to avoid pneumonectomy. This includes complex bronchoplastic and bronchovascular reconstructions when required. The justification of this approach is simply that by avoiding pneumonectomy we avoid its attendant risks while providing an equivalent cancer operation. In addition, lung-sparing procedures allow us to offer curative operations to patients with poor pulmonary function who would not otherwise tolerate the removal of an entire lung.

PREOPERATIVE ASSESSMENT AND PREPARATION

Our preoperative evaluation includes a complete history and physical examination. Special attention is focused on previous thoracic procedures and chest irradiation. The use of high-dose steroids or systemic illnesses that might interfere with bronchial anastomotic healing is noted. All patients have a chest X-ray, a chest computed axial tomography (CAT) scan, and pulmonary function testing with diffusion capacity. Patients with a diagnosis or suspicion of malignancy also have an extent-of-disease workup, which includes a bone scan and magnetic resonance imaging (MRI) of the brain when indicated.

We perform mediastinoscopy selectively in patients with malignant disease. Patients who have mediastinal adenopathy of >1.0 cm on CAT scan undergo mediastinoscopy before thoracotomy. If the mediastinoscopy is negative, we proceed with the thoracotomy. If the mediastinoscopy reveals ipsilateral N2 disease, patients are referred for preoperative chemo- or chemoradiation therapy and return later for resection. Those patients with contralateral N3 disease are referred for chemoradiation therapy and are not offered surgical resection.

ANESTHESIA

After induction of general anesthesia, all patients undergoing sleeve resection require bronchoscopy by the operating surgeon. This can be done with either a rigid or a flexible bronchoscope. Bronchoscopy allows visualization of the lesion and planning of the resection. After bronchoscopy, it is important for the surgeon to have a complete discussion with the anesthesiologist regarding the operative plan. If a right-sided sleeve resection is contemplated, a left endobronchial double-lumen tube should be placed (Fig. 7.1). If a left-sided sleeve resection is contemplated, a right endobronchial tube is placed. For sleeve pneumonectomy or a carinal sleeve resection, a sterile anesthesia circuit is required to allow direct ventilation from the surgical field.

OPERATIONS

We have performed sleeve resections through standard posterolateral incisions, serratus-sparing posterolateral incisions, and lateral incision, all of which are satisfactory for exposure and dissection.

After entry into the chest, complete exploration is carried out to rule out metastatic disease to either the pleura or lung parenchyma and to assess resectability. On both the right and left side, we begin our dissection in the anterior hilum and completely dissect out the main pulmonary artery (PA). Special care must be taken on the left side to avoid damage to the short left main PA and specifically the apical segmental arterial branch. If there is bulky disease or any difficulty is encountered with dissection, we do not hesitate to open the pericardium on either side to obtain proximal control. Next, we encircle the main PA with an umbilical tape to assure proximal control. The remaining steps are specific to the sleeve resection being performed and each will be described independently in what follows.

Right-Sided Resections

Right Upper Lobectomy Sleeve Resection: The Prototype Bronchoplastic Procedure

After proximal arterial control has been obtained, we continue our dissection superiorly and enter the plane of the right upper lobe bronchus (Fig. 7.2). The lung is retracted anteriorly, and we continue our dissection in the bifurcation between the right upper lobe bronchus and the bronchus intermedius. A "crotch" lymph node is a consistent finding in this location. This node is elevated away from the bifurcation to reveal the PA branch to the superior segment of the right lower lobe. Once this branch is identified, the posterior portion of the fissure is completed with a linear stapler. This approach avoids extensive parenchymal dissection in the fissure. The bronchus intermedius is encircled just distal to the right upper lobe take-off, and an umbilical tape is placed to aid in dividing the airway at the appropriate time. Up to this point, we have not made

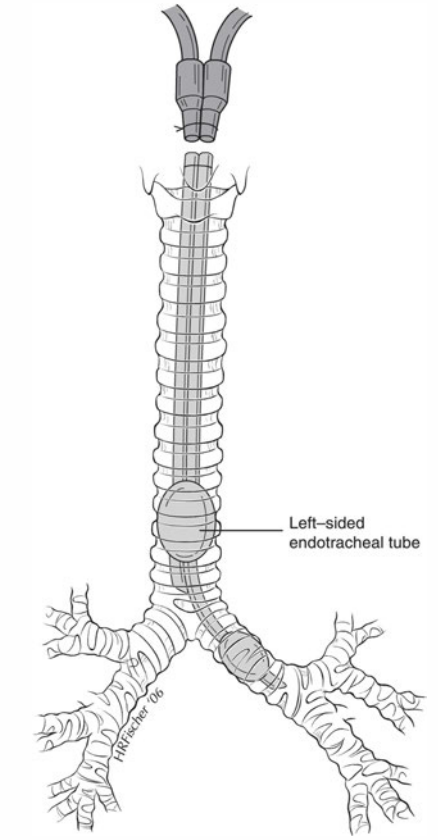


Fig. 7.1. Left-sided endotracheal tube in place for right upper lobe sleeve.

any irreversible maneuvers. A complete inspection is carried out to ensure that all diseases including nodal disease can be removed. Once complete resectability is confirmed, we begin by ligating and dividing the PA branches to the right upper lobe. Similarly, the venous drainage is divided with a vascular stapler taking care to preserve the middle lobe venous drainage. The minor fissure is completed with a linear stapler. The main stem bronchus is encircled at its origin, and an umbilical tape is placed. Before committing to the sleeve resection, it is often productive to divide the right upper lobe bronchus at its origin to see whether the tumor can be cleared with a negative bronchial resection margin. This is especially important with carcinoid tumors, in which the endobronchial component may be attached only at one point and the lesion simply pulled out leaving a clear bronchial margin. Once the bronchus has been opened, the decision to proceed with sleeve resection may be made based on the findings either grossly or microscopically.

To begin the sleeve resection, we divide the main stem bronchus with a No. 15 blade just proximal to the right upper lobe take-off. Similarly, the bronchus intermedius is divided just distal to the right upper lobe take-off (Fig. 7.3). For upper lobe sleeve resections, these cuts must be perpendicular to the long axis of the airway and placed between cartilaginous rings so as to result in a clean cut. An angled division of the bronchus is to be avoided. The proximal and distal airway margins are cut from the specimen and the true margin inked by the surgeon. We personally take the margins to the pathologist so that proper orientation can be demonstrated prior to frozen-section examination. Once we have documentation that the resection margins are free of tumor, the reconstruction is begun. Microscopic tumor present at a bronchial margin requires additional resection of the involved area or possibly pneumonectomy.

We perform the bronchial sleeve anastomosis in an interrupted manner. The key to a successful bronchial sleeve anastomosis is a pneumostatic well-approximated, tension-free repair that accounts for any size discrepancy between ends by precise suture placement. We do not take a "tuck" in the proximal airway to make up for a size discrepancy. Rather, we make up the size discrepancy along the entire circumference of the anastomosis by precise suture placement.

When performing an interrupted anastomosis, we use 4-0 oiled, absorbable, braided suture. The first suture is placed in an "outside-to-in" fashion at the junction of the cartilaginous and membranous bronchi. The suture is not tied but is secured to a suture guide. Additional sutures are placed at 2-mm intervals to complete the first half of the cartilaginous anastomosis (Fig. 7.4). Once the midpoint of the cartilaginous bronchus is reached, we begin tying the sutures starting at the corner. The surgeon "crosses" the next suture in the series to relieve tension while the assistant ties. The cartilaginous anastomosis is completed from the midpoint down to the opposite corner in a similar manner. The lung is retracted anteriorly to reveal the membranous portion of the bronchus. The membranous portion of the anastomosis is completed with interrupted sutures. The chest is filled with saline, and the anastomosis is tested to 20-cm water inflation pressure. Needle hole air leaks are ignored; however, air leaks between the cut edges of the bronchus, if small, are reinforced with simple interrupted sutures. A large area of leaking may require the entire anastomosis to be redone.

We wrap all anastomoses with intercostal muscle (Fig. 7.5), but occasionally we use an omental or pericardial fat

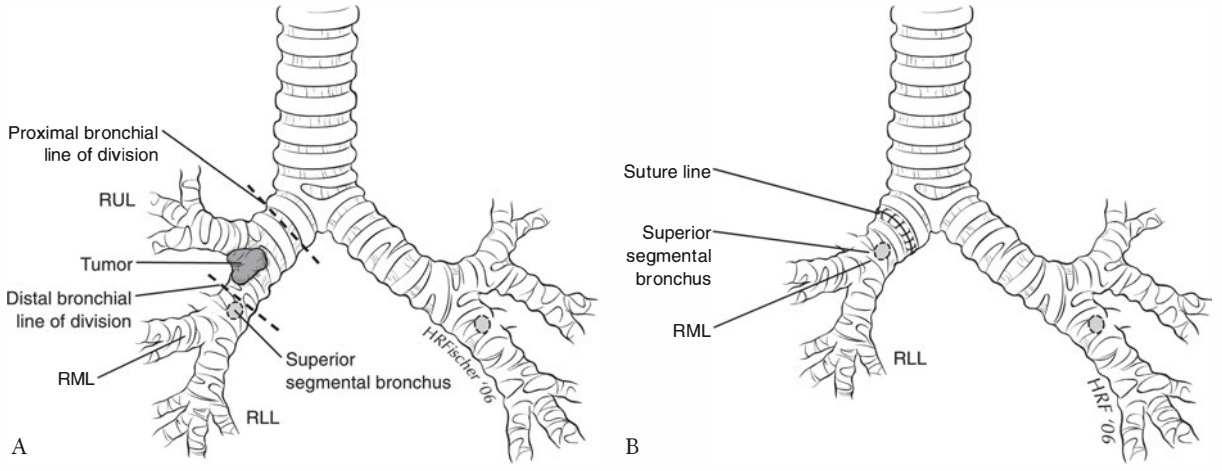


Fig.7.2. (**A**) Tumor in the right upper lobe bronchus requiring sleeve resection. (**B**) Completed bronchial resection and anastomosis. (RLL, right lower lobe; RML, right middle lobe; RUL, right upper lobe.)

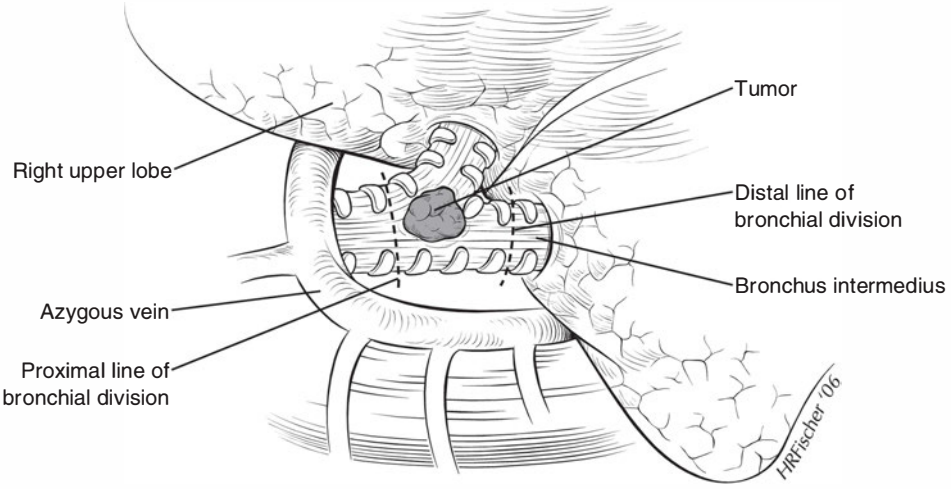


Fig. 7.3. Division of the right main stem and bronchus intermedius during right upper lobe sleeve.

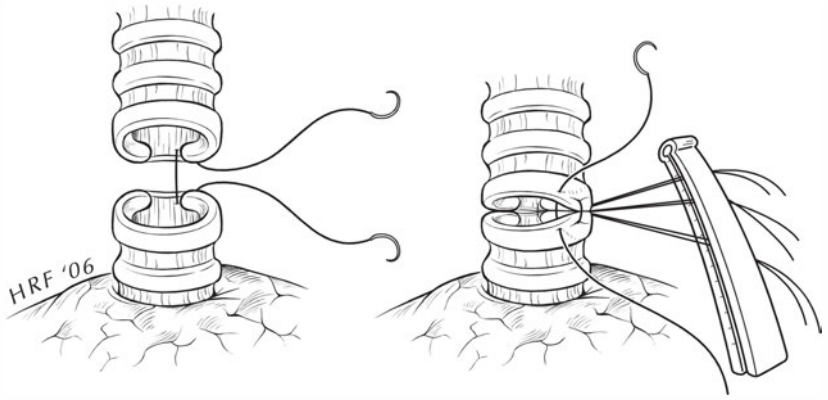


Fig. 7.4. Interrupted anastomotic technique.

flap. Although ossification of the intercostal muscle flap can occur, it does not necessarily cause problems; in fact, the bronchus has very limited intrinsic motility and a stable caliber. If the intercostal muscle flap is loosely applied around the bronchus, even if some retraction occurs, there is no reason why the hardening caused by ossification should produce stenosis. When the sleeve resection is planned preoperatively, the intercostal pedicle flap may be prepared before the insertion of the rib retractor to avoid crushing the intercostal vascular bundle, and is prepared at full thickness, encompassing a wide pleural flap. The flap is slid backward around the bronchial anastomosis, between it and the PA. The flap is then turned until its pleural side is in contact with the bronchial anastomosis, and the pleura is secured to the bronchus by interrupted sutures.

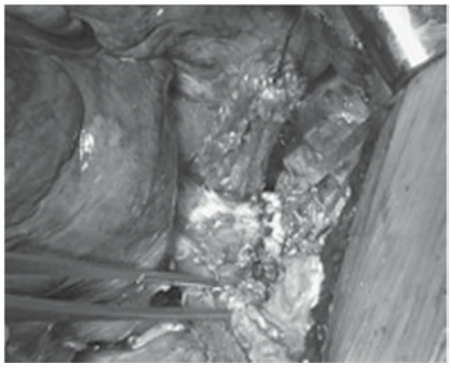


Fig. 7.5. Pericardial fat wrapped around the anastomosis.

Middle Lobe Sleeve Resection

The middle lobe sleeve resection is an infrequently performed resection. After proximal arterial control, the middle lobe vein is identified, isolated, and divided (Fig. 7.6). The bronchus to the middle lobe lies immediately posterior to the middle lobe vein. The bronchus is followed back to its origin. A right-angled clamp is placed around the bronchus intermedius, and it is divided at a location proximal to the middle lobe orifice. The division is slightly angled. The distal division is also angled to preserve the orifice to the superior segment of the lower lobe. The PA lies directly posterior and slightly superior to the bronchus, and care must be taken to avoid injury when dividing the bronchus. After division of the airway, the middle lobe arterial branch is easily visualized. The branch is ligated and divided. Next, the minor fissure and anterior portion of the major fissure are completed with firings of a linear stapler.

After confirmation of negative margins, the airway anastomosis is performed in an interrupted manner as described previously for the right upper lobe sleeve resection. In performing a middle lobe sleeve resection, special consideration must be given to the superior segmental orifice of the lower lobe. One must avoid narrowing the orifice to the superior segmental bronchus when creating an anastomosis. An intercostal muscle flap is used to wrap the anastomosis to separate it from the PA.

Bilobectomy Sleeve Resection

Bilobectomy sleeve resection is performed for an endobronchial lesion in the bronchus intermedius that extends proximally toward the upper lobe orifice (Fig. 7.7). The basic principles of proximal arterial control, microscopic negative margins, and a precise tension-free anastomosis apply. Here the right main stem bronchus is divided just proximal to the right upper lobe take-off and the right upper lobe bronchus is divided at its origin. The right upper lobe bronchus is then anastomosed to the main stem bronchus after removal of the middle and lower lobes, the so-called "Y" sleeve. Due to the reorientation of the upper lobe bronchus after removal of the middle and lower lobes, special care must be taken to avoid torsion of the bronchus at the level of the anastomosis.

Left-Sided Resections

Left Upper Lobe Sleeve Resection

Proximal arterial control is obtained with care to avoid injury to the short apical-posterior segmental branch of the left PA. We continue our dissection along the plane of the artery and identify the superior segmental branch to the lower lobe (Fig. 7.8). At this point, we complete the posterior fissure with a linear stapler. The

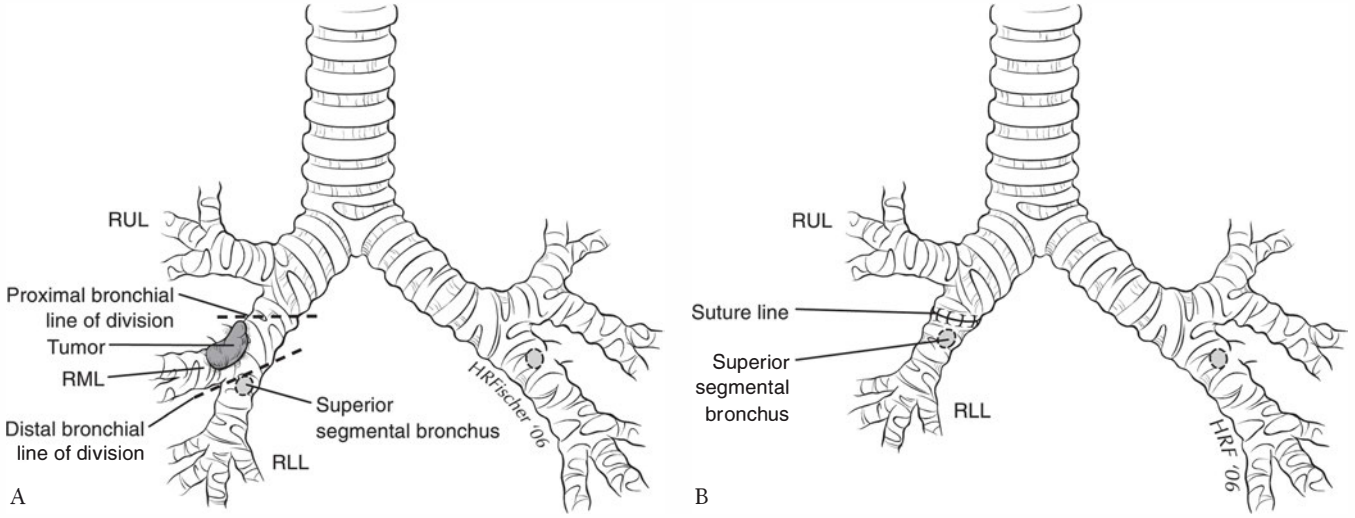


Fig. 7.6. Middle lobo sleeve. (**A**) Note that the bronchus transection is not perpendicular to the axis of the airway in order to preserve the superior segmental orifice to the lower lobe. (**B**) Completed bronchial resection and anastomosis. (RLL, right lower lobe; RML, right middle lobe; RUL, right upper lobe.)

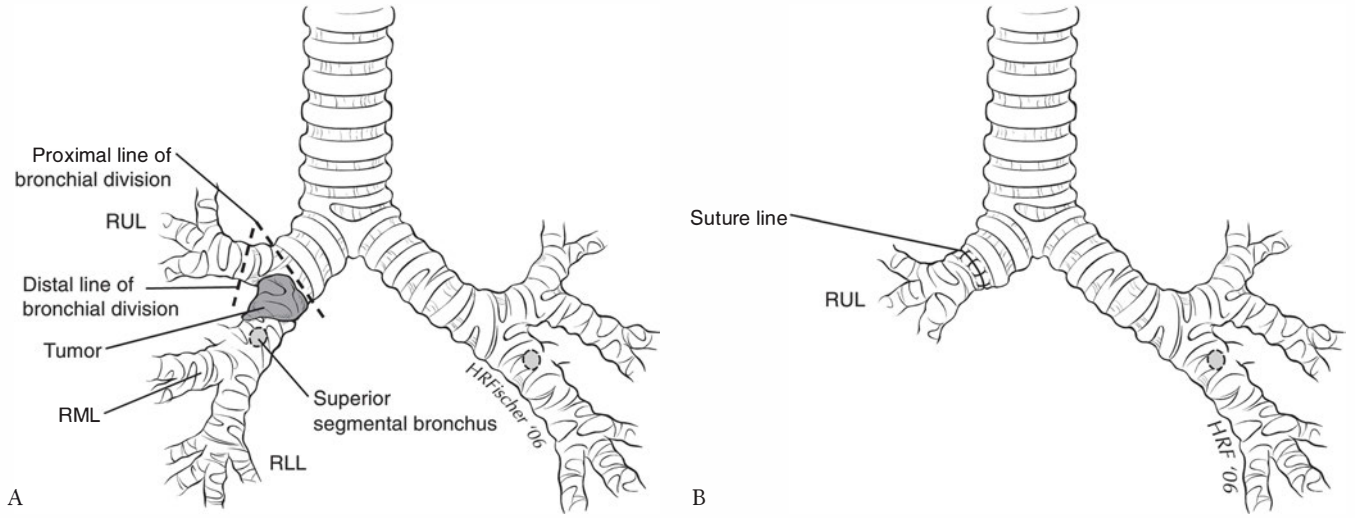


Fig.7.7. Bilobectomy sleeve. (**A**) Tumor in the bronchus intermedius that extends towards the upper lobe orifice. (**B**) Completed bronchial resection and anastomosis. (RLL, right lower lobe; RML, right middle lobe; RUL, right upper lobe.)

anterior segmental artery is ligated and divided. The lingular branches of the PA are identified, ligated, and divided. The lung is retracted posteriorly, and the upper lobe venous drainage is divided with a vascular stapler. The anterior portion of the fissure is completed with a linear stapler. The only remaining attachment to the specimen is the bronchus. The main stem bronchus is encircled proximal to the bifurcation, and an umbilical tape placed to be used as a "handle." Two 2-0 silk stay sutures are placed in the proximal left main stem and used for retraction. The main stem bronchus is divided proximal to the bifurcation, and the left lower lobe bronchus is divided at its origin. The origin of the superior segmental bronchus can be quite close to the origin of the lower lobe bronchus, and the lobar division must leave the bronchus intact without separating the superior segmental bronchus. It is possible to take the superior segment, if necessary, to achieve a complete resection and reimplant the basal segments on the main stem bronchus. The points of division of the bronchus should be perpendicular to the longitudinal axis of the airway and placed between cartilaginous rings. The margins are inked, and a frozen-section examination is performed. Once the margins are confirmed to be microscopically negative, the reconstruction is begun. We use an interrupted technique as described previously for the right upper lobe sleeve resection.

Because the left main stem bronchus is long, extensive proximal resections can be performed. This can create a technically challenging anastomosis because proximal exposure is obscured by the aortic arch. If required, the arch can be mobilized and carefully retracted to provide additional exposure. Even if a significant portion of proximal airway is removed, tension on the anastomosis is not a problem. The inferior pulmonary ligament should be released. Precise suture placement to account for

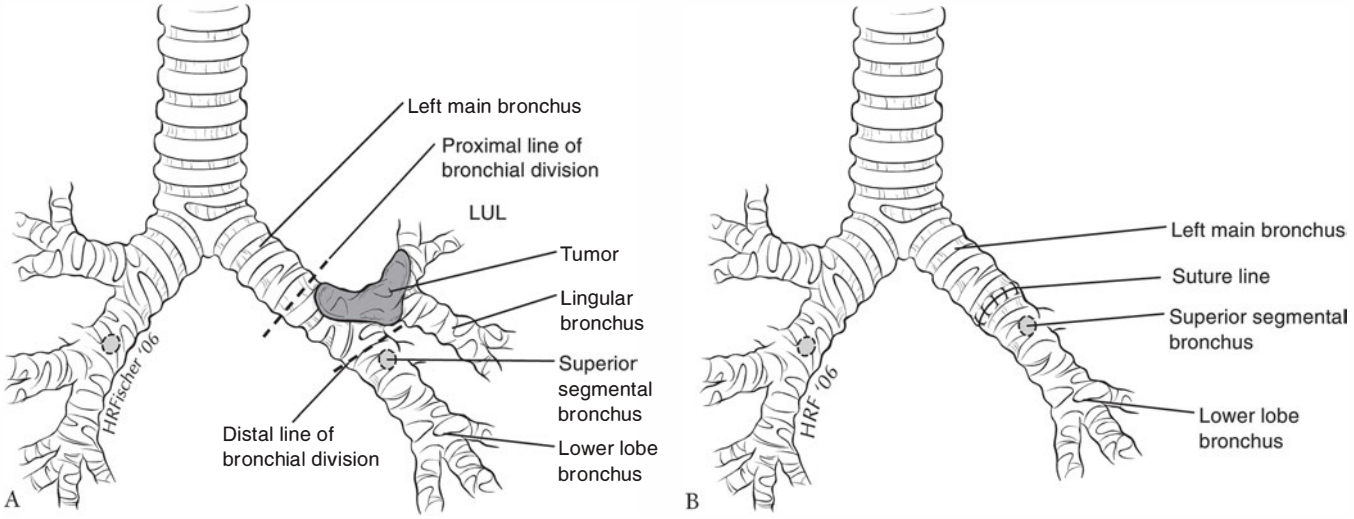


Fig. 7.8. Left upper lobe sleeve. (**A**) The tumor arises the origin of the left upper lobe. (**B**) Completed bronchial resection and anastomosis. (LUL, Left upper lobe.)

size discrepancy between the lobar bronchus and the main stem bronchus is particularly important.

Left Lower Lobectomy Sleeve Resection ("Y" Sleeve)

For lesions involving the left lower lobe orifice, with extension into the main stem bronchus but sparing the upper lobe orifice, a lower lobectomy with sleeve resection of the left upper lobe bronchus can be performed. The arterial and venous branches to the lower lobe are divided and the fissures are completed (Fig. 7.9). We dissect out and pass an umbilical tape around the left main stem and left upper lobe bronchus. Two silk stay sutures are placed in the main stem bronchus and used for retraction. The left upper lobe bronchus is divided at its origin. It is important for the division to be perpendicular to the long axis of the airway. Next, the main stem bronchus is divided proximal to the bifurcation and well proximal to the extent of the tumor. The specimen is removed from the field, and frozen-section examination of the airway margins is performed. Once the margins are confirmed to be microscopically negative, the reconstruction is completed.

The anastomosis is performed in an interrupted, as described previously for the right upper lobe sleeve resection, or in a running fashion. The lingular bronchus may arise quite proximally, and care must be taken when dividing the upper lobe bronchus to assure that the lingular bronchus remains intact. There is often a large size discrepancy between the upper lobe bronchus and the main stem. To account for the size discrepancy, precise placement of sutures is mandatory and is far preferable to taking a "tuck" in the main stem bronchus.

POSTOPERATIVE CARE

At the completion of a sleeve resection, the patient is extubated in the operating room. Postoperative pain relief is facilitated by a functioning thoracic epidural catheter. Pain control is re-evaluated periodically by the nursing staff and adjusted by the anesthesia pain service. The epidural catheter remains

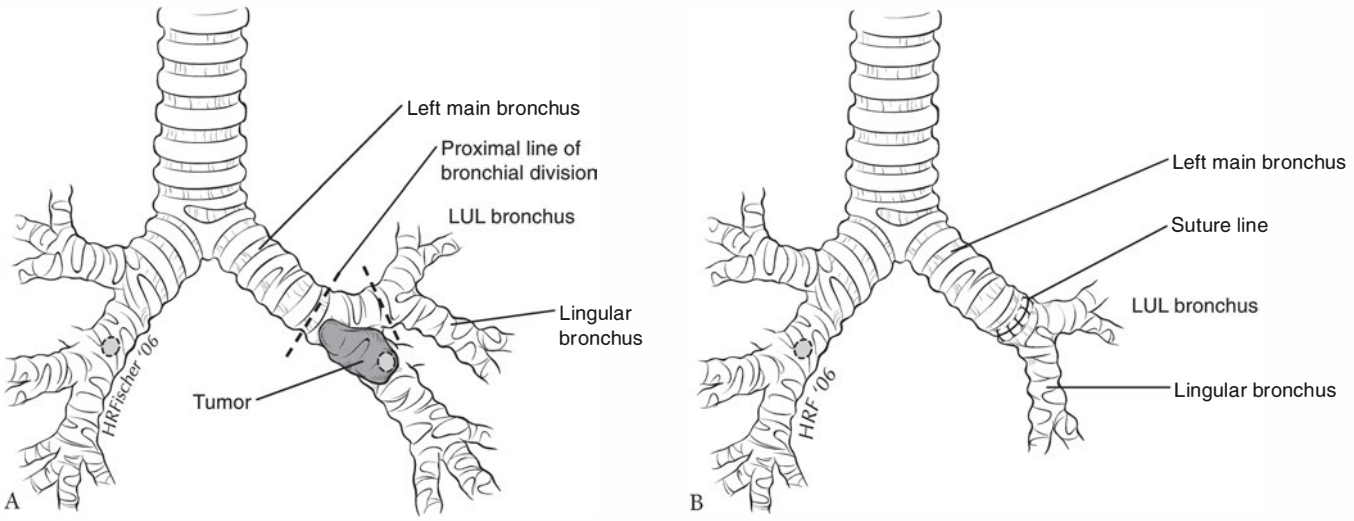


Fig. 7.9. Left lower lobe sleeve ("Y" sleeve). (**A**) The lesion involves the left lower lobe origin, with extension into the main stem bronchus. (**B**) Lower lobectomy with sleeve resection and anastomosis completed. (LUL, Left upper lobe.)

in place for the first 48 hours after operation in the majority of patients while in selected patients the epidural catheter remains in place until the chest tubes are removed. Atelectasis and subsequent pneumonia in the lung distal to the anastomosis must be avoided. This is accomplished by adequate pain relief and aggressive pulmonary toilet that may include frequent bronchoscopy if the patient is unable to clear their own secretions and consolidation occurs. We begin incentive spirometry as soon as the patient is awake and begin ambulation on postoperative day 1. The chest tube remains in place until there is no air leak, and the drainage is <200 ml/day. This usually occurs by postoperative day 3 to 5. The epidural catheter is capped 12 hours after the chest tube is removed, and oral narcotics are started. If adequate pain relief is obtained on an oral regimen, the epidural catheter is removed. Awake flexible bronchoscopy should be performed before discharge to assess the integrity of the anastomosis.

OUTCOME

The incidence of postoperative complications after sleeve lobectomy is 31.3% as reported by a recent meta-analysis. Major complications following sleeve resection include anastomotic dehiscence, empyema, and bronchovascular fistula. Fortunately, the incidence of these complications is low. We maintain a low threshold for bronchoscopy in the postoperative period. Persistent or evolving atelectasis in the lung distal to the bronchial anastomosis mandates bronchoscopy. A small partial anastomotic dehiscence (<30%) with a good pericardial fat wrap and no bronchopleural fistula can be treated conservatively, but consideration should be given to completion pneumonectomy based on the judgment of the surgeon. A complete dehiscence is caused either by anastomotic tension or more likely results from ischemia. This mandates reoperation and usually a completion pneumonectomy. An empyema can occur with or without a bronchopleural fistula. When it occurs without a bronchopleural fistula, it is handled with drainage, antibiotics, and ablation of any residual space with muscle transposition. Pleural space infection without a fistula is likely related to preoperative postobstructive pneumonia. When empyema occurs with a bronchopleural fistula from the anastomosis, a completion pneumonectomy is required, and management of the infected pneumonectomy space can be problematic.

Bronchovascular fistula presents with massive hemoptysis. This occurs when there is an anastomotic breakdown with peribronchial abscess formation, which necessitates into the adjacent PA. Under these circumstances, completion pneumonectomy is performed if this occurs in the hospital setting, which is unlikely because the time frame when this occurs usually approaches 3 weeks after operation. As expected, the mortality from this complication is very high.

The most frequent respiratory complication is pneumonia, which can be minimized by aggressive postoperative pulmonary toilet and adequate pain control. Local recurrence rates after bronchoplastic procedures are low (<5%) and similar to that achieved by pneumonectomy as long as the resection margins are free of disease. This further underscores the importance of frozen-section examination of the bronchial margins before completing the airway anastomosis. Late bronchial anastomotic strictures occasionally occur and are related either to ischemia or a healed partial anastomotic dehiscence. These usually can be managed with dilation and, if indicated, bronchial stent placement.

The mortality rate for sleeve lobectomy is slightly higher than for routine lobectomy (3%) but significantly lower than for pneumonectomy (5.7%) (Tables 7.1).

Table 7.1 Outcomes of sleeve lobectomy and pneumonectomy reported in the large series

Author (year)	N° patients (Stage I–III)	Complications (%)	Postoperative mortality (%)	Stage I 5-y (%) survival	Stage II 5-y (%) survival	StageIII 5-y (%) survival	Distant recurrence (%)	Local recurrence (%)
Sleeve lobectomy								
Gaissert ('96)	72	11	4	42	53	43	—	14
Tronc ('00)	182	16	1.6	63	48	8	11	22
Fadel ('02)	137	16	2.9	55	62	21	11	15
Terzi ('02)	150	14.5	12	60	32	22	18	5
Deslauriers ('04)	184			66	50	19		
Kim ('05)	47	74.9	6.1	88	52	8	22	22
Ludwig ('05)	116	38	4.3	57	40	22	—	—
Takeda ('06)	62	45.2	1.6	45[a]	45[a]	21.8	29	9.7
Park ('10)	102	33.4	1	58.4[b]	58.4[b]	58.4[b]	11.4	14.3
Pneumonectomy								
Gaissert ('96)	57	16	9	—	43	—	—	—
Deslauriers ('04)	996	—	5.3	50	34	22	—	35
Kim ('05)	49	44	4.1	75	36	38	20	6
Ludwig ('05)	192	26	4.6	45	42	13	—	—
Takeda ('06)	108	40.9	1.8	38[a]	38[a]	52.8	42.7	10.9
Park ('10)	103	29.5	8.6	32.1[b]	32.1[b]	32.1[b]	21.9	16.2

[a]Stage I–II; [b]Stage I–IV.

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Table 7.2 Reconstruction of the PA alone or associated with bronchus

Author	Year	N° patients	Complications (%)	Technical complications (%)	Mortality (%)	5-y survival
Rendina	1999	52	13.4	1.9	0	38.3
Shrager	2000	33	6.1	ns	ns	46.6
Okada	2000	21	ns	0	0	48[a]
Fadel	2002	11	12.4	1.4	0.7	52[a]
Chunwei	2003	21	ns	0	ns	33.3[a]
Lausberg	2005	67	ns	0	1.5	42.9[a]
Nagayasu	2006	29	27.6	ns	17.2	24.2[a]
Takeda	2006	12	ns	0	1.6	54.3[a]
Dartevelle	2007	28	22.9	0	4.1	53[a]
Venuta	2009	105	27	1	1	44

[a]Overall survival of PA and bronchial reconstruction.
ns, not significant.

Survival data reported in large series after sleeve lobectomy are comparable with those after standard lobectomy. Notably, the survival rates show better long-term survival and quality of life after sleeve lobectomy than pneumonectomy for similar lesions, suggesting that bronchoplastic resection, when technically feasible, is preferable to a pneumonectomy for the treatment of lung cancer, even in the presence of hilar and mediastinal lymph node metastases, in light of the improved pulmonary and cardiac functions, decreased morbidity and mortality, equal tumor clearance, local recurrence rate, and long-term survival.

ACKNOWLEDGMENT

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EDITOR'S COMMENTS

Dr. Rendina and coworkers have beautifully described the full spectrum of sleeve resections including those that are rarely performed. The point here is that sleeve resections can be performed on any lobe in the interest of parenchymal preservation. I agree completely with the authors that lung conservation should be the goal for any pulmonary resection as long as oncologic principles are not compromised. This makes it incumbent on the surgeon to not only have familiarity with bronchoplastic techniques but also with pulmonary artery reconstruction techniques. Important technical points to keep in mind are the absolute necessity for a clean bronchial cut perpendicular to the long axis of the bronchus for both the proximal and distal ends. Precise suture approximation making up the size discrepancy is also key because the distal bronchus almost always is significantly smaller than the proximal bronchus. Although tempting, we avoid taking a "tuck" in the larger bronchus and prefer instead to make up any discrepancy with suture bites spaced evenly around the entire circumference of the anastomosis. As opposed to the authors in most cases, I construct the anastomosis with a running technique using absorbable monofilament suture material usually of 4-0 size.

We also feel that it is important to wrap the anastomosis with a viable pedicle flap, and we prefer pericardial fat, which is attached to the bronchus. Aggressive postoperative pulmonary toilet is the key, and frequent therapeutic bronchoscopy may be required to clear secretions. Postoperative atelectasis in the reattached lobe should prompt early bronchoscopy to assure patency of the anastomosis as well as to clear any secretions. On the left side, the superior segment of the lower lobe may be taken as part of an upper lobe resection and the basal segments reimplanted on the left main stem bronchus. Depending on the size of the basal segmental orifice, there may be difficulties with secretion clearance as well as a fairly narrow lumen after the anastomosis. We routinely perform a bronchoscopy just prior to the patient's discharge to assess the healing of the anastomosis and if there is any evidence of nonhealing, we err on the side of performing a completion pneumonectomy rather than wait to have major trouble with a bronchovascular fistula and its attendant high mortality.

The authors mention that anastomotic healing problems often stem from ischemia, but they do not comment on the role of mediastinal lymph node dissection in bronchial ischemia. It is possible, though unlikely, that a complete lymph node dissection that completely cleans out the subcarinal space may contribute to bronchial anastomotic ischemia, and it is my preference to perform lymph node sampling especially in this area. I am not aware of any studies that definitively show that the bronchus can be devascularized by a lymph node dissection, but intuitively it makes sense especially with a thorough clean-out of the subcarinal space with its rich bronchial blood supply.

Bronchial sleeve resection may need to be combined with pulmonary artery resection and reconstruction especially on the left side. We do not hesitate to perform such a double-sleeve resection if that is what is required to save pulmonary parenchyma. It is our preference, and that of many others, to preferentially perform sleeve resection even when the patient's underlying pulmonary function clearly indicates that the patient could tolerate pneumonectomy.

LRK

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8 Sublobar Pulmonary Resections

Aaron M. Cheng and Douglas E. Wood

Sublobar pulmonary resections encompass all pulmonary resections of less than an anatomic lobectomy. They are subdivided into two distinct categories, anatomic, otherwise known as segmentectomy versus nonanatomic, commonly referred to as a wedge resection. Segmental resections were originally considered for patients with limited inflammatory disease, such as aspergilloma, tuberculosis, and bronchiectasis, but the indications have expanded to include metastatic disease to the lung as well as primary lung cancer in patients with poor pulmonary reserve.

It is easy to recommend sublobar resections for diagnostic procedures and benign disease but it is a more complicated decision when dealing with malignant disease. For disease that is metastatic to the lung where preservation of pulmonary parenchyma is a guiding principle, segmentectomy or wedge resection is the procedure of choice if surgery is indicated and the anatomy is amenable to sublobar resection. There is no clear evidence that more radical resection results in better survival or lower recurrence, and frequently these patients require resection of multiple lesions usually dictating smaller individual resections.

The decision to proceed with a sublobar resection for primary lung cancer is complex, and continues to evolve. The predominant indication is for patients with poor pulmonary reserve, yet this concept is ill-defined and inconsistently applied. Traditional teaching has mandated that patients require a predicted postoperative FEV_1 >0.8 to 1.0 L/s to have sufficient pulmonary reserve to maintain adequate short- and long-term functional capacity. However, two factors have contributed to thinking differently about the traditional teaching. Improvements in operative technique, anesthesia, postoperative pain management, and respiratory care have allowed successful thoracotomy and pulmonary resection even in patients with major comorbidity and/or with patients with a low preoperative FEV_1, even less than 0.5 L/s. Clearly these patients are at increased risk of postoperative pulmonary complications but it is unusual that these risks are prohibitive. These patients also have relative contraindications to radiation therapy or chemotherapy and may suffer similar or greater pulmonary complications or loss of function due to radiation when compared to surgery. In patients with major comorbidity a discrete intervention like a surgical procedure is often better tolerated than the less immediate, but insidious and progressive consequences of radiation.

The second factor changing the thinking regarding indications for surgery results from the recent experience with lung volume reduction surgery, which paradoxically often results in an increase in pulmonary function and exercise capacity when overinflated poorly functioning lung parenchyma is removed in selected emphysema patients. Patients with severe upper lobe-predominant emphysema, hyperinflation, and a tumor in the upper lobe, may actually be well served by a lobectomy or a lung reduction style of wedge resection, and actually have an improvement rather than a decrement in postoperative lung function.

The major factor that compels surgical consideration for lung cancer patients, even in those with major comorbidity, is that resection provides the only meaningful opportunity for cure. This is probably not an important factor if a patient's anticipated life expectancy from his/her other disease(s) is less than 1 to 2 years, but for the majority of patients every effort should be made to try to offer surgical therapy with curative intent. Standard lobectomy should be possible in the majority of patients with nonpulmonary comorbidity.

The Lung Cancer Study Group in a classic study conducted a randomized trial that confirmed that lobectomy is the procedure of choice for patients with lung cancer, but sublobar resection and in particular segmentectomy are a reasonable compromise for those patients who are unable to tolerate a lobectomy. However, in the past few years several investigators have suggested that sublobar resections may have similar or equal outcomes to lobectomy in select patients. Lung cancer screening using low-dose spiral computed tomographic (CT) scanning has now been shown to reduce lung cancer deaths in high-risk patients (age >55 and smoking history >30 pack-years). As screening becomes more widely available, smaller lung cancers (<2 cm) are identified. It is likely that the incidence of sublobar resections of these early stage lung cancers is likely to increase. A number of thoracic surgeons already have challenged the standard dogma that lobectomy is mandatory for all lung cancer resections arguing that subcentimeter tumors may be adequately treated by wedge resection or segmentectomy, even in patients with adequate pulmonary reserve. More recently, several single-institution studies suggested that segmentectomy or extended wedge resections may be equivalent to lobectomy in appropriately selected stage IA lung cancer patients, but definitive evidence of oncologic equivalence awaits further follow-up. Currently, a National Cancer Institute-sponsored multicenter study (CALGB-140503) is underway comparing patient outcomes between lobectomy versus sublobar resections (wedge resection or segmentectomy) in small peripheral stage IA nonsmall-cell lung cancer. The study is powered such that results of this phase III randomized clinical trial will likely answer this important question.

Low-dose CT screening also has resulted in the identification of nodules with an alveolar filling appearance rather than a solid appearance, known as ground glass opacities (GGO). The appearance of GGOs on screening CT scans has led to a renewed appreciation of the subset of lung cancer known as bronchioloalveolar carcinoma (BAC), which in its pure form is a noninvasive subset of adenocarcinoma that has little potential for lymphatic and hematogenous metastases. Yet BAC may spread within the airway resulting in a late parenchymal metastasis appropriate for further surgical resection. GGO are frequently

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BAC and may be best treated by sublobar resections since they have a low malignant potential by traditional criteria, yet may need additional pulmonary resections in the future due to the natural history of BAC. However, it is important to differentiate pure BAC from invasive adenocarcinoma with BAC features. The former may be treated by sublobar resection, but the presence of invasive carcinoma should indicate the need for a lobectomy if possible.

There are anatomic considerations that influence appropriateness of sublobar resections as well. Although exceptions always exist, generally segmentectomy and wedge resection are reserved for those patients with more peripheral and smaller tumors, a rough guideline being in the outer third of the lung parenchyma, and less than 3 cm in diameter, respectively. It is possible to consider limited resection for larger or more central tumors, but these will almost always require an anatomic segmentectomy in order to achieve adequate tumor margins. Another anatomic factor is the location of the tumor within the lung. A tumor that is close to or crosses an anatomic segment may require a bisegmentectomy or "cheating" into the adjacent segment with a staple line beyond the segmental boundary. Wedge resections are easiest (i.e., most successful) near acute lung edges, where it is possible to achieve adequate deep margins without encountering lung parenchyma that is too thick to staple. Therefore, peripheral tumors at the lung apex, lung base, or adjacent to a fissure are most amenable to an effective wedge resection.

Segmental resections are based on the principle of following the lymphatic drainage and bronchial branches of the segments resected. This provides a theoretical advantage over nonanatomic wedge resection in the treatment of primary lung cancer. However, segmental resections are also the least common type of pulmonary resection performed and are technically more challenging than lobectomy or pneumonectomy. This frequently results in wedge resections being performed as a default sublobar resection when parenchymal preservation is desired, due to inexperience with the indications for segmentectomy and lack of confidence in the technical components of segmentectomy.

Wedge resection of the lung is performed for a wide variety of indications, including lung biopsy for interstitial or infiltrative processes, excisional biopsy of a lung nodule, and definitive resection of a primary lung cancer or metastatic disease. With the current variety of standard and endoscopic staplers, wedge resection has become extremely easy and reliable. But, as a result, wedge resection is at risk of being overutilized, the technical simplicity being a seductive attraction to the surgeon with little thoracic surgical training or experience. Most of these surgeons have little or no experience with segmentectomy and so frequently will prefer a wedge resection procedure when a segmentectomy, or even a lobectomy, would be preferred for anatomic or oncologic reasons.

There are clear theoretical reasons explaining why anatomic segmentectomy may be superior to wedge resection for primary lung cancer. A segmental resection results in more reproducible deep parenchymal margins, since it extends the resection to the pulmonary hilum. Segmentectomy also incorporates the lymphatic drainage and interlobar lymph nodes and so may result in both more thorough resection as well as more accurate staging. Several studies have suggested decreased rates of local recurrence with segmentectomy but no well-designed study comparing segmentectomy to wedge resection has been performed. However, it is quite possible that the appearance of better cancer outcomes after segmentectomy may be substantially biased because of inappropriate or inadequate wedge resections being performed by less-experienced surgeons. It is reasonable to postulate that outcomes between wedge resection and segmentectomy for primary lung cancer may be similar as long as three principles are adhered to: (1) adequate sampling of N1 and N2 lymph nodes to exclude stage II and stage III disease; (2) at least a 2-cm parenchymal margin around the tumor; (3) restriction of lung cancer surgery to surgeons trained in thoracic surgical oncology who perform a high volume of pulmonary resections in order to have the experience and expertise to select patients who truly require sublobar resection, or who may benefit from segmentectomy rather than wedge resection.

Since sublobar resections are contraindicated in patients with nodal involvement, careful preoperative and intraoperative lymph node staging should be performed. Positron emission tomography (PET) adds significant accuracy to the staging accomplished with chest CT, but still suffers from false-positive and false-negative studies. Mediastinoscopy is indicated to confirm or exclude N2/3 nodal disease in cases of a positive PET scan. Our practice is to perform mediastinoscopy on every lung cancer patient during the same anesthetic as the planned pulmonary resection in order to detect all possible stage III patients prior to thoracotomy, but many would consider this optional for peripheral T1 N0 M0 tumors with a normal CT and PET in the hilum and mediastinum. At the time of thoracotomy, representative N1 and N2 lymph nodes should be sampled since stage II/III patients require lobectomy at a minimum in order to have the confidence of a complete resection.

SEGMENTECTOMY

Intimate knowledge of pulmonary anatomy, and its variants, is crucial in order to perform pulmonary segmentectomy. A complete bronchoscopic examination must be performed at the time of operation. Identification of abnormal segmental anatomy will aid in the planning of the surgical procedure; extrinsic segmental compression or presence of endobronchial tumor in the segmental orifice is a contraindication to segmental resection. Those segments that are amenable to segmentectomy include the apical, anterior, and posterior segments of the right upper lobe and the superior segment and medial basilar segment of the right lower lobe. The middle lobe is rarely considered for segmentectomy since there is little gained from a sublobar resection with this small lobe. The left lung segments amenable to segmentectomy are the apicoposterior, anterior, and lingular segments of the left upper lobe, and the superior and anteromedial basal segments of the left lower lobe. The basal segments may be resected as a unit on either side, sparing the respective superior segment that can provide significant pulmonary function and help to fill the space of the lower hemithorax.

The initial steps of the procedure involve assessment of the tumor size and location to determine the technical feasibility of segmentectomy, as well as determining any evidence of pleural, mediastinal, or nodal involvement that precludes curative sublobar resection. The anterior and apical segmental arterial branches are dissected at the medial and superior aspect of the pulmonary hilum, while the posterior, superior, and basilar segment arteries are found in the fissure (Fig. 8.1). After clearly identifying the appropriate segmental vessel it is divided using standard technique. Dissection of the superior pulmonary vein from the hilum and extending into the lung parenchyma allows identification and division of the respective vein draining the upper lobe segments, although some surgeons prefer to divide the posterior segmental vein within the fissure. The separate superior segment vein

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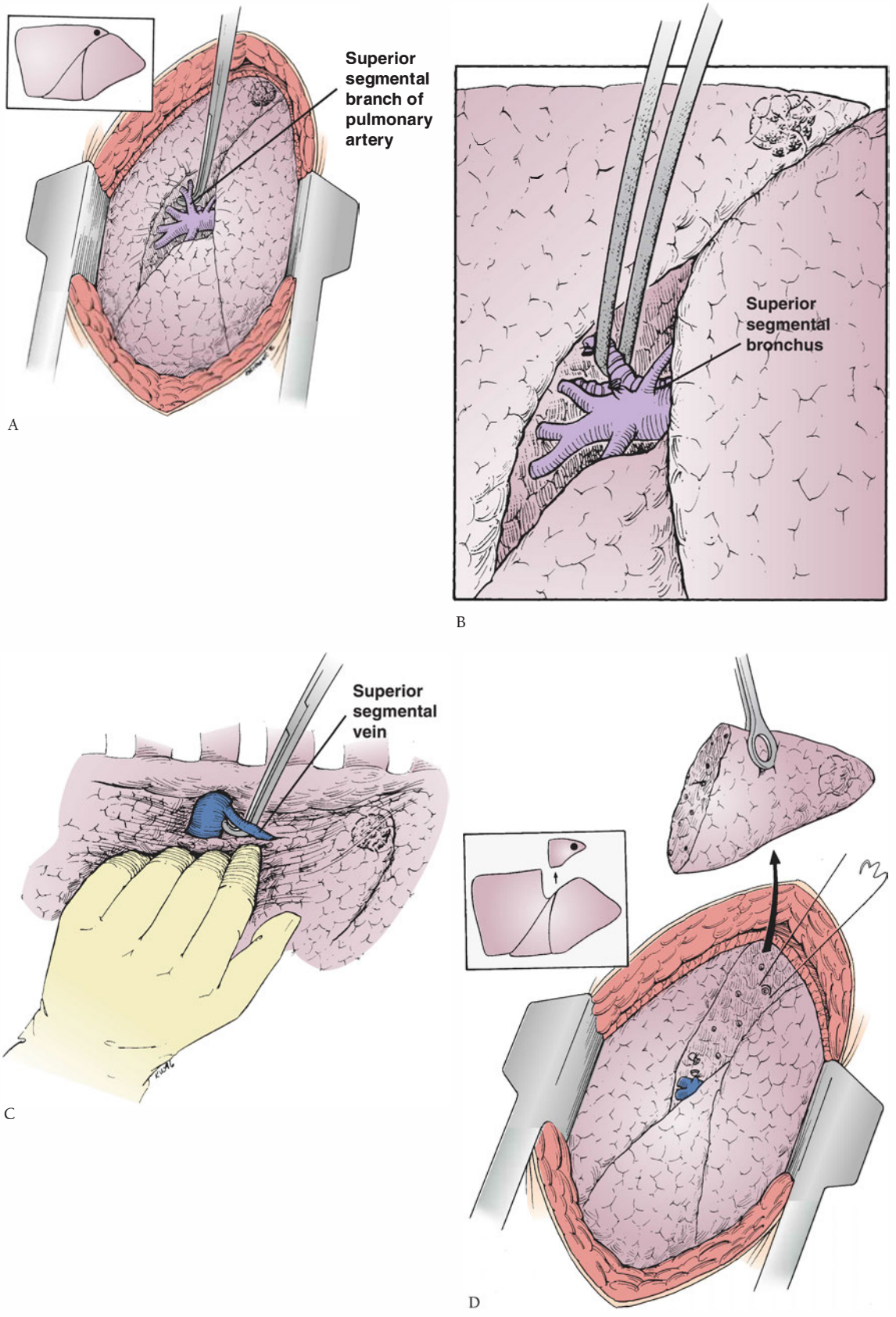


Fig. 8.1. (**A**) With the fissure open, the pulmonary arterial branch to the superior segment is dissected out. Note the tumor at the apex of the *right* lower lobe. (**B**) The superior segmental branch of the pulmonary artery has been ligated, and the superior segmental bronchus is dissected out. (**C**) The lung is retracted medially, and the superior segmental branch of the inferior pulmonary vein is dissected out. (**D**) The intersegmental plane is opened and the specimen is removed. Individual bronchioles and vessels can be ligated or cauterized.

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and basilar segment confluence is readily identified after division of the pulmonary ligament and dissection of the inferior pulmonary vein, but identification of the medial basilar and anteromedial basilar vein is more difficult and requires following the basilar branches into the lung parenchyma. The posterior aspect of the fissure should be completed for posterior, apicoposterior, and superior segmentectomies and the anterior fissure completed for the anterior, lingular, and basilar segments. No fissure division is necessary for the right apical segment. Lymph nodes encountered during vascular control and bronchial division should be resected and submitted for pathologic examination to refine intraoperative tumor staging.

The segmental bronchus is identified deep to the divided pulmonary artery. Peribronchial lymph nodes are resected separately or taken with the specimen to be resected, as part of encircling the airway and confirming that it is the appropriate bronchus leading to the segment to be resected. The bronchus can be divided at its origin with a knife and oversewn with fine absorbable sutures, or alternatively can be divided using a stapler and the stump closed with reinforcing sutures.

The historic method of parenchymal division was accomplished by simultaneous traction on the divided segmental bronchus along with finger fracture blunt dissection along the intersegmental plane defined by the venous anatomy. This technique has the advantage of avoiding restriction of the adjacent segment by a staple line, as well as clear development of adequate margins, but it is tedious to oversew and cauterize the bleeding and air leak sites on the surface of the raw lung parenchyma, and prolonged air leak is a more common postoperative complication. Therefore, stapled division of the pulmonary parenchyma is almost universally employed because surgeons are more comfortable with the technique as opposed to finger fracture and it minimizes the problems of postoperative air leak. To do this correctly, the surgeon must know the parenchymal extent of the segment very well. If there is any uncertainty, a few moments with a surgical atlas is a useful refresher. However, with the stapler it is perfectly acceptable to take a portion of the adjacent segment if this will allow a more complete resection of the tumor with a 2 cm margin. Retracting the nodule with one hand, fingers deep to the nodule on the underlying normal parenchyma, while the other hand applies and fires the stapler can help to assure and maximize an adequate margin during stapling. The surgeon has to approximate the true margin which is different in atelectatic versus inflated parenchyma, and recognize that approximately 1 cm is removed by the pathologist (the staple line) before assessing the histologic margin.

WEDGE RESECTION

The most common technique of nonanatomic pulmonary wedge resection utilizes the linear stapler. Stapling is readily available and effectively seals the lung parenchyma to minimize air leak. As mentioned above, a stapled wedge resection is most amenable to lesions in the outer third of the lung parenchyma and adjacent to an acute edge of lung at the fissure, apex, or base. These are rough guidelines however, and deeper lesions can often be successfully wedge excised, even if they do not fit the routine criteria for wedge resection. But if a deep wedge resection removes a majority of the lobe, or compromises the remaining lung parenchyma, the indications for sublobar resection should be reexamined and reconsideration given to formal lobectomy.

Although stapling is deceptively simple there are several important principles that should be followed to maximize success. First, margins are assured and maximized by the same strategy as outlined in the segmentectomy section—manually lifting the nodule away from the underlying lung while applying and firing the stapler (Fig. 8.2). Second, one must carefully judge the thickness and pliability of the lung parenchyma to be stapled. Most areas of normal lung can be successfully incorporated and sealed with a 3.5 mm or 4.8 mm staple line, but some deep areas may be thicker and not appropriate for stapling. Other patients may have underlying interstitial lung disease or inflammation that results in thickened parenchyma. Attempts to force a stapled wedge resection in these cases may result in staple line dehiscence with serious bleeding and bronchial air leak. There are no gauges to guide the thickness of lung that can be successfully stapled; so this requires surgical experience and judgment. If dehiscence does occur it should be repaired with a running absorbable monofilament suture, incorporating the faulty staple line, the edge of visceral pleura, and the raw lung parenchyma. We prefer a two-layer suture with a running horizontal mattress followed by a simple running suture.

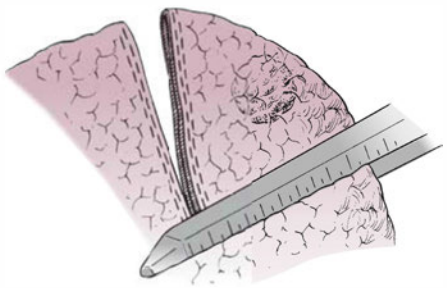


Fig. 8.2. A wedge of lung is resected using a linear stapler.

The other major technique of wedge resection is cautery or laser excision, sometimes referred to as a "precision" resection or a Perelman procedure. This technique is excellent for a benign lesion, such as a hamartoma, particularly when the nodule is deeper and not present at a lung surface. In these cases the nodule can be essentially enucleated from the surrounding lung parenchyma with very minimal loss of normal lung tissue. The second indication for cautery excision is a metastatic nodule in the lung that is deeper in the lung parenchyma or near the pulmonary hilum. The technique of cautery excision may allow successful removal of the nodule with 1 to 2 cm of parenchymal margin with anatomy that would have otherwise required a lobectomy. The third indication is a nodule surrounded by thick or stiff lung that does not have the compliance to accept the compression of a staple line. Cautery resection is rarely performed for primary lung cancer, however, since the margins are less than would normally be desired.

The technique of cautery excision is similar for the electrocautery and the Nd:YAG laser (Fig. 8.3). The desired boundary of the resection is marked on the visceral pleura with the lung partially inflated, and then progressively deepened circumferentially around the nodule with frequent examination of the margin. The excision is performed slowly to allow effective cauterization of the underlying parenchyma. When the resection is complete, the raw lung parenchyma can be oversewn with a running two-layer closure as outlined above, but the surface can also be left as a raw surface if it is superficial and hemostatic with a minimal air leak.

Video-assisted thoracic surgery (VATS), also known as thoracoscopy, can frequently be employed in wedge resections, combining a minimally invasive approach to minimize recovery and morbidity. Much of the ability to palpate the lung is lost with VATS, however, making identification of the nodule more difficult, and precluding a systematic bimanual

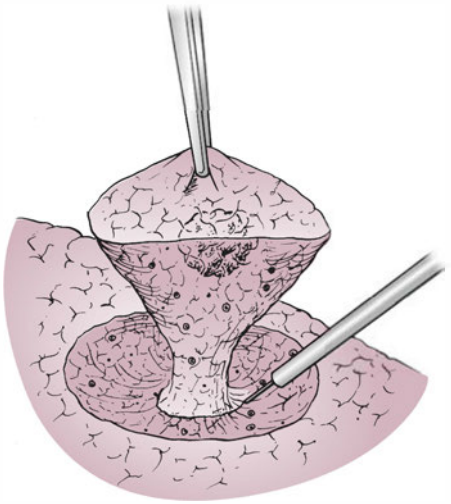


Fig. 8.3. A deep-seated nodule is delivered up into the field, and a cone of lung is resected using precision electrocautery.

palpation of the lung for other pathology. In these cases it is extremely important for the surgeon to have an ability to accurately interpret the two-dimensional CT images into a three-dimensional anatomic location to achieve a high yield of nodule identification. Then if one of the thoracoscopic ports can be placed near the nodule it is possible to palpate the lung digitally through the port to help in localization. This technique is successful in nearly all cases of peripheral lung nodules. An alternative localizing strategy is for preoperative radiologic placement of a wire guide, but this adds to the time and complexity of the surgical procedure and should not be necessary with careful planning and minimally invasive palpation. VATS wedge resection does have a limitation of the technique, however. It is much more difficult to achieve the same degree of nodule retraction with direct tactile feedback and so deep margins may be less reliable. On the other hand, this inability to hold the nodule can often result in the opposite effect, with overly large wedge resections performed to offset any uncertainty in the adequacy of the margin.

VATS is performed routinely for wedge resections, and is now also increasingly performed for standard lobectomy. However, the technical demands of a segmentectomy are still an indication for a thoracotomy at most thoracic surgery centers.

CONCLUSION

Sublobar pulmonary resections are an attractive option for benign or metastatic lung nodules, but wedge resection or segmentectomy may also be indicated in patients with primary lung cancer with poor pulmonary reserve, subcentimeter cancers, or small focal BAC. Few surgeons are familiar with the technique of anatomic pulmonary segmentectomy. This is unfortunate since segmentectomy provides a useful option for patients with poor pulmonary function and anatomy unfavorable to a wedge resection, or those with primary lung cancer where there is a desire to preserve pulmonary parenchyma. Patients undergoing sublobar resections should be carefully staged preoperatively and intraoperatively to exclude more advanced disease that is not appropriate for limited resection. Lobectomy is still the procedure of choice for nonsmall-cell lung cancer, but sublobar resections are reasonable compromise resections that are superior to the alternatives of radiation and chemotherapy.

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EDITOR'S COMMENTS

Parenchymal conservation should always be considered when approaching any lung resection. That said, the definitive resection for primary lung cancer remains lobectomy with mediastinal lymph node dissection. As the authors correctly point out, certain tumors are amenable to anatomic segmental resection with the expectation that oncologic outcome is equivalent to lobectomy. However, anatomic segmental resection should not be compared to or substituted with a nonanatomic wedge excision, a procedure that is associated with a significantly increased incidence of local recurrence and inferior survival. Wedge excision for primary lung cancer results in a compromised parenchymal margin and a complete lack of segmental lymph node staging information. Conversely, anatomic segmentectomy produces an adequate parenchymal margin and, because the segmental bronchus is taken, results in excision of the peribronchial segmental lymph nodes and thus complete staging when accompanied by mediastinal lymph node dissection.

Lingular sparing upper lobectomy on the left and superior segmentectomy on either the right or the left are the easiest segmental resections, whereas segmental resections of the right upper lobe can be quite difficult depending on the location of the lesion. When resecting the basal segments and leaving the superior segment of the lower lobe, it is important to leave the superior segment attached with the upper lobe to prevent torsion. The segmental artery is the key to segmental resection. Once the segmental artery is identified and divided the segmental bronchus becomes apparent. The segmental vein is usually taken following division of the bronchus. Once the bronchus and vessels are taken the line of parenchymal division becomes obvious. It should be underscored that segmental resection is more difficult than lobectomy because of the necessity of following the vessels and the bronchus into the parenchyma. In experienced hands segmental resection may be done using VATS techniques just as lobectomy may be done. With the advent of CT screening in individuals at high risk of developing lung cancer and the ability to recognize very small, early lung cancers it is important that thoracic surgeons be familiar with the techniques of anatomic segmentectomy. This is especially critical for those lesions formerly referred to as bronchioloalveolar carcinomas that have been reclassified as either adenocarcinomas in situ or minimally invasive adenocarcinoma with lepidic growth pattern as these patients may develop additional lesions that are best treated by resection.

LRK

9 Tracheal Resection and Reconstruction

Joel D. Cooper

The surgical management of both benign and malignant tracheal conditions is challenging, often daunting, but very rewarding because of the impact it can have on the daily life of the patient. Technical challenges are posed by both the resection and the reconstruction of the involved airway. The entire process is a logistic as much as a technical exercise, best conducted by an experienced team including radiologists, pulmonologists, thoracic surgeons, otorhinolaryngologists, thoracic anesthesiologists, skilled nurses, and respiratory therapists. The timing and the extent of resection are dictated as much by the presence or absence of comorbidities, as by the nature, location, and extent of the resection required. In carefully selected and well-prepared patients, tracheal resection and reconstruction offer a definitive treatment with excellent long-term outcomes.

Broadly speaking, there are three different types of tracheal resections, each requiring a specific operative approach and surgical technique depending on the location and extent of tracheal involvement. The most straightforward of these is a segmental resection of the trachea with end-to-end anastomosis. Resections at either end of the trachea, specifically a laryngotracheal resection at the proximal end, or a carinal resection at the distal end, may require more complicated anastomotic reconstruction and complementary release maneuvers to reduce tension.

SURGICAL ANATOMY OF THE TRACHEA

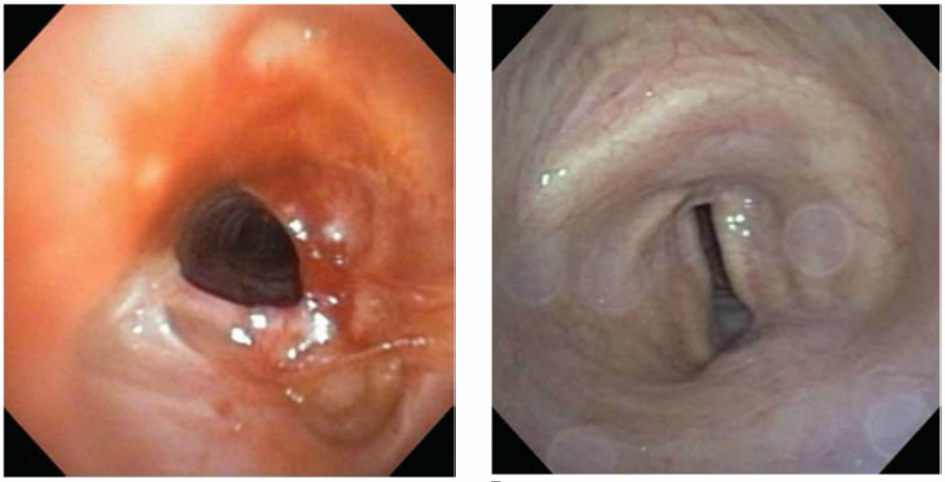
The trachea is a more or less "D"-shaped tube which, in the adult, is of fairly uniform caliber connecting the larynx above with the main bronchi below. It is supported by c-shaped cartilaginous rings and a nonsupported posterior membranous wall. The elastic properties of the cartilaginous portion provide for some degree of lengthening, such as when the head is extended, and the mobile, nonreinforced posterior membranous wall allows a certain degree of change in caliber in response to changes in pleural pressure. The trachea begins in the neck as an anterior cervical structure and terminates distally as a posterior mediastinal structure. Its blood supply is primarily a segmental one with branches from the inferior thyroid artery, the internal thoracic artery, and the bronchial arteries along with variable contributions from the subclavian and brachiocephalic arteries. The blood supply enters the trachea along either lateral aspect from which the vessels travel horizontally between cartilaginous rings. Because the blood supply to the trachea comes primarily from multiple lateral end vessels, it is essential not to circumferentially devascularize the trachea for more than 1 to 2 cm. However, complete blunt anterior and posterior dissection is well tolerated for purposes of mobilization without consequent ischemia to the trachea.

Along its entire posterior aspect, the membranous wall of the trachea is adjacent to the esophagus. Along the anterior surface of the trachea, the thyroid lies at the level of the cricoid and first tracheal ring. The left brachiocephalic artery crosses anteriorly at a variable position, which may be as high as 1 to 2 cm above the sternal notch, or a similar distance inferior to the sternal notch. The right azygos vein lies adjacent to the right lateral aspect of the distal trachea at or just above the tracheobronchial angle. The aortic arch lies in the left anterolateral position to the distal trachea, and the pericardium lies immediately anterior to the carina. The left recurrent laryngeal nerve lies in the tracheoesophageal groove throughout the length of the trachea, while the right recurrent laryngeal nerve occupies a similar position on the right side from the level of the right subclavian artery proximally. Both nerves ascend to the larynx posterior to the cricoid lamina. All of these anatomic relationships have significant implications for the approach and technique used for mobilization and resection of the trachea. In addition, in older patients with increasing kyphosis, a loss of tracheal elasticity and possible calcification may all have significant implications for the approach and extent of safe tracheal resection.

ETIOLOGY OF POTENTIALLY RESECTABLE TRACHEAL LESIONS

Indications for tracheal resection and reconstruction include both benign and malignant conditions with benign conditions far outweighing the malignant ones. The most common indication is postintubation injury to the airway followed by inflammatory conditions especially idiopathic subglottic stenosis, which occurs primarily in young women. Currently, tracheal resection for Wegener's granulomatosis, relapsing polychondritis, amyloidosis, and other inflammatory conditions is exceedingly rare.

Postintubation tracheal stenosis remains the most frequent indication for tracheal resection, though the spectrum of postintubation injuries has shifted in recent years. Formerly, postintubation stenosis at the cuff site of endotracheal tubes or tracheostomy tubes was the most common source of injury followed by stomal stenosis due to loss of the supporting cartilaginous arch at the tracheostomy stoma site. In recent years, however, postintubation stenosis at or just below the cricoid cartilage has become more common and represents a more complex problem. These injuries are usually caused by high placement of a tracheostomy tube with subsequent damage and infection at the level of the cricoid cartilage often extending proximally to within 1 cm of the vocal cords. This injury may be associated with total obliteration of the airway in the subglottic region leaving the patient dependent on a tracheostomy tube and unable to speak (Fig. 9.1). The reason for this increasing number of high airway strictures is uncertain, but the use of percutaneous tracheostomies with malpositioning of the tracheostomy site, urgent posttraumatic placement of a tracheostomy tube in patients who are now more likely to survive complex trauma



A B

Fig. 9.1. (**A**) Typical circumferential tracheal stenosis at the site of endotracheal tube cuff. The circumferential scarring and progressive airway narrowing typically presents as increasing stridor 6-8 weeks following extubation. (**B**) Typical stomal stenosis, distal to the cricoid seen at the level of the second tracheal ring due to loss of the anterior support of one or more tracheal rings. The side walls have collapsed. Such lesions may present years after the tracheostomy when aging, or co-morbidities, accentuate the limitations associated with airway narrowing. The anterior-posterior diameter is usually normal as seen here.

and the rising incidence of morbid obesity making even elective tracheostomy more difficult to site at a desirable position at the second or third tracheal ring all may contribute.

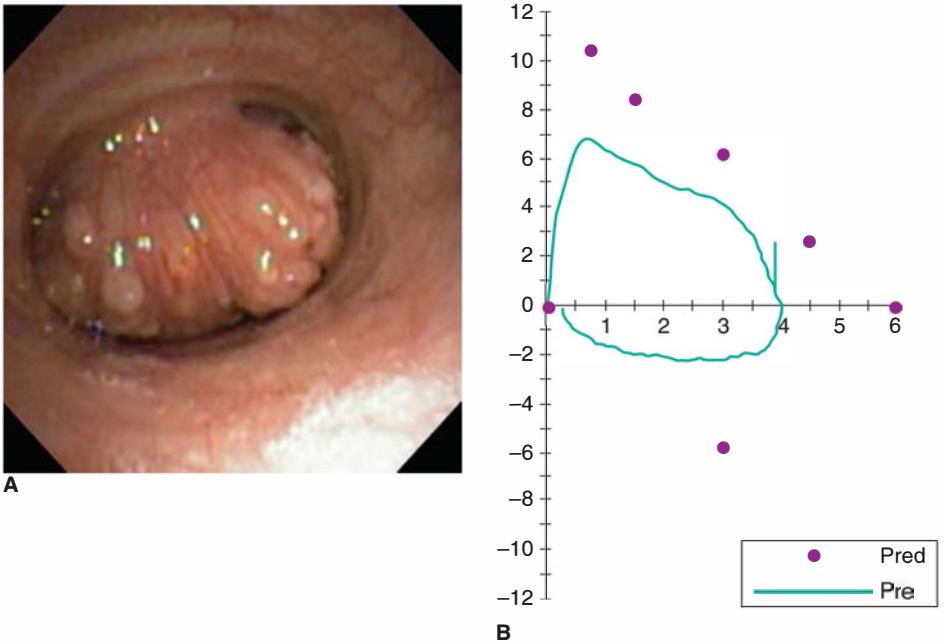
Among malignant conditions requiring tracheal resections, adenoid cystic carcinoma remains the most common. Resection for primary squamous cell carcinoma of the trachea is uncommon both because of the rarity of the condition and the commonly advanced stage of the tumor at the time of diagnosis in addition to improved results with radiotherapy as an alternate option. Tracheal resection for secondary involvement of the airway with malignant tumors arising in other sites is limited almost exclusively to resection associated with direct invasion by thyroid carcinoma.

Whatever the indication, appropriate selection of patients and judicious selection of timing of resection are essential in producing a satisfactory result. Evaluation of both the location of the lesion as well as the extent of the resection required is critical in making a decision as to whether resection is the optimal treatment and if so the type of resection required. In addition, some thought also must be given to the use of temporizing maneuvers to delay resection until the patient is in an ideal condition. The use of silicone stents such as T-tubes frequently aides in preoperative management, and the use of permanent stents as an alternative to resection may, in some cases, prove to be a reasonable strategy.

CLINICAL PRESENTATION AND DIAGNOSTIC STUDIES

The presenting symptoms of patients with tracheal obstruction vary greatly depending on the etiology of the airway compromise. Postintubation stenosis usually presents relatively early following the hospital course, which necessitated ventilator assistance in the first place. If the tracheal damage is proximal usually caused either by a traumatic intubation or injury to the cricotracheal junction from an endotracheal tube or a highly placed tracheostomy stoma, airway obstruction may first be recognized by the inability to decannulate the patient because of an inadequate upper airway. More often postintubation cuff stenosis generally presents as increasing shortness of breath with onset 4 to 8 weeks after decannulation when the full thickness damage to the trachea from the resulting inflammation and fibrosis leads to circumferential scarring and contraction (Fig. 9.2A). An exception is stomal stenosis whereby loss of the anterior supporting portion of the trachea, either due to too large a tracheostomy stoma or extension of the stoma by leverage on the tracheostomy tube by unsupported connection between the ventilator tubing and the tracheostomy, results in collapse of the lateral walls. This so-called inverted V or A-frame stenosis may first present years or even decades after the tracheostomy tube was removed (Fig. 9.2B).

Thyroid carcinoma invading the trachea may be suspected either because of stridor suggesting airway compromise or, for lesser invasion, discovered at



A B

Fig. 9.2. (**A**) Benign cartilaginous tumor (tracheopathia osteochondoplastica) in the proximal trachea of a 20-year old male with a 10 year history of asthma-like symptoms. (**B**) Flow Volume Curve showing significant inspiratory airflow limitation with relatively well-preserved expiratory flow. The dots outline the predicted, normal curve.

the time of thyroidectomy. If there is any question ideally, a bronchoscopy should be performed prior to undertaking the thyroidectomy. Squamous cell carcinomas of the trachea may present with hemoptysis or with a gradual increase in breathlessness or stridor especially with exertion. Adenoid cystic carcinomas that are usually quite slow growing may present with a history of several years of gradually increasing dyspnea on exertion ultimately resulting in shortness of breath with routine activities. A history of increasing breathlessness with stridor going back five years or more is not uncommon.

Furthermore, the degree of airflow obstruction is influenced by the location as well as the physical size of the tumor. This is illustrated in Figure 9.3A and 9.3B.

Benign tumors of the trachea may also present with a very long history of gradually increasing exertional dyspnea. Often these patients have had an extensive work-up for asthma-like symptoms. These tumors, which ultimately may fill the tracheal lumen, often are attached at a very small base or stalk, leaving the rest of the tracheal wall uninvolved. Because of the ability of the intrathoracic trachea to expand on inspiration during normal respiration, such patients may not be noticeably short of breath but may be found to have an FEV_1 of <10% of predicted because forced expiration compresses the trachea against the tumor causing the near total obstruction.

The diagnosis of postintubation stricture is usually apparent from the history and confirmed by bronchoscopic evaluation. The diagnosis of both benign and malignant tumors is also usually obvious on the basis of radiographic and bronchoscopic assessment. Conversely, the diagnosis of idiopathic subglottic stenosis is usually made based upon the typical presentation in females, with gradual progression of stenosis and the exclusion of other benign conditions such as previous intubation, Wegener's granulomatosis and relapsing polychondritis (Fig. 9.4). The diagnosis is subsequently confirmed with bronchoscopy.

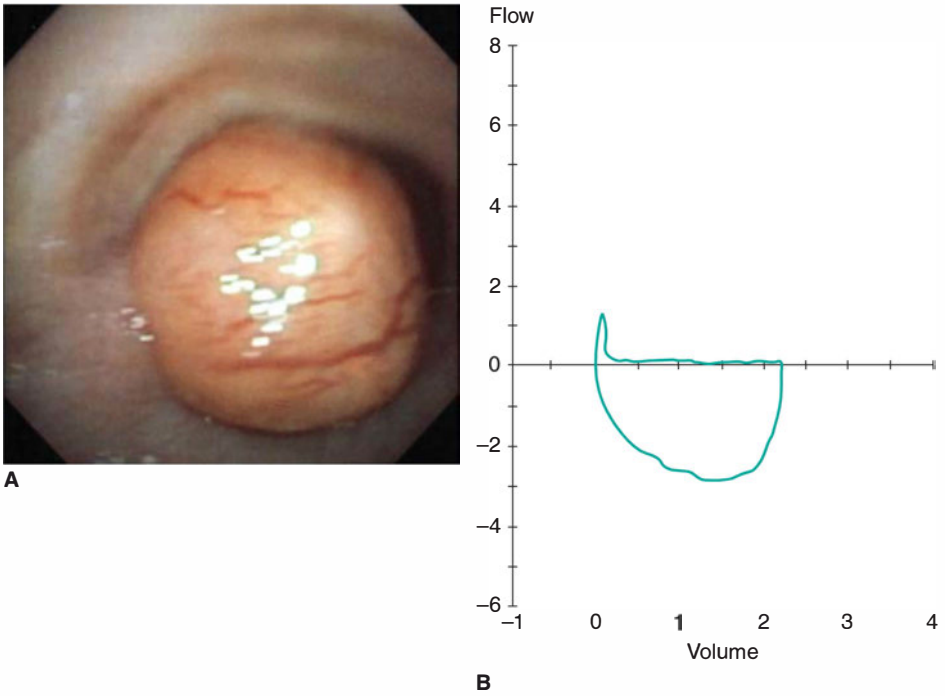


Fig. 9.3. (**A**) Polypoid tumor with mucosal stalk attached to left lateral mid-tracheal wall in 51-year old woman with 4 year history of increasing breathlessness and wheezing. (**B**) Flow Volume Loop shows almost total obstruction on forced-expiration but well preserved inspiratory flow. The FEV1 was 7% of predicted.

ASSESSMENT OF THE EXTENT OF AIRWAY INVOLVEMENT AND RESECTABILITY

Routine preoperative assessment includes a carefully obtained history, physical examination, bronchoscopic evaluation, and pulmonary function studies in addition to imaging. For patients who do not have a tracheostomy tube in place, computed tomography (CT) imaging usually precedes bronchoscopic evaluation. For patients with a tracheostomy in place, the reverse is true and at the time of CT imaging the best study is obtained if the tracheostomy tube can be temporarily removed. This involves determining in advance whether or not the patient can maintain a satisfactory airway for 30 to 60 seconds without the tracheostomy tube in place. If so, the CT scan is done with a physician in attendance. Once the patient is positioned for the scan, the tracheostomy tube is removed and immediately reinserted after the scan is completed. It is best to practice this maneuver with the patient before proceeding with the actual scan. When possible the CT scan should be done with the acquisition of images in both inspiration and expiration. Evaluation of the CT images is best done with a three-dimensional reconstruction along with the assessment of both the axial and sagittal slices.



Fig. 9.4. Typical idiopathic subglottic stenosis in a 39 year old woman with a 1 year history of increasing shortness of breath and stridor. The short circumferential stricture of the airway is centered at the level of the cricoid arch and may extend proximally to within 5-8 mm of the vocal cords and distally to the first tracheal ring.

Depending on the exact software used for the CT analysis a three-dimensional reconstruction usually defines quite nicely the location and the extent of the narrowing of the airway column. However, it does not demonstrate the actual wall thickness or consistency and it is for this reason that the evaluation of the axial sections in particular is essential. It is not uncommon for the airway wall above and below the narrowed segment to be extensively involved with calcification and scarring. In such a situation, the extent of resection will be determined not only by the length of narrowed airway but also by

the amount of tracheal wall involvement proximal and distal to it, which may have considerable influence on the ability to reconstruct the airway if one resects back to normal airway at either end. Based upon the radiologic and endoscopic assessment, a decision can be made as to the resectability as well as choice of operative approach, and the likelihood of release maneuvers that may be required in order to assure a satisfactory, relatively tension-free primary anastomosis (Fig. 9.5). The circumstances most likely to produce scarring, thickening, and calcification include trauma to the neck region, one or more previous tracheotomies, inhalation injury, or the presence of a postintubation tracheoesophageal fistula. Scarring of the airway wall beyond the segment of actual stenosis may provide significant technical difficulty at the time of resection. This is due to the loss of mobility of the airway and loss of tracheal elasticity, which ordinarily help to reduce tension on the anastomosis. Calcification makes it difficult to place sutures through the wall, and mediastinal scarring leads to fixation of the airway and difficulty in mobilization to help bring the severed margins of the resected airway together. Previous sternotomy, obesity, and kyphosis also influence resectability, the surgical approach, and the likelihood of a favorable outcome.

BRONCHOSCOPIC ASSESSMENT

Bronchoscopic evaluation by the surgeon is an essential step in determining the management and resectability of a tracheal or laryngotracheal lesion. This assessment is usually done beginning with flexible bronchoscopy following nasopharyngeal topical anesthesia with the patient mildly sedated so as to be able to assess vocal cord function. Following this, the flexible bronchoscope is advanced through the larynx. If the patient does not have critical airway stenosis, the level of sedation can be increased after assessment of the vocal cords and flexible bronchoscopic assessment continued with or without the use of a laryngeal mask to help support ventilation. Should preoperative or initial bronchoscopic visualization suggest the presence of a markedly narrowed airway in the absence of tracheostomy tube in place, a decision must be made whether or not to traverse the narrowed segment with the flexible bronchoscope out of concern for causing edema or bleeding, which could lead to a critical situation. In this circumstance, the presence of a skilled thoracic anesthesiologist, a skilled nursing team, and a wide variety of equipment appropriate for the situation is essential. This includes rigid bronchoscopes in sizes ranging from 5 mm upward, flexible bronchoscopes ranging from pediatric to therapeutic in size, appropriate-sized dilating balloons, and equipment for jet ventilation through the open, rigid bronchoscopes. It is important to avoid muscle relaxants until a safe airway has been established. This maybe greatly facilitated by the use of a laryngeal mask and the use of Dexmedetomidine (Precedex) sedation, which is relatively unique in its ability to provide sedation without causing respiratory depression. Its use is ideal in the patient requiring awake, rigid bronchoscopic dilatation of the critically narrowed airway.

Until recently, the author favored initiating dilatation of a critical airway stenosis with a No. 5 rigid pediatric bronchoscope converting to general anesthesia once the initial passage of the bronchoscope beyond the stricture had been accomplished. Increasingly, however, the use of a dilating balloon through the therapeutic bronchoscope, which itself is passed through a laryngeal mask, has proven very satisfactory and is somewhat less demanding of the skills and experience of the thoracic surgeon and anesthesiologist. The one concern with initiating dilatation with a balloon rather than with a rigid bronchoscope has been that any bleeding or edema might convert the critical airway to an obstructed airway necessitating an emergent tracheostomy. With the use of a dilating balloon having a diameter in the 6 to 8 mm range, this has



Fig. 9.5. CT Airway study on a 50-year old woman with a post-tracheostomy stricture presenting with increasing stridor 5 weeks post-extubation. (**A**) The extensive narrowing of the air column includes both the stomal site and cuff site. (**B**) Axial view showing thickness and calcification in airway wall requiring extensive resection to achieve satisfactory margins for anastomosis.

not been a problem and provides increased safety of the airway for the subsequent dilatation maneuvers. Generally, this would set the scene for converting to a general anesthesia including, if necessary, the use of paralytic agents, for further dilatation using rigid bronchoscopic techniques. The use of progressively larger rigid bronchoscopes undoubtedly provides a better dilatation than the use of progressively larger dilating balloons even if their inflated diameter is equal to or greater than that of the rigid bronchoscopes selected for dilatation (Fig. 9.6).

After the narrowed segment of trachea has been dilated, the airway should be reassessed with the flexible bronchoscope to determine the stability of the airway. Ideally, the dilatation procedure should lead to a more secure upper airway, eliminating the need for urgent tracheostomy or surgical intervention while further assessment of both the airway and the patient's suitability for resection is considered.

TIMING OF SURGICAL INTERVENTION

The appropriate timing of a tracheal resection is one of the most important factors in producing a satisfactory outcome.



Fig. 9.6. Critical post-intubation tracheal stenosis in a 90-year old woman previously undergoing percutaneous tracheostomy tube for ventilatory assistance following an automobile accident. The tracheostomy tube was in place for 8 weeks and was then removed. The patient began having increased stridor 6-8 weeks later. This bronchoschopic photo illustrates the 4 mm airway at the first-tracheal ring found at the time of patient's presentation. No tracheostomy tube was in place. The stricture was dilated followed by endotracheal tube placement and resection.

This is especially true for benign lesions as many temporizing maneuvers can be used to delay surgical resection until the patient's condition is optimal. These temporizing procedures may include repeated dilatation, use of a silicone T-tube stent, or even prolonged use of a tracheostomy tube to provide a safe airway.

As previously noted, most tracheal resections are currently done for postintubation strictures or malacia of the airway. In many cases, the airway compromise is recognized initially within a couple of months of the initial intubation, when the patient has not completely recovered from the condition (trauma, cardiac surgery, acute respiratory failure, etc.), which necessitated intubation in the first place. In such patients, it is advisable to temporize until the patient is fully active and has achieved their maximum possible overall recovery. This often means delaying definitive resection for a year or more. Any patient with potentially reversible medical conditions should have resection deferred until such time as they are deemed best equipped from a cardiopulmonary standpoint, have attained the best possible physical conditioning, and any ongoing steroid requirements for persistent underlying lung disease have been reduced or eliminated. The need for postoperative ventilatory assistance following a tracheal resection should be assiduously avoided as it is usually associated with an unsatisfactory result. Reoperation after a failed tracheal resection is clearly associated with increased risk, complexity, and an inferior result compared with an initial, successful reconstruction.

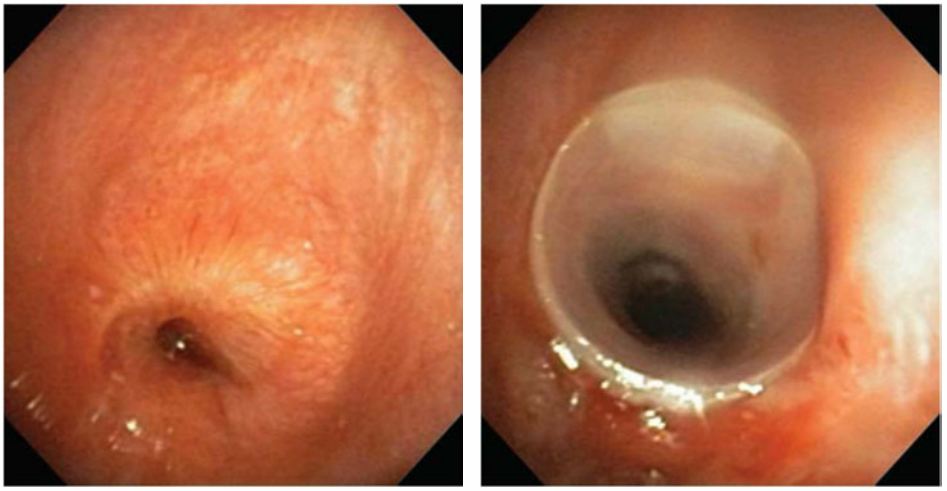
Most cases of critical stenosis can be managed for 7 to 10 days with a single dilatation using rigid bronchoscopy, after which a plan for long-term management should be formulated. If internal tracheal stents are to be used, only silicone stents or T-tubes should be utilized since expandable metal stents may cause further mucosal damage and complicate the subsequent surgical resection. If a silicone T-tube is employed, it should be used only if the patient can breathe comfortably with the horizontal (external) limb capped except for instillation of saline and mucolytic agents. If the patient is unable to breathe comfortably with the horizontal limb capped, then the upper, vertical limb will become clogged with mucous and it is preferable that a tracheostomy tube rather than a T-tube be utilized. If the patient presents with upper airway obstruction and no tracheostomy stoma is present, the stoma for a temporizing tracheotomy tube, or T-tube should be made through the damaged portion of the airway if at all possible. This will avoid damaging an additional segment of the trachea, which further complicates a subsequent resection and reconstruction.

If airway compromise, whether from a benign or malignant condition, is the patient's primary problem and the patient is otherwise suitable for resection, it still maybe advisable to improve the airway, whether by dilatation of the stricture or by endoscopic partial resection of an obstructing tumor to allow the patient to improve their exercise tolerance and improve secretional clearance to further minimize the likelihood of a postoperative pulmonary infection. Usually 3 to 4 weeks of such preparation suffices. If the patient presents with a tracheostomy tube in place, then the use of a silicone T-tube placed through the tracheostomy stoma to stent the narrowed segment of airway is often the optimum method of temporizing and can be in place for an indefinite period. In some situations, leaving a T-tube in place for 6 to 9 months before its removal may lead to a stable widely patent airway, thus eliminating the need for a resection. In the majority of such cases, however, temporary stenting alone is not sufficient and a resection is usually required (Fig. 9.7).

PRINCIPLES OF TRACHEAL RESECTION

As previously noted, tracheal resections can, in general, be divided into three anatomic categories: those requiring a segmental trachea resection with end-to-end to tracheal anastomosis; those involving a resection of the upper trachea and cricoid region with a resulting laryngotracheal anastomosis; those with resection of the distal trachea including the carina with anastomosis between the distal trachea and one or both main bronchi. This latter resection is usually indicated for malignant tumors and is dealt with in Chapter 10.

For all surgical resections and reconstructions of tracheal lesions, the principles are the same. The limits of tracheal resection must extend to healthy, normal airway, whenever possible. Compromise of this principle increases the possibility of an unsatisfactory outcome with persistent or recurrent stenosis. Circumferential dissection of the trachea more than a centimeter or so beyond the resected ends should be avoided so as not to jeopardize the tracheal blood supply. However, blunt dissection for considerable distance along the anterior wall of the trachea, and posteriorly between the membranous wall of the trachea and



A B

Fig. 9.7. (**A**) 27 year-old man exposed to flash fire of his face from acetylene torch welding accident. Endotracheal tube was in place for several days and then removed. Several weeks later the patient developed onset of increasing stridor. Initial treatment was tracheostomy. (**B**) 10 days later a silicone T-tube stent was placed through the trachesostomy site. The upper limb of T-tube extends to the subglottic region allowing normal vocal cord closure above it. The T-tube was left in place pending subsequent laryngotracheal resection.

the esophagus usually is not a problem. Finally, undue tension on the anastomosis should be avoided with and tension-relieving maneuvers employed when necessary. These include the customary postoperative maintenance of partial flexion of the head with a tethering suture between the under surface of the chin and the skin anterior to the manubrium, a suprahyoid release of the larynx, and the right hilar intrapericardial release, particularly useful for distal tracheal and carinal resections. With all of these maneuvers, up to one-half of the trachea maybe resected in some cases, usually in younger patients, without preexisting scarring from a tracheostomy, and with good flexibility of the head and normal tracheal elasticity.

One of the more serious complications that can be incurred during tracheal resection involves injury to the recurrent laryngeal nerves. For benign conditions, keeping the dissection on the cartilaginous tracheal wall will prevent injury to the recurrent laryngeal nerves without the necessity of actually identifying them or dissecting them out.

ANESTHETIC EQUIPMENT FOR TRACHEAL RESECTION

The management of the airway during tracheal resection and reconstruction requires constant communication between the surgeon and an experienced anesthesiologist. The available equipment should include a laryngeal mask, a series of endotracheal tubes ranging from size 5 upwards; a high-frequency jet ventilator set at 15 to 20 lbs per square inch of pressure at a rate of 100 to 120 cycles per minute; appropriate jet catheters 1½ to 2 times the length of the endotracheal tube, with the distal side holes cutoff; a set of sterile anesthetic tubing to be passed from the operative field to the anesthesia machine for ventilation across the surgical field; a sterile flexible armored endotracheal tube, size 6 or 7 to be used when intubating the distal, severed, airway across the operative field; and Parker endotracheal tubes (endotracheal tubes [Parker Medical, Highlands Ranch, CO] with a curled distal tip, to avoid injury to the vocal cords, useful for reintubating the patient through the mouth, if necessary during the conduct of the operation). The sterile draping should be done in a manner, which allows the anesthetist to visualize the operative field as much as possible.

INCISIONS

The surgical approach is dictated by the location and extent of the tracheal damage, and whether or not the resection is for benign or malignant conditions. For most lesions involving the subglottic region or upper half of the trachea, a cervical incision is usually adequate. In a patient with reasonable neck extension and normal elasticity of the trachea, benign lesions even at the mid-tracheal level can usually be managed through the cervical incision. The addition of an upper partial sternotomy may be required in some instances, especially in older patients with lack of good head extension, kyphosis, or fixation of the trachea due to scarring from an old tracheostomy site. Malignant lesions involving the distal third, and benign lesions extending to within several rings of the carina may be accessed either by a right thoracotomy or through a median sternotomy approach. The author favors the trans-sternal approach for benign lesions but for malignant tumors of the distal trachea, where posterior exposure to dissect the trachea away from the esophagus and other mediastinal structures may be important, the right thoracotomy approach is preferable. For lesions that require a very extensive resection, either for a benign condition or in some cases of adenoid cystic carcinoma, a combined median sternotomy and cervical incision provides the opportunity for both the suprahyoid cervical release and the inferior, right hilar, release.

VENTILATOR MANAGEMENT DURING TRACHEAL RESECTION AND RECONSTRUCTION

Prior to induction of general anesthesia, topical anesthesia to the nasopharynx is administered and bronchoscopic assessment of vocal cord function is carried out as well as visualization of the damaged segment of the trachea. If there is a tight stricture present, it is dilated with gradually increased sizes of rigid bronchoscopes to allow passage of an endotracheal tube, preferably a size 5 or larger.

CERVICAL TRACHEAL RESECTION WITH END-TO-END ANASTOMOSIS

The patient is positioned supine with an inflatable pillow beneath the shoulders (Fig. 9.8). The head is extended as much as possible. A curved-collar incision is made one to two fingerbreadths above the sternal notch and superior and inferior subplatysmal flaps are raised. The strap muscles are separated in the midline. Blunt dissection separating the more superficial sternohyoid muscle from the underlying sternothyroid muscle on either side may improve the ability to retract the strap muscles out of the way.

If a tracheostomy stoma is present, the collar incision is made at the level of the stoma and an ellipse of skin is taken with the stoma. The anterior surface of the

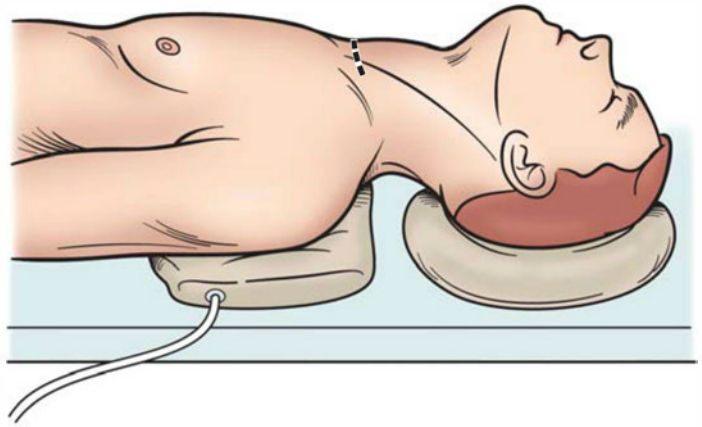


Fig. 9.8. The patient is positioned supine with an inflatable pillow beneath the shoulder. The head, which is positioned at the very upper end of the table, is supported by a ring cushion and is extended.

trachea is exposed below the stoma and blunt finger dissection is carried down as far as possible towards the carina being especially careful not to injure the innominate artery which, if high-riding, may be above the sternal notch. The anterior surface of the trachea is then cleared superior to the tracheostomy stoma, up to the cricoid cartilage.

If no current or previous tracheostomy stoma site is present following separation of the strap muscles, the anterior surface of the trachea is cleared with blunt finger dissection from the larynx down to the distal trachea.

In the case of a cuff stricture caused by a previous tracheostomy, the identification of the stricture site is usually obvious from the external scarring and narrowing of the trachea. In some instances, especially with strictures caused by previous, brief, endotracheal intubation the damage to the tracheal wall at the site of the stricture may not be obvious from external inspection of the trachea. In this case, a flexible bronchoscope is placed down the endotracheal tube, the endotracheal tube is withdrawn to the subglottic position, the stricture is endoscopically identified, and a 25-gauge needle is passed by the surgeon through the anterior tracheal wall into the tracheal lumen as a reference point to determine the exact position of the stricture, which is then marked with placement of a stitch through the anterior tracheal wall.

Traction sutures placed in a midlateral position on either side are helpful not only for the subsequent reduction of tension when doing the anastomosis but also for rotating and elevating the trachea from side to side when dissecting out the damaged section of the trachea. These are full thickness sutures of either 2-0 silk or 2-0 polyglactin (vicryl) placed around one or two rings. These are placed beyond the anticipated limits of resection, and then tied down to the trachea to prevent back-and-forth sawing of the sutures through the wall of the trachea as they are being manipulated.

The lateral dissection of the trachea is then carried out at the level of the lesion, staying very close to the tracheal wall to avoid injury to the recurrent laryngeal nerves. It is not necessary to actually identify the nerves if the dissection is kept on the tracheal wall. The lateral dissection should be done only for the distance that is necessary to encompass the anticipated segment of trachea to be resected. The use of a fine-tipped bipolar cautery for cauterizing blood vessels close to the trachea is useful to help avoid injury to the adjacent recurrent laryngeal nerves. Once the trachea has been divided and the ends are trimmed back, further lateral dissection can be done if necessary.

Division of the trachea can be done in one of two ways. Formerly, the recommendation was to completely encircle the trachea separating the membranous wall of the trachea from the underlying esophagus just distal to the segment of tracheal damage where the scarring is less and the ability to safely separate the trachea from the esophagus is easiest. Often this separation is difficult and there is concern about injury either to the underlying esophagus or damage to the membranous wall of the trachea at a site that might otherwise be suitable for the anastomosis. For benign lesions, division of the trachea at the site of the stenosis is now favored. Using this technique, the trachea is dissected laterally on either side of the lesion down to the junction between the cartilaginous and membranous wall on either side. Then the cartilaginous portion of the trachea is transected transversely sparing the membranous wall.

Once the anterior wall of the trachea has been incised, the endotracheal tube is withdrawn until its tip is visible in the airway and then a heavy traction suture is passed through the tip of the endotracheal tube to facilitate its re-advancement later in the procedure. A high-frequency jet catheter is inserted through the endotracheal tube from above, and the tip is passed into the distal trachea for continued ventilation during completion of the tracheal transection. This improves the visualization of the posterior wall of the airway (Fig. 9.9A). Alternatively, a sterile endotracheal tube can be inserted into the distal trachea and attached to sterile corrugated anesthesia tubing, which has previously been passed through the operative field to the anesthetist. In this situation, periodic removal and replacement of the endotracheal tube is used for intermittent ventilation until the trachea has been completely divided and the distal portion mobilized.

The membranous wall is then transected from the inside with a scalpel, tenting the trachea up with the traction sutures. Once transected, the membranous wall of the trachea is separated from the esophagus at either end of the divided trachea. Following this, the trachea is trimmed back proximally and distally to reach satisfactory margins for the primary anastomosis. A small bougie (e.g., size 36 Maloney dilator) may be placed in advance into the esophagus to help identify the esophagus and facilitate separation of the back wall of the trachea from the esophagus.

Once the resection has taken place and the sutures are to be placed in the posterior membranous wall, a jet ventilation catheter can be placed across the operative field into the distal trachea end and used for jet ventilation (Fig. 9.9B). This gives good visualization of both ends of the membranous trachea while the sutures are placed without having to repeatedly insert and remove an endotracheal tube into the distal trachea.

The posterior wall of the anastomosis involving the membranous trachea is carried out first using 4-0 vicryl sutures. The technique proposed by Grillo involves placement of the sutures with the knot to be tied on the outside of the lumen after all of the other anastomotic sutures have been placed and tied. I prefer placement of the sutures with knots on the inside, tying all of the membranous wall sutures initially, using the superior and inferior traction sutures to take tension off the anastomosis

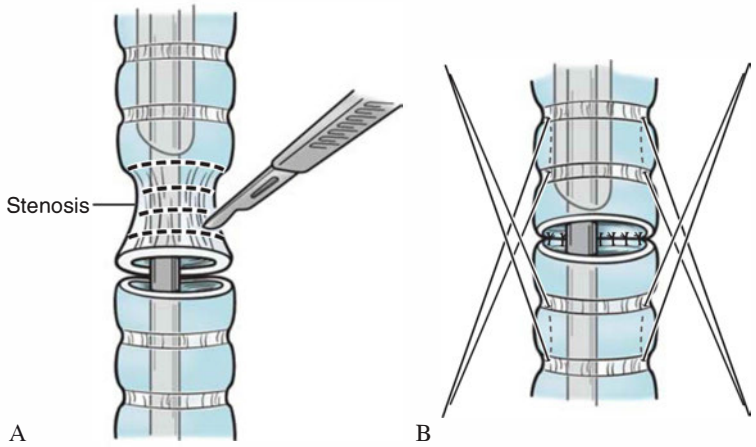


Fig. 9.9. (**A**) After the cartilaginous portion of the trachea has been transected, the endotracheal tube is partially withdrawn so that its tip is proximal to the lesion for improved visualization of the airway. The high frequency jet-catheter is passed down the endo-tracheal tube across the field and into the distal trachea for continued ventilation while the membranous wall of the trachea is transected. Then, in the case of a benign stricture, serial transverse incisions are made through the damaged segment until reaching a level satisfactory for anastomosis and the membranous wall is again transected at that level, completing the resection. (**B**) Following resection, the posterior (membranous) portion of the anastomosis is completed with interrupted absorbable sutures using the jet-catheter for continued ventilation.

as they are tied. It may also help to reduce tension by deflating the inflatable cushion under the shoulders, and asking the anesthesiologist to temporarily flex the head somewhat.

For the placement of the lateral and anterior cartilaginous anastomotic sutures, a jet catheter passed through the endotracheal tube from above and across the partially completed anastomosis, into the distal trachea allows for excellent visualization for the placement of the sutures. Alternatively, an endotracheal tube can be inserted directly into the distal trachea and intermittently removed.

The cartilaginous sutures, using 4-0 polyglactin (vicryl), are placed with knots to be tied on the outside. They are placed sequentially working from the posterior to the anterior portion of the trachea on either side and are left untied until all have been placed.

After the sutures have been placed into the cartilaginous portion of the anastomosis on either side the endotracheal tube, which had been withdrawn into the larynx, can be advanced into the distal trachea to provide ventilatory support while the remaining sutures are placed. Alternatively, the jet catheter can be used for this portion, but once the anastomosis has been nearly completed, ventilation through the jet catheter is unsatisfactory as most of the ventilatory volume from the jet catheter involves entrained air through the open trachea. Once the opening of the tracheal wall is closed, it is best to ventilate the patient with the endotracheal tube advanced into the distal trachea with its cuff placed well beyond the anastomosis.

Once the anastomotic sutures have been placed and are ready to be tied, the inflatable bag is deflated, the extended head brought forward by the anesthesiologist, and the proximal and distal traction sutures on either side are pulled together to avoid tension when tying the sutures.

In most cases, the traction sutures are usually removed following completion of the anastomosis. In cases where tension on the anastomosis is a concern, the superior and inferior traction sutures on either side can be tied to each other and left in place.

The anastomosis should be covered, if possible, with surrounding tissue. Often the thyroid isthmus has been divided as part of the dissection, and the isthmus can then be reconnected over the anastomotic site. Otherwise, the strap muscles can be used.

A ¼-inch Penrose drain is placed in subcutaneous space on either side and brought out through the corner of the incision as a blood drain. This is removed on the first postoperative day.

At the end of the procedure, the endotracheal tube is withdrawn and replaced with a laryngeal mask allowing flexible bronchoscopy through the laryngeal mask to assess the anastomosis and also allowing for visualization of vocal cord function as the patient awakens from the anesthesia. The operative field remains sterile at this time so that if a protective tracheostomy is deemed necessary, it can be performed below the anastomosis. The need for this is very uncommon with segmental trachea resections but may be necessary after complex laryngotracheal resections, as subsequently described.

Following closure of the wound, a No. 2 monofilament polypropylene suture is placed transversally through the submental skin and then passed transversally deeply through the skin in front of the manubrium. This tethering suture, designed to protect the patient from inadvertent hyperextension in the postoperative period, is placed while the patient is still under light anesthesia but left untied until the patient is breathing spontaneously. It is then tied with the head in slight flexion.

SUBGLOTTIC RESECTION AND LARYNGOTRACHEAL ANASTOMOSIS

Resection of the subglottic airway is most commonly required for idiopathic subglottic stenosis and for postintubation injury affecting the region of the cricoid and upper trachea. It may also be required for the treatment of traumatic injuries including cricoid fractures and laryngotracheal separation and for occasional tumors involving the cricoid cartilage.

For this procedure, the anastomosis often must be placed within several millimeters of the vocal cords. Avoiding injury to the recurrent laryngeal nerves is accomplished by resection of the anterior portion of the cricoid cartilage and reaming out of the posterior cricoid plate, or lamina, with preservation of its posterior perichondrium as the nerves are located posterior to the posterior cricoids lamina. The most common source of postintubation injury to the subglottic area is a high tracheostomy site placed inadvertently through, or just inferior to, the cricoid cartilage, or placed there during the performance of an emergency tracheostomy in a life-threatening situation. In addition, such a high placement can occur during an elective tracheostomy in a patient with adverse anatomy such as significant cervical kyphosis, obesity, or other conditions limiting access to the trachea at the level of the second or third rings. As a result of the high tracheostomy placement, the cricoid arch is either directly injured or subsequently eroded into by the tracheostomy tube. With such an injury caused by a high tracheostomy, it is not uncommon for patients to present with a tracheostomy tube in place accompanied by complete obliteration of the subglottic airway just above the tube (Fig. 9.10). In such cases, it may be possible to reestablish an airway



Fig. 9.10. This 23-year old male suffered a spontaneous sub-arachnoid hemorrhage and was intubated with an endotracheal tube for several days. Following discharge to a rehabilitation center he developed increasing stridor and subsequently underwent emergency tracheostomy. Subsequent assessment, shown here, demonstrated complete obliteration of the subglottic airway. The patient underwent laryngotracheal resection with primary anastomosis.

using rigid bronchoscopic techniques and dilatation followed by placement of a silicone T-tube to temporarily stent the subglottic stricture. This allows the patient to speak and breathe through the upper airway permitting humidification of inspired air. Just as a highly placed tracheostomy can lead to considerable inflammatory changes in the subglottic region, so too can repeated application of laser ablation and endoluminal dilatation in a patient with idiopathic subglottic stenosis. This may prevent or significantly complicate subsequent surgical management of the subglottic stricture. An occasional patient with idiopathic subglottic stenosis may be managed with periodic dilatation, steroid injection, and mitomycin injection, but the natural history of the condition is one of decreasing intervals between the need for repeated dilatation. Such patients should be considered for surgical resection after no more than two or three attempts at endoscopic management.

Operative exposure for a laryngotracheal resection is similar to that for standard cervico tracheal resection. The operative field should be prepped and draped up to the chin, as exposure of the hyoid bone may be necessary for a complementary suprahyoid laryngeal release procedure. If a tracheostomy tube is in place, and there is no airway passage remaining through the strictured airway to allow intubation through the mouth, then an armored endotracheal tube is substituted for the tracheostomy tube and prepped into the field. Following exposure of the larynx and anterior trachea, the lateral portion of the trachea at the level of the first tracheal ring is dissected back to the membranous wall of the trachea on either side taking great care to stay directly on the tracheal cartilage to avoid injury to the recurrent laryngeal nerves. Only bipolar cautery should be used in this region. A horizontal incision is made through the perichondrium of the cricoid ring from one mid-lateral position to the other and the perichondrium peeled back proximally and distally to expose the cartilage. It is not uncommon for the cartilage to be calcified in such patients. The anterior half of the cricoid is then removed, saving intact for the moment the perichondrium behind the cricoid ring. Following this, the anterior wall of the airway is incised with a scalpel generally through the bed of the cricoid which is also likely an area of dense scarring and airway narrowing (Fig. 9.11). The transaction of the airway is then carried down in an oblique manner on either side to reach the junction between the inferior rim of the cricoid lamina and the membranous wall of the first tracheal ring just inferior to the cricoids lumina. This gives good exposure of the posterior tracheal wall, allowing it to be carefully transected with a scalpel, from the inside of the partially divided airway.

In our early experience, the recurrent laryngeal nerves were carefully identified at this point, as they traveled cephalad from the tracheoesophageal groove on either side to ascend behind the posterior cricoid lamina. More recently, however, no attempt is made to identify the nerves, and the dissection of the lateral tracheal wall and transection of the posterior wall is done carefully to keep dissection posterior to the cricotracheal junction to a minimum. Once the trachea has thus been completely transected, upward traction on the distal cut end of the trachea permits careful dissection of the trachea from the esophagus. In most cases, when there has been an existing tracheotomy in place at the time of the operation, the tracheostomy site in the trachea is excised as well. However, in some cases, the stoma may be left in place if there is sufficient trachea proximal to the stoma to permit a satisfactory anastomosis without

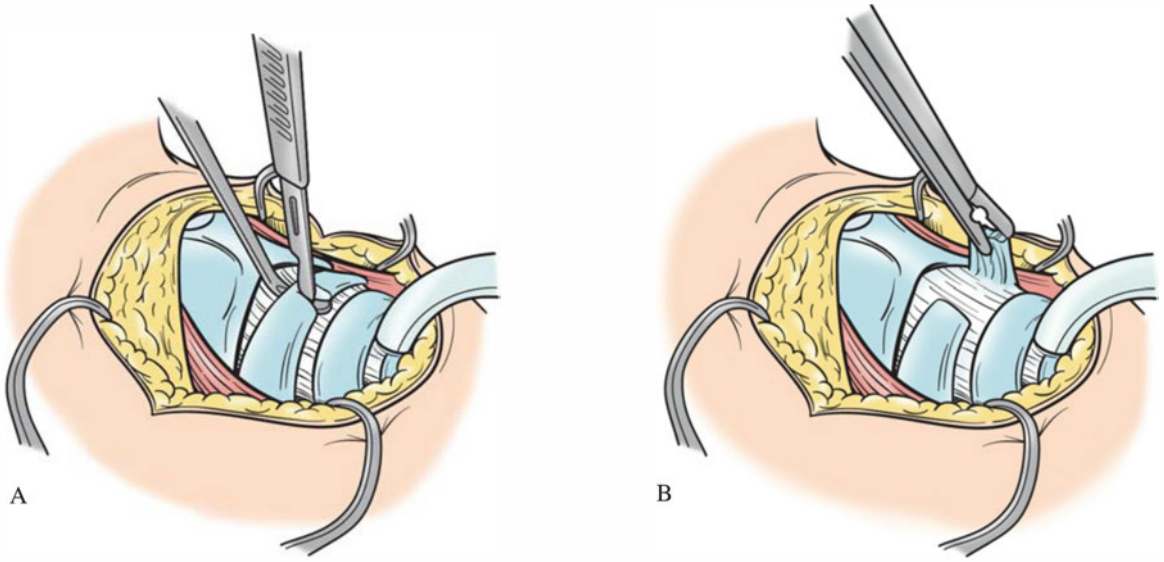


Fig. 9.11. (**A**) The cricoids ring is transected vertically in the mid-line. (**B**) The cricoid ring is then resected back to the mid-lateral position, or further, on either side using a pituitary rongeur.

the need to resect the additional tracheal length necessary to excise the segment of the trachea containing the stoma.

Once airway division has occurred, the posterior cricoid lamina is reamed out between its anterior and posterior perichondrial layers. This can be done with a pituitary rongeur, a dental burr, or a curette depending on the circumstances. This removal of a portion of the posterior cricoid lamina basically provides a recess behind the airway mucosa and perichondrium, which can be carried up for as much as 1½ to 2 cm (Fig. 9.12). Any advancement beyond this point risks injury to the cricoarytenoid joints, which are on the cephalad border of the cricoid lamina. The anterior airway, essentially the cricothyroid membrane, and the posterior membrane, the mucoperichondrial flap provided by the coring out of the posterior cricoid lamina, are then sharply trimmed with a scalpel up to the point where the tissue is as healthy as possible, still permitting a primary anastomosis just inferior to the vocal cords. Clearly, the vocal cords present an upper limit to such excision and to avoid injury to them, it may not be possible to excise back to entirely normal tissue especially if the injury extends up into the conus elastica below the superior borders of the vocal cords.

Exposure for the posterior dissection of the subglottic airway and placement of the posterior anastomotic sutures may be facilitated with the use of an anterior midline laryngofissure. Ordinarily, a complete anterior laryngofissure is carried out, but for this procedure an incision of the lower two-thirds of the laryngeal cartilage, preserving the anterior vocal process, may be sufficient to avoid injury to the anterior vocal cord attachment while still providing excellent exposure (Fig. 9.13).

Once both the upper and lower resection margins have been defined and prepared, the amount of tension on the anastomosis is evaluated by deflation of the inflatable cushion behind the shoulders, flexion of the head by the anesthesiologist, and upward traction of the trachea using the lateral traction sutures. If it appears that the tension on the anastomosis will be excessive, then a suprahyoid laryngeal release is employed as subsequently described. If it is apparent from the outset that adequate resection of the subglottic stenosis will likely result in significant tension, then the laryngeal release can be done prior to the division of the airway.

The tracheal end of the anastomosis is usually done utilizing the full circumference of the trachea; however, if there is excessive discrepancy between the subglottic airway and the trachea, the tips of the cartilaginous ring at the end of the divided trachea can be brought together with one or two side-to-side stitches imbricating the membranous wall and creating a complete cartilaginous ring, as described by *Pearson* in the initial report of laryngotracheal resection procedure (Fig. 9.14). *Grillo* subsequently described a modification in which the uppermost cartilaginous ring of the divided trachea was excised, leaving the associated membranous portion of this segment intact thus creating a posterior mucosal flap for anastomosis to the posterior laryngeal mucosa while the remainder of the tracheal portion of the anastomosis is done through the cartilaginous portion of the uppermost tracheal cartilage.



Fig. 9.13. Using an oscillating saw, or a scalpel, a vertical incision is made in the midline from the inferior edge of the laryngeal cartilage up to or through, the anterior vocal commissure. A small, HEISS retractor is utilized to spread the cartilage giving excellent exposure to the posterior wall for a high subglottic anastomosis. The laryngeal cartilage is then re-approximated and the anterior portion of the anastomosis between the tracheal and the laryngeal cartilage is then completed.

If the posterior wall of the anastomosis on the laryngeal side involves thickened or inflamed tissue, or is well up into the larynx, the posterior portion of the anastomosis may be carried out with fine stainless steel wire (34 gauge) to provide a strong, inert suture line in this narrow portion of the upper airway. This reduces the chances or postoperative granulation tissue that may form if absorbable sutures are utilized. Alternatively, absorbable sutures (4-0 vicryl) may be placed in a manner such that the knots are to be on the outside of the lumen but tied from the inside, as described by *Montgomery*, reducing the likelihood of granulation tissue.

When the anterior wall of the airway at the laryngeal end is being prepared, it may be trimmed back to the inferior border of the laryngeal cartilage and the laryngeal cartilage used for the anastomosis. However, preserving a small fringe of the cricothyroid membrane, inferior to the laryngeal cartilage, may facilitate matching up the size of the tracheal end of the anastomosis with the laryngeal end of the anastomosis.

After the posterior sutures have been placed and subsequently tied, the lateral sutures on either side are then placed and tied. The endotracheal tube can be advanced into the distal trachea and the remaining sutures placed and tied using 4-0 absorbable vicryl sutures.

As previously described following cervical tracheal resection with end-to-end anastomosis, it is useful to substitute a laryngeal mask for the endotracheal tube

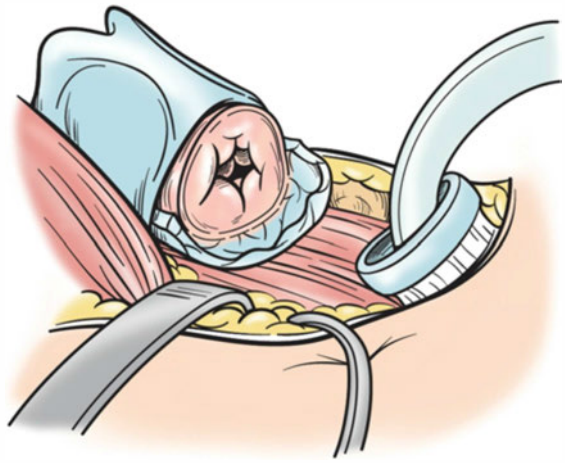


Fig. 9.12. The posterior plate, or lamina, of the cricoid cartilage is reamed out undermining the airway mucosa anteriorly but preserving a thin rim of cartilage and perichondrium posteriorly to avoid injury to the underlying recurrent nerves. This excavation of the cartilage is usually carried out with a pituitary rongeur or dental burr.

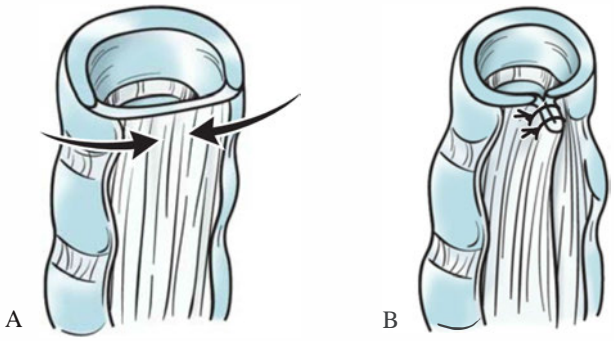


Fig. 9.14. If the circumference of the trachea needs to be reduced to match that of the proximal, subglottic resection margin, this is best accomplished by plication of the membranous trachea bringing the ends of the cartilage together with one or two sutures to plicate the membranous wall.

after the anastomosis has been completed and the wound is being closed. This permits the assessment of vocal cord function and of anastomotic patency. If there is any concern about the safety of the airway, a mini-tracheostomy or small cuffed tracheostomy may be placed below the airway anastomosis. Rarely, following completion of the anastomosis, a silicone T-tube may be placed through a tracheostomy stoma with the upper limb of the T-tube extending through the anastomotic region to a point just below the vocal cords. This is done if, having extended the proximal resection as high as safely possible, it is recognized that the mucosa at the proximal end of the anastomosis is thickened, scarred, or inflamed.

The "guardian" chin suture is placed but left untied until the patient is fully awake, the airway is found to be satisfactory, and the patient is about to leave the operating room.

RESECTION OF DISTAL TRACHEAL LESIONS

For resection of a distal tracheal lesion, either a median sternotomy or a right posterolateral thoracotomy approach may be used. For benign conditions such as stricture or for a malignant tumor that appears confined to the airway without significant extratracheal extension, the median sternotomy provides excellent exposure and may prove technically less demanding than a posterolateral thoracotomy. For bulky tumors or tumors that may have a posterior extension involving the esophagus, a right thoracotomy is preferred. Both approaches permit a hilar release procedure, when necessary to reduce tension on the anastomosis. For the trans-sternal approach, a single-lumen endotracheal tube is usually satisfactory with the availability of a sterile set of tubing on the operative field for intubation of the distal trachea or one of the main bronchi. In addition, provision for jet ventilation through the endotracheal tube from above or across the operative field should be made. If a right thoracotomy approach is chosen, then a left-sided double lumen tube should be employed if the airway lumen will allow its passage. Otherwise, a long endotracheal tube directed into the proximal left main bronchus under bronchoscopic control should be placed. In this situation, inflating the cuff will provide for left lung ventilation with isolation of the right lung, while deflation of the cuff and packing the pharynx with a wet gauze provides for direct ventilation of the left lung and indirect ventilation of the right lung. Following dissection of the airway via a right thoracotomy, it is important to keep in mind that when encircling the distal trachea care must be taken to avoid injury to the left recurrent laryngeal nerve, which may be difficult to visualize. Keeping the dissection on the wall of the trachea and as close to the carina as possible will help avoid injury to the nerve.

If a right thoracotomy is being used for the resection of a malignant tumor, the operation often begins with a mediastinoscopy and assessment of the extent of the tumor and any possible spread to mediastinal lymph nodes, which may contraindicate proceeding with the resection. If the mediastinoscopy findings support proceeding with the tracheal resection, then it is helpful to dissect the left recurrent laryngeal nerve away from the lateral wall of the distal trachea and tracheobronchial angle with blunt dissection creating a free-plane between the trachea and the nerve. This greatly facilitates safe encirclement of the distal trachea through the subsequent right thoracotomy exposure.

When the sternotomy approach is used, the sutures for the posterior, membranous wall of the anastomosis are the first to be placed and tied followed by advancement around both sides of the cartilaginous trachea and finally completing the anterior portion of the anastomosis. When done through the right thoracotomy, the "far" sutures, involving the left lateral trachea wall, are the first to be placed and subsequently tied with knots on the inside followed by the remainder of the anastomosis.

RELEASE PROCEDURES FOR DECREASING TENSION ANASTOMOSIS

Suprahyoid Release

Release of the superior attachments of the larynx, as part of a tracheal resection, may increase the limits of safe resection by providing additional "length" and thus avoiding undue tension on the suture line. This maneuver is most commonly used for laryngotracheal and cervico tracheal resections if there appears that there will be significant tension at the level of the anastomosis. It is usually required if 5 cm or more of trachea is to be resected. It should be performed, in general, prior to the completion of the anastomosis since the release will help determine how much of the airway can be resected without undue tension. Most commonly during the resection, the inferior and superior margins are progressively extended intermittently pulling the two ends of the airway together to estimate anastomotic tension before deciding the balance between further excision versus the risk of increasing anastomotic tension and the complications, which may result. For the laryngeal release, the suprahyoid procedure described by *Montgomery* is preferred as it is associated with a lesser risk of postoperative pharyngeal dysfunction and aspiration than the infrahyoid release.

Montgomery's original description of this release utilized a large "U"-shaped skin incision with creation of a large skin flap extending upward from the inferior aspect of the incision just above the sternum notch up almost to the mandible on either side. This is the same incision that is ordinarily used for laryngectomy. We have found such a flap unnecessary and instead utilize a small separate transverse incision made directly over the hyoid bone (Fig. 9.15A). It is important, however, to dissect the subplatysmal plane between the initial inferior cervical incision and the hyoid incision in order to make certain that the larynx is completely released from its subcutaneous attachments to assure maximum descent following the release.

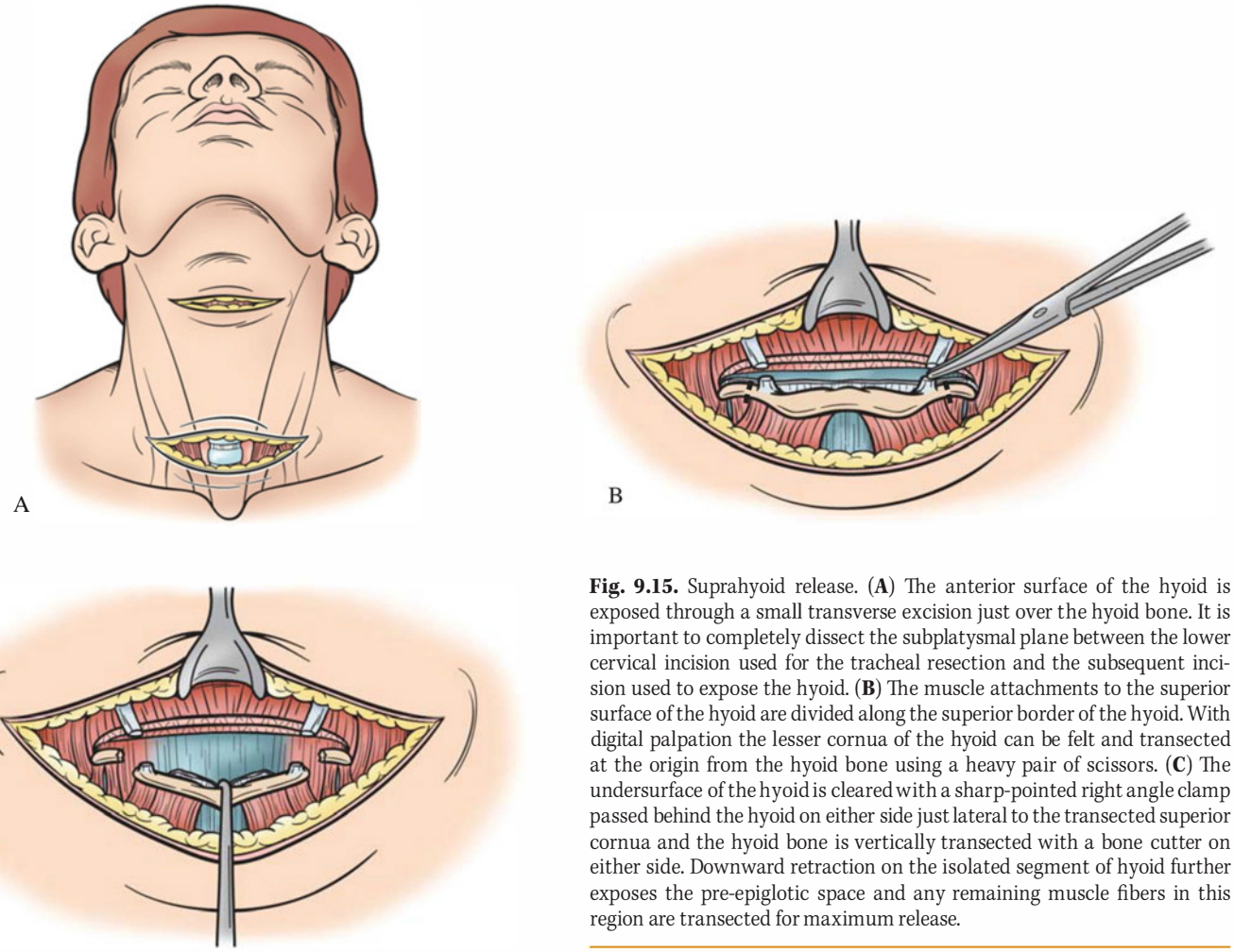


Fig. 9.15. Suprahyoid release. (**A**) The anterior surface of the hyoid is exposed through a small transverse excision just over the hyoid bone. It is important to completely dissect the subplatysmal plane between the lower cervical incision used for the tracheal resection and the subsequent incision used to expose the hyoid. (**B**) The muscle attachments to the superior surface of the hyoid are divided along the superior border of the hyoid. With digital palpation the lesser cornua of the hyoid can be felt and transected at the origin from the hyoid bone using a heavy pair of scissors. (**C**) The undersurface of the hyoid is cleared with a sharp-pointed right angle clamp passed behind the hyoid on either side just lateral to the transected superior cornua and the hyoid bone is vertically transected with a bone cutter on either side. Downward retraction on the isolated segment of hyoid further exposes the pre-epiglotic space and any remaining muscle fibers in this region are transected for maximum release.

After the hyoid bone has been exposed, a needle-point cautery is used to divide the muscle attachments from the superior surface of the hyoid bone from a point just lateral to the inferior cornu on one side to a similar point on the other side. These muscle attachments include the mylohyoid, geniohyoid, and genioglossus muscles. The lesser cornua of the hyoid bone are identified by palpation and cut from their attachment to the hyoid bone. These maneuvers expose the pre-epiglottic space. The hyoid bone is then transected with a small bone cutter on either side just lateral to the lesser cornua (Fig. 9.15B). With downward traction on the mobilized anterior segment of the hyoid, the pre-epiglottic space is further dissected to give maximum release taking care not to breach the mucosa (Fig. 9.15C).

Intrathoracic (Right Hilar) Release

The right hilar release maneuver is most commonly employed in conjunction with a major resection of the mid or distal trachea or carina but may also rarely be used for an airway anastomosis following a right upper lobe sleeve resection or a bilobectomy with reattachment of the lower lobe bronchus to the right main bronchus.

The hilar release can be performed through the median sternotomy or through a right thoracotomy approach. The inferior pulmonary ligament is mobilized and the pericardium anterior to the inferior pulmonary vein is incised about 1 cm anterior to the vein. This incision is carried inferiorly in a curved manner toward a point just inferior to the pulmonary vein. A similar incision is made posterior to the inferior pulmonary vein and joins the previously made anterior incision to form a U-shaped incision around the lower half of the pulmonary vein. The actual release procedure involves intrapericardial incision of the fibrous connection between the inferior vena cava and the pericardium. This connection is the distal extension of the interatrial groove. Using a Kocher clamp the midpoint of the cut edge of the pericardium below the vein is grasped, and using scissors inside the pericardium, the septum between the inferior vena cava and the inside of the pericardium is carefully incised down toward the diaphragm as far as possible. This release then allows the right lung and its attachment to the atria to be retracted upward (Fig. 9.16). The pericardial release can be carried more superiorly by extending the pericardial incisions superiorly toward the pulmonary artery and when used to its maximum extent may involve freeing up the right pulmonary artery from its pericardial attachments as well.

POSTOPERATIVE MANAGEMENT

Following completion of a tracheal resection, the patient is allowed to awaken and is extubated while in the operating room. During this time, the instrument table should remain sterile, a flexible and rigid bronchoscope should be ready,

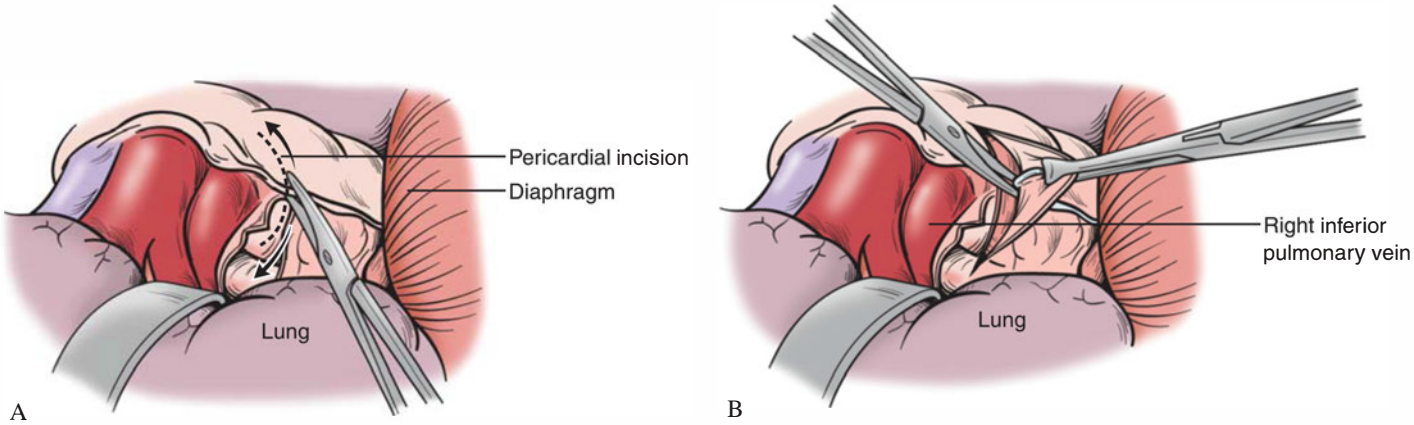


Fig. 9.16. Right Hilar Release. A semi-circular incision is made in the pericardium just below the inferior pulmonary vein. The incision begins anterior to the vein, curves around its inferior edge, and continues upward for 2 cm posterior to the vein. This incision crosses the fibrous connection between the inferior vena cava and the pericardium which is a downward extension of the inter-atrial groove. A Kocher clamp is used to grasp the distal cut edge of the pericardium just inferior to the vein. As the pericardium is lifted away from the atria, the fibrous connection is well visualized and is divided with scissors from the cut edge of the pericardium down toward the diaphragm within the pericardial sack.

and a tracheostomy kit should also kept readily available. As previously noted, the endotracheal tube is removed and a laryngeal mask is substituted as the patient is emerging from anesthesia. Through this mask a flexible bronchoscope is utilized to assess the anastomosis, clear the secretions from the airway, and assess vocal cord function as the patient awakens. A decision is then made as to whether or not a protective tracheostomy is required. If so, a small endotracheal tube can be inserted with the cuff placed distal to the anastomosis or ventilation can be maintained with a laryngeal mask. If a tracheostomy is to be inserted, this is ideally done with a small opening in the trachea 1 to 2 cm distal to the anastomosis. If the procedure has been very long and complicated and there is a question of needing ventilatory support postoperatively, then a cuffed tracheostomy tube is utilized. Otherwise, a noncuffed, or a so-called, "mini-tracheostomy" tube is utilized for airway protection.

The patient should be observed for the first few hours in the postoperative recovery area, readily accessible for periodic observation by the surgeon and anesthesiology staff before transfer to the nursing unit. The patient is initially placed in the supine position with the head of the bed elevated to reduce edema. To minimize edema a dose of methyl prednisone sodium succinate, 125 to 250 mg, may be administered intravenously toward the end of the operation. Diuretics may also be employed to reduce postoperative edema, and a tank of heliox (30% O_2 to 70% He) should be available for administration through a mask, if there is stridor. Nebulized racemic epinephrine may also be administered via face mask in the recovery room.

Oral intake should be withheld until at least the second postoperative day assuming that both vocal cords are working normally. If there is vocal cord paralysis, a longer period without oral intake may be necessary to minimize the risk of aspiration that is particularly likely to occur when the combination of vocal cord palsy, laryngeal edema, and restriction of laryngeal elevation secondary to the resection coexist. Oral intake is generally begun with Jell-O, which is more easy for the patient to handle than liquids, and which, if aspirated, is nonirritating and easily coughed up. Once the patient has demonstrated the ability to swallow Jell-O without difficulty the diet is advanced. The patient is usually ambulatory by the first postoperative day. The chin stitch is left in place for 1 week and removed prior to the patient's discharge. In the absence of any concern regarding upper airway compromise, postoperative bronchoscopy is not routinely employed prior to discharge.

COMPLICATIONS

The most common complications following tracheal resection include vocal cord dysfunction, superficial wound infection, and anastomotic complications that include both early separation of the anastomosis and late recurrent stenosis. Obstructing granulation tissue is not uncommon and may be treated with endoscopic cauterization, bronchoscopic debridement, or balloon dilatation.

The wound should be left undressed starting on the first postoperative day and inspected once or twice daily. At the earliest signs of wound infection, the neck incision should be partially opened and packed to avoid deep infection, which can affect the suture line. Wright et al. reviewed the results from the Massachusetts General Hospital following tracheal resection in 901 cases over a 20-year period primarily mainly comprising the experience of *Grillo*. Successful results were reported in 95% of patients with anastomotic complications occurring in 9% and with an overall perioperative mortality of 1.2%. The risk factors found to be associated with anastomotic complications included diabetes, the need for reoperation, resections >4 cm, the need for a tracheostomy prior to operation, and laryngotracheal resection. Anastomotic complications, especially stenosis and granulation tissue, may resolve with repeated dilatation but if a tight stricture ultimately results, then insertion of a silicone T-tube or the use of a tracheostomy tube distal to the anastomosis probably should be placed. If anastomotic narrowing of significant degree occurs in the first month or two injections of steroids seems to be of limited value, though one or more anastomotic dilatations coupled with a course of oral steroids for several weeks, may allow for resolution. In our experience, the most challenging and complicated procedures tend to be in those patients with postintubation injury to the subglottic region resulting in total or near-total

obliteration of the airway at the level of the cricoid cartilage. Such patients have multiple risk factors, as identified by *Wright*, including major comorbidities associated with the original need for the ventilator assistance that led to the airway injury in the first place. Most of these patients have had a tracheostomy tube placed to provide a safe airway in advance of the laryngotracheal resection. In more than half of such patients, a suprahyoid laryngeal release was required and a protective tracheostomy was also placed at the completion of the resection. One patient required a T-tube. In this challenging group, 11 of 14 patients had an excellent outcome with one postoperative death.

Another group of patients at higher than usual risk for postoperative complications are those with idiopathic subglottic stenosis who have been subjected to repeated dilatation and/or laser procedures resulting in extensive scarring in the subglottic region. It is for this reason that laryngotracheal resection is advised after one or two endoscopic procedures at most have been attempted, especially if the interval between the intervention and the subsequent recurrence of symptoms is limited.

CONCLUSION

Postintubation stricture is the most common indication for tracheal resection. This is especially true if there has been traumatic initial intubation, emergency tracheostomy, or a high tracheostomy, which is adjacent to, or involves the cricoid cartilage either by inadvertent malpositioning or because the weight of the patient, curvature of the spine, or other physical factors make it difficult or impossible to place a tracheostomy tube in the usual position.

Resection of postintubation strictures should be delayed until the patient is in optimal condition using such temporizing maneuvers as repeat dilations, insertion of a T-tube, or use of a tracheostomy tube. The importance of a skilled thoracic anesthesiologist as a member of the operative team cannot be overemphasized. Generally, excellent outcomes with low morbidity and mortality rate can be achieved.

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EDITOR'S COMMENTS

The average thoracic surgeon during an entire career will likely see very few patients requiring tracheal resection and thus expertise is hard to develop. A few centers and, for that matter, a few surgeons have dominated this area and we must pay homage to both the late Hermes Grillo and to F. Griffith Pearson for their significant contributions to this field. Dr. Cooper himself also has made significant contributions to surgery of the trachea having worked with both Drs. Grillo and Pearson.

The diagnosis of a tracheal obstructing lesion often is delayed with the patient being told either by their primary care physician or a pulmonologist they have "asthma." The key is recognizing that these patients have a wheeze with inspiration, a finding that differentiates the problem from reversible airways disease. A simple flow-volume loop essentially makes the diagnosis, and patients should not have to experience almost complete airway obstruction before a diagnosis is made. Patients who present with an obstructing lesion can be temporized by simple debulking of the tumor done with the rigid bronchoscope. This assumes, of course, that the thoracic surgeon is facile with rigid bronchoscopy, an art that I am afraid is not being taught to any great extent in most residency programs today.

Tracheal stenosis resulting from prolonged endotracheal intubation is usually related to ischemia caused by an overinflated tracheal cuff. More attention is being paid to cuff inflation and we are more aggressive about performing tracheostomy in the patient requiring prolonged ventilator support so fewer of these lesions are being seen. Essentially, all of these are located in the upper to mid-trachea and can be approached via a cervical incision. As pointed out, care must be taken to preserve the segmental blood supply to the trachea by only circumferentially mobilizing in the area to be resected. Often the stenosed area must be defined bronchoscopically and a needle placed for localization to guide the resection. Sufficient trachea must be resected so that normal tracheal lumen is present. The inexperienced tracheal surgeon may be hesitant to take the amount of trachea required because of fear of not being able to put the airway back together so a stenotic portion is left, a result that is doomed to failure. Any surgeon who undertakes a tracheal resection must be familiar with the various release procedures as well as limits of resection.

Subglottic tracheal stenosis requires special expertise and really should only be undertaken by those experienced in these resections. Often the collaboration with a head and neck surgeon facilitates the dissection especially if a laryngofissure is required for optimal exposure. Injury to one or both recurrent laryngeal nerves can be disastrous and must be avoided at all costs but adequate resection must be accomplished as a revision procedure is fraught with a higher incidence of morbidity. Working in the subglottic area is not for the faint of heart.

Distal tracheal resection, as noted, may be approached via right thoracotomy or median sternotomy and the decision rests with surgeon experience as well as lesion location. Both approaches allow excellent exposure of the carina but in my

(*continued*)

EDITOR'S COMMENTS (*continued*)

experience the right thoracotomy offers somewhat better exposure to the proximal left main bronchus.

I favor using an armored tube and sterile anesthesia tubing ventilating periodically once the airway is divided over the use of jet ventilation. There is usually ample time dictated by the oxygen saturation level to allow for suture placement during the period when the patient is not being ventilated. Too many times I have seen the jet ventilation catheter propel itself out of the airway for me to be comfortable with that technique.

The placement of the chin stitch remains an important adjunct preventing the patient from significantly extending the neck. The stitch should be loosely placed only serving as a reminder, not something punitive.

LRK

10 Carinal Resection

Elizabeth A. David and Garrett L. Walsh

INTRODUCTION

Carinal resections are some of the most technically challenging procedures in thoracic surgery and should be approached with great attention to detail. The preoperative evaluation should include a complete medical workup, oncologic staging, and careful operative planning from the surgical and anesthesia perspectives. Of paramount importance is the creation of a secure, well-vascularized anastomosis under minimal tension. A vast majority of carinal resections are performed for malignancies, but occasionally benign conditions can result in critical airway obstruction that mandates a carinal resection. Despite the high incidence of morbidity associated with these procedures, 1-year survival for malignant disease as high as 75% has been reported. Survival is significantly shorter in patients with unresectable lesions and is usually measured only in months.

PATIENT EVALUATION

The workup of a patient with a carinal mass is not dissimilar from the workup for routine lung cancer patients. Involvement of the carina by lung cancer histology is considered a T4 lesion. Careful attention to a complete oncologic staging as well as appropriate medical and cardiopulmonary clearance is mandatory. Patients should undergo cardiac clearance and pulmonary function testing especially in situations in which concomitant pulmonary parenchymal resection will be required. Consideration of comorbid conditions and assessment of nutritional status is required. Many patients with tracheal lesions that result in respiratory compromise are placed on steroids during their diagnostic workup and it is important to wean the patients either completely off steroids or to as low a dose as possible prior to the elective resection and reconstruction. Smoking cessation is also important to optimize healing of the airway anastomoses and facilitate secretion clearance in the postoperative period. Common diagnoses associated with carinal resections are listed in Table 10.1.

Computed tomographic (CT) scans are an essential part of the evaluation of tracheal lesions in patients who may require a carinal resection. CT provides information regarding the estimated length of the tracheal wall involvement and the endoluminal and extraluminal extent of disease. CT imaging can evaluate the size, shape, and number of mediastinal lymph nodes in the paratracheal, subcarinal, and supraclavicular stations. Pulmonary nodules suspicious for metastatic disease can also be detected. If available, 3D reconstructions based on CT images may help with operative planning (Fig. 10.1A and 10.1B). We use total body positron emission tomography (PET) fused with CT (PET/CT) and magnetic resonance imaging (MRI) of the brain to complete the staging workup for distant metastatic disease. The finding of distant metastatic disease, of course, is a contraindication to resection.

Bronchoscopy is a mandatory component of the evaluation of patients being considered for a carinal resection. Flexible bronchoscopy can be used to biopsy the lesion as well as assess the proximal and distal extent of the airway involvement. Rigid bronchoscopy using the tip of the bronchoscope to core through the tumor is useful to debulk obstructing lesions in a patient who presents with critical airway obstruction. This can quickly reestablish a patent airway with minimal bleeding in most cases. Lasers, argon plasma coagulators, and electrocautery techniques can also be used particularly for the more vascular tumors to control any bleeding if this should be encountered. Rigid bronchoscopy is also helpful to perform detailed measurement of the lesion, as well as assessing tumor fixation to the tracheal wall and overall tracheal mobility in relation to other mediastinal structures.

Mediastinal staging is required in these patients. N2 or N3 disease is difficult to characterize because of the central nature of these lesions, but we usually assign laterality based on the predominant involvement of the main stem bronchi. Patients demonstrating involvement of the mediastinal lymph nodes most commonly are treated with nonsurgical modalities (T4N2/N3 stage IIIB disease).

Endobronchial ultrasound (EBUS) offers additional diagnostic information in these patients. We use EBUS to obtain samples of lymph nodes for staging purposes at levels 2, 4, 7, and 11. In some cases we have also employed radial probe ultrasound to assess both the depth and extent of tracheal wall invasion.

We reserve the use of mediastinoscopy at the time of formal resection and favor EBUS for diagnostic purposes in the preoperative period. We do not recommend mediastinoscopy and a delayed carinal resection, as scar tissue that may compromise tracheal mobility at the time of resection will develop in the pretracheal plane in the time between the two procedures.

ANESTHETIC MANAGEMENT

A successful carinal resection requires excellent communication between the thoracic surgeon and an experienced thoracic anesthesiologist. For the safe conduct of the procedure, the anesthesiologist must be facile with multiple modes of ventilation, the performance of intraoperative bronchoscopy, and be committed to the planned extubation of the patient in the operating room suite immediately at the completion of the procedure. Immediate postprocedure extubation should be the goal of all team members; therefore, short-acting agents and careful fluid management are particularly important especially in situations where lung parenchyma resection is also planned, as in a carinal pneumonectomy.

The airway should be managed initially with an extra-long reinforced single-lumen tube that is placed with fiberoptic guidance. This will minimize trauma to

Table 10.1 Common Diagnoses Requiring Carinal Resection

Malignant	Benign
Squamous cell carcinoma	Traumatic injury
Adenoid cystic carcinoma	Inflammatory stricture
Adenocarcinoma	
Mucoepidermoid carcinoma	
Carcinoid	

the airway and to the tumor itself so as to avoid hemorrhage into the distal airway. The extra-long tube (usually #7) provides a significant advantage in that it can be positioned in the trachea or bronchoscopically guided into the left or right main stem bronchus depending on the tumor location and the operative approach. The use of a double-lumen tube is not recommended as the tube is bulky and not easily maneuvered intraoperatively.

Prior to any incision in the airway, a focused discussion and simulation of the airway management between the surgeon and the anesthesiologist will ensure patient safety. A sterile reinforced endotracheal (ET) tube with extension tubing and appropriate and tested connectors is passed from the surgical field to allow for cross-table ventilation, prior to an airway incision. Once the airway has been opened, the sterile tube is placed into one of the main stem bronchi for ventilation. Throughout the resection and creation of the anastomosis it may be necessary to intermittently remove the ET tube from the airway to facilitate suture placement. During these periods of intermittent ventilation, hand ventilation by the anesthesiologist may be the best way to maintain oxygen saturation, prevent hypercarbia, and avoid barotrauma to the ventilated lung. In patients who do not tolerate intermittent ventilation, it may be necessary to use high frequency jet ventilation techniques. The surgeon and anesthesiologist, however, must be aware of the risks of air trapping and barotrauma associated with this method of ventilation.

In patients who present emergently with near-obstructing tracheal lesions, the use of cardiopulmonary bypass (CPB) or extracorporeal membrane oxygenation (ECMO) systems has been described. Perfusion cannulas can be placed under local anesthesia prior to the induction of the general anesthetic in these patients who present in extremis. Centers who have described these techniques show comparable morbidity and mortality to more traditional approaches. The use of rigid bronchoscopy to core out and debulk the tumor, however, in most cases will eliminate the need and risks of CPB and will convert an emergent procedure into an elective resection. This is a lifesaving skill that all general thoracic surgeons need to acquire.



Fig. 10.1. (**A**) Coronal computed tomographic (CT) chest image demonstrating a carinal mass. (**B**) Three-dimensional reconstruction of CT chest of carinal mass.

OPERATIVE TECHNIQUE

Intraoperative Evaluation and Approaches

At the time of the resection, bronchoscopy is used to reassess the lesion and its resectability based on the total length of

required airway resection. In cases of cervical tracheal resections, it may be possible to resect up to 5 cm of the airway. We do not recommend greater than a 4-cm resection for more distal lesions, as this may result in excessive tension on the anastomosis. For lesions approached through a right thoracotomy, mediastinoscopy can be done at the time of the resection. This serves several purposes. First, mediastinal lymph nodes can be assessed by frozen section analysis. Second, the pretracheal tissue plane can be mobilized up to the level of the cricoid cartilage, a move that facilitates tracheal mobilization. Additionally, the left recurrent laryngeal nerve can be identified and mobilized away from the left side of the trachea, which may minimize the risk of nerve injury when the trachea is approached from the right chest.

Tumors in the distal trachea, right main stem bronchus, and left main stem bronchus can be accessed through both the right chest and via the midline. The operative approaches to these tumors, therefore, are typically via a right posterolateral thoracotomy through the fourth or fifth intercostal space or through a median sternotomy. Although not contraindicated, approaching these tumors from the left chest can be challenging because of the location of the aortic arch and left pulmonary artery (LPA), both of which require mobilization to expose the carina. The left recurrent laryngeal nerve as it courses around the ligamentum arteriosum must be protected from injury and stretch during the mobilization of these structures.

Exposing the trachea through a median sternotomy requires mobilization of the ascending aorta to the patient's left, the right pulmonary artery (RPA) caudally, and the superior vena cava (SVC) to the patient's right. If at any time during the operation the patient becomes hypoxic, hypercarbic, or hypotensive, several physiological factors may be contributing to the clinical deterioration. Venous return may be compromised by excessive SVC retraction, cardiac output may be impeded by aortic retraction, and shunting may occur when the right lung is ventilated but the RPA is compressed resulting in a complete ventilation/perfusion mismatch. It may be necessary to adjust the retractors if hemodynamic or ventilatory compromise develops.

The initial dissection should include clearance of the subcarinal space of lymphatic tissue to facilitate exposure of the carina. Preservation of the blood supply to the lateral aspects of the main stem bronchi must be considered during this lymphadenectomy. If the lymph nodes have been previously sampled by EBUS, mediastinoscopy may not be required for cases approached with a median sternotomy. Blunt anterior dissection along the trachea, right main stem bronchus (RMB), and left main stem bronchus (LMB) is accomplished with ease through a median sternotomy. Mobilization of the membranous trachea can also be achieved with this approach, but posterior mobilization should take place after the airway has been transected. This mobilization is necessary to free the esophagus and right vagal nerve from the posterior aspect of the airway; however this dissection must be done judiciously to avoid injury and devascularization of the membranous trachea, which is perfused through small shared, esophageal branches. Preservation of the soft tissues along the lateral aspect of the trachea, RMB, and LMB is key to preserving blood supply to the surgical anastomoses; therefore blunt dissection and mobilization must be restricted to the anterior and posterior aspects of the airway (Figs. 10.2 and 10.3).

From a right thoracotomy, the division of the azygos vein can facilitate exposure to the trachea, RMB, and LMB. It is rarely necessary to retract the RPA when the approach is from the right side of the chest, therefore problems with shunt physiology during intermittent ventilation are less frequently encountered (Fig. 10.4; Table 10.2).

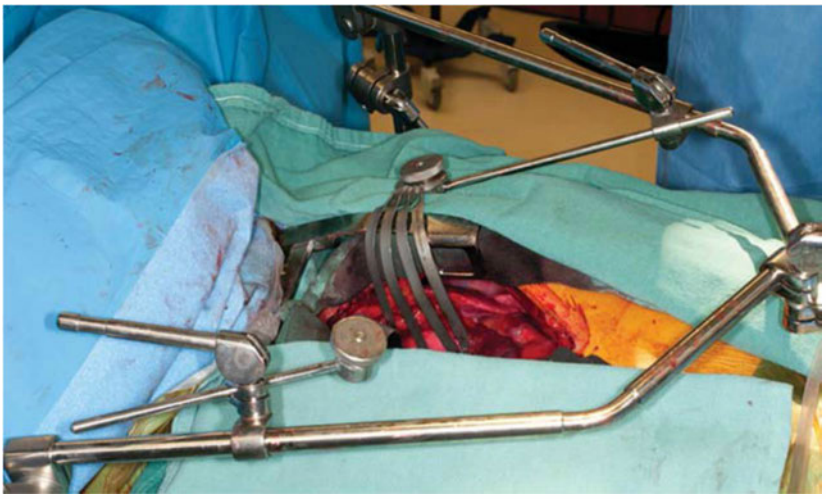


Fig. 10.2. Retractor setup for median sternotomy. A small Finochietto is placed in the superior aspect of the wound to assist with sternal retraction. The Thompson retractor is used to retract the great vessels and allow access to the trachea.

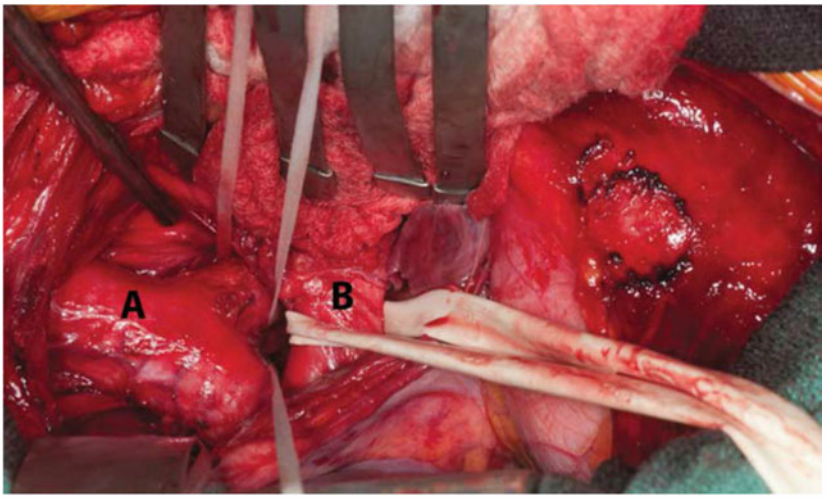


Fig. 10.3. View of trachea from median sternotomy, from the surgeon's view. Trachea is marked "*A*" and right pulmonary artery is marked "*B*." Umbilical tapes isolate the right and left main stem bronchi. A Penrose drain is used to retract the right pulmonary artery. The aorta and superior vena cava are retracted out of view.

Release Maneuvers

The surgical tenants of preserved blood supply and minimal anastomotic tension are critical for healing of airway anastomoses. Several release maneuvers have been described as adjuncts for airway surgery including neck flexion, suprahyoid release, mobilization of the pretracheal fascia, hilar release, and division of the inferior pulmonary ligament. All of these maneuvers, with the exception of the suprahyoid release, are beneficial in the setting of a carinal resection. The last three are the most important to minimizing tension on the tracheal anastomosis. The hilar release is described as a U-shaped incision in the inferior pericardium at the level of the inferior pulmonary vein, which allows a cephalad shift of all of the hilar structures. It is important to remember to release the raphe on the lateral aspect of the inferior vena cava (IVC), which will permit further hilar movement. The pericardial incision must be placed immediately posterior to the phrenic nerve coursing along the superior pulmonary vein and around the inferior pulmonary vein. Direct or cautery injury to the phrenic nerve must be avoided at all costs (Fig. 10.5).

Table 10.2 Operative Approaches to Carinal Resection

Right thoracotomy	Median sternotomy	Left thoracotomy
• Access to trachea, RMB, and LMB	• Access to both right- and left-sided pulmonary parenchyma	• Limited by aorta, LPA
• Cannot access left pulmonary parenchyma or inferior pulmonary ligament	• Can divide bilateral inferior pulmonary ligaments	• Close proximity to recurrent laryngeal nerve
• May require mediastinoscopy to mobilize pretracheal plane	• No need for mediastinoscopy	• Limited access to right-sided airway and pulmonary parenchyma
• Requires division of azygos vein	• Release retraction on RPA, if shunting occurs intraoperatively	• May require mediastinoscopy to mobilize pretracheal plane
• Can harvest intercostal muscle flap	• Cannot harvest intercostal muscle flap	• Cannot divide inferior pulmonary ligament on right side
	• Can harvest omentum or pericardial fat pad	• Can harvest intercostal muscle flap

Resection

Prior to the division of the airway, it is important to ensure that the airways have been mobilized anteriorly and posteriorly with preservation of the lateral blood supply and that the hilar release maneuvers have been completed. We identify our point of division either by palpation of the lesion or using bronchoscopic guidance. A needle can be placed through the tracheal wall and visualized bronchoscopically from the inside of the tracheal lumen to determine the optimal placement of the initial incision in the airway. Once the line of division is decided, we ensure that all equipment and team members are prepared for cross-table ventilation. Ventilation is stopped and the trachea is divided with a knife and the lesion is visualized. One must never enter an airway with electrocautery as an airway fire can result because of the high oxygen concentration. We place lateral airway traction sutures completely around a cartilaginous ring using 2-0 Vicryl or polydioxanone suture (PDS) to prevent the suture from pulling through with airway manipulation. The RMB and LMB are then divided. The specimen is removed and cross-table ventilation is initiated by placement of the reinforced ET tube into the RMB or LMB (Fig. 10.6). The specimen can be sent for frozen section analysis of

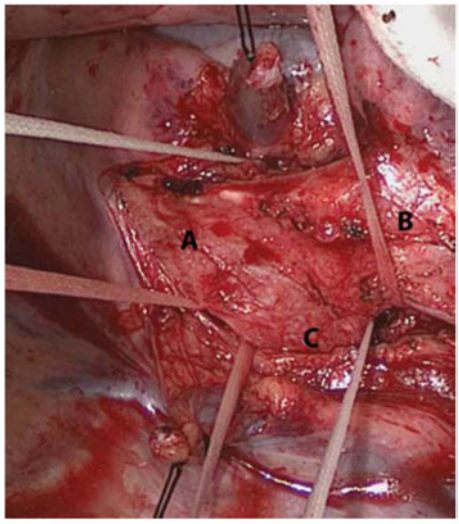


Fig. 10.4. View of trachea from *right* thoracotomy from surgeon's side. "*A*" trachea, "*B*" right main stem bronchus, "*C*" left main stem bronchus. The azygos vein has been divided to facilitate dissection and mobilization.

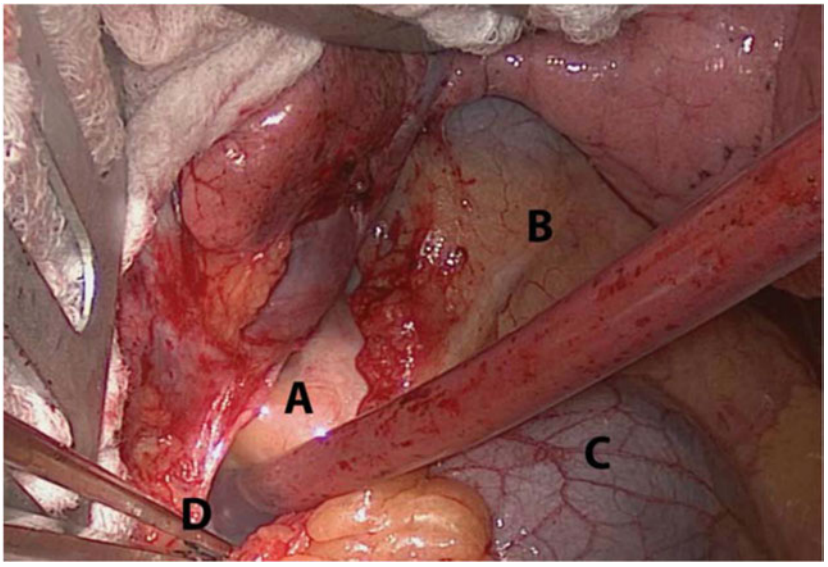


Fig. 10.5. Inferior view of hilar release. A U-shaped incision is created in the pericardium around the pulmonary veins. The lateral raphe from the inferior vena cava is released. Both of these maneuvers permit additional hilar mobility to minimize the anastomotic tension. "*A*" IVC, "*B*" Phrenic nerve, "*C*" Pericardium-covered RV, "*D*" incised pericardium.

the margins or alternatively the initial resection cuts are placed very close to the tumor and three additional cartilaginous rings are removed and sent separately for margins from the trachea, RMB, and LMB (Fig. 10.7). The surgeon must always be cognizant of the importance of leaving sufficient airways intact to permit a safe reconstruction yet the attainment of a negative margin is the goal, if possible.

The patient should be physiologically stable to allow a detailed and controlled assessment of pathologic margins to determine if additional airway resection is necessary depending on histology and length of airway defect. In cases of adenoid cystic carcinoma, gross tumor resection and reconstruction of the airways are the goals of the resection. It is not uncommon for this histology to track submucosally and have microscopic positive margins. Patients with positive margins can be treated with adjuvant radiation therapy with excellent long-term survival.

Reconstruction

Once the specimen has been removed and margins have been assessed, the operative focus must shift to creating an airway anastomosis that is well vascularized and under minimal tension. The previously placed lateral traction sutures are used to position the airway and facilitate suture placement. Size mismatches and the tracheobronchial angles must be considered when creating the anastomosis. Intraoperative bronchoscopy may help to avoid the creation of tracheobronchial anastomoses with acute angles that may be difficult to appreciate from the external view. In cases of size discrepancy, the bronchial end should be allowed to intussuscept into the trachea. The anastomosis should be created with interrupted sutures of 3-0 or 4-0 Vicryl or PDS appropriately and evenly spaced. When using Vicryl sutures, we use a drop of sterile mineral oil to lubricate the suture to minimize tissue drag and potential abrasive injury to the delicate tissues, especially the membranous airway. A great deal of surgical literature emphasizes the importance of placing knots on the outside of the lumen. We have not experienced any consequences when placing knots on the inside of the lumen with these absorbable sutures. The integrity of the back wall of the anastomosis is critical because, unlike vascular structures that can be manipulated after the completion of the anastomosis, the airway becomes relatively fixed and it is difficult to rotate or visualize the sites of leaks from the back wall suture line (Fig. 10.9). For this reason we favor tying the knots on the inside of the back wall, which allows for easier suture management, better visualization of the apposition of the mucosal surfaces, and complete assurance that the knot is well seated. In cases where the anatomy is favorable, the membranous portion of the anastomosis may be sewn with a running PDS suture. The anterior wall is generally always performed with interrupted sutures with knots tied on the outside. The end result should be an airway anastomosis that is widely patent, oriented well for ventilation and secretion clearance, and under minimal tension with secure knots. The larger traction sutures are also tied at the angles to help minimize tension on the finer sutures.

In cases of carinal resection, the reconstruction options are several. The RMB and LMB can be joined to create a neocarina, which is then anastomosed to the trachea directly (Fig. 10.8). Alternatively, the RMB can be anastomosed to the trachea directly and the LMB can be anastomosed to the cartilaginous portion of the bronchus intermedius if the length permits. After completion of these anastomoses, we recommend a couple of intraoperative maneuvers to evaluate the integrity of the anastomosis including bronchoscopy and testing for pneumostasis by ventilating under saline (Figs. 10.9 and 10.10).

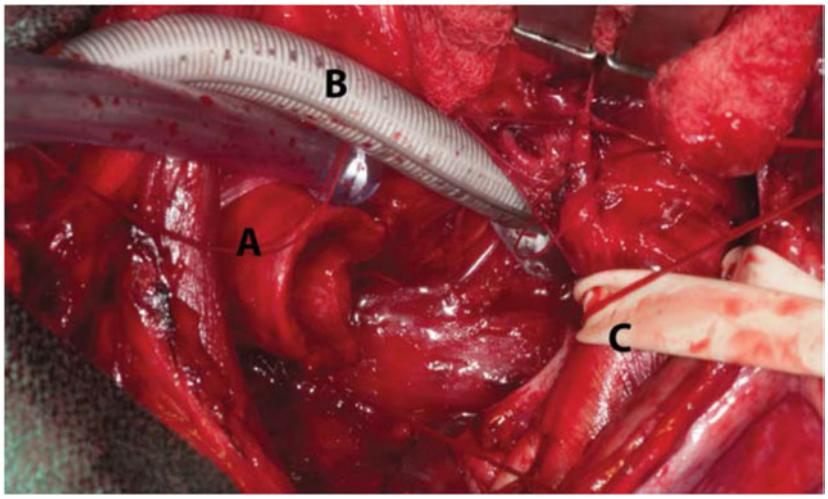


Fig. 10.6. View from median sternotomy. Divided trachea is marked with "*A*." Carina has been resected. Cross-table ventilation is established through the *left* main stem bronchus "*B*." "*C*" marks the right pulmonary artery that is retracted caudally.

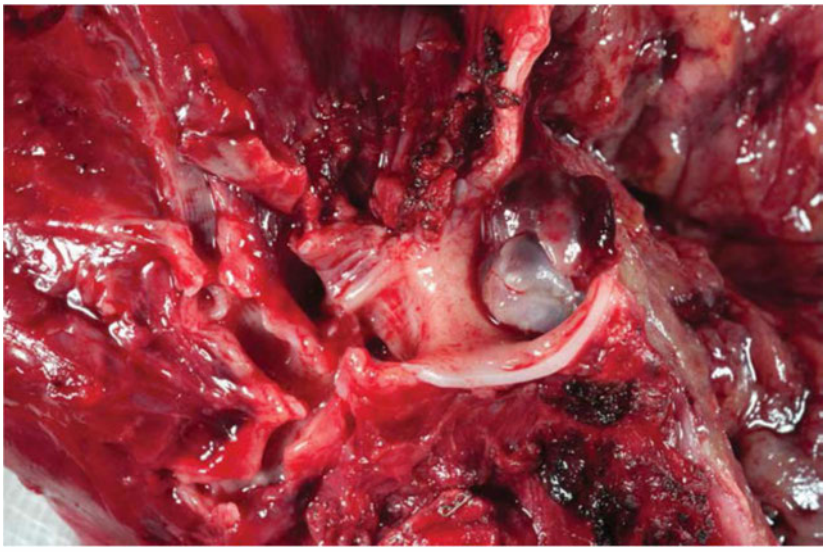


Fig. 10.7. Gross pathology specimen of a right carinal pneumonectomy for a central carcinoid tumor.

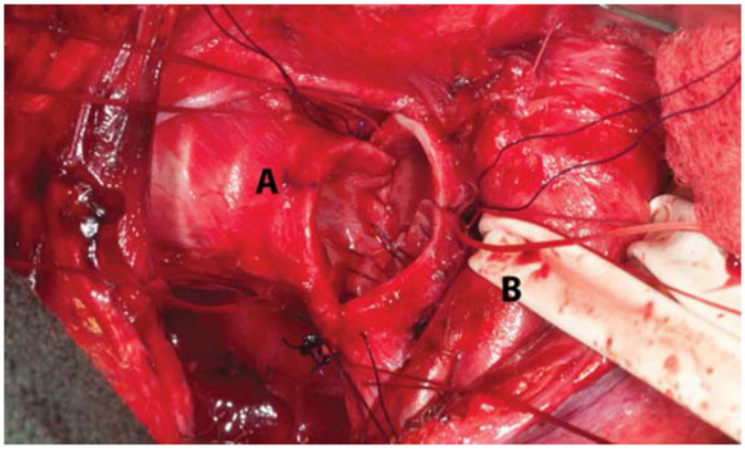


Fig. 10.8. View from median sternotomy. Partially completed anastomosis. A neocarina has been created between the right and left main stem bronchi and is being connected to the trachea. "*A*" trachea, "*B*" right pulmonary artery.

extubation is important to avoid positive pressure on the newly created airway anastomoses.

Aggressive pulmonary toilet with chest physiotherapy, bronchodilators, secretion clearance, and adequate pain control is essential. There should be a low threshold to use flexible bronchoscopy to facilitate pulmonary toileting in patients who are having a difficult time clearing secretions, often secondary to the transient interruption of the mucociliary clearance mechanism. We do not use routine bronchoscopy in patients who are doing well with a clear chest radiograph, but we do perform bronchoscopy prior to discharge to assess the healing of the anastomosis.

Buttress

Anastomotic integrity is of paramount importance during carinal resection as dehiscence can be fatal. We recommend soft tissue buttressing of the airway anastomoses in all cases. We use intercostal muscle flaps for cases approached from a thoracotomy but an omental tissue transfer or a pericardial fat/thymus pad when a median sternotomy is used. All of these soft tissue flaps can be harvested through the operative incision with low morbidity. The flaps should be positioned in such a way as to provide a tissue barrier between the sutures and the adjacent structures (esophagus, pulmonary artery, and aorta). When using an intercostal muscle flap it is important not to wrap the muscle 360 degrees around the airway anastomosis as anastomotic stenosis can result secondary to calcification and ossification of this particular flap which is harvested with the periosteum intact. Omental and pericardial flaps can be wrapped circumferentially around the anastomosis without concern. Pleural flaps are not used routinely as they provide minimal tissue coverage (Fig. 10.11A and 10.11B).

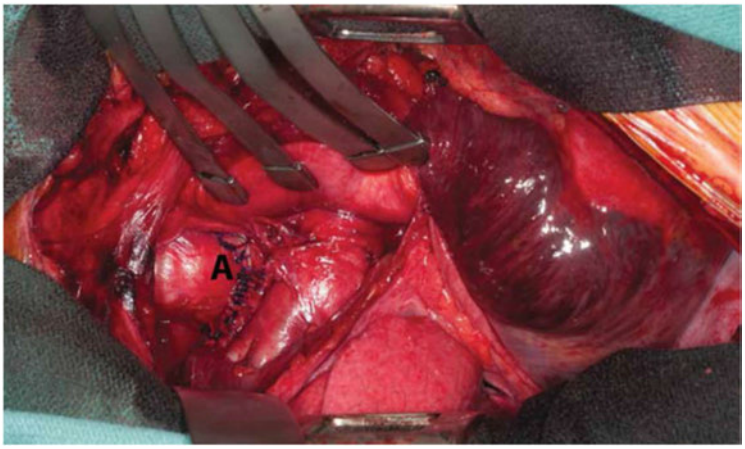


Fig. 10.9. View from median sternotomy. "*A*" marks the trachea. Completed anastomosis between distal trachea and neocarina created from right and left main stem bronchi.

POSTOPERATIVE MANAGEMENT

At the conclusion of the operation we change the ET tube to a laryngeal mask airway (LMA), which permits suction bronchoscopy, a final assessment of the anastomosis and confirmation of movement of the vocal cords as the patient is awakening. Use of an LMA permits a smooth anesthetic reversal, minimal coughing, and therefore minimal pressure on the anastomosis. As mentioned earlier, postprocedure

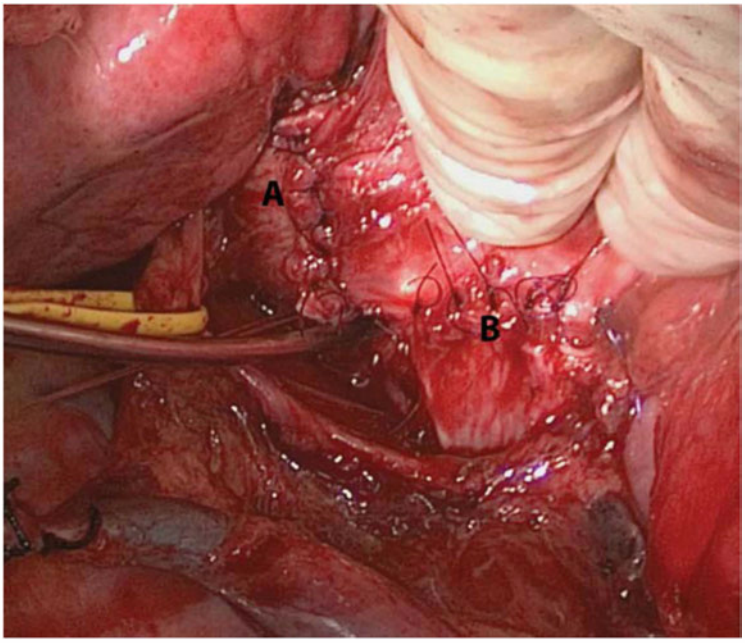


Fig. 10.10. View from right thoracotomy. Completed anastomosis between distal trachea and right main stem bronchus (**A**). Completed anastomosis between left main stem bronchus and bronchus intermedius (**B**).

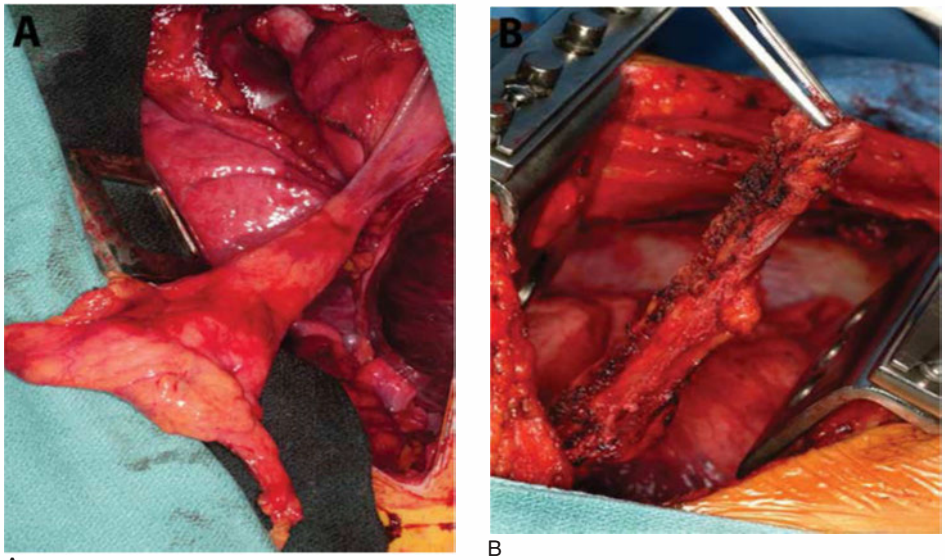


Fig. 10.11. (**A**) Mobilized pericardial fat pad to be used as a pedicled flap to provide a buttress for the anastomosis. (**B**) Intercostal muscle flap for use as a pedicled flap to buttress the airway anastomosis.

Postoperative complications after carinal resection include atrial dysrhythmias, atelectasis, pneumonia, respiratory failure, recurrent laryngeal nerve paresis/paralysis, anastomotic ischemia, structuring, or partial or frank dehiscence. Nasogastric tubes can be considered in patients who have undergone a major pulmonary resection in combination with airway reconstruction, as aspiration in this setting can be fatal. However, we do not routinely use them in patients who have parenchymal-sparing airway resections and have good function of both vocal cords to protect their airway. We are conservative in initiating an oral diet until postoperative ileus has completely resolved.

If an anastomotic stricture does develop, many of these stenoses are amenable to early dilation with a rigid bronchoscope and placement of a removable silicon stent. This management is similar to that used to deal with an airway stricture following a lung transplant.

OUTCOMES

While carinal resections are technically demanding, excellent clinical outcomes can be achieved in high-volume centers. Thirty-day mortality rates range from 2% to 5% in most series, but can be as high as 10%. Perioperative morbidity approaches 40% in experienced centers, similar to most major general thoracic procedures. Early anastomotic complications are associated with a high mortality and must be avoided. If there is a suspicion of early anastomotic dehiscence (fever, continuous air leak, hemoptysis), bronchoscopy should be performed and there should be a low threshold for reexploration and revision of the anastomosis, if necessary.

The overall survival for patients with malignant histologies undergoing carinal resection is approximately 25% to 50% at 5 years. The need for postoperative mechanical ventilation and development of anastomotic complications are predictors of decreased overall survival on univariate analysis. Nodal status and degree of endobronchial tumor extension are predictors for decreased disease-free survival on multivariate analysis. Survival at 5 years in patients with involved mediastinal lymph nodes (N2) is 7% versus 37% for patients who are N0/N1. Locoregional control is the best option for long-term survival in patients with primary malignancies involving the carina; however, resection must be thoughtfully considered in patients with known mediastinal nodal disease since long-term survival is so poor.

FUTURE DIRECTION

Although carinal resection has been well established as a treatment modality for airway tumors since the 1950s, we have made slow progress to overcome the limitation of 4-cm resections for safe reconstructions. Tracheal and carinal replacement with aortic allografts may provide one method of overcoming the 4-cm barrier. The allografts, some fresh and some cryopreserved, require silicone stents to provide rigidity. Extensive soft tissue flap coverage including pectoralis major muscle, omentum, or pericardial fat pad is necessary to prevent fistulization but no immunosuppression is required. Early results are promising in animal and human studies.

Complete tracheal replacement with a tissue-engineered nanocomposite graft has also been successfully performed in humans. The nanocomposite scaffolding is seeded with autologous stem cells for 36 hours prior to implantation into the recipient. No immunosuppression is required. Early results are promising, but at the time of this writing this procedure has not been performed in the United States. This may provide an excellent alternative therapy for patients who have tumors that extend beyond the 4 to 5 cm length restrictions of present surgical techniques.

SUMMARY

Carinal resection is a high-risk procedure that offers acceptable clinical outcomes for patients who are appropriate candidates. Preoperatively, patient factors such as smoking and steroid cessation and optimization of physiologic and nutritional parameters help minimize perioperative morbidity and mortality. For malignant lesions, every effort must be made to exclude distant metastatic disease and the surgeon must weigh the benefit of resection in patients with regional nodal metastasis, as the long-term outcomes are quite poor. Once patient factors are optimized, technical aspects of the operation must include preservation of the lateral blood supply of the airways and utilization of release maneuvers to allow for the creation of an anastomosis under minimal tension. Immediate postoperative extubation in the operating room, aggressive pulmonary toilet, and early ambulation are crucial components to achieving a successful outcome for these patients.

ACKNOWLEDGMENTS

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EDITOR'S COMMENTS

As the authors correctly point out, perhaps the single most important determinant of successful outcome following carinal resection is patient selection. Resectable tumors involving the carina are extremely rare as most of the squamous carcinomas have significant extraluminal extension that precludes resection. Adenocystic carcinomas are more likely to present as resectable lesions but because of tracking along submucosal nerves the likelihood of achieving a negative resection margin is low, but with postoperative radiation therapy the long-term results remain acceptable. Mediastinal nodal involvement is associated with very poor long-term results and likely the operation should not be offered.

Whether one chooses a right thoracotomy approach or median sternotomy depends somewhat on the location of the lesion and whether or not parenchymal resection is required. Lesions involving the carina and the left main bronchus present a particular challenge since approaching the carina from the left is extremely difficult. These lesions are best approached via sternotomy with left pneumonectomy able to be carried out in addition to allowing for reconstruction with anastomosis of the right main bronchus to the distal trachea. Alternatively, a right thoracotomy allows for excellent carinal mobilization and resection but if the left lung needs to be removed a left thoracotomy would also be required.

Reconstructing the airway following carinal resection can be challenging when one lung is not removed. Because of the aortic arch there is limited mobility of the left main bronchus and depending on the length of distal trachea resected trying to reconstruct the airway by fashioning a "neocarina" can be quite difficult. The more common reconstruction involves a distal trachea to right main bronchus anastomosis with the left main bronchus attached to the lateral cartilaginous portion of the right main bronchus. An anastomosis that is under minimal tension is critical for success. Anastomotic dehiscence is a disastrous complication and commonly results in mortality. If there is any question in the early postoperative course regarding anastomotic integrity, a bronchoscopy needs to be performed and potentially reoperation with repair of the anastomosis. Aggressive secretion management in the postoperative period is also critically important. Bottom line: these are high-risk, technically challenging surgical procedures with a high likelihood of major morbidity. It probably is safe to say that carinal resections should be limited to those centers that have significant experience in dealing with surgery of the airway.

LRK

11 Video-Assisted Thoracoscopic and Robotic-Assisted Pulmonary Resections

Eugenio Pompeo, Thomas A. D'Amico, and Abbas E. Abbas

INTRODUCTION

Video-assisted thoracic surgery (VATS), also termed thoracoscopic surgery, refers to minimally invasive chest surgery that avoids rib spreading and rib resection and relies entirely on cameras and video technology for visualization. Thoracoscopic techniques may be applied to a variety of thoracic procedures, but thoracoscopic pulmonary resections are the most commonly performed. Despite the potential advantages of minimally invasive surgery, only approximately 30% of major pulmonary resections are currently completed using the thoracoscopic technique. The strategies and outcomes of thoracoscopic pulmonary resections are described, with a focus on anatomic procedures.

THORACOSCOPIC STRATEGIES

Thoracoscopic techniques may be applied to virtually all pulmonary resections, from wedge resection to pneumonectomy. The indications for thoracoscopic pulmonary resection are similar to those for resections performed using an open approach. The only absolute contraindication to thoracoscopic resections is the inability to achieve a complete resection or reconstruction. Relative contraindications include inability to maintain single-lung ventilation and tumor size, which may preclude a minimally invasive approach to extraction. Prior thoracic radiation, prior thoracotomy, prior resections, and adhesions are no longer considered absolute contraindications, and resection in these patients using minimally invasive techniques can be performed by experienced surgeons.

Several surgical strategies have been employed to perform an anatomic lobectomy, utilizing a variety of incisions to facilitate dissection. We believe that the thoracoscopic strategy described below provides for equivalent oncologic results compared to an open strategy and minimizes perioperative and postoperative complications.

BASIC PRINCIPLES

Single-lung ventilation is required and may be achieved with a dual-lumen endotracheal tube or a bronchial blocker. The patient is placed in the lateral decubitus position. It is helpful to limit the tidal volume to increase the space within the thorax, and pressure-controlled ventilation is preferred. Most thoracoscopic anatomic resections may be performed via two or three incisions, and the overwhelming majority in our experience has been performed with only two incisions. Wedge resections should require no more than two incisions.

The placement of port incisions vary according to the preference and experience of the surgeon. In general, the port positions are the same whether an upper, lower, or middle lobectomy is performed, as this strategy places the access incision over the major fissure. The first port, placed in the 7th intercostal space in the midaxillary line, is used predominantly for camera placement and, ultimately, chest tube placement. The second incision (4 to 6 cm) is placed in the 5th or 6th intercostal space in the anterior axillary line. This site is chosen, in part, to allow easy access to hilar structures and to allow for extraction of the specimen (Fig. 11.1).

Instrumentation is important when performing thoracoscopic pulmonary resection, including the use of a 30-degree videoscope and long, curved instruments to improve retraction and dissection. High-definition video equipment improves visualization for difficult dissections. Linear staplers are used to control and divide lung parenchyma, vessels, and bronchus.

Once access to the chest has been achieved, thoracoscopic examination is undertaken. The lung parenchyma is assessed for presence of a mass, additional disease, metastatic disease, and adhesions. For most anatomic procedures (described in detail below), the specific pulmonary vein is the first structure to be divided, and complete hilar dissection usually performed prior to parenchymal division. Advantages of this hilar dissection technique include prevention of postoperative air leaks and minimization of back and forth retraction: dissection for an upper lobe, for example, begins anteriorly and progresses in the posterior direction only.

Most of the hilar dissection may be performed bluntly, with either a dissecting instrument or a thoracoscopic suction device, which also keeps the field dry during dissection. At the conclusion of the dissection, the fissure is completed with the stapling device and the specimen is removed using a protective bag. Mediastinal lymphadenectomy is subsequently performed, although this may be done prior to hilar dissection at the surgeon's discretion.

In addition to the dissection strategy which varies according to the lobe being resected, the surgeon should have a planned strategy for conversion if bleeding is encountered or if there is failure to progress with the dissection thoracoscopically. Usually, conversion is best achieved by extending the anterior access incision to an anterior thoracotomy, as opposed to a separate posterolateral thoracotomy. It is important to note that most of the bleeding encountered can be controlled with direct pressure using a sponge stick, and that conversion need not be performed emergently.

WEDGE RESECTIONS

Thoracoscopic wedge resections may be performed for various indications, both diagnostic and therapeutic. In general, wedge resections are technically straightforward, and the most difficult aspect of the procedure may be the localization of the nodule to be resected. Localization may



Fig. 11.1. Port placement. Camera Port: 1.5 cm incision in the 8th intercostal space. Access Incision: 4.5 cm incision anteriorly in the 5th intercostal space. *Dotted line*, Anterior superior iliac crest.

be particularly challenging when the tumor is less than 1 cm in diameter and not immediately subpleural, or when numerous wedge resections are required, such as for pulmonary metastasectomy. Tumor localization techniques include local injection of radiotracer, wire hook and coil markers, radiopaque markers using intraoperative fluoroscopy, and navigational bronchoscopy. However, other investigators have demonstrated that successful localization may be achieved in the majority of patients without these techniques.

Metastasectomy has become the standard of care for patients with secondary lung tumors. Complete macroscopic resection is the most important prognostic factor since patients with untreated metastatic disease having a median survival of less than 10 months and a 5-year survival rate of less than 5%. Although still controversial, thoracoscopic techniques are becoming more frequently utilized for the resection of pulmonary metastases, with equivalent efficacy and superior quality-of-life outcomes, as compared to thoracotomy.

LOBECTOMY

Right Upper Lobectomy

Once the right hemithorax has been entered, the lung is retracted medially and dissection along the posterior pleura is carried out at the level of the bronchial bifurcation, which will facilitate bronchial dissection later from the anterior approach. The lung is then reflected posterior to allow dissection of the superior pulmonary vein. Although uncommon, the presence of a common pulmonary vein must be excluded. Dissection is performed to identify the bifurcation of the upper and middle lobe veins. Once the upper lobe vein has been clearly identified, it is circumferentially dissected free and divided with a vascular stapler. This reveals the underlying pulmonary artery. In a similar fashion, the pulmonary arteries to the upper lobe are mobilized and divided, beginning with the truncus anterior. The last structure to be dissected is usually the bronchus; however, occasionally the bronchus is divided prior to dissection of the posterior ascending artery. After dividing the bronchus, the fissures are developed and divided using stapling devices and the specimen is extracted from the chest in a protective bag.

Left Upper Lobectomy

Thoracoscopic left upper lobectomy is performed in a similar fashion to that on the right. Posterior dissection is undertaken first to divide the pleural reflection and to expose the left pulmonary artery as it emerges under the aorta and to identify the posterior artery; as with the right upper lobe, this posterior dissection will greatly facilitate the completion of the hilar dissection from the anterior approach. With the lung retracted posteriorly, dissection is used to identify both pulmonary veins (to ascertain that a common pulmonary vein is not present). The superior pulmonary vein is then encircled and divided, revealing the underlying pulmonary artery and upper lobe bronchus. Dissection of the lymph nodes between the cephalad aspect of the bronchus and the arterial trunk (to the anterior and apical segments) will facilitate the ultimate arterial dissection. The branches of the arterial trunk can now be individually exposed and divided, followed by division of the posterior branch. Bronchial dissection and division is now easily accomplished, followed by division of the lingular arteries. Finally, the major fissure is divided with the stapling device, and the lobe is removed in a protective specimen bag.

Left and Right Lower Lobectomy

There are two basic strategies for lower lobectomy, both of which begin with division of the inferior pulmonary ligament, followed by dissection and division of the inferior pulmonary vein. The preferred method does not involve dissection within the fissure (which is stapled last, as with upper lobectomy). After dividing the vein, attention is directed to the bronchus by retracting the lobe cranially, a perspective not obtained via thoracotomy. A plane is created between the bronchus and the artery by dissecting close to the bronchus, which is then divided. For right lower lobectomy, this dissection is begun at the bifurcation with the middle lobe bronchus, which must be preserved. For left lower lobectomy, the dissection is undertaken at the bronchial bifurcation, after identifying the lingular bronchus. After division of the bronchus, the arterial trunk is then encircled and divided, although it is sometimes easier to divide the branches to the superior and basilar segments individually. Ultimately, the fissure is divided and the specimen removed in a protective specimen bag.

The alternative method involves opening the fissure, and stapling the lower lobe pulmonary arterial trunk, followed by stapling of the bronchus. If this method is employed, there may be an advantage to stapling the fissure after careful dissection in the arterial plane, as opposed to dissecting the fissure bluntly, which is often done in open procedures.

Middle Lobectomy

Unlike other lobectomy procedures, the strategy for middle lobectomy begins with opening the major fissure. This is not performed to expose the pulmonary vessels; rather, it allows passage of the stapler to ligate the middle lobe pulmonary vein. The bronchus is then dissected and stapled, with identification and preservation of the bronchus intermedius. The middle lobe pulmonary artery is then stapled. Finally, the horizontal fissure is stapled and the lobe is removed in a protective specimen bag.

OUTCOMES WITH THORACOSCOPIC LOBECTOMY

Recently, single-and multiinstitutional studies have demonstrated that thoracoscopic lobectomy is an accepted oncologic procedure for patients with early stage lung cancer, and is recommended in treatment guidelines for nonsmall-cell lung cancer (NSCLC). Thoracoscopic lobectomy has been demonstrated to have better outcomes compared to conventional thoracotomy,

including shorter length of stay, shorter chest tube duration, decreased postoperative pain, improved preservation of pulmonary function, reduced inflammatory response, shorter recovery time, lower cost, and better compliance with adjuvant chemotherapy when required. In addition, it has been demonstrated that thoracoscopic lobectomy is a safer procedure than lobectomy by thoracotomy, as it is associated with fewer postoperative complications.

Using a prospective database, the outcomes of patients who underwent lobectomy at Duke from 1999 to 2009 were analyzed with respect to postoperative complications. Propensity-matched groups were analyzed based on preoperative variables and stage. Of the 1,079 patients in the study, 697 underwent thoracoscopic lobectomy and 382 underwent lobectomy by thoracotomy. In the overall analysis, thoracoscopic lobectomy was associated with a lower incidence of prolonged air leak ($P = 0.0004$), atrial fibrillation ($P = 0.01$), atelectasis ($P = 0.0001$), transfusion ($P = 0.0001$), pneumonia ($P = 0.001$), sepsis ($P = 0.008$), renal failure ($P = 0.003$), and death ($P = 0.003$). In the propensity-matched analysis based on preoperative variables, comparing 284 patients in each group, 196 patients (69%) who underwent thoracoscopic lobectomy had no complications, versus 144 patients (51%) who underwent thoracotomy ($P = 0.0001$). In addition, thoracoscopic lobectomy was associated with fewer prolonged air leaks (13% vs. 19%; $P = 0.05$), a lower incidence of atrial fibrillation (13% vs. 21%; $P = 0.01$), less atelectasis (5% vs. 12%; $P = 0.006$), fewer transfusions (4% vs. 13%; $P = 0.002$), less pneumonia (5% vs. 10%; $P = 0.05$), less renal failure (1.4% vs. 5%; $P = 0.02$), shorter chest tube duration (median 3 vs. 4 days; $P < 0.0001$), and shorter length of hospital stay (median 4 vs. 5 days; $P < 0.0001$).

Similar results were obtained when the Society of Thoracic Surgeons (STS) database was analyzed by Paul and colleagues. All patients undergoing lobectomy as the primary procedure via thoracoscopy or thoracotomy were identified in the STS database from 2002 to 2007. After exclusions, 6,323 patients were identified: 5,042 thoracotomy and 1,281 thoracoscopy. A propensity analysis was performed, incorporating preoperative variables, and the incidence of postoperative complications was compared. Matching based on propensity scores produced 1,281 patients in each group for analysis of postoperative outcomes. After VATS lobectomy, 945 patients (73.8%) had no complications, compared to 847 patients (65.3%) who had lobectomy via thoracotomy ($P < 0.0001$). Compared to open lobectomy, VATS lobectomywas associated with a lower incidence of arrhythmias ($N = 93$ [7.3%] vs. 147 [11.5%]; $P = 0.0004$), reintubation ($N = 18$ [1.4%] vs. 40 [3.1%]; $P = 0.0046$), and blood transfusion ($N = 31$ [2.4%] vs. $N = 60$ [4.7%]; $P = 0.0028$), as well as a shorter length of stay (4.0 vs. 6.0 days; $P < 0.0001$) and chest tube duration (3.0 vs. 4.0 days; $P < 0.0001$). There was no difference in operative mortality between the two groups.

Finally, Berry and colleagues reported a recent analysis of high-risk patients over 70 years of age. During the study period, 338 patients older than 70 years (mean age 75.7 $\pm$ 0.2) underwent lobectomy (219 thoracoscopy, 119 thoracotomy). Operative mortality was 3.8% (13 patients) and morbidity was 47% (159 patients). Patients with at least one complication had increased length of stay (8.3 $\pm$ 0.6 vs. 3.8 $\pm$ 0.1 days; $P < 0.0001$) and mortality (6.9% [11 of 159] vs. 1.1% [2 of 179]; $P = 0.008$). Significant predictors of morbidity by multivariable analysis included age (odds ratio 1.09; $P = 0.01$) and thoracotomy as surgical approach (odds ratio 2.21; $P = 0.004$). Thoracotomy remained a significant predictor of morbidity when the propensity to undergo thoracoscopy was considered (odds ratio 4.9; $P = 0.002$).

SEGMENTECTOMY

As experience with thoracoscopic lobectomy increases, minimally invasive strategies are being more readily applied to more complex cases and surgical interventions, including segmentectomy. A recent review of the STS database demonstrates that segmentectomy is performed in approximately 5% of pulmonary resections at institutions contributing to the database. Thoracoscopic segmentectomy is defined as a sublobar resection of one or more anatomic pulmonary segments using a completely minimally invasive approach. As with thoracoscopic lobectomy, visualization is dependent on video monitors and rib spreading is avoided. Thoracoscopic segmentectomy employs anatomic resection, with individual vessel ligation. Hilar and mediastinal lymph node dissection are a standard part of the procedure.

Strategy for Thoracoscopic Segmentectomy

Thoracoscopic segmentectomy may be used for resection of primary NSCLC, pulmonary metastases, and benign conditions that are best treated with anatomic resection. Although lobectomy is considered the standard of care for most patients with NSCLC, certain patients may benefit incrementally from sublobar anatomic resection, such as patients with restrictive lung disease, patients who have undergone previous resection(s), and patients with small tumors in specific segments, such as the superior segment, the lingula, or the upper divisions of the left upper lobe. In addition, for selected patients with early stage NSCLC, segmentectomy may be safely performed for small (<2 cm) peripheral lesions, providing acceptable resection margins, superior survival compared to wedge resection, and equivalent survival as compared to lobectomy.

Although the thoracoscopic approach may be employed for any anatomic segmental resection, the most commonly performed segmental resections are lingula-sparing left upper lobectomy, lingulectomy, superior segmentectomy, and basilar segmentectomy. Contraindications to thoracoscopic segmentectomy include the inability to achieve complete resection with segmentectomy: in patients with lung cancer, the parenchymal margin should be at least the diameter of the tumor.

The operative strategy for thoracoscopic segmentectomy is similar to that of thoracoscopic lobectomy described above. In general, segmentectomy is performed with division of segmental vessels and bronchi in a manner similar to the open approach. The approach to thoracoscopic segmentectomy begins with ligation of the segmental pulmonary vein, followed by either the bronchus or artery, depending on the segment. The parenchymal excision is accomplished by stapling in intersegmental fissures. Bi- or trisegmentectomy is performed when tumors are close to intersegmental fissures. All specimens are removed in a protective bag.

Technique for Thoracoscopic Segmentectomy

Lingula-Sparing Left Upper Lobectomy (S1–3)

This segmentectomy is performed by dividing the pleural reflection posteriorly, beginning over the left pulmonary artery near the fissure and extending the pleural division cranially to the aortopulmonary window. Attention is then turned anteriorly, pleural dissection is continued to complete the circumferential pleural division of the hilum. The upper lobe proper branches of the left superior pulmonary vein are then encircled and divided with a linear stapler, sparing the lingular vein. This maneuver (and all stapling for this segment) is facilitated by moving the camera to the anterior

access incision so that the stapler may be introduced through the "camera" incision.

Reflection of the distal stapled vein allows identification of the apical-posterioarterial and bronchial segmental branches to the upper division. The space between the arterial branches and the upper lobe bronchus is developed, providing exposure for stapling the anterior and apical branches of the left upper lobe artery. The previous posterior dissection greatly facilitates this maneuver. Dissection of the left upper lobe bronchus is then initiated at the bifurcation with the lingular branch, where there is often a small lymph node. The bifurcation of the lingular bronchus is usually behind the intact lingular vein, but care must be taken that the entire left upper lobe bronchus is not dissected.

Once the bronchus is divided, an anterior segmental arterial branch within the fissure will be unveiled. This arterial branch can be dissected and stapled. Parenchymal division with the stapler completes the segmentectomy. This is facilitated by grasping the stapled bronchial stump to provide retraction of the upper division away from the hilum. In some patients, a visible segmental fissure or demarcation is present; however, hilar anatomic landmarks are always present to identify the correct line of division. When the segmental anatomy is not clear, temporary reinflation of the lingula distinguishes the atelectactic upper division for parenchymal resection.

Lingulectomy (S4–5)

Unlike most lobar and sublobar thoracoscopic resections, lingular segmentectomy is initiated by opening the oblique fissure, as with middle lobectomy. Once the fissure is open, the upper lobe is retracted posteriorly, exposing the lingular vein. The vein is encircled and divided with a stapler, exposing the bifurcation of the upper lobe division and lingular bronchi. The lingular bronchus is encircled and stapled, exposing the lingular arterial branches, which are then stapled and divided as well. The segmentectomy is completed with parenchymal division. As with other thoracoscopic anatomic resections, this is facilitated by grasping the stapled bronchial stump to provide retraction of the lingula away from the hilum. In some patients, a visible segmental fissure or demarcation is present; however, hilar anatomic landmarks are always present to identify the correct line of division. When the segmental anatomy is not clear, temporary reinflation of the upper division distinguishes the atelectactic lingula for parenchymal resection.

Superior Segmentectomy (S6)

As with most anatomic thoracoscopic resections, division of the pleura posterior facilitates the procedure. The inferior pulmonary ligament is divided with electrocautery to the level of the inferior pulmonary vein, and the division of the posterior pleural reflection is continued cranially to the pulmonary artery. The inferior pulmonary vein is then identified and dissection on its posterior surface undertaken to expose the superior segmental vein. Once the vein is encircled and stapled, the superior segmental bronchus is exposed; there is often a small lymph node at this bifurcation, which should be removed for oncologic purposes as well as to improve exposure. The segmental bronchus is then divided, which exposes the segmental artery. The artery may at that point be stapled; alternatively, the segmental artery may be approached by opening the posterior aspect of the oblique fissure. The segmentectomy is completed by dividing the parenchyma with the stapler.

Basilar Segmentectomy (S7–10)

As with superior segmentectomy (S6), basilar segmentectomy is initiated with division of the inferior pulmonary ligament and the posterior pleural reflection to expose the inferior pulmonary vein, including the bifurcation of the superior segmental and basilar segmental veins. Anterior dissection is then undertaken to encircle the basilar segmental veins, which may be stapled. Superior retraction of the lung exposes the lower lobe bronchus, and the basilar segmental bronchi may then be dissected using this approach. Once again, thorough posterior dissection, in this case between the bronchus and the artery, greatly facilitates this strategy. Once the basilar segmental bronchi are stapled, the basilar segmental arteries are similarly divided, followed by division of the anterior portion of the major fissure. Finally, the parenchymal resection is performed.

Alternatively, after division of the basilar segmental veins, the anterior portion of oblique fissure may be opened to expose the basilar segmental arteries. Division of the arteries exposes the bronchi, which are similarly stapled and divided, sparing the superior segmental vessels. The parenchymal resection is then completed with the stapler.

Posterior Segment of the Right Upper Lobe (S–2)

Although posterior segmentectomy is an uncommon procedure for NSCLC, as retention of the apical and anterior segments is less likely to result in preserved pulmonary function, this procedure may be beneficial in patients with relatively central pulmonary metastases. In these patients, large wedge resections may compromise more pulmonary parenchyma than a precise segmentectomy. Dissection is once again begun posteriorly, to divide the pleural reflection, to expose the posterior aspect of the right upper lobe bronchus, and to begin to open the posterior portion of the oblique fissure. At this point, identification and division of the posterior ascending artery may be performed. Further dissection within the fissure exposes the posterior segmental vein as it courses underneath the horizontal and oblique fissures. Once the vein is divided, the parenchyma of the posterior segment is reflected off of the interlobar pulmonary artery, providing visualization of the anterior trunk and the bifurcation of the posterior segmental bronchus, which is then stapled. There is often a substantial lymph node or nodes in this location, and complete dissection should be performed. The parenchymal division is then performed with the stapler.

PNEUMONECTOMY

Experience with thoracoscopic pneumonectomy is limited, and the advantages conferred on minimally invasive strategies for lobectomy and segmentectomy may not be necessarily conferred on pneumonectomy. Nevertheless, it is reasonable to pursue investigation of this technique, including feasibility, safety, and outcomes. In a recent review, the outcomes of 67 patients who underwent pneumonectomy for malignancy were analyzed, including 32 patients in whom thoracoscopic pneumonectomy was attempted. Patients in the thoracoscopic group had shorter lengths of stay in the hospital and less operative blood loss; however, the 8 patients who were converted to thoracotomy had significantly more operative blood loss. The complication rates were similar among thoracoscopic, converted, and open groups. While this study does demonstrate safety and feasibility, further studies are required to determine outcome advantages.

AWAKE THORACOSCOPIC SURGERY

Despite the notable advantages of VATS over thoracotomy for pulmonary resections, use of general anesthesia, which is still considered mandatory for VATS, can be associated

with several potentially adverse effects. As a consequence, the concept of awake VATS is emerging in an attempt to minimize the surgical trauma of VATS. Initial series, employing awake thoracic surgery for nonresectional indications, demonstrated feasibility and cost effectiveness. Furthermore, preliminary data suggest that awake VATS is associated with a reduced perioperative stress hormone response and better preserved lymphocyte activity, which might also improve outcomes.

Awake Resection of Indeterminate Pulmonary Nodules

Awake VATS, performed with thoracic epidural anesthesia (TEA) and sedation, has been successfully applied for removal of indeterminate pulmonary nodules. The indication is the need to obtain a rapid, histologically proven diagnosis of nodule in patients in whom further surgical resection is not indicated. Eligibility criteria include radiologic findings of a nodule less than 3 cm in maximal size at CT and localized in the peripheral one-third of the lung. The presence of anxiety, which interferes with full patient cooperation in the operating room, is considered a potential contraindication, but this can be controlled in most instances by sedation.

In a small randomized comparison of VATS wedge resection performed with TEA or with general anesthesia plus TEA, technical feasibility was equivalent in the study groups, whereas the perioperative arterial oxygenation, the need for prolonged nursing care, and hospital stay were better in the awake group.

Awake Pulmonary Metastasectomy

Since the majority of pulmonary metastases are located peripherally, VATS has been advocated as a viable option particularly in patients with solitary lesions. As discussed above, potential advantages of VATS over open approaches include less pain, lower morbidity rate, and shorter hospital stay. Recently, awake VATS metastasectomy through TEA has been proposed in order to minimize surgical stress. In a cohort of 14 patients undergoing awake VATS metastasectomy, the procedure was demonstrated to be feasible and safe, accomplished under TEA in all patients with no operative mortality or major morbidity. Hospital stay was significantly shorter than in a control group operated through general anesthesia while intermediate-term results and survival were similar.

Awake Resection of Lung Cancer

Resection of lung cancer represents the most provocative indication for awake VATS, and to date both lobectomies and pneumonectomies have been performed through TEA in fully awake patients. Proposed inclusion criteria for awake resection of lung cancer include peripheral stage I lesions, age older than 75 years, poor pulmonary function, or other comorbidity leading to consider the patient at high risk for anatomic resection or for general anesthesia. In addition, selected patients with peripheral lesions previously considered medically inoperable, who already underwent percutaneous radiofrequency ablation or stereotactic radiofrequency ablation, can be eligible for an awake lung resection.

ROBOTIC-ASSISTED THORACIC SURGERY

Abbas E. Abbas

As noted in this chapter the advent of endoscopic VATS, allowed for the performance of anatomic lobectomies with complete mediastinal lymph node dissection through small ports instead of a large thoracotomy and has become the goal of many thoracic surgeons. As noted by Dr. Pompeo and Dr. D'Amico, the technique has been shown to have equal or even better outcomes compared to open thoracotomy when performed by surgeons facile and experienced in this technique. However, many factors have made the adoption of VATS lobectomy less than universal as currently less than one-third of anatomic pulmonary resections are accomplished with a VATS approach. These factors include the inadequacy of precise dissection of the vascular and mediastinal structures within the thoracic cavity due to the limited maneuverability of the instruments and limitations of the two-dimensional view of the operative field. In addition, there is the concern for the loss of immediate control of the pulmonary hilum in case of bleeding. For most surgeons mastering VATS lobectomy is also associated with a prolonged learning curve. Robotic-assisted thoracic surgery (RATS) may offer an alternative to address some of these concerns. Robotic instruments are wristed and have 360 degrees of freedom, mimicking maneuvers done with the human wrist during open surgery. The image seen by the surgeon is high definition and three-dimensional with 10 times optical zooming and is a significant improvement over that available with VATS. This makes RATS quite ideal for fine dissection in a confined space such as the pleural cavity. However, there are also certain concerns associated with RATS. There is a lack of tactile feedback that must be compensated for by visual cues. In addition, the concern for loss of control may be greater than with VATS, as the surgeon is not even at the field while performing the procedure. It is therefore important to have an experienced surgical team at the table that is adept at dealing with different scenarios in the execution of this operation, including emergency conversion to thoracotomy for bleeding.

The procedures described for robotic lung surgery have included "hybrid" techniques with robotic assistance to open or thoracoscopic surgery. Examples of these techniques are those where the robot is used during the dissection phase of the lobectomy, including dissection of the mediastinal, hilar, and subcarinal structures. For the subsequent vascular, parenchymal, and bronchial division along with removal of the specimen from the chest cavity, standard VATS lobectomy techniques were used with the robot undocked and the surgeon scrubbed into the surgical field. More recently, the totally endoscopic and completely portal (TECP) approach has become the standard technique for most surgeons who practice robotic thoracic surgery. This technique mimics the exposure and approach of a thoracotomy, as most of the ports for the camera and robotic arms are along the same intercostal space (ICS). It can be performed with either three or four arms depending on surgeon preference.

Our own approach to lobectomy is a totally endoscopic four-arm technique using the da Vinci Surgical System (Intuitive Surgical, Inc. Sunnyvale, CA). After induction of general anesthesia, intubation, and bronchoscopy, a bronchial blocker or double-lumen tube is inserted. The patient is then placed in the lateral decubitus position and the bed is flexed at the level of the 7th ICS (Fig. 11.2). The robot is then brought in directly over the head of the patient.

Five ports are then inserted. The first is a 12 mm port at the 9th ICS for CO_2 insufflation and assistance. Another four ports are placed ranging from 5 to 8.5 mm, attempting to place all in one ICS, usually the 7th or 8th ICS depending on the patient's body habitus (Fig. 11.4). These ports are placed with guidance from a thoracoscope placed through the assistant port and will be at or close to the midclavicular line, anterior axillary line, posterior axillary line, and paraspinally. In general, the ports should be placed about a hand's breadth or 9 to 10 cm apart to avoid external collision between the arms. The robot is then brought in from the head

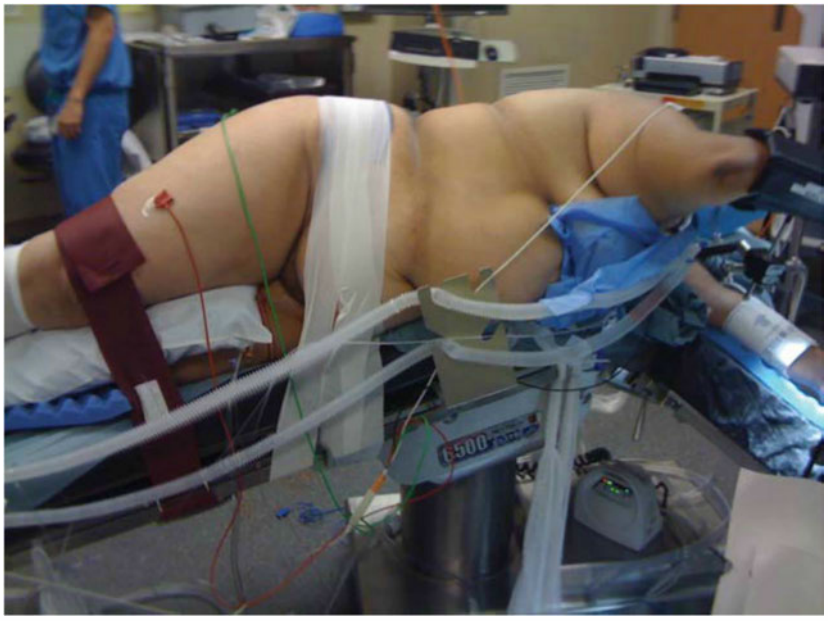


Fig. 11.2. Patient positioned for robotic thoracic surgery. The robot is brought in from the head of the table forcing the anesthesia team to be at the foot of the bed.

of the table and the surgeon moves to the console (Fig. 11.3).

Arm #3 is used mainly for retraction using a 5-mm lung-grasping instrument and is placed posterolateral to either the left or the right arm depending on the side of the lobectomy. The remaining two arms are both 8 mm and are used for the majority of the dissection. For right-handed surgeons, the left arm usually uses a Cadiere forceps for gentle traction, whereas the right arm uses an instrument with an energy source such as the Maryland forceps with bipolar cautery. A left-handed surgeon can simply reverse the instruments. A variety of other instruments including a monopolar hook cautery, monopolar scissors, and a wristed suction dissector are also available.

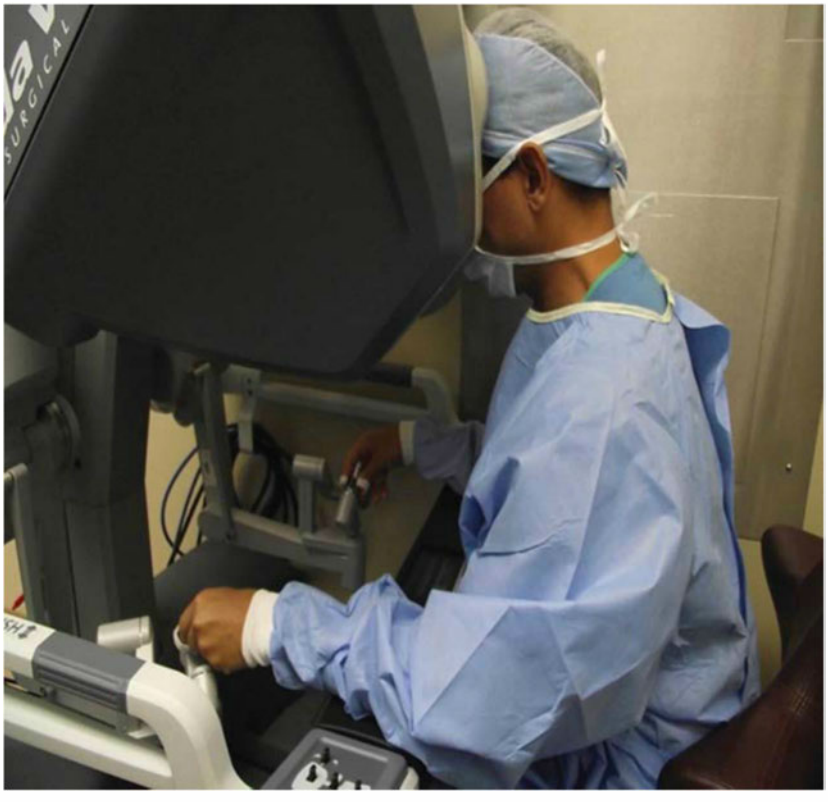


Fig. 11.3. Surgeon at the console. Three-dimensional visualization with magnification allows for precise dissection of bronchovascular structures. It is key to have an experienced assistant at the operating table working with the surgeon at the console.

Dissection is begun by dividing the inferior pulmonary ligament and incising the mediastinal pleura circumferentially around the hilum. In the process, a complete mediastinal lymph node dissection is performed. Following this, the approach to lobectomy is the same as that for the open procedure. After dissection and removal of the specific N1 nodes, sequential division of the pulmonary vein, pulmonary arterial branches, bronchus, and fissure is undertaken. The order is sometimes changed to accommodate for individual anatomic variations. The lobe is finally completely detached and placed in a surgical pouch for removal from the assistant port after converting it to a 3 to 5 cm subcostal incision through the diaphragm and below the 10th rib as has previously been described by Dylewski et al. Others have described creating a minithoracotomy utility incision for extraction of the lobe.

SUMMARY

Minimally invasive pulmonary resection has become an accepted, safe, and oncologically sound procedure. A number of studies have demonstrated that the use of thoracoscopic lobectomy is associated with less postoperative pain, reduced length of stay, better compliance with adjuvant therapy, and fewer postoperative complications. Although the thoracoscopic strategy may be difficult to learn, it is increasingly becoming the preferred method of anatomic lobectomy. RATS may address a number of the limitations presented by VATS lobectomy. The ability to have "wristed" instruments with 360 degrees of freedom allows one to essentially recreate an open operation within the confines of the closed chest. All of the port incisions for docking the robot occur along a single intercostal space as in an open thoracotomy. The three-dimensional visualization with magnification allows for precise dissection of bronchovascular structures and new and improved instruments are continually being introduced. These include the recently developed linear staplers for use with the robotic arms. In light of the outcome of the National Lung Screening Trial, the use of thoracoscopic or robotic procedures is certain to increase. In the future, minimally invasive strategies will be more commonly used in the management of locally advanced lung cancer, including patients with chest wall tumors, patients

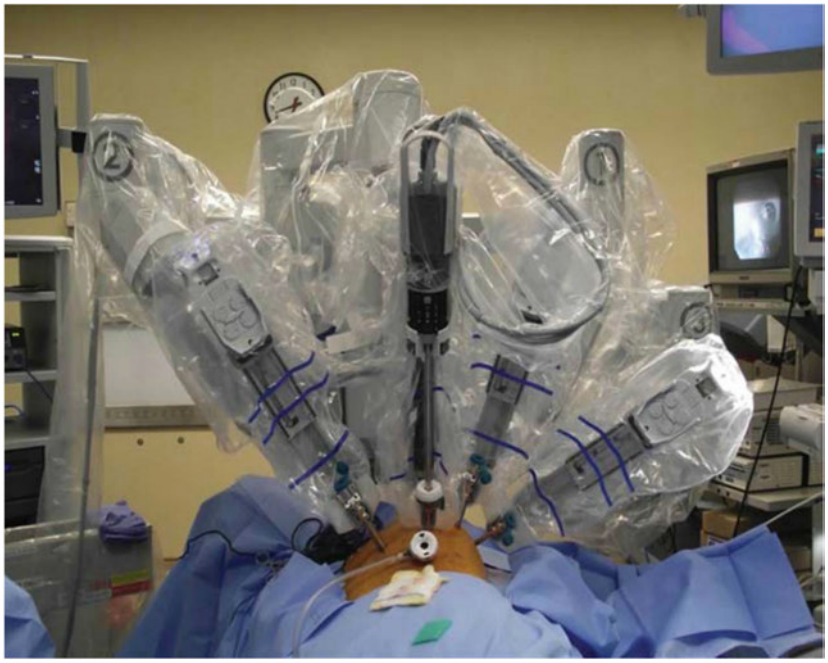


Fig. 11.4. The robotic arms docked within the patient. All working ports are placed along a single interspace, most commonly the 7th.

after induction therapy, and patients that require pneumonectomy.

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EDITOR'S COMMENTS

This is a critically important chapter in that it represents the state of the art in minimally invasive thoracic surgery, specifically pulmonary resection. Dr. D'Amico has been at the forefront of the development of VATS lobectomy and is one of the most experienced practitioners of the procedure. It must be stressed that the learning curve is prolonged and newly minted surgeons would be well advised to be intimately familiar with open surgical techniques prior to embarking on complex resections utilizing a VATS approach.

Response to significant intraoperative hemorrhage remains one of my major concerns with any minimally invasive approach. Though it happens only infrequently, bleeding from a pulmonary arterial branch does occur during the performance of a pulmonary resection. When the chest is open often just digital pressure suffices to staunch the bleeding, but if the hemorrhage is significant it is fairly straightforward to obtain proximal control of the main pulmonary artery if it had not already been done as part of the routine. The authors point out that pressure on the bleeding area with a sponge stick usually suffices to control bleeding but with limited visualization and the inability to put a hand in the chest complete control may be difficult to achieve. Sufficient control of significant bleeding must be accomplished to allow time to open the chest in at least some of the situations that may be encountered. In addition, repairing a rent in the pulmonary artery is best and most safely accomplished when there is proximal control of bleeding. Blindly trying to put a stitch in a bleeding pulmonary artery usually results in further disaster. Though no surgeon likes to talk about it, there have been intraoperative deaths resulting from massive hemorrhage during VATS lobectomy that never would have had a fatal outcome if the procedure had been done open. This is not to say that VATS is any more dangerous, only that a contingency plan for a disaster should be well thought out.

The authors have nicely described the technique for the various anatomic resections noting that dividing the vein first facilitates the rest of the procedure. I differ with their description of middle lobectomy in that it is not necessary, or recommended, to dissect in the fissure to identify the artery. Middle lobectomy may be expeditiously accomplished by identifying and dividing the middle lobe vein at the hilum as the initial maneuver. Following division of the middle lobe vein, the middle lobe bronchus is easily visualized and can be divided nicely revealing the middle lobe artery or arteries. Once the arterial branch or branches are divided the minor fissure and oblique fissure may be taken with a linear stapler to complete the resection. The same approach may be employed for a lingular segmentectomy again avoiding the messy dissection within the fissure.

Having for a long time been a nonbeliever I have become a convert to the use of the robot for pulmonary resection and, in fact, feel that it is a better operation than the VATS procedure. This occurred once I observed an experienced robotic surgeon in action and saw some of the currently available instruments. The introduction of the robotic linear staplers will be a major advance that I believe will, and should, convert more surgeons to a robotic approach. It truly is "intuitive" and really is an extension of the open procedure yet done within the confines of the chest. Abbas nicely describes his technique and elaborates specifically how he removes the lobe from the chest. The use of the robotic arms avoids the levering that occurs between the thoracoscopic instruments and the rib edges during VATS procedures likely further attenuating postoperative pain. Some would argue that the additional expense incurred by a robotic procedure outweighs the potential advantage over the VATS approach but in hospitals where multiple specialties are using the robot, the expense is diffused somewhat by being spread across multiple users. Whether VATS or robotics, the decision must be based on the experience and comfort of the surgeon. Both techniques appear to be a benefit to the end-user, the patient, and that really is what matters most.

LRK

12 Management of Pneumothorax and Bullous Disease

Stephen D. Cassivi and Claude Deschamps

The pleural space is a potential space during normal conditions with the visceral pleura directly in apposition to the parietal pleura. Pneumothorax is defined as the presence of air in the pleural space and can be due to a number of causes. The etiology and volume of the pneumothorax and the resultant intrapleural pressure and condition of the underlying lung play a role in determining the clinical severity.

This chapter outlines the anatomy and basic physiology of the pleural space. The pathophysiology of the various etiologies of pneumothoraces is discussed, as are the diagnosis and management options. Special attention is directed to the particular condition of bullous disease.



ANATOMY

The pleural space is lined by the visceral and parietal pleurae (Fig. 12.1). The visceral pleura is a thin layer (usually one cell thick) intimately covering the outer surface of the lung. It adheres to the underlying alveolar walls of the lung parenchyma via connective tissue made up of elastic fibers. There is, therefore, no true cleavage plane between the visceral pleura and the lung parenchyma that it envelops. The visceral pleura has no somatic innervation.

The parietal pleura is a more complex serous membrane. It lines the inside of the chest wall, the diaphragm, and the mediastinum and is attached to these by a fibrous and connective tissue layer known as the endothoracic fascia. Between the endothoracic fascia and the parietal pleura is a dissection plane that allows the parietal pleura to be stripped off of the chest wall and other structures. It is thickest and most substantial along the chest wall, overlying the ribs, and thinnest as it covers the mediastinal structures and beneath the sternum. The parietal pleura is innervated by somatic, sympathetic, and parasympathetic nerve fibers via the intercostal nerves.

PHYSIOLOGY

The physiology of the pleural space is relatively straightforward, although dynamic. Functional residual capacity is the measure of lung volume with the patient at rest after normal exhalation. In this state, the elastic and retractive nature of the chest wall and lung pull the parietal and visceral pleurae away from one another, thus creating a negative intrapleural pressure usually in the range of −2 to −5 cm H_2O. During inspiration, the outward chest wall and diaphragmatic forces counteracting the normal elastic recoil of the lung parenchyma can create intrapleural pressures of −20 to −35 cm H_2O. Gravity also exerts an influence on this negative intrapleural pressure. In the upright position, the apex has a greater negative intrapleural pressure than the base of the lung in the region of the costophrenic sulci (0.25 cm H_2O/cm of height). This phenomenon may contribute to some degree to creating increased distention of alveoli in the apex and a greater predisposition to spontaneous pneumothoraces by rupture of apical blebs.

As a consequence of having more oxygen consumed than carbon dioxide produced during the respiratory cycle (respiratory quotient <1), there is a resultant partial pressure gradient between the gases in the venous blood and those of the arterial system and pleural space. This gradient, usually between 54 and 72 cm H_2O, ensures against spontaneous gas formation in the pleural space as long as the intrapleural pressures do not become less than −72 cm H_2O. On a more practical level, this also explains how intrapleural air, as in the case of a pneumothorax, can be gradually reabsorbed by diffusion into the venous circulation.

Pleural gases can also be affected by barometric pressure. Whereas the relative proportions of gases do not change with variation in atmospheric pressure, there can be a significant change in the volume of these gases. Boyle's law states that at constant temperature, for a given mass of gas, pressure p multiplied by volume V is a constant:

$$pV = c$$

Thus, the volume change in a gas is inversely proportional to the change in atmospheric pressure. This has a number of clinically significant consequences when considering pneumothoraces. First, although a person with a pneumothorax being transported by airplane is likely to be in a pressurized cabin, one can expect the barometric pressure to decrease, with a resultant proportional increase in the volume of their pneumothorax if there is not a path of egress for the intrapleural gas such as provided by a chest tube. Second, a clinician whose practice is located at a higher altitude (with lower atmospheric pressure) can expect a slower resolution of pneumothoraces by resorptive diffusion alone than that seen by a colleague practicing closer to sea level.

ETIOLOGY AND PATHOPHYSIOLOGY

Primary spontaneous pneumothorax is the most common cause of pneumothoraces and has an estimated overall incidence of 5 to 10 cases per 100,000 per year. It occurs predominantly in young, healthy men, with an incidence in this group as high as 1 in 500 per year, and is due in most cases to rupture of apical subpleural blebs in otherwise normal lungs (Fig. 12.2). The pathogenesis of these apical blebs is unclear, although it is postulated that higher transpulmonary pressures at the apex lead to greater alveolar distending pressures. The resultant rupture of alveoli traps air between the internal and external elastic membranes of the visceral pleura. It is noted that these types of pneumothoraces most often occur in tall, thin individuals, many of whom are smokers. The lifetime risk of developing a pneumothorax in an otherwise healthy man is estimated at 0.1%, whereas it is as high as 12% in one who is a smoker. There may also be an association with connective

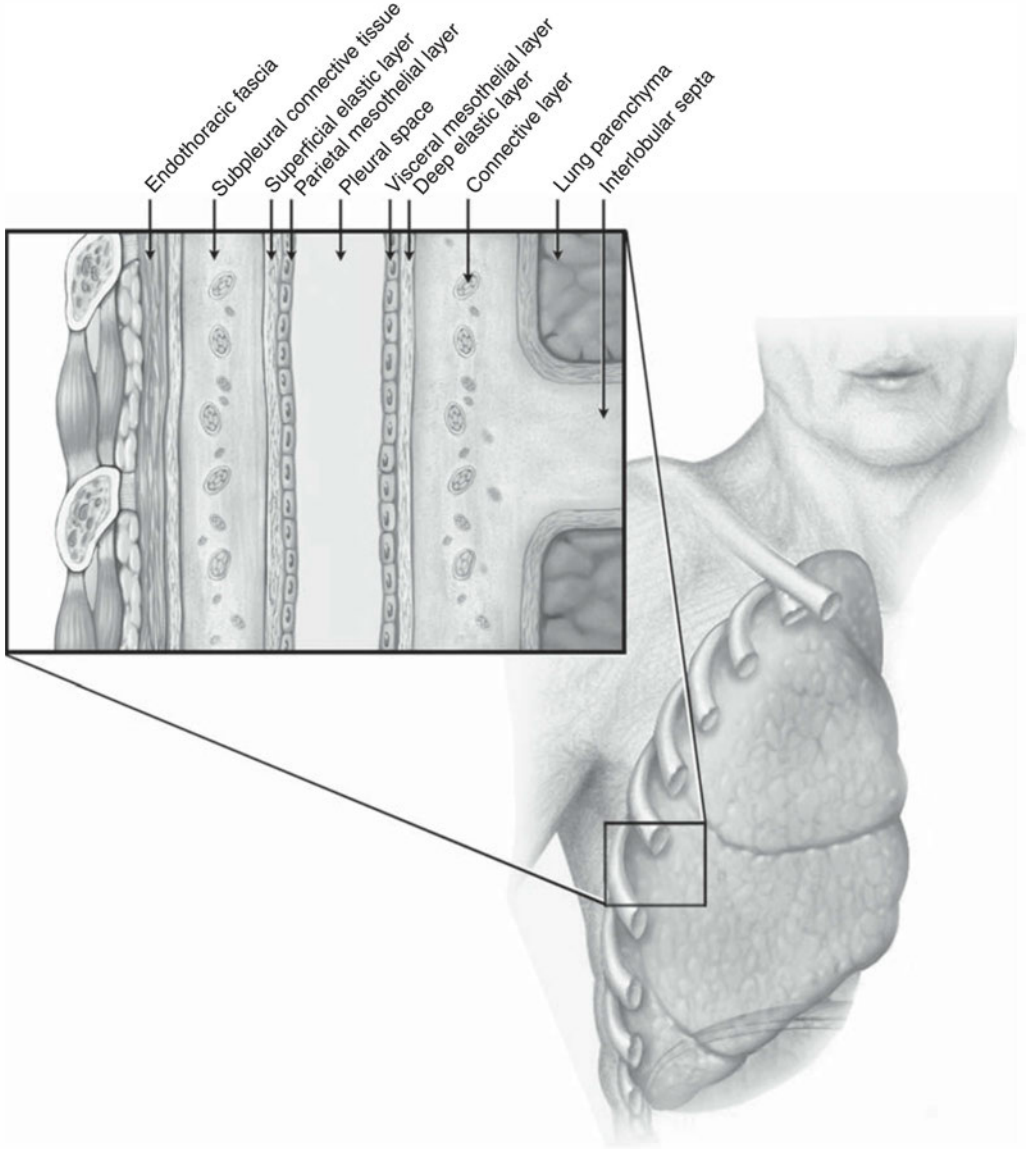


Fig. 12.1. Anatomy of the pleural surfaces.

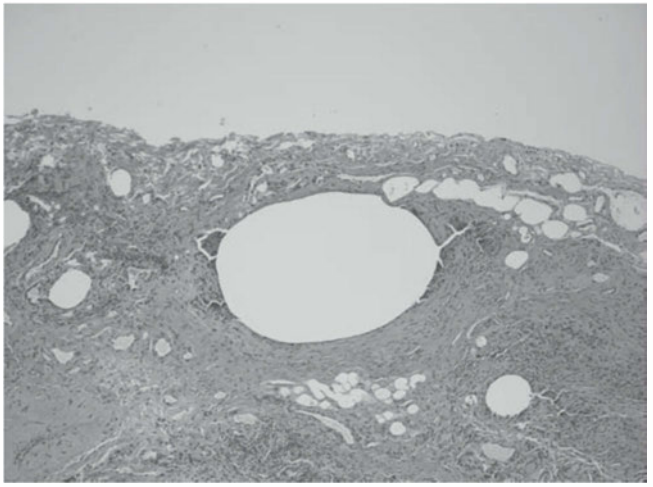


Fig. 12.2. Photomicrograph of apical resection for recurrent primary spontaneous pneumothoraces demonstrating subpleural bleb.

tissue disorders such as Marfan syndrome. A familial form of spontaneous pneumothorax has been described with autosomal-dominant inheritance and incomplete penetrance.

Spontaneous pneumothorax can also occur as a result of underlying lung disease, in which case it is referred to as a secondary spontaneous pneumothorax. There are various pulmonary disease states leading to secondary pneumothoraces (Table 12.1). Most cases are related to bullous-type disease, with hyperinflation leading ultimately to rupture and subsequent pulmonary parenchymal collapse. Diseases characterized by cystic lesions in the pulmonary parenchyma such as cystic fibrosis and lymphangioleiomyomatosis can also lead to spontaneous rupture and pneumothorax. Malignant

Table 12.1 Etiologies of Pneumothorax

Spontaneous	
Primary	
Rupture of subpleural bleb	
Secondary	
Bullous disease	Emphysema, chronic obstructive pulmonary disease (COPD), α-1 antitrypsin deficiency emphysema
Cystic disease	Cystic fibrosis, lymphangioleiomyomatosis
Malignancy	Primary lung cancer, lung metastases (especially osteogenic sarcoma), after chemotherapy
Catamenial	
Infectious	*Pneumocystis carinii* pneumonia, acquired immunodeficiency syndrome (AIDS), severe acute respiratory syndrome (SARS)
Eosinophilic granuloma	
Connective tissue disorders	Marfan syndrome
Acquired	
Iatrogenic	
Needle puncture	Transthoracic needle aspiration/biopsy, thoracentesis, central venous line placement
Transbronchial biopsy	
Laparoscopic surgery	
Traumatic	
Blunt	
Penetrating	
Barotrauma	Ventilator induced

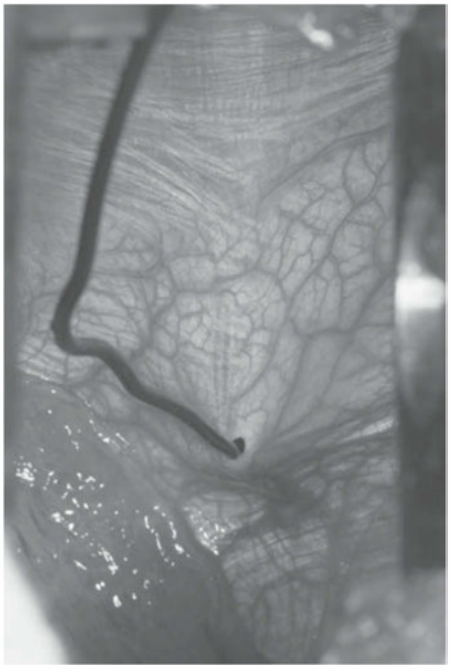


Fig. 12.3. Diaphragmatic surface demonstrating a probe passing through a transdiaphragmatic fenestration.

pulmonary nodules, both primary and metastatic, usually located subpleurally, can rupture as they expand and result in a spontaneous pneumothorax.

Spontaneous pneumothoraces that occur and recur in relation to the menstrual cycle are referred to as catamenial pneumothoraces. These episodes occur within 72 hours before or after the onset of menstruation. Three distinct mechanisms have been proposed based on metastatic, hormonal, and anatomic models. The metastatic model hypothesizes migration of endometrial tissue via the peritoneal cavity through transdiaphragmatic lymphatic channels, via diaphragmatic fenestrations, or hematogenously into the pleural space. An alternative theory suggests that endometrial tissue may be deposited in the chest cavity during embryonal development. Endometrial deposits have been identified in the pleural space in 13% to 62.5% of these cases. Hormonally regulated monthly sloughing of endometrial tissue is believed to result in pleural irritation, causing chest pain, and subpleural breaches, causing air leaks. The hormonal hypothesis suggests that high serum levels of prostaglandin F_2 at ovulation leads to vasospasm, associated ischemia with tissue injury, and alveolar rupture. The anatomic model for catamenial pneumothorax is based on the influx of air into the pleural space from the peritoneal cavity via diaphragmatic fenestrations (Fig. 12.3).

Infectious diseases of the lung in their many forms may result in secondary spontaneous pneumothorax. This is most notable in the immunocompromised host such as individuals infected with the human immunodeficiency virus (HIV) and those with manifestations of acquired immunodeficiency syndrome. Pneumothorax occurs in 1% to 2% of hospitalized patients with HIV and is associated with 34% mortality. The most common etiologic agent in these cases is *Pneumocystis carinii*. Other virulent or opportunistic infections are also causally related to the occurrence of spontaneous pneumothoraces, including most recently severe acute respiratory syndrome (SARS).

Pneumothoraces may also be incited by iatrogenic or traumatic events. Penetration of the pleural space by a needle during a diagnostic or therapeutic procedure, whether deliberate or accidental, may result in a so-called acquired pneumothorax. This is usually due to a breach or laceration of the visceral pleura. This can occur during procedures such as transthoracic needle aspiration, central line placement in the neck, and thoracentesis. In the latter procedure, recent studies have shown that the incidence of iatrogenic pneumothorax can be significantly reduced by using ultrasound guidance.

DIAGNOSIS

The most common presenting symptom of spontaneous pneumothorax is ipsilateral pleuritic chest pain of relatively sudden onset. Dyspnea may or may not be a prominent symptom and is usually proportional to the amount of lung collapse. Ipsilateral decreased breath sounds and hyperresonance on percussion are usually noted. In the case of a tension pneumothorax, there may also be deviation of the trachea to the contralateral side, tachycardia, hypotension, and diaphoresis. Although they are useful in suggesting the diagnosis of pneumothorax, clinical history and physical examination are not generally reliable in predicting the volume of underlying lung collapse.

Standard posteroanterior (PA) chest radiographs (CXR) remain the standard for the diagnosis of spontaneous pneumothorax in the clinically stable

patient. If the diagnosis is suspected but is not confirmed by a standard upright CXR taken on inspiration, a CXR taken either during expiration or in the lateral decubitus position may accentuate the pneumothorax and facilitate diagnosis. Computed tomography (CT) scans are the most sensitive and specific imaging modality for the diagnosis of pneumothoraces and can be especially useful in differentiating such a diagnosis from that of a large emphysematous bullae.

A large body of literature exists positing many methods of estimating the volume of the pneumothorax as a percentage of the pleural space volume. None of the methods has been universally accepted. CT scans are much more precise for such estimates because they take into account the nonuniform collapse that usually characterizes spontaneous pneumothoraces. However, the clinical usefulness of a precise measurement of pneumothorax size is debatable. Most often treatment decisions are made on the basis of a relative estimate of pneumothorax size, its evolution over time, and, more important, the clinical status of the patient.

TREATMENT

The most important factor to consider in the management of a patient with a pneumothorax is the clinical status. An unstable patient, whether due to the hemodynamic compromise resulting from a tension pneumothorax or the respiratory compromise of a clinically significant simple pneumothorax, requires urgent treatment. This can be accomplished, in the case of a tension pneumothorax, by immediate insertion of a 14-gauge angiocatheter in the ipsilateral mid-clavicular line in the second interspace. This is followed by an expeditious insertion of an ipsilateral chest tube to evacuate the remaining air and re-expand the underlying lung.

There are a number of treatment options for patients with pneumothoraces, either primary or secondary (Table 12.2). In the clinically stable patient with a spontaneous pneumothorax, an evaluation should first be made of his or her level of dyspnea or breathlessness as well as the relative size of the pneumothorax (Fig. 12.4). In a patient with little or no breathing difficulties and a small pneumothorax (<20%), it is reasonable to observe the patient with a repeat CXR in approximately 6 hours. If the pneumothorax has not progressed and the patient has not developed worsening symptoms, further observation as an outpatient with return in 24 to 48 hours for reassessment by clinical exam and repeat CXR usually is all that is necessary.

The rate of resolution of a spontaneous pneumothorax by reabsorption is between 1.25% and 1.8% per day. Using high concentrations of oxygen in the inspired gas in an effort to create a gradient of nitrogen favoring reabsorption of intrapleural gas has been shown to increase the resolution of pneumothoraces by up to four times the rate on room air. However, hospitalization for the sole purpose of providing high-flow oxygen in an effort to hasten the resolution of the pneumothorax is likely unnecessary and is not advocated.

Table 12.2 Treatment Options for Pneumothorax

Observation	
Needle aspiration	
Chest tube	
Pleurodesis	
Thoracoscopy	With wedge resection
	With pleurodesis
	Mechanical
	Chemical
	With pleurectomy
Thoracotomy	With wedge resection
	With pleurodesis
	Mechanical
	Chemical
	With pleurectomy

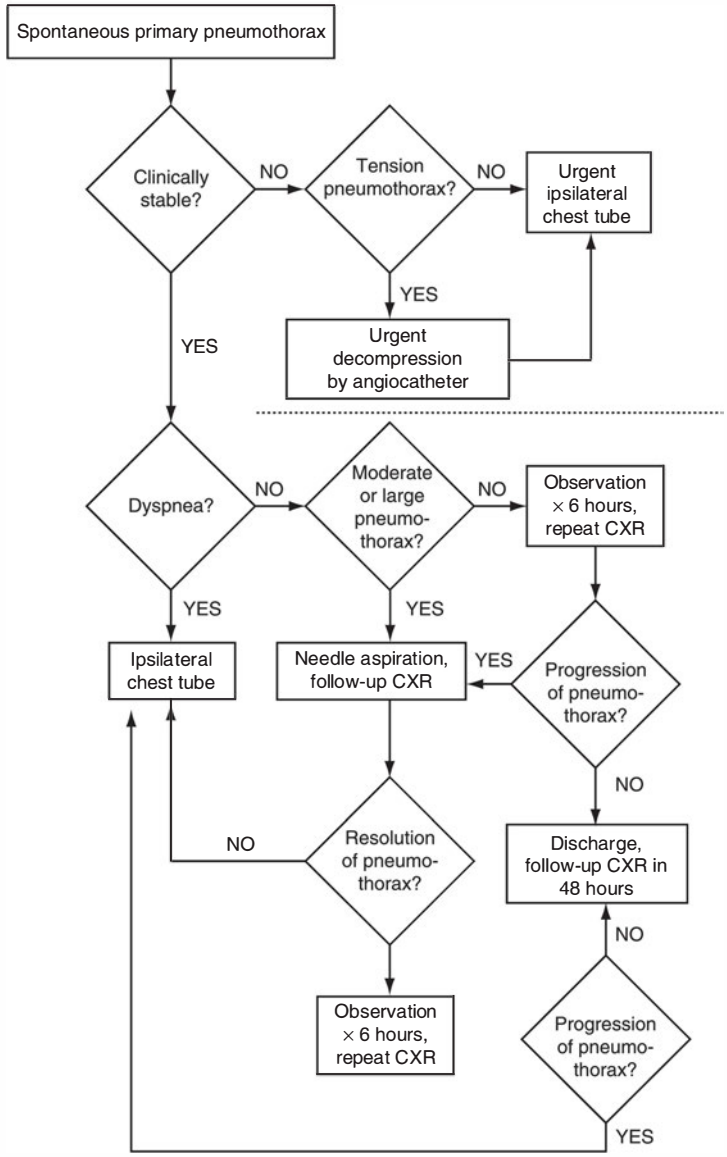


Fig. 12.4. Algorithm for the treatment of spontaneous pneumothoraces (CXR, chest X-ray).

In patients with few respiratory symptoms and moderate-to-large pneumothorax, there is growing evidence that a trial of needle aspiration may be warranted. The consensus guidelines from both the American College of Chest Physicians and the British Thoracic Society advocate an initial trial of needle aspiration in these cases. These recommendations are based on studies showing initial aspiration to be as effective as chest tube management in resolving pneumothoraces in patients with minimal symptoms. Recurrence rates were also equal in both groups. The most recent of these studies also shows that repeat attempts at needle aspiration after an initial failed attempt are of no use. In those instances, a chest tube is warranted.

Needle aspiration of a pneumothorax can be accomplished by using the widely available disposable thoracentesis kits. Local anesthetic is used to anesthetize the skin and subcutaneous tissues in the area of the second interspace in the ipsilateral midclavicular line. The small-bore catheter, loaded on the aspirating needle, is guided carefully into the pleural space just above the upper edge of the third rib. Suction on the syringe is applied as the needle is inserted, and once air is aspirated, the catheter can be advanced over the needle, which is ultimately withdrawn. With a syringe and three-way stopcock or one-way valve attached to the catheter, air is aspirated until no more can be evacuated. The catheter is then removed, and a postprocedure CXR is performed to assess the adequacy of the aspiration. If the pneumothorax has resolved, a delayed CXR can be obtained at 6 hours to confirm resolution. The patient can then be followed as an outpatient and reassessed in 24 to 48 hours with a clinical examination and repeat CXR. If, however, on the initial postaspiration or the delayed CXR, there remains a moderate-to-large pneumothorax, insertion of a chest tube is warranted. Failure to resolve a pneumothorax after needle aspiration either on initial postaspiration or delayed chest films is likely due to a persistent air leak.

Chest tube drainage is usually advocated for large, asymptomatic or smaller, symptomatic pneumothoraces. The technique has been well described. Proper functioning of the tube requires deliberate directing of the tube in the desired position. For the most part, chest tubes are inserted in the anterior or mid-axillary line in the fourth or fifth interspace and should be guided posteriorly and cephalad. A tube positioned in this manner will drain both air and fluid satisfactorily in the vast majority of cases. The usual pitfall of a tube placed anteriorly is that it will often find its way into the major fissure and cease to function adequately. Failure to adequately resolve a pneumothorax after chest tube insertion should be investigated with a view to ensuring correct positioning. If this is not clear by CXR, a CT scan should be obtained and in most cases will confirm proper or improper tube location.

Acquired pneumothoraces due to blunt, penetrating, or ventilator trauma almost always require chest tube insertion as initial management. These injuries are usually more severe, and symptoms are not likely to resolve with mere observation or simple needle aspiration.

Most spontaneous primary pneumothoraces can be managed effectively by conservative measures: observation, needle aspiration, and/or chest tube drainage. The adjunctive use of chemical pleurodesis (talc, tetracycline, or other pleural sclerosant) is usually not necessary and is less effective than surgical alternatives as discussed later in this chapter. Chemical pleurodesis should only be used in cases of treatment failure where the patient is either unwilling or unable to undergo surgical repair.

OPERATIVE MANAGEMENT OF PNEUMOTHORACES

Operative intervention for first occurrences of primary spontaneous pneumothorax is controversial because most resolve with conservative, nonoperative management, and a large number do not recur. However, recurrence of a primary spontaneous pneumothorax does occur, and the risk of recurrence within 4 years has been reported as high as 54%, with increased risk being associated with smoking, increased height, male gender, and age >60 years. The risk of recurrence of a secondary pneumothorax is even higher and is associated with risk factors such as increasing age, pulmonary fibrosis, and emphysema.

The indications for operative intervention for the definitive treatment of pneumothorax are given in Table 12.3. The principle objectives of surgery in these cases are to resect the blebs or bullae and to obliterate the space to avoid further recurrences. The former can usually be accomplished by stapled wedge resection, and the latter can be approached by several means. Pleural space obliteration can be achieved by pleurodesis, either chemical or mechanical, or by parietal pleurectomy.

The findings at the time of operation are variable. In the case of secondary pneumothorax, the underlying disease is the major determinant of operative findings. In primary pneumothorax, the majority have blebs or bullae, usually located in the apex of the upper lobe and sometimes also in the superior segment of the lower lobe. The finding of abnormalities, however, is not universal. A classification of pleural findings has been suggested in which stage I refers to no discernible abnormalities, stage II refers to pleuropulmonary adhesions, stage III refers to blebs/bullae <2 cm, and stage IV refers to bullae >2 cm.

Table 12.3 Indications for Operative Intervention for Pneumothorax Treatment

Persistent air leak (>3–5 days) or failure of lung to fully re-expand with chest tube drainage
Hemopneumothorax
Recurrent ipsilateral pneumothorax
Bilateral spontaneous pneumothorax
First occurrence of contralateral pneumothorax
At-risk activities/professions (i.e., pilots, divers)
Poor access to medical treatment or follow-up

Open thoracotomy via a transaxillary or posterolateral incision is certainly possible with excellent effectiveness (recurrence rates <0.5%) and low morbidity (3% to 11%). In most cases in which surgical intervention is indicated, a thoracoscopic approach can also be used and tends to be the procedure of choice. A standard three-port approach is usually used with single-lung ventilation being obtained by the way of a double-lumen endotracheal tube. The deflated lung is examined, and if blebs are found, these are excised by stapled wedge resection (Fig. 12.5). This is followed by a procedure to obliterate the space by either pleurectomy or pleurodesis. Parietal pleurectomy appears to be the most effective way of achieving pleural symphysis and preventing further recurrences. Pleurectomy is usually accomplished in the apex and carried down to the level of the third or fourth rib or the azygous vein.

If no blebs are found (stage I), after a negative search for other pathology, pleurectomy is usually performed to obliterate the space. A recent retrospective review demonstrated reduced recurrences with the addition of an apical wedge resection. The addition of an apical staple line on the lung may encourage pleural adhesion and obliteration of the space when accompanying an apical parietal pleurectomy. This is our preferred approach as well.

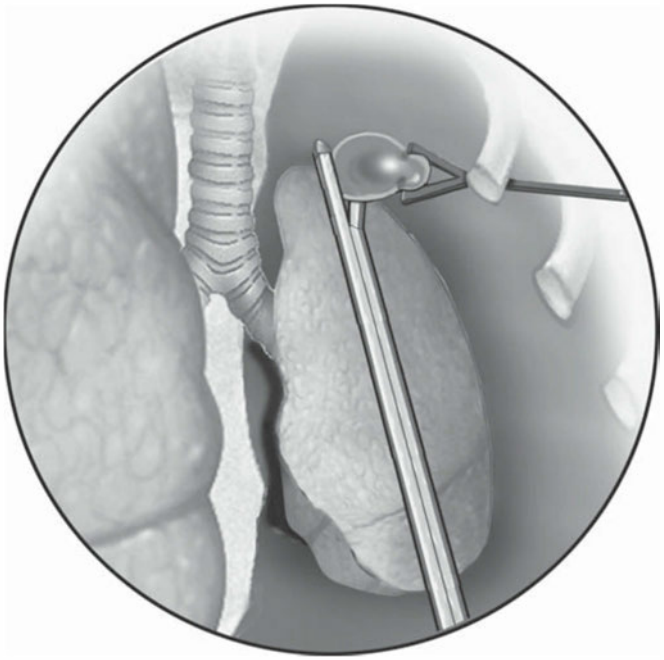


Fig. 12.5. Wedge resection of apical blebs found at thoracoscopy.

SPECIAL CONSIDERATIONS

Bullous Disease

Whereas blebs are defined as small (<2 cm) subpleural air collections contained within the visceral pleura, bullae are larger air pockets usually resulting from alveolar wall destruction. A practical classification system was developed to divide bullous disease into four groups (Table 12.4). Bullae can be associated with any form of emphysema.

Although the mechanism of secondary pneumothorax in bullous disease is for the most part similar to those with primary spontaneous pneumothoraces due to subpleural blebs, the underlying emphysema that usually accompanies the bullous disease accounts for the increased severity of the symptoms. These emphysematous patients often have such poor pulmonary reserve that symptoms related to a pneumothorax can be quite marked and severe.

Table 12.4 Classification of Bullous Lung Disease

Group	Number of bullae	Underlying lung pathology
I	Single	Normal
II	Multiple	Normal
III	Multiple	Emphysema
IV	Multiple	Other lung diseases

From: DeVries WC, Wolfe WG. Surg Clin N Am 1980;60:851-866.

One of the more common problems encountered after chest tube placement is the development of a prolonged air leak. In an effort to avoid the increased morbidity and mortality of operative intervention in these patients, conservative management may be attempted with prolonged pleural drainage with or without chemical (talc) pleurodesis. Operative intervention is sometimes chosen or becomes necessary to definitively treat the persistent air leak. Bullectomy by stapled resection or ligation is the usual procedure used with concomitant parietal pleurectomy or mechanical/abrasive pleurodesis. When possible, this is preferably done by thoracoscopy in an attempt to minimize the postoperative respiratory complications due to the incisional pain of thoracotomy.

Another option in the occasional poor-risk patients who is deemed unable to tolerate surgical resection is the Monaldi approach of intracavitary drainage. This was initially proposed as a treatment of tuberculous pulmonary abscesses and most recently advocated by Goldstraw and colleagues at the Royal Brompton Hospital. In this procedure, a small portion of rib overlying the bulla is excised subperiosteally. The pleurae and bulla are incised within a purse-string suture encompassing bulla wall and both parietal and visceral pleurae. A Foley catheter is inserted through this incision, and the purse-string suture is secured overtop of the Foley catheter balloon, which is within the bulla cavity. The catheter is connected to a usual chest drain suction device with underwater seal. This is supplemented by a talc pleurodesis of the bulla cavity and the intrapleural space.

Cystic Fibrosis

There is an approximate 10% incidence of pneumothorax in patients with cystic fibrosis. Due to the nature of their underlying lung disease, these patients may experience severe symptoms that sometimes culminate in a fatal episode. Conservative therapy with or without a chest tube may be the initial treatment of choice for these patients. This alternative is usually undertaken in an effort to avoid pleurodesis in a patient who may subsequently be a candidate for lung transplantation. It should be noted that pleurodesis, although it may increase the technical difficulty at the time of explanting the diseased lungs, does not in and of itself constitute a contraindication to lung transplantation.

Catamenial Pneumothorax

There are many theories as to the etiology of catamenial pneumothorax, the most common manifestation of intrathoracic endometriosis. Diaphragmatic fenestrations often are encountered in these patients. Catamenial pneumothorax can often be treated thoracoscopically with wedge resection of visible endometrial implants and direct suture closure of diaphragmatic fenestrations in addition to either mechanical pleurodesis or parietal pleurectomy. Nonsurgical options include hormonal suppression with gonadotropin-releasing hormone agonists such as leuprolide, oral contraceptives, and bilateral salpingo-oophorectomy.

In a prospective study, only one of eight patients who underwent surgical repair of diaphragmatic defects suffered a recurrence during a mean follow-up period of 6.6 months (range 2 to 15 months). The retrospective analysis of 10 patients with catamenial pneumothorax revealed no recurrences during a mean follow-up of 33 months (range 12 to 48 months) in the five patients treated by diaphragmatic repair. These data suggest that surgical repair of diaphragmatic defects, often requiring only simple suture closure, is associated with an excellent therapeutic outcome.

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EDITOR'S COMMENTS

When we talk about spontaneous pneumothorax, it is necessary, as the authors point out, to distinguish primary spontaneous pneumothorax from secondary pneumothorax because the treatment and approach differ for the two entities. The usual patient who presents with a primary spontaneous pneumothorax is a young man commonly with a characteristic body habitus. These young men tend to be tall and thin and may have a family history of pneumothorax. The initial management needs to be individualized, but many of these patients can be managed with simple aspiration via the second intercostal space and a repeat chest radiograph to confirm the absence of a reaccumulation of air. Even if an air leak is suspected, initial management may consist of placing a small cannula connected to a Heimlich (one-way) valve and outpatient management. For a first primary spontaneous pneumothorax, we proceed to operation only if the air leak lasts >48 hours or if the patient's occupation makes travel to remote areas likely. The chance of a recurrence approaches 30%, and thus there is a 70% chance that the patient will never have another pneumothorax. Either a recurrence on the ipsilateral side or a contralateral pneumothorax is an indication for operation. If the second pneumothorax occurs on the contralateral side, consideration should be given to operation on both sides. The operation may be done thoracoscopically or open, but I prefer a "hybrid" operation that involves a small transaxillary incision with visualization afforded by placement of the videothoracoscope through the chest tube site. Graspers and a stapling device may be placed via the transaxillary incision because the interspace, usually the second, is quite wide. Apical bulla may be stapled and mechanical pleural abrasion, which is our preference, carried out. I tend to avoid apical pleurectomy because of the increased risk of bleeding, but this procedure may be readily carried out via the transaxillary incision.

Secondary spontaneous pneumothorax, usually occurring in the patient with significant emphysema and potentially bullous disease, requires a different approach than the one just discussed. Most of these patients are poor operative candidates, and conservative management with chest tube drainage often is the preferred approach. Thoracoscopic localization of the air leak often is difficult, and frequently the outcome of an operation is the creation of additional air leaks. If a residual space is present despite adequate chest tube placement and there is continued air leak, the mobilization and transposition of a serratus anterior muscle flap into the space may be the treatment of choice, with the obliteration of the space likely to effect closure of the air leak. The approach to the patient with a secondary spontaneous pneumothorax must be individualized based on the patient's underlying clinical status and intrathoracic findings as demonstrated on a CT scan.

I would be remiss in not expanding on the issue of tension pneumothorax. The term tension pneumothorax is often misused based on a lack of understanding of the underlying physiology. The simple loss of the normal negative intrathoracic pressure leads to the entry of air into the hemithorax. This may or may not be accompanied by an ongoing parenchymal air leak. If the air leak is small, air accumulates in the pleural space and often in the soft tissue, that is, there is subcutaneous emphysema. A significant air leak usually caused by blunt or, more likely, penetrating trauma can lead to an accumulation of enough air in the chest that the mediastinum shifts and venous return to the heart is compromised. This is the definition of a "tension" pneumothorax. An air leak in a patient on positive-pressure ventilation frequently may be accompanied by tension physiology and requires emergent decompression of the pleural space with a needle placed in the second intercostal space followed by placement of a chest tube. It is the hemodynamic compromise that not only characterizes a tension pneumothorax but also threatens the patient's life, so emergent recognition and appropriate management are key.

The key to treating giant bullous disease is differentiating a giant bulla from advanced emphysematous changes that results in hyperlucency that may mimic the appearance of a giant bulla. In evaluating a giant bulla, it is critically important to determine if there is a significant amount of compressed lung parenchyma

(*continued*)

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EDITOR'S COMMENTS (*continued*)

that, with excision of the bulla, may re-expand to completely fill the residual space. A CT scan is the imaging modality of choice to evaluate the amount of underlying lung compressed by a bulla and provides the information on which to base a decision as to whether operation is likely to benefit the patient. Significant improvement in pulmonary function and relief of dyspnea may occur in the well-selected patient where the excision of a bulla allows for functioning lung to re-expand and participate in gas exchange. Simply resecting emphysematous destroyed lung is unlikely to be of benefit unless the changes are heterogeneous and localized to the upper lobe in which case the procedure is termed lung volume reduction.

LRK

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13 Diagnostic Evaluation of Anterior Mediastinal Masses and Clinical and Surgical Approach to Thymic Tumors via Sternotomy

Usman Ahmad and Frank C. Detterbeck

INTRODUCTION

Mediastinal tumors are relatively rare and can be of various origins. Their appearance is often variable, and there is overlap among different tumors. Therefore, it takes many years before one has seen enough of most types to be able to recognize patterns and develop an efficient clinical approach. This chapter presents a practical and logical approach for the workup of anterior mediastinal masses with a focus on thymic tumors, which are the most common and are the primary focus for thoracic surgeons.

The incidence of mediastinal tumors is difficult to define. This is due in part to the inclusion of benign cysts and lesions in some series and not in others. Probably most important, it is due to the inclusion of varying numbers of patients with lymphomas. Although about 50% of Hodgkin disease (HD) and 20% of non-Hodgkin lymphomas (NHL) involve the mediastinum, only about 3% of HD and 6% of NHL involve the mediastinum exclusively. While inclusion of the latter groups as mediastinal tumors seems legitimate given the diagnostic issues they present, inclusion of the former group seems inappropriate if there are enlarged lymph nodes elsewhere, such as in the neck or axilla, which are more easily accessible for biopsy.

CLASSIFICATION OF THE MEDIASTINUM

Several classification schemes for mediastinal compartments have been used. The scheme proposed by Shields is recommended and is currently used most frequently. It is based on anatomic landmarks and is easiest to translate to a computed tomography (CT) scan. This definition describes an anterior compartment, a visceral compartment, and a bilateral paravertebral compartment (Fig. 13.1). The anterior compartment extends between the posterior aspect of the sternum and the anterior surface of the pericardium and the great vessels. The visceral compartment includes the heart, great vessels, trachea, and esophagus, and is bounded posteriorly by the anterior spinal ligament (anterior surface of the vertebral bodies). The paravertebral compartment extends from this boundary to the posterior ribcage.

GENERAL APPROACH TO PATIENTS WITH ANTERIOR MEDIASTINAL TUMORS

Most of the literature regarding mediastinal tumors has been in the form of retrospective series of patients with a specific known diagnosis, and reported on characteristics of the patients. This is contrary to the clinician's needs, who already knows about the clinical characteristics of an individual patient in question and is trying to ascertain the diagnosis. This chapter therefore starts with clinical features that are clearly defined (age, gender, location of the lesion), and some features that may be well defined (duration of symptoms, associated conditions or features, particular radiographic characteristics). Additional features may be clinically occult, but can be brought out by appropriate testing (laboratory results).

The approach begins with a history and physical exam. The degree and duration of symptoms and the presence of associated diseases can provide important clues to the diagnosis. This is supplemented with a CT scan of the chest. For mediastinal masses the CT scan should always be done with intravenous contrast in order to better define the relationship of the mass to the normal (vascular) mediastinal structures. The chest radiograph (CXR) and CT allow the tumor to be assigned to one of the mediastinal compartments. Although many tumors may overlap somewhat and not be completely restricted to a single compartment, in general the epicenter of the lesion or the bulk of the mass clearly falls within one of the mediastinal areas.

The anterior mediastinum is the most common site of mediastinal tumors, and generally presents the greatest clinical challenges in making a presumptive diagnosis. Overall, data from large series show the approximate relative proportion of tumors that occur in the anterior mediastinum to be thymoma 35%, benign thymic lesions 5%, lymphoma 25% (HD 13%, NHL 12%), benign teratoma 10%, malignant germ cell 10% (seminoma 4%, nonseminomatous germ cell tumor [NSGCT] 7%), and thyroid and other endocrine tumors 15%.

Assessing the clinical characteristics and presentation and defining which mediastinal compartment a tumor is in is an important step in the strategy of how to approach these patients. Despite the variety of tumors, a reliable clinical diagnosis often can be made using the combination of demographic factors, symptoms, and radiologic findings. In many cases, a presumptive clinical diagnosis can be made with sufficient confidence to justify proceeding with a treatment plan without further confirmation.

Patients over Age 40

The proportion of anterior mediastinal tumor types by decades of age for men and women over age 40 is shown in Figure 13.2A and 13.2B. Small differences by age and gender exist, but overall thymoma is the most likely tumor type. With increasing age, substernal thyroid goiters also account for a substantial proportion. Thyroid masses are generally easily and reliably recognized by a characteristic radiographic appearance (high density), continuity with the thyroid gland, and extension posterior to the great

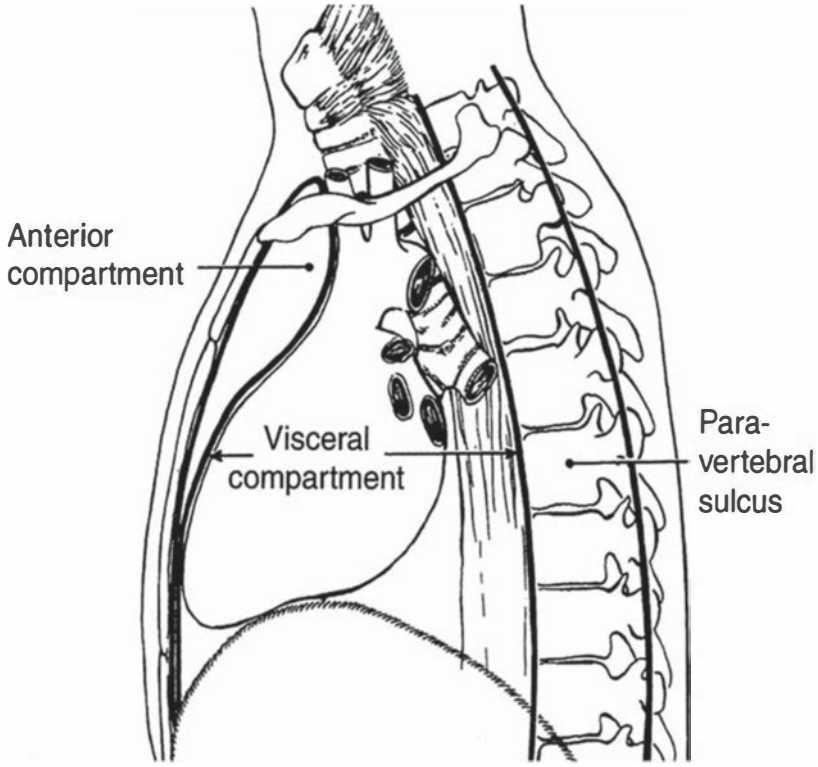


Fig. 13.1. Classification of regions of the mediastinum.

vessels. In rare instances, an iodine 133 scintigraphy scan can be useful, although it must be remembered that an iodine scan is likely to have decreased sensitivity when performed within approximately 4 weeks of a CT scan involving iodinated contrast.

Many patients (30% to 50%) with a thymoma will have an associated parathymic condition (e.g., myasthenia gravis [MG], hypogammaglobulinemia, pure red cell dyscrasia), which is virtually pathognomonic that an anterior mediastinal mass is a thymoma. In general, tissue confirmation of a thymoma or substernal goiter is not needed prior to resection.

The small remaining subset of patients (especially men and those aged 40 to 49) includes a variety of relatively rare tumors that require histologic confirmation for diagnosis. Preferably, the initial approach is a needle biopsy, but because of the variety and rarity of these tumors this may or may not be sufficient to reliably establish the diagnosis.

Women Aged 10 to 39

In women aged 10 to 39, the most prevalent tumor is HD or mediastinal large cell NHL (MLC-NHL) lymphoma (Fig. 13.3). Many have constitutional symptoms suggestive of lymphoma. In addition, although the epicenter of the mass is in the anterior mediastinum, there is often a fairly characteristic enlargement of multiple lymph nodes in the middle mediastinum and neck. Therefore, a clinical diagnosis of HD or MLC-NHL can be made quite reliably in a large proportion of women aged 10 to 39. However, this must be confirmed by a tissue biopsy. Although one may attempt a needle biopsy first, in most institutions this will be insufficient for full lymphoma characterization and a surgical biopsy will be needed (see later section on biopsy procedures).

A substantial portion of the remaining patients have a thymoma, especially those over age 20. Once again, due to the frequent occurrence of parathymic conditions that are essentially pathognomonic for thymoma, many of these patients can be very reliably identified on clinical and radiographic grounds.

The other tumor seen relatively infrequently compared to thymoma or lymphoma is a benign teratoma that can be easily identified based on radiographic and clinical features. Most are asymptomatic and when present, symptoms are typically long-standing (29% chest pain, 25% cough, 22% dyspnea, 9% pleural effusion, 1% superior vena cava [SVC] syndrome in 305 patients). The key radiographic feature is a focus of fat density, which is present in about 50%. These tumors are usually rounded with sharp margins, appear well encapsulated and cystic, and contain material of variable density, especially fat, soft tissue, and cartilage. Although 25% have calcification, less than 10% have areas resembling a bone or a tooth. Only 5% of benign teratomas are not in the anterior mediastinum (usually posterior).

Among the remaining patients, a small percentage has substernal thyroid goiter (easily recognized on a CT). In the 20 to 29 age group, about 5% of tumors in women are primary mediastinal germ cell tumors and in this group checking for elevation of alpha fetoprotein (AFP) and β–human chorionic gonadotropin (β–HCG) can be worthwhile (especially if the presentation is atypical for the more common lesions). Especially in the 10 to 19 age group, lymphoblastic NHL must be considered. These typically exhibit fulminant growth, rapid onset of symptoms, present with a large inhomogeneous mediastinal mass and often a pleural effusion. Cytologic findings are quite characteristic and sufficient to establish the diagnosis.

Men Aged 10 to 39

For men aged 10 to 39 there is no dominant tumor type (Fig. 13.3B); however, strong clues can usually be obtained from a clinical history. The rapidity of the onset of symptoms is usually the best marker. Features that are truly pathognomonic, however, are limited to a markedly elevated serum AFP or β-HCG levels, which are quite consistently found only in NSGCT and MG and establish an anterior mediastinal mass as a thymoma.

A rapid onset of symptoms (over days to several weeks) strongly suggests either NSGCT or lymphoblastic non-Hodgkin lymphoma (LB-NHL). Radiographically a bulky, inhomogeneous mass is usually seen in both, sometimes with areas of necrosis or hemorrhage. LB-NHL is more common in ages 10 to 20, and is often accompanied by a pleural effusion, "B" symptoms (fever, night sweats, weight loss), and an elevated serum lactate dehydrogenase (LDH) level. With NSGCT, however, pulmonary metastases are often seen but pleural effusions are uncommon.

The diagnosis of NSGCT is made easily and reliably by demonstrating markedly elevated serum AFP or β-HCG levels. This is present in 90% of NSGCT. A needle biopsy to confirm the diagnosis is not needed in experienced centers when the presentation is typical, as further subtyping of the tumor does not alter the treatment approach. The diagnosis of lymphoblastic NHL is usually easily made from a fine-needle aspiration of the primary mass or a pleural effusion due

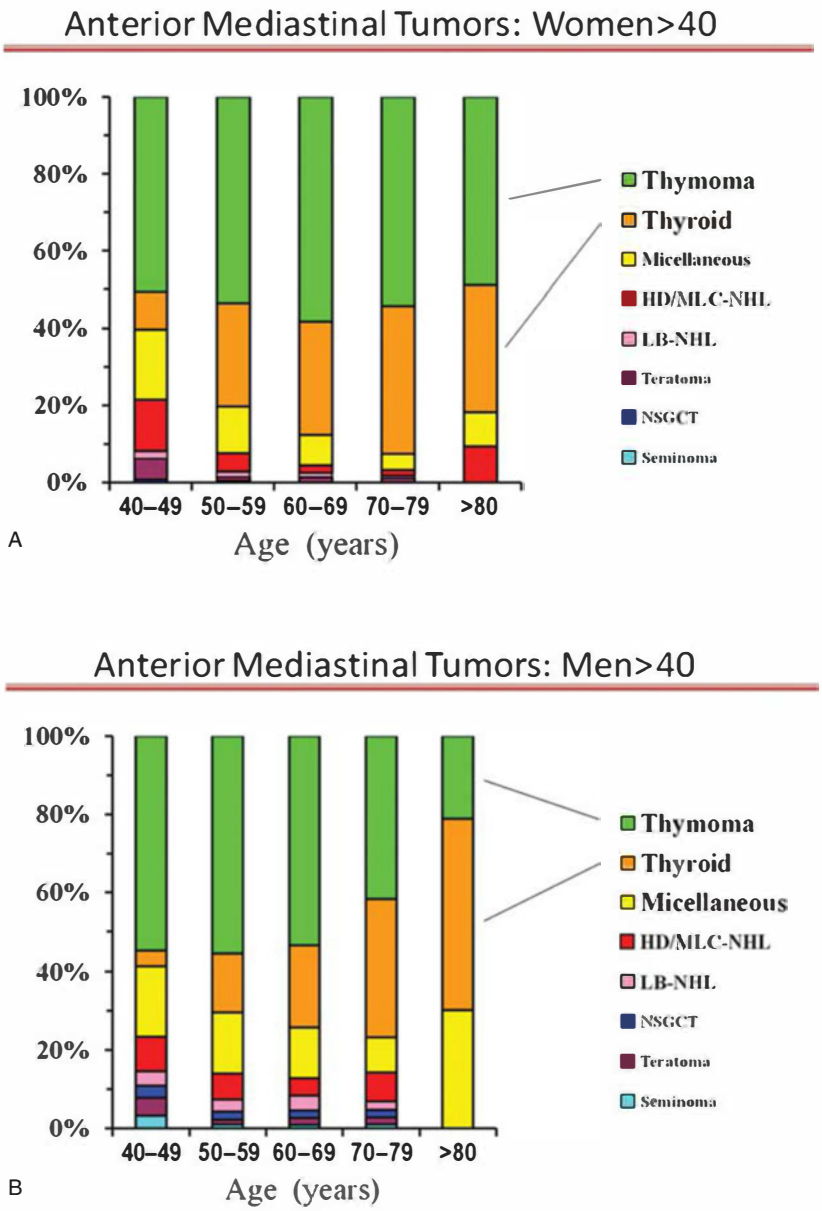


Fig. 13.2. Anterior mediastinal tumors in patients over age 40 proportion of tumor types by decades of age: (**A**) women and (**B**) men. HD/MLC-NHL, Hodgkin disease/mediastinal large cell non-Hodgkin lymphoma; LB-NHL, lymphoblastic non-Hodgkin lymphoma; NSGCT, nonseminomatous germ cell tumor.

to the presence of characteristic cytologic features. In addition, approximately 50% of patients have bone marrow involvement.

An intermediate onset of symptoms (over many weeks to months) suggests a seminoma, HL or MLC-NHL. Radiographically, these two lymphoma types usually involve multiple enlarged lymph nodes or a multilobulated mass. Frequently nodal enlargement is also seen in cervical, hilar, or upper abdominal nodes in HD, whereas the vast majority of MLC-NHL involves only the mediastinum (sometimes with pleural or pericardial effusion). In both lymphoma types, B symptoms are often present. Seminomas, on the other hand, are usually homogeneous or slightly inhomogeneous with smooth pushing margins and only rarely are accompanied by a pleural effusion but often have associated pulmonary metastases.

About 10% of seminoma patients have a low grade elevation of serum β-HCG, whereas AFP is almost always normal. An elevated LDH level is common, but this is found in lymphoma as well. A core needle biopsy usually establishes the diagnosis of seminoma and is sometimes sufficient to establish the diagnosis of lymphoma, but usually more tissue is needed (i.e., a surgical biopsy) in order to complete the subtyping that is important in selecting the appropriate therapy. Both HD and MLC-NHL often have prominent sclerosis, so that a needle biopsy typically yields only a small amount of actual tumor tissue.

In general, when symptoms are present, teratomas should be resected, although they are benign. So-called malignant degeneration of a benign teratoma has only been reported in secondary benign teratomas (residual after chemotherapy for a malignant germ call tumor), and marked growth resulting in mediastinal compression has also only been reported in this context. Therefore, an asymptomatic benign primary teratoma may be left alone. A discrete mediastinal mass suggestive of thymoma should be resected, especially in young patients. Thymic hyperplasia does not necessarily need to be resected but in the presence of MG thymectomy often is the treatment of choice. Making the distinction between a discrete thymic mass and thymic hyperplasia is often difficult. No biopsy is needed prior to resection of these lesions.

Children Aged 0 to 9

The proportion of anterior mediastinal tumor types in children aged 0 to 9 are shown in Figure 13.4. There is no dominant tumor type. A rapid onset of symptoms suggests a LB-NHL, an intermediate onset suggests HD/MLC-NHL, and more chronic symptoms a thymoma or teratoma. The same specific clinical symptoms, radiographic features, and laboratory tests as discussed in the previous section also apply to this age group. The selection of what confirmatory test to pursue is also the same.

THYMIC TUMORS

Clinical Presentation

Thymomas are relatively rare and usually indolent tumors that present with vague and subtle symptoms. Thymomas have been reported from ages 8 to 94 years. A broad peak is noted from 35 to 70 years. Patients with MG tend to present at a slightly younger age. While one-third of patients with thymoma present with MG, another one-third are asymptomatic. Approximately 40% of patients with thymoma present with local symptoms, while 30% have systemic symptoms. Cough, dyspnea, and chest pain are the most common symptoms. Dyspnea can result from local compression as well as from the neuromuscular effects of MG. Parathymic syndromes are common and aid in diagnosis (e.g., MG in 30%, pure red cell aplasia, and hypogammaglobulinemia in 2% to 5% each).

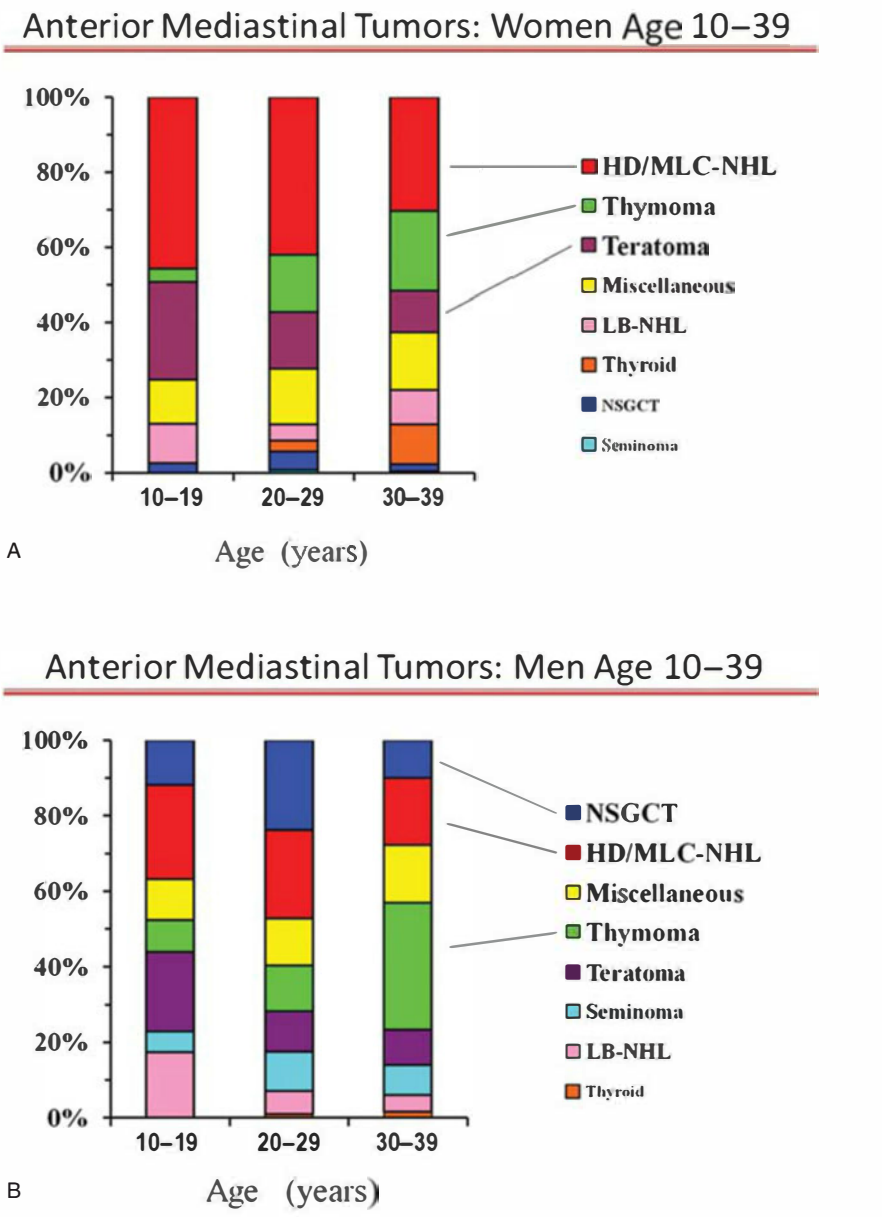


Fig. 13.3. Anterior mediastinal tumors in patients age 10 to 39: proportion of tumor types by decades of age: (**A**) women and (**B**) men. HD/MLC-NHL, Hodgkin disease/mediastinal large cell non-Hodgkin lymphoma; LB-NHL, lymphoblastic non-Hodgkin lymphoma; NSGCT, nonseminomatous germ cell tumor.

Imaging Characteristics

Radiographically, CT scan has high sensitivity (97%) in detecting these tumors. Intravenous contrast is helpful in determining vascular anatomy and relationship with the tumor. Thymomas typically appear as well-defined round or oval masses, generally anterior to the great vessels but can wrap around these structures. Curvilinear calcifications are seen in 10% to 20% of patients. Extension into mediastinal fat or pleura can be suggested on CT, however, is unreliable. The presence of smooth or lobulated contour, homogeneous enhancement, absence of areas of low attenuation, absence of pericardial or pleural effusion, and absence of calcification in the tumor favor the presence of a thymoma or a well-differentiated thymic carcinoma. A recent study showed that tumors that are greater than 7 cm in greatest diameter, infiltrate mediastinal fat, and have lobular appearance are more likely to be stage III/IV thymomas. Tumor size had the most robust association with advanced stage. In addition, pleural nodules were associated with stage IV disease.

Diagnosis

Using the combination of symptoms, associated conditions and radiologic findings, most thymomas can be reliably differentiated from other anterior mediastinal masses. A clinical diagnosis of thymoma is sufficient to plan operation for localized tumors. Tissue diagnosis is recommended for extensive tumors that require neoadjuvant chemotherapy or if a nonoperative approach is planned. Sometimes it is difficult to differentiate thymoma from lymphoma due to lack of symptoms and equivocal radiologic findings; in this case open surgical biopsy is recommended.

Tissue diagnosis can be achieved through FNA or open biopsy (anterior parasternal mediastinotomy, thoracoscopy). The success rate is higher with open biopsy (90%), as compared to FNA (62%). Standards for performance and reporting of microscopic findings of needle biopsies of suspected thymomas have been established and should be followed. The suspicion of tumor seeding at biopsy site or needle tract is not substantiated by evidence or the experience at major centers where this has been commonly performed.

Histologic and Stage Classification

Despite their indolent behavior, and frequent lack of malignant cytologic features, all thymomas (regardless of initial stage or histologic type) have been noted to develop extrathymic invasion, local recurrence, and metastasis after resection, and therefore should not be labeled as "benign" tumors. The majority of thymic tumors have nonmalignant-appearing thymic epithelial cells mixed with variable proportions of lymphocytes. A small proportion of cases has cells with malignant features and is labeled as thymic carcinoma. These tumors account for less than 10% of cases in surgical series and about 15% overall and have more aggressive local and systemic behavior and worse prognosis. The World Health Organization (WHO) classifies thymic tumors as type A, AB, B1, B2, B3, and thymic carcinoma. However, tumor heterogeneity is common, and application of the WHO classification has been found to be associated with significant interobserver variability.

Multiple staging schemes have been proposed; the Masaoka classification with Koga modification is endorsed as the current international standard by the International Thymic Malignancy Interest Group (ITMIG). The scheme (Table 13.1) is focused on local extension of the primary tumor with less emphasis on nodal involvement (this is the natural history of majority of thymomas, while thymic carcinomas can have nodal spread).

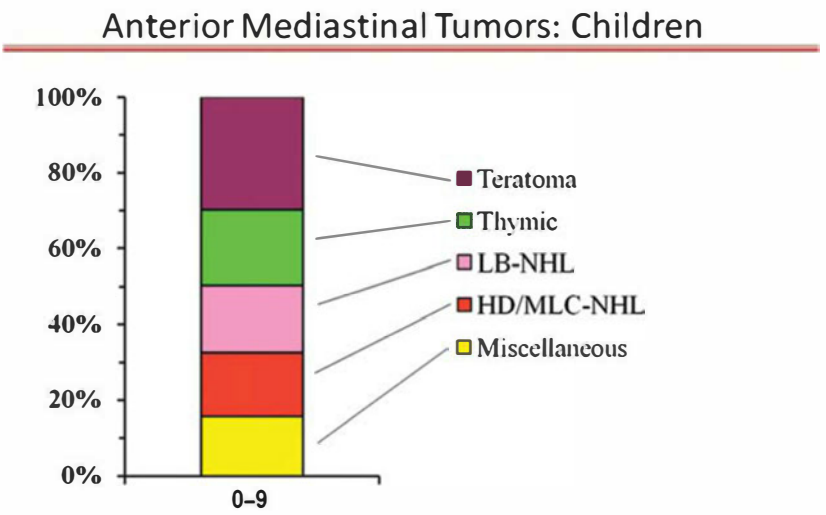


Fig. 13.4. Anterior mediastinal tumors in children under age 10: proportion of tumor types by years of age. HD/MLC-NHL, Hodgkin disease/mediastinal large cell non-Hodgkin lymphoma; LB-NHL, lymphoblastic non-Hodgkin lymphoma.

In general, approximately 40% of thymomas present as stage I lesions, 25% each at stages II and III, 10% at stage IVa, and 1% to 2% at stage IVb. Invasion into mediastinal tissue (stage II, III) is present in 50% of thymomas, with pleural invasion being most commonly followed by pulmonary and pericardial invasion. Approximately 30% of these cases have involvement of the innominate vein or SVC and 20% have phrenic nerve involvement. Direct extension is also seen into the aorta and pulmonary artery (11%) and chest wall (8%).

Surgical Resection

Perioperative care should involve specific anesthetic considerations and neurologic consultation in the case of MG. Preoperative medical optimization is critical. Plasmapheresis has been shown to improve postoperative recovery and outcomes in myasthenic patients. Postoperative complications have been reported in approximately 19% (range 7% to 32 %) of cases in series including patients with and without MG. These mostly include pneumonia, respiratory complications, and exacerbation of myasthenia. With modern perioperative care operative mortality is less than 1%.

The standard surgical approach for resection of a thymoma is a median sternotomy because of the following factors: (1) all thymomas are malignant tumors; (2) it is critically important to perform a complete resection; and (3) it remains to be demonstrated that the excellent results with open resection can be duplicated by other approaches. The standard approach also involves a total thymectomy, although the evidence and rationale for this are weak. Especially in patients with MG, an extended thymectomy (includes en-bloc resection of thymus gland and surrounding areolar tissue and fat) is recommended due to frequently found ectopic rests of thymic tissue in the surrounding mediastinal fat, although the data are only suggestive of a benefit with this approach.

A complete resection has consistently been found to be of major prognostic importance in the treatment of thymomas of every stage. This means that surgeons should make every effort necessary during the resection to achieve this, with no hesitation to resect additional structures. Furthermore, it means that a surgeon should not embark on an operation if there is the possibility of needing to resect a structure he/she is not prepared to deal with (e.g., chest wall, SVC). The opportunity to cure a patient of a malignant disease is often lost because of an intraoperative compromise resulting from a lack of understanding of the nature of the thymoma and limited anticipation of potential intraoperative findings. There should be a low threshold to refer the occasional patient with a higher stage thymoma to a larger center if there is a possibility of encountering situations beyond an institution's routine experience.

Standards for exploration of the role of a minimally invasive approach to resection of a thymoma have been published. A minimally invasive approach for what may be a thymoma should only be done in experienced centers, should not compromise the oncologic principles (complete, en-bloc resection of the thymus and any potentially involved adjacent structures), should avoid disruption of the specimen, mark the orientation and the location of any potentially questionable margins on the specimen, and be willing to convert to an open procedure without hesitation if any oncologic principles are compromised.

Operative Technique

After performing a standard sternotomy and placement of sternal retractor, the thymus gland is exposed and an assessment of tumor burden and involvement of extrathymic sites is performed. It is often easiest to begin with the right lower pole of the thymus; this is mobilized away from the pericardium and diaphragm. Even in the absence of gross tumor involvement, the fat pad lying in this region should be resected with the gland. All fatty tissue is removed from the surface of the pericardium using blunt dissection. The right pleura reflection is opened and the phrenic nerve is identified. The pleura is incised starting at the inferior portion of the right hilum toward the apex of the chest staying about 1 cm anterior to the phrenic nerve. The pleura is typically resected as the lateral most margin of dissection, along with the adjacent fatty tissue, which is swept medially and anteriorly by blunt dissection, separating it from the nerve. It is useful to place a marking stitch on the mediastinal pleural surface because it can be very difficult for the pathologist otherwise to identify it.

If there is any suspicion of involvement of the lung or pericardium, a portion of these tissues should be resected en bloc with the tumor. One should be very hesitant to categorize such abutment as adherence and to sharply separate the tissues. Furthermore, frozen section examination is notoriously inaccurate for thymoma invasion and should be used with caution. There is little downside to performing a wedge resection of the lung or resection of the anterior pericardium.

In case of unilateral abutment of the tumor against the phrenic nerve, the nerve should be resected unless there is significant preoperative respiratory impairment. If the phrenic nerve is encased unilaterally it should be resected (usually it is already not functioning). If both nerves are involved, one nerve must be preserved, either by sharply dissecting the skeletonized nerve from the tumor or rarely by performing a subtotal tumor resection (in which case one should have probably not attempted resection in the first place). However, there

Table 13.1 ITMIG Definition of Details of the Masaoka-Koga Staging System

Stage	Definition
I	Grossly and microscopically completely encapsulated tumor
	This includes tumors with invasion into but not through the capsule, or
	Tumors in which the capsule is missing but without invasion into surrounding tissues
IIa	Microscopic transcapsular invasion
	Microscopic transcapsular invasion (not grossly appreciated)
b	Macroscopic invasion into thymic or surrounding fatty tissue, or grossly adherent to but not breaking through mediastinal pleura or pericardium
	Gross visual tumor extension into normal thymus or perithymic fat surrounding the thymoma (microscopically confirmed), or …
	Adherence to pleura or pericardium making removal of these structures necessary during resection, with microscopic confirmation of perithymic invasion (but without microscopic extension into or through the mediastinal pleura or into the fibrous layer of the pericardium)
III	Macroscopic invasion into neighboring organ (i.e., pericardium, great vessel, or lung)
	This includes extension of the primary tumor to any of the following tissues:
	Microscopic involvement of mediastinal pleura (either partial or penetrating the elastin layer);
	Microscopic involvement of the pericardium (either partial in the fibrous layer or penetrating through to the serosal layer);
	Microscopically confirmed direct penetration into the outer elastin layer of the visceral pleura or into the lung parenchyma;
	Invasion into the phrenic or vagus nerves (microscopically confirmed, adherence alone is not sufficient);
	Invasion into or penetration through major vascular structures (microscopically confirmed);
	Adherence (i.e., fibrous attachment) of lung or adjacent organs only if there is mediastinal pleural or pericardial invasion (microscopically confirmed).
IVa	Pleural or pericardial metastases
	Microscopically confirmed nodules, separate from the primary tumor, involving the visceral or parietal pleural surfaces, or the pericardial or epicardial surfaces,
b	Lymphogenous or hematogenous metastasis
	Any nodal involvement (e.g., anterior mediastinal, intrathoracic, low/anterior cervical nodes, any other extrathoracic nodes)
	Distant metastases (i.e., extrathoracic and outside the cervical perithymic region) or pulmonary parenchymal nodules (not a pleural implant)

ITMIG, International Thymic Malignancy Interest Group.

is limited data that substantiates these recommendations. In a series of 18 stage III patients, Yano et al. reported a lower tumor recurrence rate after phrenic nerve resection (as well as reduced vital capacity and forced expiratory volume in 1 second). However, no difference in overall survival was noted in the two groups.

Dissection is continued cephalad, dissecting under the insertion of the strap muscles on the undersurface of the manubrium. Tissue is swept medially, taking care to not inadvertently injure the right internal mammary vein. As this is continued cephalad the right upper pole of the thymus begins to emerge. It is dissected from the surrounding fat and traced toward the thyroid using blunt dissection. Generally, the upper pole attachments thin out and break off; otherwise they can be ligated close to the thyroid gland. The free upper pole is then reflected downward and the upper mediastinal fat surrounding the innominate vein is carefully dissected to free up the gland. During this dissection, the thymic artery is ligated. Small venous tributaries draining into the innominate vein are also ligated. A similar technique is then used to dissect the left side of the gland. The dissected tissue can be lifted up or inverted to dissect the underlying mediastinal fat. The pleural spaces should be inspected, and appropriate nodal stations sampled if not already resected and marked. The sternum is approximated using steel wires, after placement of chest tubes.

Standards have been established by ITMIG to promote better surgical outcomes and to better define pathologic reporting of results. The main principles are marking and orientation of the specimen, clear communication with the pathologist, and a systematic approach to nodes and adjacent structures, and documentation. During dissection, any areas of concern (close margin, tissue disruption) should be marked immediately on the specimen as well as in the patient. The resected specimen should be oriented by the surgeon, and given to the pathologist on a mediastinal board or diagram. Several aspects of the specimen should be routinely identified (tissue adjacent to the innominate vein, pericardium, and the mediastinal pleura). Areas of intraoperative partial disruption of the specimen should be marked and communicated to the pathologist so that there is no misrepresentation of such an area as a positive margin. A sketch or digital photo of the specimen and marking sutures is encouraged (Fig. 13.5).

Any mediastinal or surrounding lymph nodes that carry clinical suspicion of tumor involvement (firm, enlarged, or FDG avid) should be resected and appropriately marked. For small encapsulated tumors, resection of anterior mediastinal and adjacent nodes is encouraged. For stage III and IVa tumors, an anterior mediastinal lymphadenectomy is recommended and a systematic sampling of intrathoracic nodal stations is encouraged. For thymic carcinoma at least a systematic sampling is recommended of anterior mediastinal, intrathoracic, supraclavicular, and low cervical nodes.

Handling the Specimen

Marking the specimen during the course of resection, as noted above, is important. Orientation of the specimen by the surgeon and communication with the pathologist is critical to obtaining a pathology report that can be interpreted clearly. Standard areas that should be marked especially for large tumors, include "surface adjacent to pericardium and innominate vein," "surface adjacent to SVC," and "surface adjacent to pleura," if these structures are not resected en bloc. If marking stitches are used, they

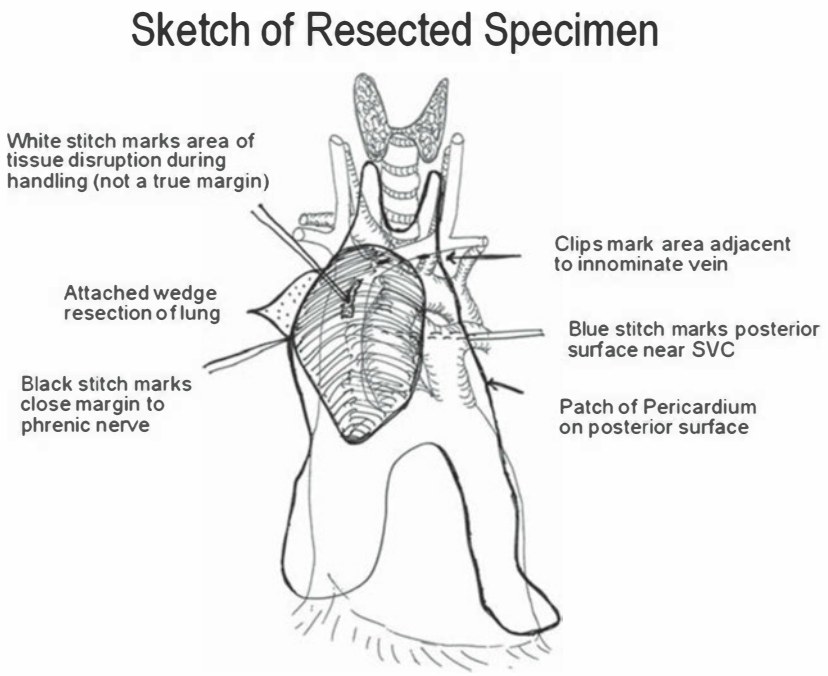


Fig. 13.5. Sketch of the oriented specimen on a mediastinal diagram depicting marking stitches and additional resected structures diagrammatically, which minimizes subsequent confusion and aids communication.

should be placed deep into the specimen to avoid tearing the loose areolar tissue on the surface. Area of potentially positive margins must be marked and communicated to the pathologists, as well as areas that do not represent a true margin, but in which the tissue overlying the tumor is either absent or has been disrupted during the dissection.

There are also standards for how the pathologist should process and report the findings. Surfaces should be identified with different color inks or a similar technique. Because of the heterogeneity of thymoma, at least five sections should be taken to improve the reliability of defining the histologic type. There are also standards for the pathologists to follow in reporting the status of the capsule, the margins, and the distance to the margins that address issues specific to thymoma that have created confusion in the past.

Survival after Resection

Various outcome measures have been reported, which cause fairly dramatic differences and limit the ability to compare past publications; standards have been established to promote comparisons and collaboration. Because many thymomas are relatively indolent, both 5- and 10-year outcomes should ideally be reported (however, 5-year outcomes are sufficient for thymic carcinoma).

Overall, 10-year survival rates of 90% for stage I and 70% for stage II thymomas are typical. The vast majority of these cases is amenable to complete resection. The majority of deaths in patients with stage I or II thymoma is due to unrelated causes. Overall survival with or without complete resection is 55% for stage III and 35% for stage IVa; however, R0 resections result in longer survival. The most important factors that predict survival are tumor stage and completeness of resection. The presence of MG is not an independent prognostic factor; however, MG may play a role in detection of tumors at an early stage due to the presence of symptoms.

Recurrence Rates

Given the fact that recurrence is not always associated with death and vice versa, the best outcome measure for thymic malignancies is probably freedom from recurrence. Resected stage I tumors have average recurrence rate of 3%, which increases to 16% for stage II and 26% for stage III. Variable recurrence rates are noted for stage IV tumors. The mean time to recurrence was reported as 10 years for stage I and 3 years for stages II to IV.

Local recurrence is more common than extrathoracic recurrences. However, what has been classified as a local recurrence has varied among studies; a standard definition for future reports has been developed. A pleural and/or pericardial nodule is the most common recurrence pattern. Distant metastases are seen most commonly in liver and bone.

Factors associated with a lower rate of recurrence are Masaoka stage I and II, histologic type (especially thymoma vs. thymic carcinoma), smaller tumor, and a complete resection.

Preoperative Therapy

There are no randomized trials involving preoperative chemotherapy; however, retrospective studies have shown promising results. Several series have consistently reported increased R0 resection rates after preoperative chemotherapy (72%), as compared to surgery alone (50% for stage III, 25% for stage IV). Chemotherapy resulted in radiographic objective response rate of approximately 90%, and pathologic complete response rate of approximately 20%. In one series, downstaging (stage III→II) was reported in 16% of patients. Preoperative chemotherapy should be considered for clinical stage III/IVa tumors to increase their likelihood of R0 resection. Chemotherapy has also been used for recurrent thymomas followed by resection or radiation therapy (RT) as a curative-intent strategy.

Most chemotherapy regimens are cisplatin based and are well tolerated. It is recommended that resection be performed within 8 weeks of preoperative chemotherapy. If the patient is not a surgical candidate and curative-intent radiation is planned, it should be initiated within 6 weeks of chemotherapy.

Adjuvant Radiation Therapy

The role of adjuvant RT to the operative field is controversial and various practices have been reported. Since most recurrences are local (intrathoracic), it may seem intuitive to add radiation; however, the recurrent tumor is not always in the surgical bed, but may present as nodules on the parietal pleura and pericardium.

Interpretation of the reported data on adjuvant RT is difficult because of limited number of patients, studies spanning many decades, and frequent reporting of completely and incompletely resected patients together. The recurrence rate for completely resected stage I tumors is low and the data do not suggest this is influenced by adjuvant RT. Therefore, in general, adjuvant radiation is not employed for

completely resected stage I thymomas. For completely resected stage II–III tumors, the data do not clearly indicate a difference in recurrence; in fact in the largest study higher recurrence rates were noted after RT (presumably because the patients getting radiation were considered at higher risk). Several systematic reviews and meta analyses have reported similar findings.

RT seems to lower recurrence rates in the case of incompletely resected stage III and IV tumors. In particular, RT was noted to decrease mediastinal recurrence after subtotal resection.

RESECTION OF NONTHYMIC ANTERIOR MEDIASTINAL TUMORS

Resection of nonthymic anterior mediastinal masses can be performed using similar principles. Some anterior mediastinal tumors, such as lymphoma, are treated with chemotherapy and do not require resection. Nonthymic tumors do not require a complete thymectomy as described for thymomas. Surgical excision must however entail resection of all involved structures, such as pericardium, lung, SVC, and so on, just as described for thymic tumors. Limited resection can be safely undertaken for smaller masses through partial sternotomy.

SUMMARY

Thymoma is the most common anterior mediastinal tumor followed by thyroid masses in patients over age 40. The most common anterior mediastinal tumor in women 10 to 39 years old is lymphoma followed by thymoma. In men aged 10 to 39, there is no predominant tumor type; however specific symptoms, chronicity of symptoms, radiographic, laboratory, and pathologic findings can help diagnose the lesion.

Thymomas are indolent, malignant tumors that have a propensity for local spread. Approximately one-third of patients with thymoma have MG. CT scan is highly sensitive in diagnosis and certain features can predict the presence of advanced stage disease. Thymic carcinoma is a rare tumor type that is characterized by malignant histologic features and aggressive tumor invasion. The Masaoka-Koga staging scheme is most commonly used and is based on local tumor invasion.

Excellent 5- and 10-year survival rates are noted for completely resected early stage thymomas. Complete resection is the best prognostic factor for these tumors. While stage I/II tumors undergo primary surgery, preoperative chemotherapy appears to increase the chances of complete resection for stage III/IVa tumors. Postoperative radiation can be considered for patients with residual disease.

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EDITOR'S COMMENTS

Compared to lung tumors mediastinal tumors are rare indeed and the average thoracic surgeon may see only a few in any given year. As the authors point out the two most common lesions seen in the anterior mediastinum are thymoma and lymphoma with the incidence of each varying according to the age of the patient. Thymoma is a rare tumor in children while lymphoma is significantly more common. Differentiating between lymphoma and other lesions remains a critical distinction since surgical resection does not contribute to outcome for lymphoma.

For any lesion other than a well-encapsulated one, it is hard to criticize the surgeon who chooses to pursue an open incisional biopsy either via anterior mediastinotomy or videothoracoscopy. Anterior mediastinotomy can be performed as an extrapleural procedure thus eliminating even the small possibility of intrapleural seeding. Remaining extrapleural is facilitated by removing a portion of the second costal cartilage and simply sweeping the pleural reflection laterally as one approaches the lesion which can be easily visualized. I prefer to use a mediastinoscopy biopsy forceps to sample the lesion, taking care to know exactly where the aorta and pulmonary artery are located. The anesthetic management of patients with large mediastinal masses requires special consideration. Especially in those patients with a mass that has rapidly increased in size, usually a large B-cell lymphoma, the airway may be compromised when the patient is placed supine as airway compression from the tumor may occur. In these situations it is safest to keep the patient in the semi recumbent position and proceed with an awake intubation. These situations are unusual but must be kept in mind.

The authors refer to thymomas as "malignant." I prefer to use the terms "encapsulated" and "invasive" when referring to these lesions and avoid the term "malignant." It is difficult to put these lesions into the category of malignancy based on their indolent natural history but we do treat the invasive lesions as if they were "malignant." Complete resection is the gold standard and, as the authors point out, the surgeon needs to be prepared to take en bloc whatever structures are involved by the lesion. These include portions of the pericardium, lung, superior vena cava, and the innominate vein. If pleural lesions are noted at the time of resection these should also be taken with a parietal pleurectomy. If a phrenic nerve is involved it should be taken if by doing so a complete resection will be accomplished. For locally advanced lesions consideration should be given to obtaining a tissue diagnosis and treating with neoadjuvant chemotherapy prior to surgical resection as there is evidence that what might have been less than an R0 resection may indeed be one following induction therapy. Total thymectomy and not just tumor resection should always be carried out.

The decision to pursue postoperative radiation therapy following resection of a thymoma should be made jointly by the surgeon and the radiation oncologist. The incidence of local recurrence does not seem to be different when patients with resected stage II lesions received postoperative radiation therapy compared to those who receive resection alone.

The decision to resect a substernal goiter needs to be based on patient factors, specifically the patient's age and whether the patient is symptomatic specifically with airway compromise. Essentially all of these lesions, though appearing to be well within the mediastinum, may be resected via a cervical incision and only very rarely is sternotomy necessary.

Germ cell tumors, in particular nonseminomatous lesions, are initially treated with induction chemotherapy followed by resection to remove all residual gross disease. The histologic findings in the treated lesion determine prognosis as well as the need for additional therapy. Findings range from complete necrosis with no viable cells to mature teratoma to the persistence of tumor with sarcomatous elements. Again the intent of the surgeon should be to accomplish a complete resection. Pure seminomas, though rare, may be treated with primary radiation therapy. These are more commonly seen in testis tumors. Primary mediastinal germ cell tumors in women are exceedingly rare.

LRK

14 Transcervical Thymectomy

Larry R. Kaiser

INTRODUCTION

With so much emphasis today being placed on new minimally invasive techniques, it is easy to forget that some minimally invasive procedures have been around for years but seemingly never referred to as such. In a sense, one can make a strong case that transhiatal esophagectomy is a minimally invasive operation since it avoids a chest incision and we have been doing that procedure for many years, thanks to Mark Orringer popularizing it back in the 1970s. Other procedures have been "minimized" with smaller incisions, muscle sparing, and, of course, with the use of endoscopes.

Removal of the nontumorous thymus gland for myasthenia gravis classically involved a median sternotomy and despite the efforts of some to utilize a so-called mini sternotomy, at least a portion of the sternum is still split. Jaretski in New York described even a more "maximal" approach utilizing a neck incision combined with a sternotomy making the case that aberrant "rests" of thymus occurred in locations that even a standard sternotomy might miss (Fig. 14.1). It occurred at least to a few surgeons that performing a sternotomy to remove a normal thymus gland was overkill. Working in the same city, Papatestas resurrected the transcervical approach to thymectomy during the 1970s and 1980s with results, relative to myasthenic symptoms, similar to those obtained via the much larger and more morbid operation. Cooper made a major modification to the transcervical approach by designing a retractor that instead of strictly relying on blind blunt dissection, as practiced by Papatestas, allowed for direct visualization of the anterior mediastinum to assure that all of the thymus gland was removed. Both pleural reflections could be visualized; the inferior extent of the gland and the extension of the gland into the aortopulmonary window could also be directly visualized. The Cooper thymectomy retractor allowed for an extended transcervical thymectomy, an approach more predictable and reliable.

Recognizing that a median sternotomy is a big operation with significant attendant morbidity simply to remove a normal structure that easily dissects away from surrounding structures with blunt dissection further made the case for transcervical thymectomy. However, there remained a significant group of vocal opponents to the procedure based on their contention that a transcervical approach failed to remove the entire thymus gland and certainly would miss rest of the gland that, according to Jaretski, commonly occurred in aberrant locations. The objection fundamentally was based on the contention that complete removal of the entire thymus gland was absolutely necessary if one was to achieve a complete remission of myasthenic symptoms. Whether or not that contention is valid still remains an open question. This has been further called into question since in at least one study it has been shown that in patients with ectopic thymic rests, even when removed, the incidence of complete remission remains significantly inferior to that seen in those who do not have ectopic gland. Thus, in a setting in which no procedure results in 100% remission rates, we need to look critically at the type of operation and specifically at the risk-benefit ratio involved in removing what is essentially a normal structure. It has been our hypothesis that transcervical thymectomy should be the preferred approach for the removal of the thymus gland in patients with myasthenia gravis. We believe that the risk-benefit ratio is favorable enough to make the case that essentially all patients with myasthenia should be offered thymectomy via this approach. Even those neurologists who are concerned about subjecting their patients to a surgical procedure often are swayed when they are informed about the minimally invasive nature of this procedure and the results when compared with the open procedure.

DESCRIPTION OF THE PROCEDURE

No specific preoperative preparation is required prior to proceeding with transcervical thymectomy. As opposed to thymectomy via the standard sternotomy approach, plasmapheresis is not necessary even for those patients with significant symptoms. It has been our practice to allow the attending neurologist to decide whether plasmapheresis should be done and if they wish to proceed the exchanges should be done the week prior to the planned operative procedure. We have patients take their usual medication on the morning of the operative procedure, specifically their anticholinesterase inhibitor and, if still taking it, the corticosteroid. We attempt to wean patients to no more than 10 or at most 20 mg of prednisone prior to operation.

The patient is positioned supine with an inflatable bag behind the shoulders which, when inflated, allows for maximal hyperextension of the neck (Fig. 14.2). In the office during the preoperative evaluation, we assess the degree of neck extension recognizing that in those patients with limitation to extension the procedure is somewhat more difficult with respect to visualization. The skin overlying the neck and chest is prepared in the usual manner and the appropriate draping is carried out. A small transverse incision is made as low as possible in the midline of the neck at the level of the sternal notch and carried down through the subcutaneous tissue and through the platysma down to the level of the strap muscles. It has been my preference to make this incision with a scalpel as opposed to using an energy device. Once the strap muscles are visualized, the midline raphe is identified and incised using the scissors. The midline is opened superiorly as high as possible and inferiorly down to the level of the sternal notch. The ligament traversing the sternal notch is incised with the electrocautery. The sternohyoid and sternothyroid muscles on one side are grasped by the assistant and again

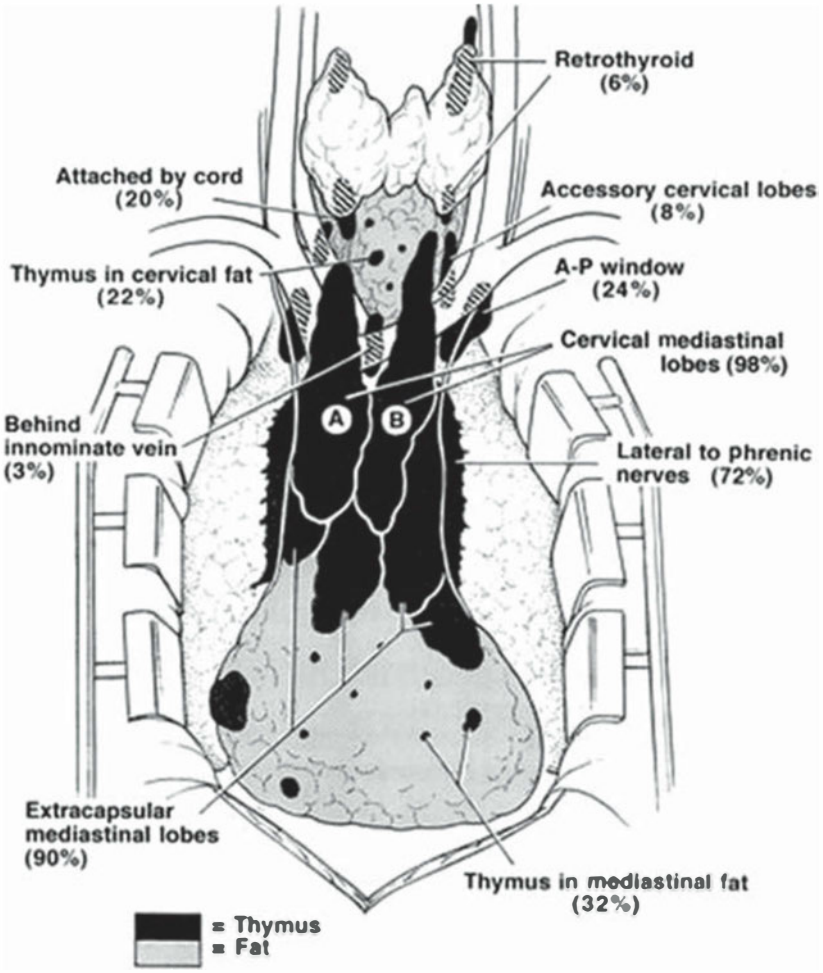


Fig. 14.1. Potential locations of aberrant thymus gland as defined by Jaretzki utilizing a combined cervicomediastinal approach. (From Jaretzki A, Barohn RJ, Ernstoff RM, et al. Myasthenia gravis: recommendations for clinical research standards. Ann Thorac Surg 2000;70:327-334.)

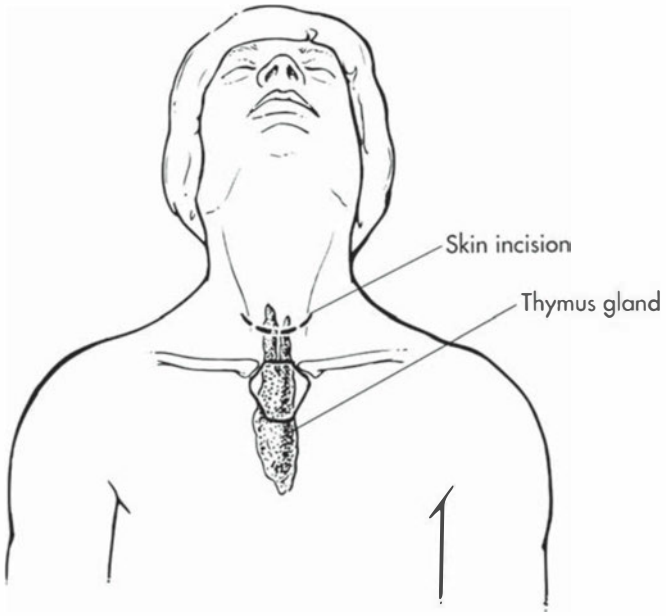


Fig. 14.2. The patient is positioned supine with the neck hyperextended. Incision is made at the level of the sternal notch. (Fig. 32.1 from Kaiser LR. Atlas of General Thoracic Surgery. WB Saunders Co; 1997.)

using the scissors dissection is carried closely along the posterior border of the sternothyroid muscle reflecting the muscle carefully away from the underlying tissue, which will be the capsule of the thymus gland. The gland has a definite salmon pink color that easily differentiates it from the surrounding cervical fat and in addition the capsule defines the gland as a separate structure. The gland is followed superiorly to its proximal extent where a small arterial branch will be encountered. A clip is placed and the gland elevated anteriorly while dissecting it away from surrounding structures both medially and laterally. A silk tie is placed at the superior most aspect of the pole of the gland to be used as a "handle" to elevate the gland and apply countertraction (Fig. 14.3). The gland is followed inferiorly to where it intersects with the opposite lobe. As described above, the opposite lobe is reflected away from the strap muscles, followed to its termination superiorly and then dissected away from the surrounding structures moving inferiorly.

As the gland is mobilized from superior to inferior, the innominate vein comes into view at the level of the sternal notch. The gland is carefully lifted anteriorly as it is bluntly dissected off the innominate vein using a peanut sponge. Venous tributaries from the gland to the innominate vein are visualized and are either clipped and divided or ligated and divided (Fig. 14.4). The number and the size of venous tributaries vary significantly from patient to patient but are easily visualized as the gland is elevated and the vein pushed down with the peanut sponge. After all of the venous tributaries have been divided, the gland is completely free of the innominate vein and now using a ball sponge in a ring forceps the gland is bluntly reflected off the pericardium as far inferiorly as possible. With the ball sponge, the gland is bluntly freed away from the sternum anteriorly.

At this point, with the gland separated from the innominate vein following division of the venous tributaries, the Cooper thymectomy retractor is placed. Clamps for the retractor arms are placed over the drapes onto the operating table and the arms are inserted and secured. The retractor blade is placed behind the sternal notch and lifted as far as possible before securing it thus almost lifting the patient off the table. Army-Navy retractors are placed in the lateral aspect of the wound and secured to the retractor with penrose drains as shown (Fig. 14.5). The inflatable bag is deflated to optimize the retraction further facilitating the view into the

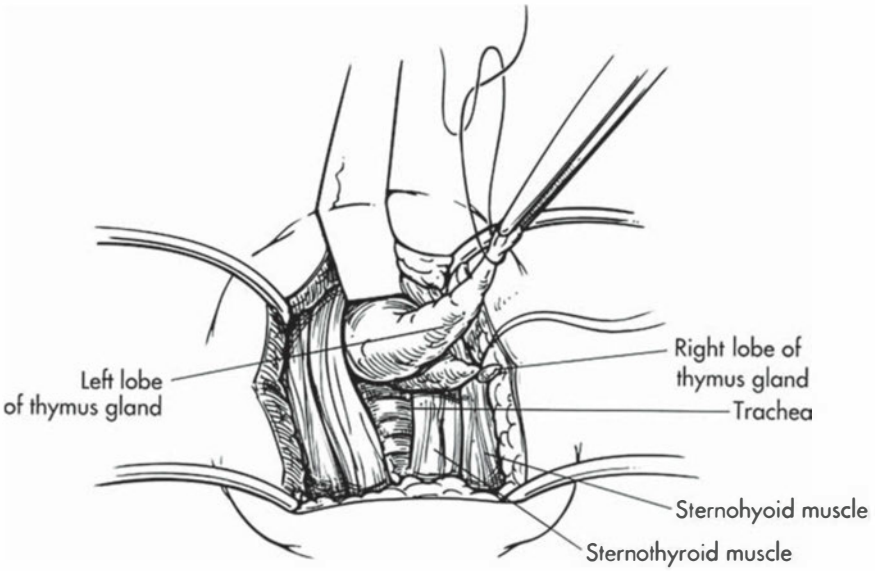


Fig. 14.3. The gland is elevated anteriorly away from surrounding tissue. A silk ligature is placed on the superior aspect of each pole to be used to apply countertraction. (Fig. 32.2 from Kaiser LR. Atlas of General Thoracic Surgery. WB Saunders Co; 1997.)

mediastinum. Under direct visualization, the right lobe of the gland is bluntly freed away from the right pleural reflection using the ball sponge and the gland is further freed off the pericardium to its termination inferiorly and brought over toward the left side (Fig. 14.6). Once at the inferior extent of the gland is visualized, it can be elevated and brought from inferior to superior. The left lobe of the gland is bluntly mobilized away from the left pleural reflection and followed inferiorly toward the aortopulmonary window. The gland often extends fairly far inferior and must be followed to its termination. This is the most difficult part of the procedure but care must be taken to remove the entire gland. One should be aware that the phrenic nerve can be easily damaged either with the cautery or if sharp dissection is used. Recall that the nerve courses anteriorly at its superior extent in the mediastinum and thus can be easily injured if care is not taken. The nerve usually may be visualized.

By completely freeing up the right lobe of the gland and using it as countertraction, the entire left lobe may be mobilized and elevated from inferior to superior. The entire gland is now completely freed away from the pericardium and brought up superiorly for delivery out of the wound. Occasionally, one or more venous tributaries not previously recognized may need to be divided to completely free up the gland and remove it. If a pleural space has been entered during the dissection, it is easily recognized by ballooning of a pleural reflection with ventilation. A thorough inspection should be made to assure hemostasis, and occasionally a small bleeder may be found in the mediastinal or pleural fat. Care should be exercised in using the electrocautery to avoid thermal injury to the phrenic nerve on either side.

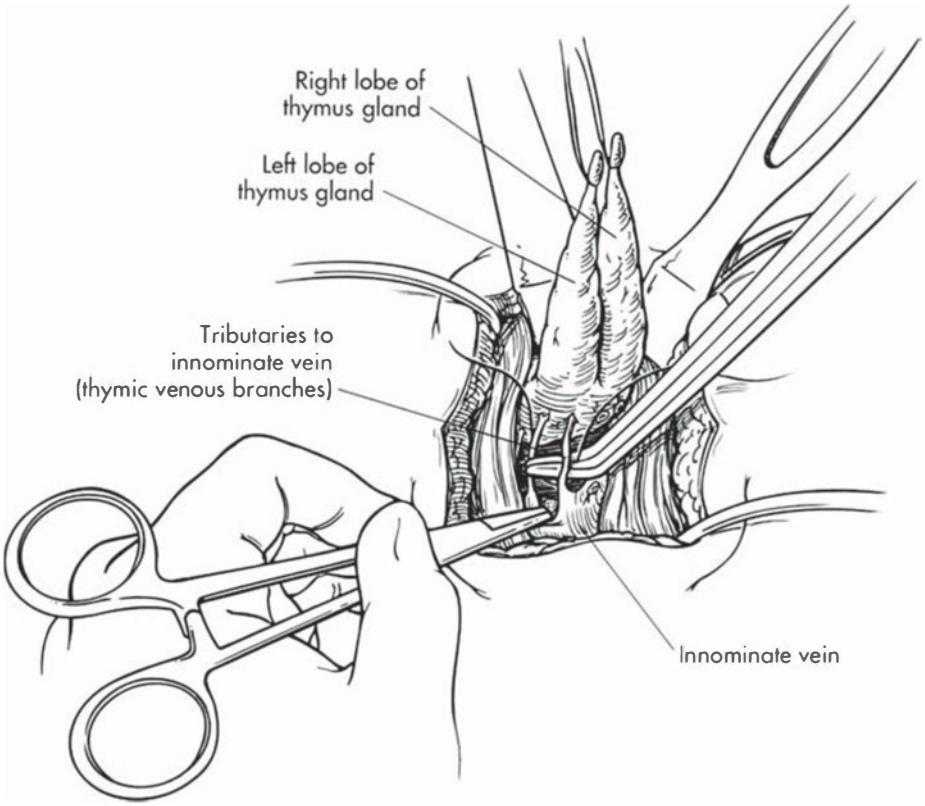


Fig. 14.4. As the gland is elevated, the innominate vein is easily visualized and venous tributaries, which can be quite variable in location, number, and size, are either clipped or ligated and divided to completely free the gland away from the vein. (Fig. 32.3 from Kaiser LR. Atlas of General Thoracic Surgery. WB Saunders Co; 1997.)

If a pleural space has been entered simply placing a red rubber tube through the pleural rent and bringing it out through the neck allows the air to be removed via a valsalva maneuver just prior to completing the skin closure. The wound is closed in layers and the skin closed with a subcutaneous suture to avoid having to remove sutures. The patient is awakened and extubated in the operating room and then brought to the postanesthesia recovery unit. A chest radiograph is performed to assure no significant residual pneumothorax if a pleural space has been entered. There is no need for any further treatment since there is no ongoing air leak. Myasthenia medications are resumed within a couple of hours and the patient may be discharged home the same day with mild analgesics to be used as needed.

Patients experience mild-to-moderate incisional pain and some have some neck pain from the hyperextension. The pain usually subsides within 2 to 3 days. Patients are reminded that myasthenic symptoms relief occurs over time and the time course may be quite variable with the maximum response not seen in some until two or more

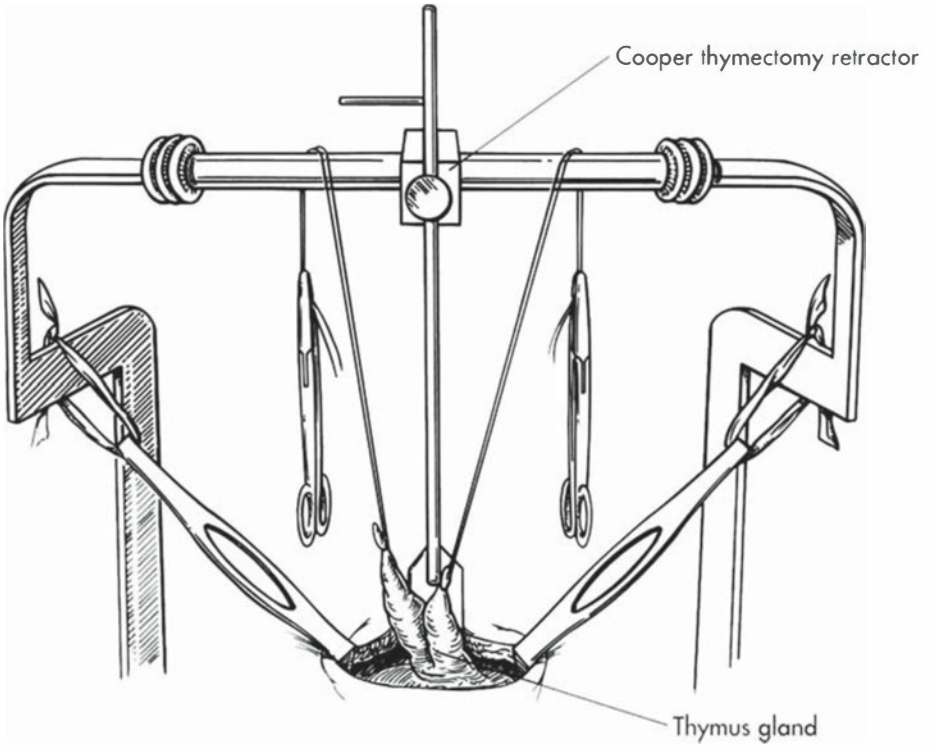


Fig. 14.5. The Cooper thymectomy retractor in place with army-navy retractors held in place with penrose drains. (Fig. 32.4 from Kaiser LR. Atlas of General Thoracic Surgery. WB Saunders, Co; 1997.)

years after thymectomy. The neurologist adjusts medication based on symptoms.

When interpreting the results following thymectomy for myasthenia gravis, one needs to assess the number of complete responses defined as the alleviation of all symptoms with the patient on no medication. That being said, some neurologists prefer to leave their patients on small doses of corticosteroids for the duration. As mentioned above, the number of complete responders increases over time, and there is no predictable time course to a response thus it is critically important when evaluating results in a series to look at the Kaplan-Meier probability of achieving a complete response. We can expect that somewhere between 40% and 50% of patients will achieve a complete response over time, another 40% a partial response with a significant reduction in medication required and only about 10% or 20% fail to achieve any response.

In our series of now over 200 patients with a median follow up of 54 months, the complete response rate is slightly over 40% with another 40% achieving a significant partial response. Looking at the

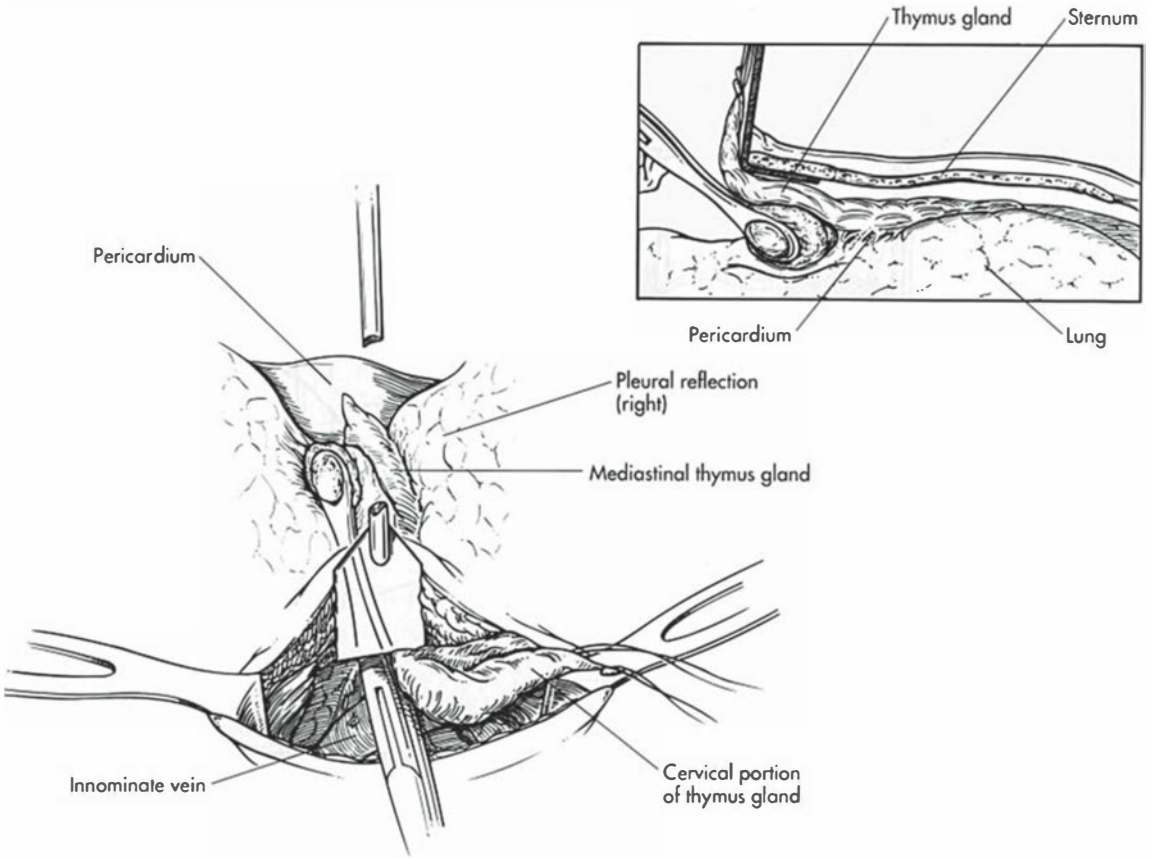


Fig. 14.6. Using the ball sponge the gland is freed away from the pericardium to its inferior extent. The gland is also mobilized away from both the right and left pleural reflections. (Fig. 32.5 from Kaiser LR. Atlas of General Thoracic Surgery. WB Saunders, Co; 1997.)

Kaplan-Meier probability of achieving a complete response, there is a 45% complete response rate at 72 months. The only factor that was statistically significant in terms of predicting response was the preoperative Osserman class. The less severe the disease (lower Osserman class) the more likely to achieve a complete response. Fewer than 20% of patients, when last followed, had no improvement. Interestingly, in our initial series only about 10% of patients remained unimproved but as we gained experience we became less selective in our approach operating on some patients where visualization was severely limited by the inability to extend the neck or by obesity. Morbidity for our series was 7% with only one patient experiencing an injury to a recurrent laryngeal nerve. We observed two pleural effusions, two pneumothoraces, and two wound infections. One patient developed pneumonia.

Cooper looked at the long-term outcome in 52 patients undergoing transcervical thymectomy between 1977 and 1986 with a mean follow-up of 8.4 ± 6.1 years and noted a mean decrease from a preoperative Osserman grade of 2.7 to 0.4 at last follow-up. [1]

CONCLUSIONS

Extended transcervical thymectomy achieves Kaplan-Meier complete response rates (45%) very similar to those achieved either by sternotomy or combined cervicosternotomy approach ("maximal" thymectomy) (50%). It is likely that remission rates would be equivalent or better depending on patient selection for transcervical thymectomy. Patients with minimal or no neck extension and obese patients are not ideal candidates for the transcervical approach. Despite the objection by some that transcervical thymectomy is, by definition, going to leave residual thymus gland, at least one study shows that patients with ectopic thymic tissue, even when removed, have decreased remission rates compared with those found not to have any ectopic thymus. Thus, it must be concluded that total removal of all thymic tissue should be the goal but it is likely that small amounts of residual gland may also allow for patients to achieve a complete remission over time.

The transcervical approach may also have applicability for other procedures in the anterior mediastinum. We have used this approach in combination with the endocrine surgeons when an aberrant parathyroid gland is located in the mediastinum. The thymectomy using the Cooper retractor may be done at the same time as the parathyroidectomy precluding a second operation. Complete removal of the thymus gland under direct vision, as opposed to removing it blindly, usually results in identification of the aberrant gland. In addition, we have used the transcervical approach to remove encapsulated thymomas up to approximately 4 cm in size. However, we have no hesitation in opening the sternum if the tumor appears to be invading a surrounding structure or the phrenic nerve on either side is at risk. As long as the tumor is not disrupted and can be completely removed and we do not violate oncologic principals, we feel that the transcervical approach is reasonable. We also have, on multiple occasions, used the approach to obtain a biopsy of an unknown anterior mediastinal mass in lieu of a parasternal mediastinotomy.

Transcervical thymectomy is a hard operation to teach because it essentially is a single operator procedure with only a limited part time view by a trainee. The use of video equipment may facilitate training but most centers do relatively few thymectomies in any given year, thus making it even more difficult to reinforce the technical details. In most centers, with only a few exceptions, median sternotomy, either complete or partial, remains the preferred approach to thymectomy VATS thymectomy is utilized except some, but this operation requires the placement of a double lumen endobronchial tube, lateral incisions that require one to traverse the pleural reflection to get to the mediastinum and limited access to the neck. Others have described the use of the robot for thymectomy taking what essentially is a simple procedure, transcervical, and making it significantly more complex and costly. In this era of minimally invasive surgery, transcervical thymectomy, a throwback to the past, remains a true minimally invasive procedure that requires no expensive equipment.

SUGGESTED READINGS

Bril V, Kojic J, Ilse W, Cooper JD. Long term clinical outcome after transcervical thymectomy for myasthenia gravis. Ann Thorac Surg 1998;65:1520-1522.

Jaretzki III A, Barohn RJ, Ernstoff RM, et al. Myasthenia gravis: recommendations for clinical research standards. Ann Thorac Surg 2000;70:327-334.

Ozdemir N, Kara M, Dikman E, et al. Predictors of clinical outcome following extended thymectomy in myasthenia gravis. Eur J Cardiothorac Surg 2003;23:233-237.

EDITOR'S COMMENTS

Dr. Kaiser certainly makes a convincing case for transcervical thymectomy. He is quite correct in avoidance of sternotomy. This can be very helpful if this operation can be done correctly. The fact that many patients done either way do not get complete response certainly makes it much more preferable to do the operation through a smaller incision.

Dr. Kaiser also makes a case that in difficult situations partial or full sternotomy is certainly an appropriate way to do this operation. I think this approach is important and Dr. Kaiser and his group have great expertise at doing this procedure safely.

ILK

15 Resection of Posterior Mediastinal Lesions

Joseph B. Shrager

The posterior mediastinum is bounded anteriorly by the posterior pericardium and extends posteriorly to the chest wall and laterally to include the costovertebral sulci. It contains the descending thoracic aorta, inferior vena cava and azygous veins, the sympathetic chains, and origins of the intercostal nerves at their nerve roots, and it is generally defined to include the esophagus and associated vagi as well. Most anatomic systems consider only lesions that are caudal to the fourth thoracic vertebral body to be within the posterior mediastinum, with more cephalad lesions resting within the superior mediastinum.

The majority of posterior mediastinal masses occurring in adults are benign. These lesions can be usefully classified according to whether they are cystic or solid on radiographic evaluation. Cystic masses in this region typically represent *bronchogenic cysts* (Fig. 15.1) or *esophageal duplication cysts*, whereas solid masses are most commonly *benign neurogenic tumors* (Figs. 15.2 and 15.3; e.g., schwannomas, neurofibromas, or ganglioneuromas). These neural tumors usually arise from the sympathetic chain or the proximal intercostal nerves, but sometimes the exact anatomical origin does not even become clear intraoperatively. Occasionally, one comes across a patient with a pheochromocytoma or paraganglioma, which arise from the randomly located mediastinal paraganglionic cells and may secrete hormones. At initial office evaluation, one should be alert to the presence of hypertension or palpitations that should dictate measurement of urine metanephrines. Esophageal leiomyomas (benign intramuscular tumors within the esophageal wall) are also generally grouped among the posterior mediastinal lesions. The approach to these lesions and to esophageal duplication cysts differs somewhat from the approach to lesions that are unassociated with the esophagus.

In many cases, posterior mediastinal masses come to light as asymptomatic radiographic abnormalities. They may uncommonly, however, present with signs of infection (in the case of infected cysts), or with dysphagia, chest pain, respiratory complaints, or neurological changes (e.g., Horner's syndrome from tumors involving the upper sympathetic trunk; lower extremity symptoms from dumbbell tumors) resulting from mass effect on adjacent structures. Esophageal lesions can, of course, cause dysphagia but they too are often asymptomatic.

At present, because of the growing availability of less morbid, minimally invasive approaches to posterior mediastinal masses, most authors recommend resection even when lesions are asymptomatic. I believe that the recommendation about whether to proceed with resection of a posterior mediastinal mass needs to be individualized. Clearly, a symptomatic lesion is best managed by resection, except in unusual circumstances. When an asymptomatic lesion has all of the radiographic characteristics of a benign cyst or tumor (i.e., smooth margins, simple-appearing cyst material, minimally positron emission tomography PET) positive if PET has been done), then the age and general medical condition of the patient should be the key considerations. Since these lesions do generally grow—albeit at a fairly slow rate—it is likely that a young patient will eventually develop symptoms due to impingement on surrounding structures or infection of a cyst. Patients under the age of 60, then, who are otherwise good surgical candidates, are best managed by resection. For older patients, or those who have substantial comorbidities, it is perfectly reasonable to follow these benign-appearing, asymptomatic lesions with radiographic studies and operate only if they begin to grow dramatically, the patient develops symptoms, or a change in radiographic appearance suggests malignancy. In any case, there is no urgency to remove these cysts and tumors. The significance of the very few reports of development of malignancy within a benign posterior mediastinal mass is likely overstated.

SURGICAL PRINCIPLES

Video-Assisted Thoracoscopic Surgery vs Thoracotomy

Resection of posterior mediastinal masses may be accomplished by means of either video-assisted thoracoscopic surgery (VATS) or thoracotomy. The procedure, apart from the incisions, is essentially the same with either approach, and the goal is complete resection. With some exceptions, VATS is considered preferable to thoracotomy in this setting. It has been well established that VATS results in less postoperative pain and quicker functional recovery. Some surgeons argue that a VATS approach may be more likely to leave a patient with microscopic residual disease. In our experience and that of others, however, recurrences of these lesions are very rare after VATS excision. Given the low recurrence rate and the fact that these masses are almost always benign, we believe that the risk–benefit ratio favors VATS in most cases.

There are, however, several *circumstances in which thoracotomy is indicated from the outset.* A suggestion of malignancy (in particular, frank invasion of surrounding structures) on preoperative imaging mandates exploration and resection by thoracotomy; in this situation, the potential consequences of positive margins justify the more aggressive approach. The presence of active infection within a cyst is a relative indication for thoracotomy, in that it can cause disruption of normal tissue planes and thereby render VATS dissection more difficult and possibly hazardous. Masses larger than approximately 6 cm also call for an open approach in my opinion: such lesions are typically more difficult to mobilize safely from surrounding structures than smaller lesions, they are more likely to be malignant (though this is still rare), and their removal between the ribs is likely to necessitate substantial rib spreading, which may negate the benefit of true VATS. Another approach is to attempt

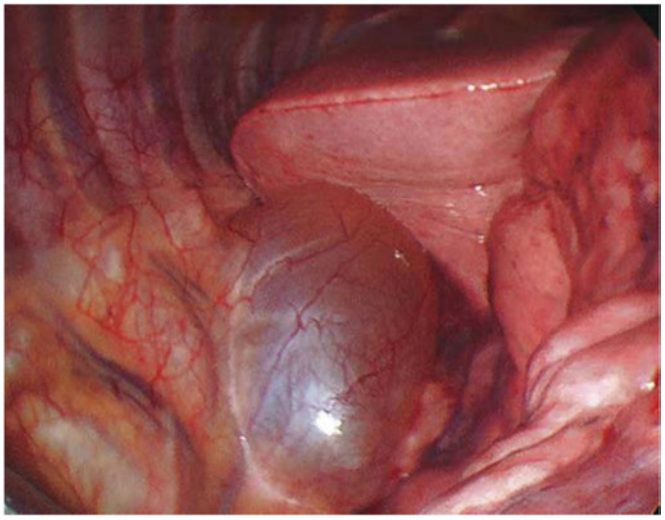


Fig. 15.1. Intraoperative view of a typical benign bronchogenic cyst of the posterior mediastinum.

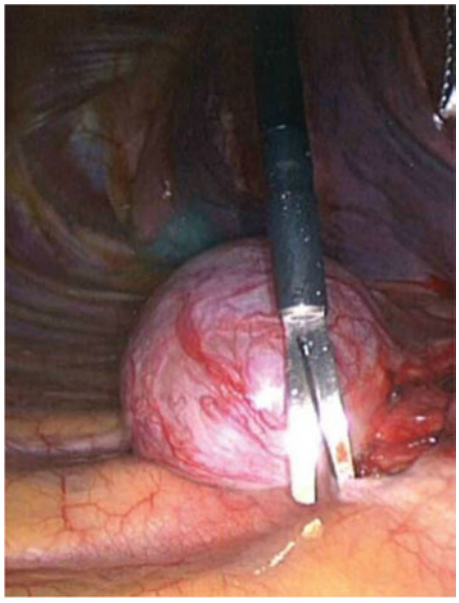


Fig. 15.2. Intraoperative view of a typical benign neurogenic tumor in the costovertebral sulcus.

VATS for these larger lesions, but to have a low threshold to convert to thoracotomy if difficulty is encountered. If the dissection can be completed by VATS, one can then typically remove larger tumors by resecting a small portion of the rib without sacrificing an intercostal nerve. It is likely that this will result in less pain and earlier recovery than a standard thoracotomy with rib spreading, but this has to my knowledge never been studied.

Other Preoperative Issues

Any patient with a centrally located cyst should undergo bronchoscopy to rule out the rare occurrence of a communication with the bronchial tree. This may be suggested on computed tomography (CT) scans by the presence of an air–fluid level. If a communication is identified, strong consideration should be made to proceed with thoracotomy rather than VATS. *When a cyst arises from or abuts the esophagus*, the possibility of a communication between the cyst and the esophageal lumen should be similarly investigated. To rule out this phenomenon, I obtain barium esophagography during the preoperative workup, followed by esophagoscopy in the operating room prior to the start of the operation. If a communication is identified, thoracotomy is performed. After excision of an esophageal duplication cyst with a communication, reapproximation of the esophageal mucosa is a paramount consideration; in my view, this is still most reliably carried out through an open approach.

In cases of suspected *leiomyoma* of the esophagus, preoperative investigation with esophagoscopy should be done to confirm the presence of intact overlying mucosa, which is virtually pathognomonic of this disease. If the mucosa is intact, the possibility of malignancy is essentially ruled out. Simultaneously, endoscopic ultrasonography may be performed to establish the depth to which the esophageal wall is involved. With a preoperative diagnosis of probable leiomyoma, VATS is the approach of choice in our practice.

The so-called *dumbbell neurogenic tumors* (tumors that invade the neural foramen and have a spinal canal component) are special cases (Fig. 15.4). Any solid mass in the costovertebral sulcus that cannot be clearly separated on CT imaging from the neural foramen should be evaluated by means of magnetic resonance imaging (MRI). Although invasion of the neural foramen by tumor is not in itself an indication for thoracotomy, it does necessitate a combined approach with neurosurgical involvement for the intraspinal portion of the procedure. Several versions of such an approach have been described, including a posterior approach via costotransversectomy or extension of a midline incision into a posterolateral thoracotomy, through which both the intraspinal and intrathoracic components of the tumor



Fig. 15.3. Computed tomographic image of a typical benign neurogenic tumor in the costovertebral sulcus.

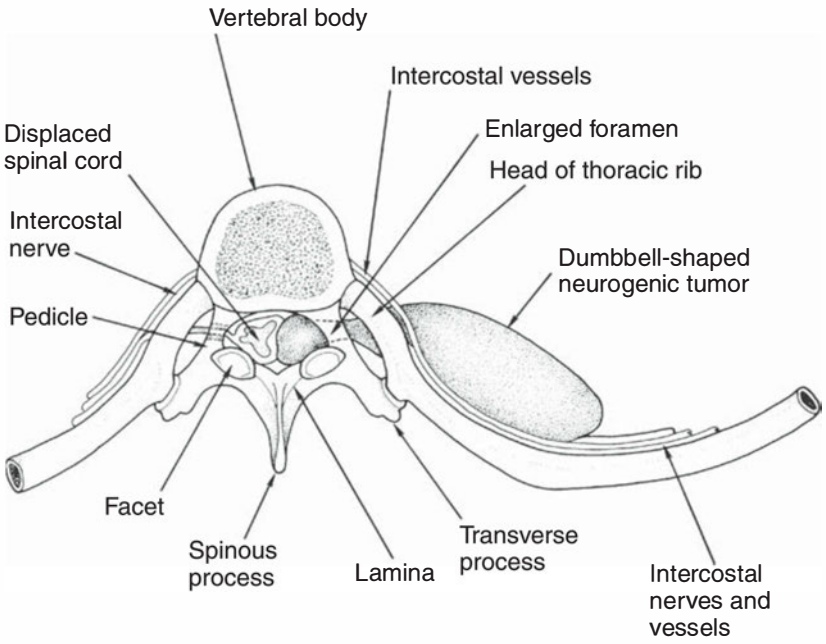


Fig. 15.4. A neurogenic dumbbell tumor showing compression of the spinal cord, enlarged intervertebral foramen, and a large posterior mediastinal tumor component.

can be resected. We prefer to perform the following approach: under a single anesthetic, the neurosurgeons first resect the intraspinal component (laminectomy and intervertebral foraminotomy), then the patient is repositioned to lateral decubitus and we carry out the remainder of the procedure (generally via VATS). Failure to diagnose a dumbbell tumor preoperatively and plan an appropriate combined operation may lead to inadvertently cutting through tumor from within the chest. This can result in tumor hemorrhage within the spinal canal and spinal cord compression with disastrous consequences.

Patients with functioning paragangliomas or pheochromocytomas, if hypertensive, should receive approximately 2 weeks of α-adrenergic blockade and volume loading, followed by β blockade. Clear discussion about the intraoperative anesthetic and anti hypertensive plan should be undertaken between the surgeon and the anesthesiologist in these cases before incision.

There is in the vast majority of cases no advantage to preoperative needle biopsy of posterior mediastinal lesions, although this can usually be readily performed either transthoracically, or via the transesophageal route in the case of periesophageal masses. Only in cases of invasive malignancy might this alter the therapeutic approach. For example, extremely large or invasive-appearing lesions that turn out to be sarcomas may be best treated by preoperative chemotherapy and/or radiation. It is therefore appropriate to obtain a needle biopsy in the unusual cases when these features are present.

Although VATS is often an excellent approach to posterior mediastinal lesions, it must be emphasized that one should never hesitate to convert a VATS procedure to a thoracotomy if required. Accordingly, informed consent to undergo thoracotomy should be sought before operation from all patients being treated for posterior mediastinal lesions, even when VATS is the intended approach. Further, any patient with a tumor encroaching upon the neural foramen should understand preoperatively that there is a very rare possibility of spinal cord compromise from the operation, as well as of cerebrospinal fluid (CSF) leak.

SURGICAL TECHNIQUES

Video-Assisted Thoracoscopic Surgery Resection of Neurogenic Tumors of the Posterior Mediastinum

Resection of a solid neurogenic tumor of the posterior mediastinum that *does not* invade the neural foramen (Fig. 15.3) proceeds as follows. The intraoperative photographs are selected from several separate VATS operations that the author has performed.

Step 1: intubation and endoscopy. The patient is intubated with a double-lumen endotracheal tube to allow single-lung ventilation. Preoperative bronchoscopy (for cystic lesions) or esophagoscopy (for lesions abutting the esophagus) is performed as indicated (see above).

Step 2: patient positioning and placement of ports. The patient is placed in the lateral decubitus position and stabilized with a bean bag so that the operating table can safely be tilted as much as 45 degree anteriorly. With this degree of tilt, the lung will almost always fall away from the field of vision; thus, there is usually no need to place an additional port for a lung retractor.

The port for the scope is placed through an incision in approximately the anterior axillary line at the level of the mass; if it is placed much more anteriorly than the anterior axillary line, the surgeon's view of these posterior lesions may be obscured by the lung. The two working ports are placed through separate incisions also in approximately the anterior axillary line, made as far cephalad and caudad as possible. Sometimes, the placement of an alternative upper working port posterior to the scapula is advantageous. The main working instruments are an endoscopic scissors-cautery, a ring clamp, an endoscopic peanut dissector, a Maryland dissector, a long right-angle clamp, a hook-cautery, and an endoscopic clip applier. I prefer a 5 mm, 30 degree thoracoscope. The 30 degree lens provides much greater versatility and visualization around to the "far side" of lesions than a 0 degree lens, and a 10 mm scope provides no advantage over 5 mm to justify the increased chance of intercostal nerve injury. I prefer to use 2 cm incisions for the working ports, without gas insufflation, to allow a variety of instruments to be passed without difficulty.

Step 3: incision of pleura. The parietal pleura is incised, with a margin of approximately 1 cm circumferentially around the mass (Fig. 15.5). The pleura can be tented up with the aid of the right-angle clamp or the Maryland dissector to separate it from the underlying structures. This separation allows the use of the electrocautery, which provides hemostasis while protecting the underlying esophagus, vagus and intercostal nerves, and azygos vein. This dissection can also be done with the hook-cautery to both create the lift from underlying structures and incise the tissue. This dissection and all subsequent work are facilitated by placing gentle traction on the mass with a sponge stick or, for smaller masses, by grasping the entire mass within a ring clamp to allow manipulation. This sort of manipulation must be minimized in the case of a

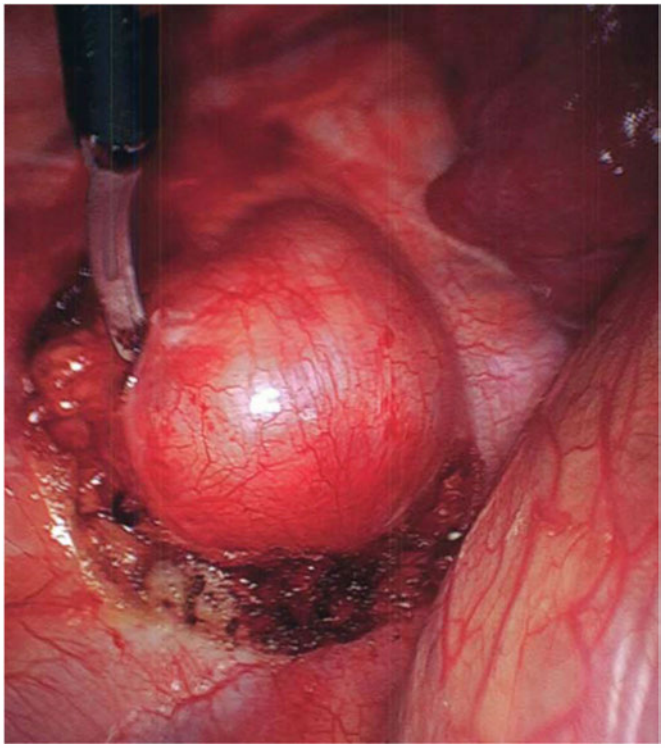


Fig. 15.5. The pleura is incised circumferentially, approximately 1 cm from the tumor.

functioning paraganglioma/pheochromocytoma to avoid marked hypertension.

Step 4: dissection of soft tissue attachments. Once the pleura has been incised circumferentially, the soft tissue attachments are further dissected bluntly with the endoscopic peanut dissector. Attachments that are relatively thick or vascular are controlled with the hook-cautery when remote from surrounding structures, or by double-clipping and division when closer to danger. Often, one needs to use cautery to dissect the tumor and associated periosteum cleanly off the surface of one or two ribs above or below the tumor (Fig. 15.6). By progressively dissecting in a circular motion around the tumor, dividing all attachments to the mass, one moves progressively closer to the source of the tumor, most commonly from an intercostal nerve. The attachment to the nerve is typically more easily and safely addressed when most of the other attachments have already been divided. When the mass has been mobilized off of all other surrounding structures, gentle dissection at the medial and lateral margins of the tumor, just inferior to the associated rib, will identify the intercostal bundle (or sympathetic chain) that is the source of the lesion (Fig. 15.7).

Step 5: division of source intercostal bundle. The source intercostal bundle, first lateral to the tumor, is mobilized, doubly clipped, and divided (Fig. 15.8). Once this has been accomplished, further blunt and cautery dissection are performed until the nerve root emerging from the neural foramen and the associated intercostal vessels are the last remaining attachments (Fig. 15.9). If the tumor originates from the sympathetic chain, the chain is clipped above and below the tumor, and the intercostal bundle is spared if it is clearly uninvolved.

Step 6: removal of specimen. After dividing any remaining soft tissue attachments, the "live" end of the intercostal bundle, medial to the tumor, is dissected and then doubly clipped and divided (Fig. 15.10), and the mass is removed in an endoscopic bag (Fig. 15.11). The typical appearance of the operative field at the end of the procedure is shown in Figure 15.12.

Step 7: drainage. A 24F chest tube is positioned posteriorly at the apex.

Potential Problems and Their Avoidance

Care must be taken to ensure that only very gentle traction is exerted on a mass adjacent to the neural foramen. Overzealous traction can cause tearing of the nerve root proximal to the extraspinal extent of the dura, and this tearing can lead to a CSF leak, which most often becomes evident only postoperatively (in the form of persistent clear chest tube output). The diagnosis of CSF leakage can be confirmed by measuring the β_2-transferrin level in the fluid. If CSF leakage is confirmed, reoperation with a neurosurgeon is mandatory; the site of the leak is repaired and buttressed with vascularized tissue.

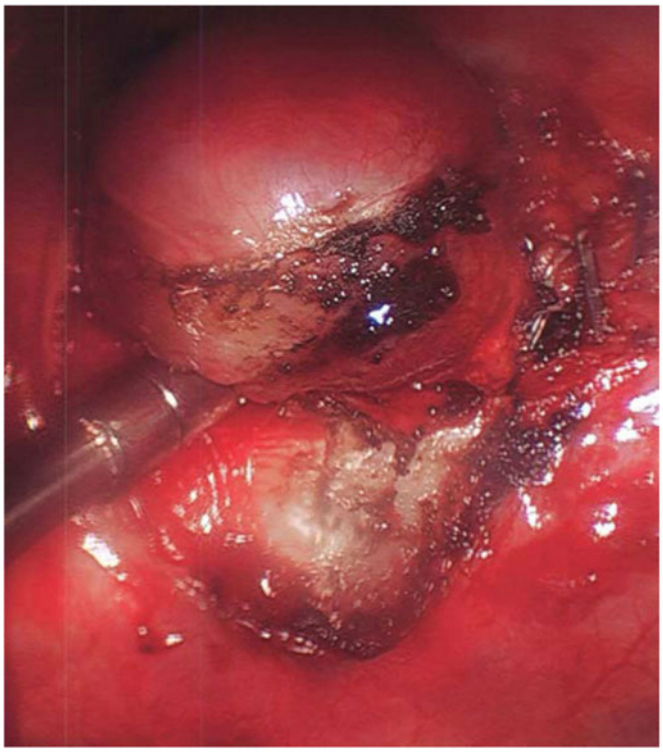


Fig. 15.6. As additional attachments are dissected, one often must cauterize directly on the periosteum of surrounding ribs to assure a clean resection, as shown here.

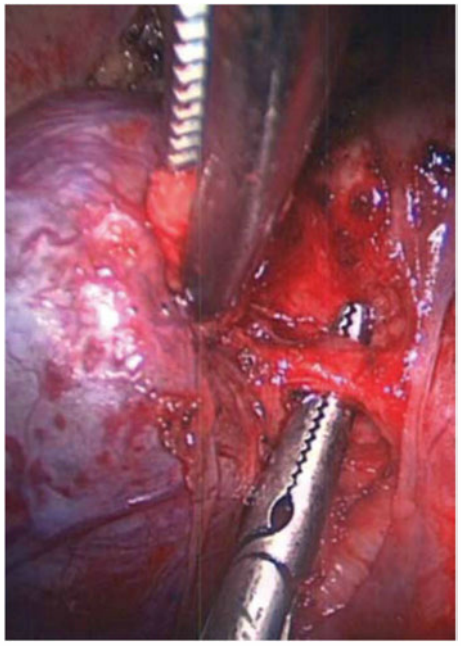


Fig. 15.7. The associated intercostal bundle is isolated next.

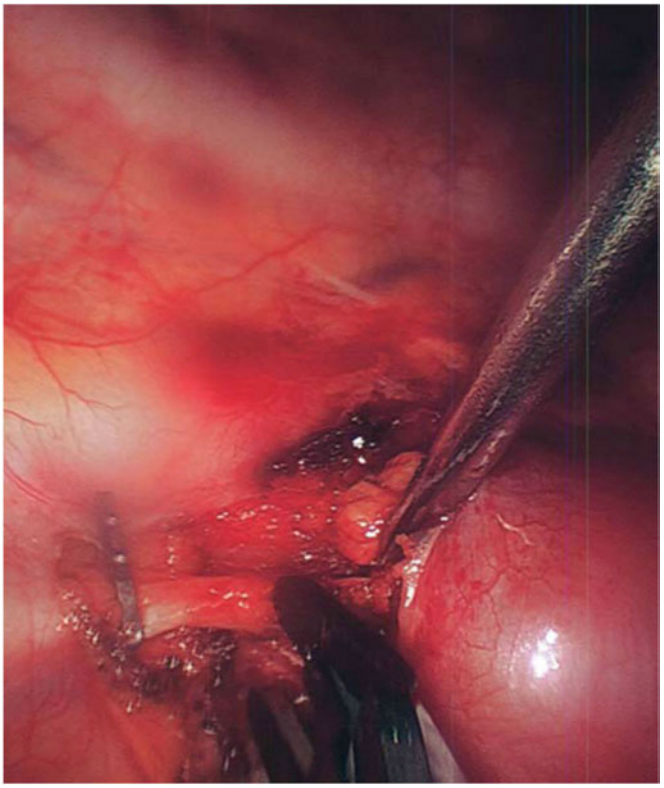


Fig. 15.8. The bundle lateral to the tumor is clipped first, prior to division, as shown here.

After resection of a tumor at the costovertebral sulcus, regular neurologic examinations of the lower extremities are indicated. Tamponade with hemostatic agents should never be employed for bleeding at the neural foramen as doing so can result in an intraspinal hematoma with subsequent cord compression. Careful use of the electrocautery at the bony margins of the foramen or watchful waiting is preferable. If hemostasis cannot be achieved with these measures, a neurosurgical consultation should be obtained. In the event of oozing from the vicinity of a foramen that is not easily controlled, there should be no hesitation in converting a VATS procedure to an open procedure.

In a minority of patients, clipping and division of an intercostal nerve result in substantial intercostal neuralgia after the procedure; the possibility that this may occur must be discussed with the patient preoperatively. Many patients who undergo division of a lower thoracic intercostal nerve that supplies an upper abdominal dermatome notice minor postoperative bulging of the ipsilateral abdomen in the area supplied by that nerve.

Video-Assisted Thoracoscopic Surgery Resection of a Benign Cyst of the Posterior Mediastinum

Resection of a benign cystic mass of the posterior mediastinum closely resembles resection of a neurogenic tumor (see steps above). One starts by incising the pleura

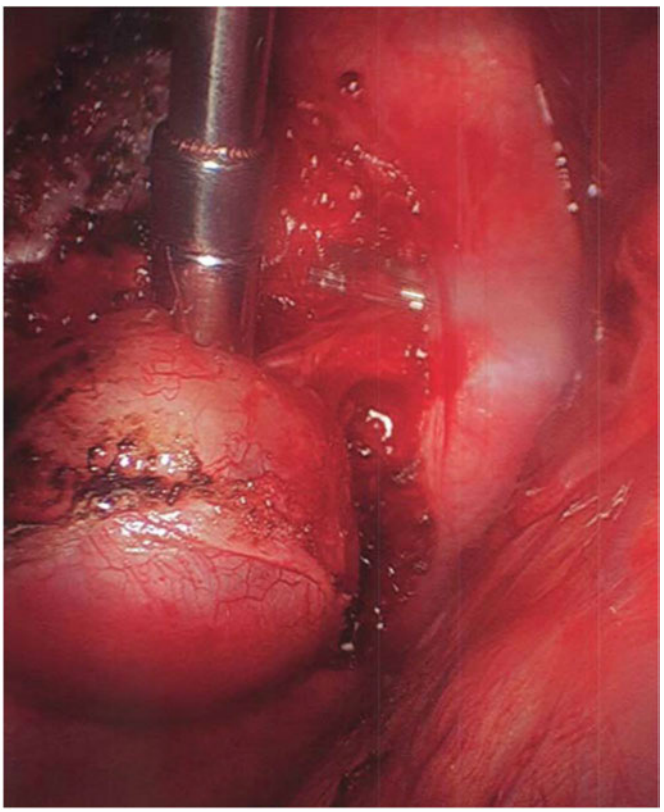


Fig. 15.9. Division of all other remaining attachments is typically carried out next, until the bundle medial to the tumor is all that is left.

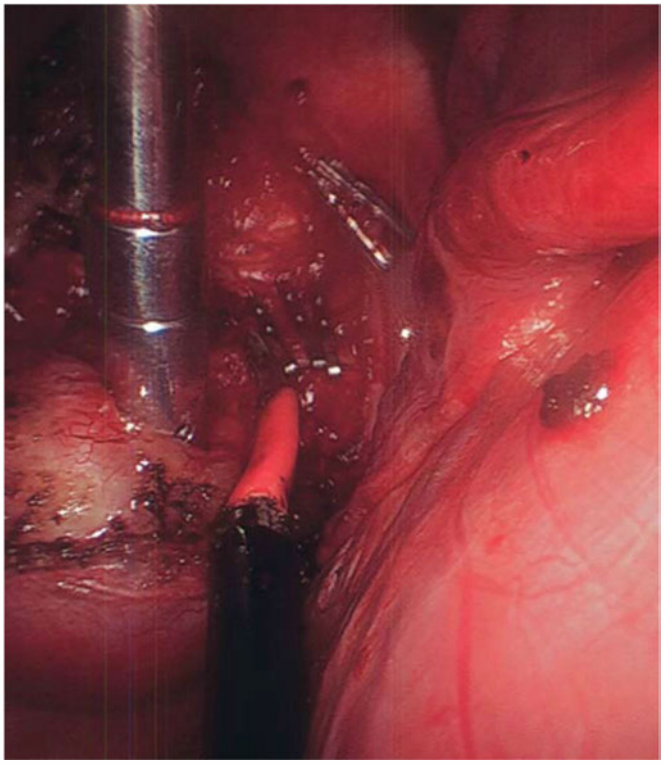


Fig. 15.10. At least two clips are placed, as shown here, on the source intercostal bundle medial to the tumor, prior to division.

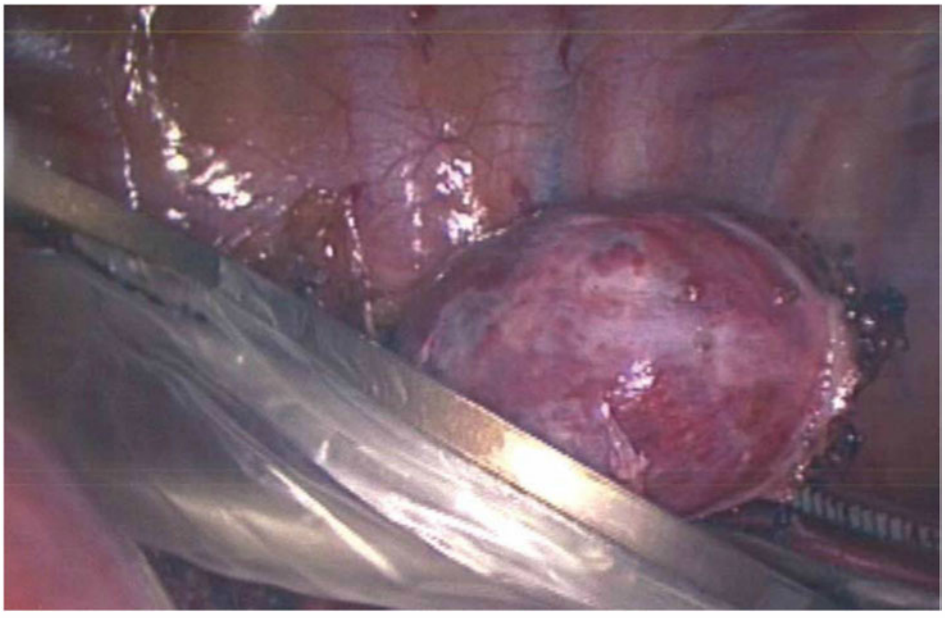


Fig. 15.11. The specimen is bagged and removed.

circumferentially, then dissects and divides attachments progressively, circumferentially, until the cyst has been completely mobilized. The procedures are typically simpler since there is no intercostal bundle to be concerned with.

Potential Problems and Their Avoidance

In the initial stages of dissection of a benign cyst of the posterior mediastinum, care should be taken *not* to rupture the cyst; initial mobilization from surrounding structures is far easier when the cyst wall remains under tension (Fig. 15.13). Occasionally, a portion of the cyst wall is found to be inseparable from an important mediastinal structure such as the esophagus or the membranous wall of the airway and cannot be removed safely. This is more common in the case of active or recent infection of the cyst. In these situations, I will intentionally rupture the cyst once that part of the wall is all that remains to be mobilized. I then resect all of the nonadherent portion of the cyst wall and ablate the residual adherent cyst wall with electrocautery to destroy any potential remaining secretory tissue. In rare cases, I have left as much as 35% of the wall of a bronchogenic cyst in place, and I have never seen a recurrence to date. It is my recommendation that if more than approximately 35% of the cyst must be left in place, conversion to thoracotomy should be considered to prevent recurrence.

Resection of Esophageal Leiomyomata and Duplication Cysts

In addition to the steps described above for resection of a neurogenic tumor, there are several special maneuvers that facilitate resection of esophageal intramural masses, such as leiomyomata and duplication cysts.

1. After incising the pleura, the longitudinal esophageal muscle fibers that overlie the mass are separated bluntly or with the electrocautery in the line of the fibers. These fibers are often markedly attenuated as a result of the expansion of the mass.
2. Blunt dissection with an endoscopic peanut dissector allows careful, progressive mobilization of the mass, first from the muscle layer and then from the underlying mucosa. Gentle traction on the mass aids exposure during this portion of the procedure. A suture may be placed into the tumor to facilitate this retraction. Having an assistant place the endoscope within the esophageal lumen to distend and illuminate the mucosa also may be helpful at this stage. Once the mass has been completely resected, it is sent for pathologic examination. Even horseshoe-type leiomyomata can usually be successfully resected by this technique.
3. After resection, the esophagus is distended by insufflating air from above through the endoscope while the distal esophagus is occluded with a sponge stick. The air-filled esophagus is then submerged in saline, and the area of the resection is examined for air leakage.

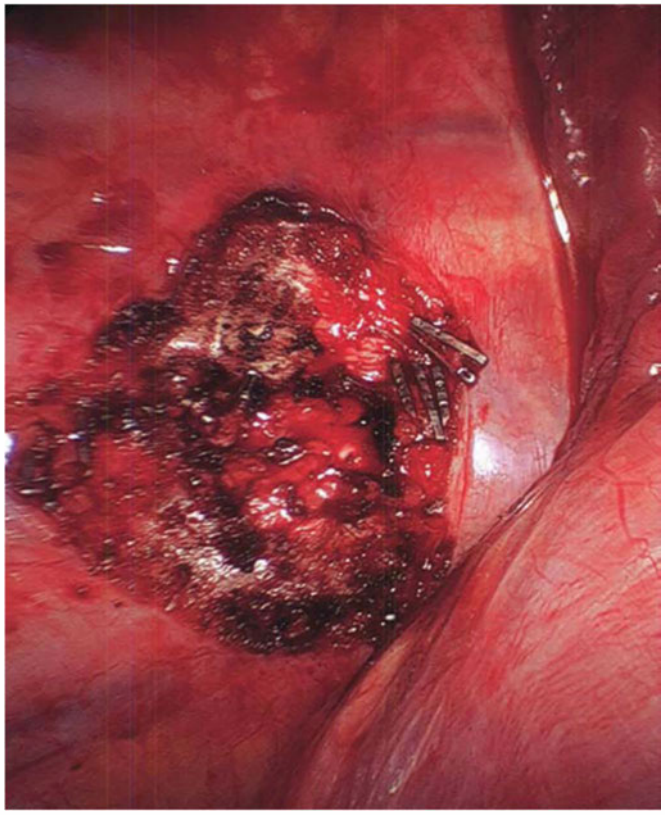


Fig. 15.12. The typical appearance of the prior bed of the tumor, following resection.

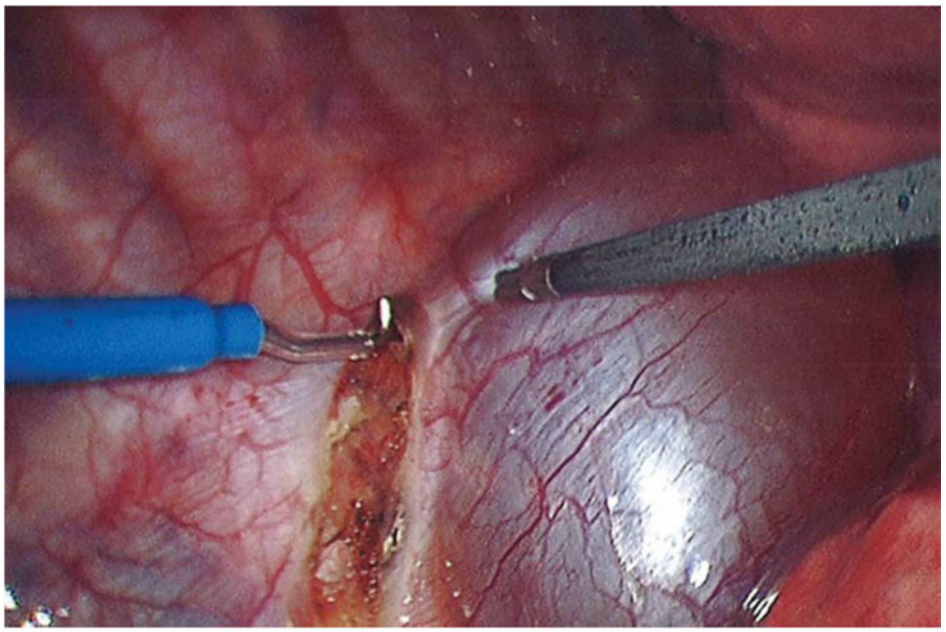


Fig. 15.13. Dissection of posterior mediastinal cysts away from surrounding structures is facilitated by keeping the cyst wall intact for as long as possible during the resection.

Potential Problems and Their Avoidance
The muscular defect in the esophageal wall must be closed after resection to assure that an esophageal diverticulum does not develop. Such closure may be accomplished by means of thoracoscopic suturing.

Frequently, duplication cysts are more adherent to the underlying esophageal mucosa than leiomyomata are, and transillumination of the esophageal wall helps define the plane at which blunt dissection should be performed. Where the cyst wall becomes difficult to separate from the mucosa, a small amount of the wall may be left in place if, in the surgeon's judgment, attempting to remove all of it might lead to a breach in the mucosa. In my opinion, if the mucosa is breached, a thoracotomy should be performed to assure precise mucosal and muscle layer repair. All patients should have a gastrografin/barium swallow to insure mucosal integrity prior to advancing diet on postoperative day 2.

RESULTS

There should be essentially zero recurrences if the principles and techniques described above are followed in the resection of benign posterior mediastinal masses and cysts. Patients are generally discharged on postoperative day 2. The severe complications of spinal cord compression, CSF leak, and esophageal leak should never occur when attention is paid to the details of the procedure. The most common ill effect, in my experience, is intercostal neuralgia when the intercostal nerve requires clipping and division. Since there is no known way to prevent this when the nerve must be sacrificed, patients need to be informed of the possibility, and this possibility needs to be weighed in the decision about whether to proceed with surgical resection or follow these lesions radiographically.

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EDITOR'S COMMENTS

As Shrager points out most of the lesions that occur within the posterior mediastinum are benign and many can simply be followed as opposed to resected. When a decision is made to resect I agree with Shrager that VATS is the preferred approach. The exposure is excellent and once the overlying pleura is incised most of these lesions are easily mobilized. Care must be taken to avoid undo traction on the intercostal nerve so as to avoid avulsion with tearing of the dura and a resultant CSF leak. The extent of the dural sheath on an intercostal nerve varies, so care should be taken to make sure that a clip is placed prior to dividing a nerve if one is close to the neural foramen. When dissecting at the level of the neural foramen one encounters a rich vascular network that may be difficult to control. I avoid the use of monopolar cautery at or near the foramen and rely on bipolar cautery to control this sometimes troublesome bleeding.

As the author points out the occasional cystic lesion that has become infected can be quite adherent to surrounding structures including the esophagus. I completely agree with him that in those circumstances it not only is acceptable but likely safest to leave a portion of the cyst wall intact as opposed to potentially causing damage to another structure.

One cannot stress enough the care that must be taken with the so-called dumbbell lesions, those benign, usually Schwannomas, that have both an extraspinal and intraspinal components. If there is any question of an intraspinal component I obtain an MRI scan and based on the results plan the surgical procedure accordingly. Interestingly, though we often talk about these lesions they are quite rare but when encountered require a collaborative approach between the neurosurgeon and the thoracic surgeons. As Shrager points out, the intraspinal component should be dealt with first followed immediately by excision of the intrathoracic component. Whether one approaches the intrathoracic component by an extension of the posterior incision or repositions the patient and does a separate approach likely makes little difference. Again, a VATS approach is our favored one for the intrathoracic component. Malignant lesions of the posterior mediastinum are extremely rare as are the hormone-producing lesions.

LRK

16 Antireflux Surgery

John G. Hunter and Jeffrey S. Fronza

INTRODUCTION

Peptic esophageal injury was first described in the late 19th century, but became more frequently recognized with the advent of upper gastrointestinal endoscopy. The association of peptic esophageal injury and gastroesophageal reflux disease (GERD) was demonstrated in the 1930s. It was not until the association of hiatal hernia and GERD was recognized in the 1950s, however, that antireflux surgery emerged. The 360 degree fundoplication to treat GERD was reported in 1955 by Rudolf Nissen, a Swiss surgeon. Nissen's fundoplication created a long, super competent lower esophageal sphincter (LES), which in turn frequently led to postoperative bloating, dysphagia, inability to belch, and flatulence. In the 1980s, Donahue and DeMeester perfected the short, floppy Nissen fundoplication by dividing the short gastric and retrogastric attachments, shortening the wrap to 1 to 2 cm, and performing the wrap around a large Maloney dilator. By better allowing gas to vent from the stomach, this modification avoided most of the unwanted side effects while still controlling pathologic reflux.

In 1992, the *New England Journal of Medicine* published a seminal study from the Veterans Administration that showed open Nissen fundoplication to be superior to continuous medical therapy in patients with complicated GERD. That same year, the field of antireflux surgery was revolutionized when Bernard Dallemagne of Belgium and Alfred Cuschieri of Scotland performed the first laparoscopic Nissen fundoplication (LNF). Between 1990 and 1997, the annual rate of LNF more than doubled. Rates have decreased since that time for a variety of reasons, including improper technique and an unacceptable rate of recurrent GERD mainly due to inexperience on the part of the surgeon. In spite of this, LNF remains the gold-standard treatment of GERD with multiple studies demonstrating control of reflux at 10 years in 90% of patients. Continued successful surgical correction of GERD requires both adherence to proper surgical technique and appropriate patient selection.

INDICATIONS

GERD is the most common disease of the alimentary tract requiring medical or surgical treatment. It affects 7% of adults in the United States on a daily basis and >30% on a regular basis. In addition, 5% to 15% of these individuals experience serious complications of GERD such as esophageal stricture or Barrett's esophagus. Typical symptoms of GERD include heartburn, regurgitation, and dysphagia. Atypical symptoms include cough, hoarseness, wheezing, and chest pain. Nonoperative treatment of GERD involves lifestyle modifications such as weight loss, avoidance of food intake for several hours prior to retiring to bed, elimination of food and medications that promote LES dysfunction, cessation of smoking, and elevation of the head of the bed. Many patients require pharmacologic treatment such as daily use of proton-pump inhibitors (PPIs). Quality-of-life assessments performed on patients with severe GERD reveal scores similar to patients with chronic congestive heart failure, emphasizing the debilitating effect of this disease.

Patients whose symptoms are refractory to PPIs are candidates for LNF, as are those whose symptoms are controlled on PPIs, but who prefer to avoid life-long need for medication. Studies have demonstrated LNF to be cost-effective in young patients with severe GERD. Those with predominant respiratory symptoms are another group best served with LNF as only 50% have symptom control with medication. That being said, those patients responsive to PPIs and with typical symptoms of GERD have the best outcomes from operation. The current recommendation is to consider operation for heartburn patients responsive to medication, but with frequent troublesome symptoms poorly treated by PPIs, such as regurgitation and respiratory symptoms. Patients with large hiatal hernias are unlikely to have their symptoms of postprandial chest pain and regurgitation adequately treated with PPIs.

A relative contraindication to LNF is morbid obesity (BMI >40), as these patients have an increased risk of recurrent GERD and are better served with a roux-en-Y gastric bypass to not only control their GERD but also for optimal weight loss.

PRINCIPLES OF ANTIREFLUX SURGERY

Prior to offering an antireflux operation, a thorough preoperative evaluation must be completed. In a patient with typical or atypical symptoms of GERD, the diagnosis is confirmed by endoscopic evidence of erosive esophagitis, peptic stricture, or Barrett's esophagus or by a 24-hour ambulatory esophageal pH study revealing a pH of <4 for >4.2% of the study period. Even with an abnormal pH study, an upper endoscopy is essential to document sequelae of GERD and to rule out other pathology masquerading as GERD. Finally, esophageal manometry assesses esophageal motility. Previously, it was thought that ineffective esophageal motility (>60% ineffective swallows or <40 mmHg mean distal esophageal contraction pressure) was an indication for partial fundoplication, but it is clear now that impaired esophageal bolus clearance after Nissen fundoplication is only a risk in the presence of aperistalsis. Video barium swallow is indicated to evaluate an associated hiatal hernia and/or a shortened esophagus. A nuclear medicine gastric emptying study is recommended in patients with diabetes or symptoms of nausea, vomiting, and early satiety as those found to have delayed emptying will benefit from pyloroplasty at the time of LNF.

The objective of any antireflux operation is to reconstruct a LES that has been rendered mechanically defective due to hypotension, inappropriate relaxation, and/or intrathoracic displacement. Guiding surgical principles include the establishment of an adequate intra-abdominal length of esophagus (>3 cm), reduction and repair of an associated hiatal

hernia, and tension-free construction of an appropriate fundoplication. All fundoplications should restore the acuteness of the angle of His and calibrate the fundoplication diameter to be loose around an 18 to 20 mm esophageal dilator. There is consensus within the surgical community that a laparoscopic approach is preferred in the vast majority of patients undergoing fundoplication. Thoracotomy is indicated only in specific reoperative settings, where a hostile upper abdomen makes a transthoracic approach appealing.

PATIENT PREPARATION

Patients should be maintained on PPIs preoperatively to minimize esophagitis. Peptic strictures should be dilated preoperatively. Anticoagulants and nonsteroidal anti-inflammatory agents are discontinued. Glucose control should be optimized, and cardiopulmonary disease should be thoroughly evaluated with appropriate consultation and testing preoperatively. The patient should be *nil per os* the night prior to surgery. Perioperative antibiotics are unnecessary as antireflux surgery is a clean operation.

SURGICAL TECHNIQUES

Laparoscopic Nissen Fundoplication

LNF is generally performed in the supine position with the legs extended and abducted 30 to 60 degrees so that the surgeon may operate between the legs. Low lithotomy is a less desirable position as peroneal nerve injury, deep venous thrombosis, and patient movement while in reverse Trendelenburg position is more frequent. Some surgeons prefer the patient in the supine position, operating from either the left or the right side. This setup, however, results in poor ergonomics with resultant chronic shoulder injuries in the surgeon and the need to address the hiatus obliquely. Clearly, optimal positioning favors a split-leg position. The assistant standing to the left of the patient holds the camera with the left hand and assists with the right hand. The monitor is placed over the patients head ideally fixed on a ceiling-mounted boom (Fig. 16.1).

LNF is performed with a five-port technique. The operation begins by insufflating the abdomen with a Veress needle placed above the umbilicus. After obtaining pneumoperitoneum, an 11-mm trocar is placed 15 cm below the costosternal junction slightly to the left of the midline. By placing the port to the left of midline, and obliquely entering the abdomen—aiming toward the esophageal hiatus—the surgeon minimizes the risk of port-site hernia, stays out of the fatty ligamentum teres, stays further away from the left lobe of the liver, and more accurately aligns the laparoscope with the trajectory of the esophagus (right to left) as it traverses the diaphragm. A 45-degree laparoscope is introduced and a brief abdominal survey is performed. All other trocars are placed under direct vision and all sites are infiltrated with local anesthesia before an incision is made. The second 11 mm trocar is introduced along the left costal margin 12 cm from the base of the sternum. The 11 mm trocar facilitates the passage of suture and is useful for the placement of endoscopic clips when necessary. The third trocar placed is 5 mm, generally located 8 cm farther down the left costal margin than the second trocar but not further lateral than the anterior axillary line. A Nathanson liver retractor is introduced through the track of a cutting 5 mm port that has been placed and removed just inferior and left of xiphoid. The liver retractor is connected to a mechanical arm fixed to the right side of the operating table. The optimal position of the liver retractor allows access to the anterior arch of the hiatus as well as thoroughly defining the right crus of the diaphragm. The patient is then placed in steep reverse Trendelenburg position. The fifth trocar is used for the surgeon's left hand and is placed along the right costal margin just inferior to the edge of the retracted left lobe of the liver. The point at which this trocar is placed is determined by internal, not external, anatomy, and therefore is variable in its

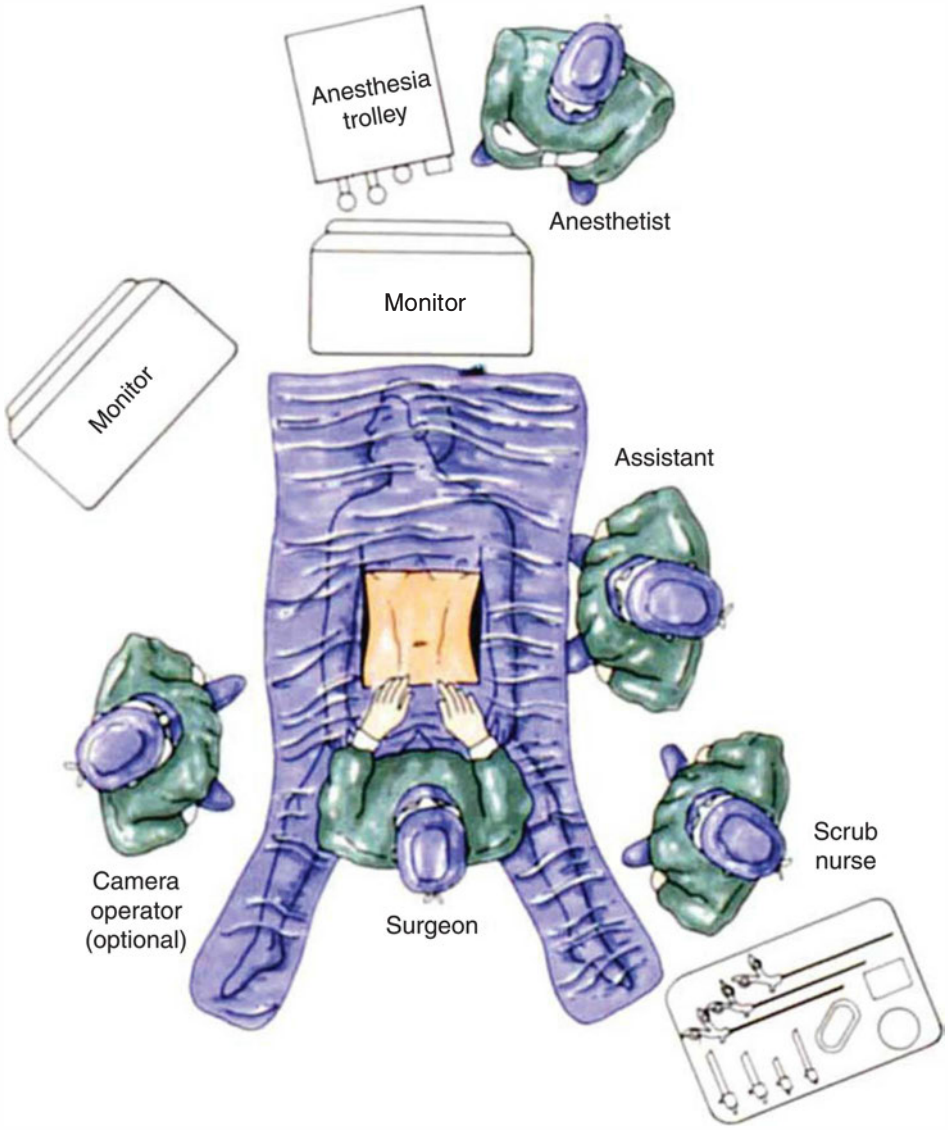


Fig. 16.1. Operating room set-up and patient positioning.

superficial projection. It is generally desirable to have this trocar as high and as wide as possible without going into the left lobe of the liver or achieving placement to the right side of the falciform ligament.

Dissection commences by the assistant grasping the epiphrenic fat pad and retracting inferiorly and slightly toward the patient's left. This reduces any hiatal hernia and places the phreno esophageal membrane on tension. Next, the surgeon exposes the right crus of the diaphragm by opening the gastro hepatic ligament above and below the hepatic branch of the vagus (Fig. 16.2). While it is desirable to preserve the hepatic branch of the vagus, many surgeons routinely divide it to obtain better access to the right crus. A blunt atraumatic grasper is placed beneath the phrenoesophageal ligament to establish the plane between the esophagus and the diaphragm. This layer is divided with ultrasonic energy (Fig. 16.3). The dissection proceeds around the arch of the hiatus and down the left pillar of the diaphragm as low as is easily visualized. Access to the mediastinum is obtained by placing two blunt graspers (closed) between the esophagus and the right crus of the diaphragm, and then spreading horizontally. This is repeated on the left side of the esophagus. The anterior vagus nerve is generally affixed to the esophagus, and the posterior vagus nerve is not visible at this time. Dissection down the right crus of the diaphragm continues until the surgeon reaches its base where the left crus is visualized emerging from posterior to the esophagus. This completes the preliminary phase of the crural dissection.

The next phase of dissection includes the division of the short gastric arteries, and posterior mobilization of the stomach. The light cord of the laparoscope is rotated 30 degrees counterclockwise (11 o'clock position), and the surgeon grasps the greater curvature of the stomach at the level of the inferior pole of the spleen. The assistant grabs the greater omentum adjacent to the greater curvature of the stomach. The surgeon retracts the stomach to the patient's right and posteriorly while the assistant elevates the omentum anteriorly and to the patients left. The ultrasonic shears are introduced and entry to the lesser sac is gained by incising the greater omentum adjacent to the greater curvature of the stomach (Fig. 16.4). Several applications of the harmonic scalpel may be necessary to gain access to the lesser sac in heavier patients. It is important that the short gastric vessels are visualized and completely occluded with the ultrasonic shears before the application of energy. Incomplete division of short gastric vessels results in troublesome bleeding. As the dissection moves cephalad, the surgeon keeps moving the retraction point higher along the greater curvature, generally placing one jaw of the atraumatic grasper on the anterior wall of the stomach and one jaw on the posterior aspect of the stomach. The assistant places one jaw of their grasper in the lesser sac, and one jaw in the greater sac to optimally retract the greater omentum.

When one approaches the superior pole of the spleen, a different retraction strategy is required. Incision of the peritoneum at the superior pole of the spleen widens the splenogastric omentum into several layers. To adequately expose this remote corner of the abdominal cavity, both surgeon and first assistant retract on the stomach. The surgeon grasps the fundus of the stomach on the anterior surface and the assistant grasps the fundus of the stomach on the posterior surface. The surgeon retracts anteriorly and to the right. The assistant retracts posteriorly and to the right. This opens up the field at the superior pole of the spleen. If a fatty greater omentum fills the lesser sac to the superior of the spleen, obscuring the field of dissection, it may be necessary to reef up and retract the omentum inferiorly with a 90 cm 2-0 polypropylene suture that is brought out of the abdominal wall through the trocar in the left anterior axillary line. One should not proceed with dissection of this area without adequate exposure of the anterior wall of the stomach, the posterior wall of the stomach, and the superior pole of the spleen. Generally, the light cord

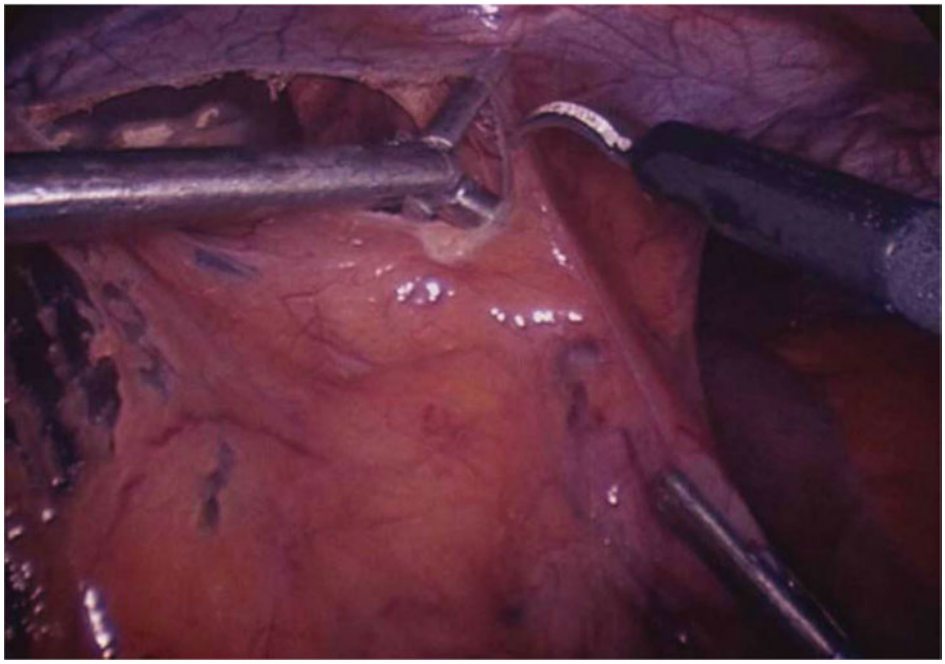


Fig. 16.3. Opening the phrenoesophageal membrane.



Fig. 16.2. Identifying and preserving the hepatic branch of the vagus.

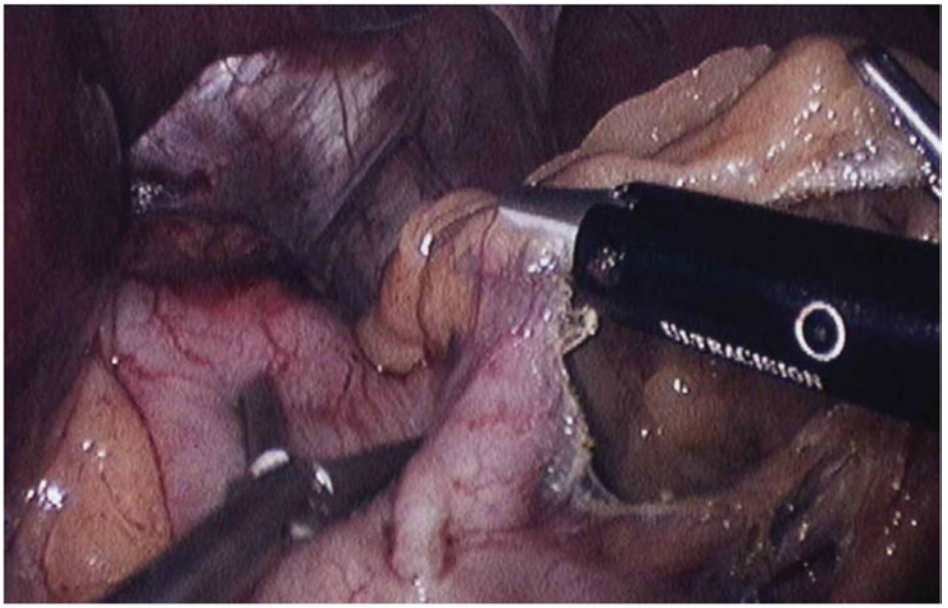


Fig. 16.4. Mobilization of the greater curvature.

of the laparoscope is rotated back to the 12 o'clock position or to the 1 o'clock position to visualize this region.

The next point of dissection is the posterior pancreaticogastric fold, which contains the posterior gastric artery. This fold is identified by medial retraction on the posterior body of the stomach, lower on the stomach than was necessary to expose the tip of the spleen. Medial and occasionally anterior retraction of this posterior wall lifts the posterior stomach away from the posterior retroperitoneum, exposing a consistent band from the superior surface of the pancreas (near the splenic artery) to the posterior aspect of the stomach (Fig. 16.5). This band is divided with the harmonic scalpel, moving in a cephalad direction toward the base of the left crus of the diaphragm. The posterior gastric artery encountered in this bundle is taken with the ultrasonic shears. When the base of the left diaphragmatic crus is reached, dissection then proceeds anteriorly along the left crus of the diaphragm dividing all attachments between the left crus and the gastroesophageal (GE) junction. The best retraction for this region often requires the surgeon to grab the epiphrenic fat pad and retract anteriorly and to the right while the assistant maintains posteromedial retraction on the posterior wall of the stomach. The mediastinum to the left of the esophagus is thus entered. When the left crus is followed posteriorly, one reaches the right crus. If this maneuver is difficult, it is sometimes desirable to return to the right side of the esophagus and follow the right crus of the diaphragm further posteriorly. The posterior window behind the esophagus is not achieved by blunt dissection or passage of a blunt instrument, but by division of the peritoneal and neurovascular attachments between the diaphragm and the GE junction around the entire hiatal aperture. At this point, the posterior vagus nerve becomes visible from either the left or the right side of the esophagus, lying adjacent to the posterior esophagus.

A Penrose drain is placed around the GE junction. For most dissections, a 4 cm length of ¼ inch Penrose drain is appropriate. The two ends of the drain are held by the assistant anteriorly while the surgeon places an endo loop on the drain to hold the ends together. The assistant then replaces the atraumatic grasper with a more aggressive grasper, which can be ratcheted down and retracts the GE junction caudally and slightly to the patient's left. This allows access to the mediastinum on the right side of the esophagus. The surgeon mobilizes the distal esophagus from the posterior mediastinum with a combination of blunt dissection and ultrasonic shears. Most periesophageal attachments can be divided bluntly between two graspers without risking thermal injury to either vagus nerve.

The extent of mediastinal dissection varies as the only requirement is to achieve a tension-free intra-abdominal esophageal length of 3 cm. The best way to assess tension-free intra-abdominal length is to remove the assistant's grasper from the Penrose drain and perform a trial closure of the diaphragm posteriorly with two graspers. A dissecting instrument or ruler may be used to measure the length of esophagus remaining below the diaphragm at rest.

Once it has been determined that there is adequate length of intra-abdominal esophagus, attention is directed toward closure of the diaphragmatic hiatus with nonabsorbable braided suture on an atraumatic round needle. Sutures are placed 5 to

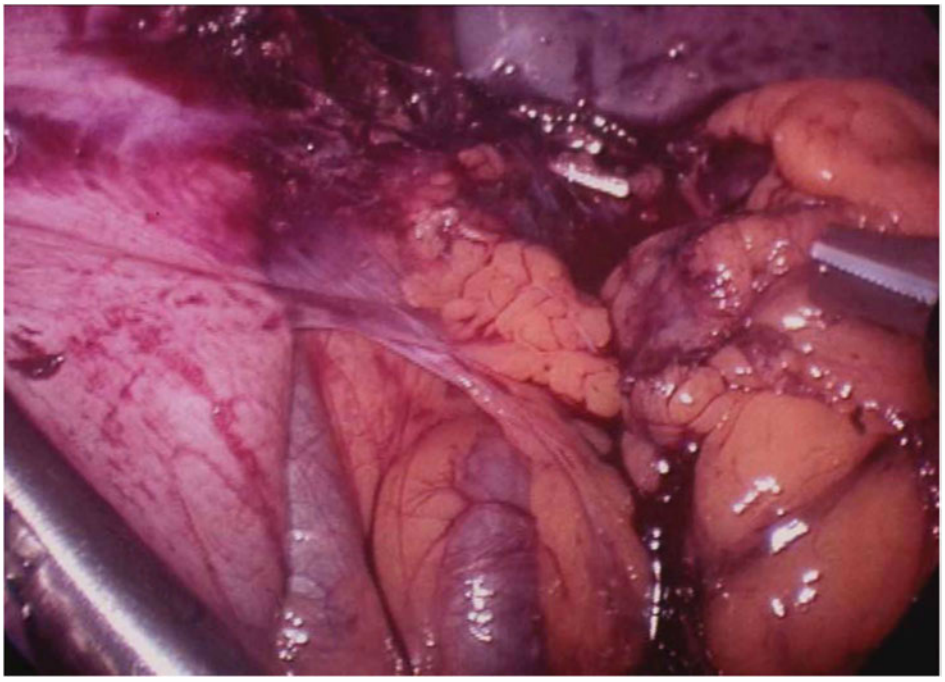


Fig. 16.5. The pancreaticogastric fold.

10 mm apart, require deep bites of the left and right crura of the diaphragm, and are tied intracorporeally. Because crural failure has been noted in many fundoplication procedures, we use 1 cm square Teflon-felt pledgets to buttress the closure (Fig. 16.6).

Upon completion of the crural closure, an atraumatic grasper is placed from right to left behind the GE junction, and the posterior aspect of the gastric fundus is grasped and pulled behind the esophagus. To avoid twisting the wrap, it is important that the posterior wall is the portion grasped. The posterior wall is then walked hand-over-hand backwards down toward the angle of His to make sure that a point high on the fundus has been chosen. A "shoe-shine" maneuver is performed by sliding the two proposed points of fixation back and forth behind the esophagus (Fig. 16.7).

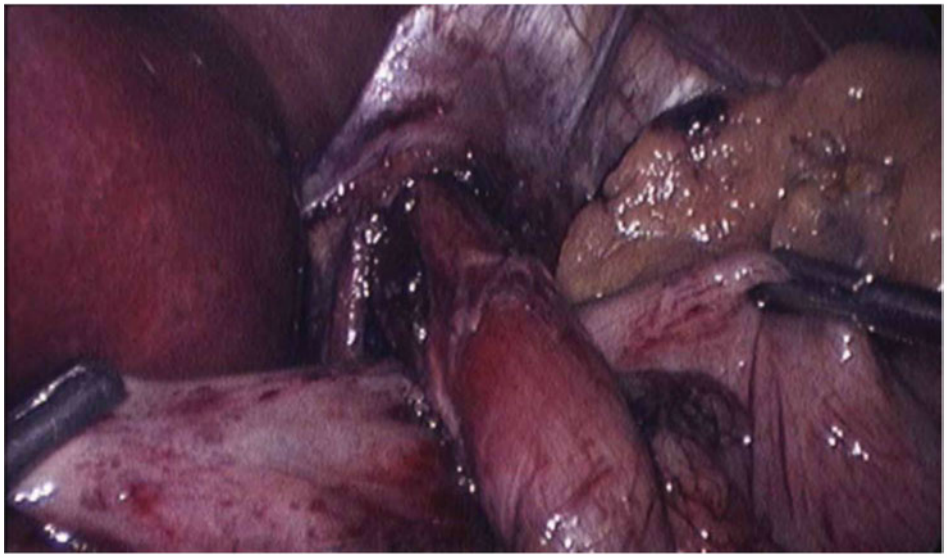


Fig. 16.7. The "shoe-shine maneuver."

At this point, the orogastric tube is removed and a large Maloney dilator (54F to 60F) is passed into the stomach. This maneuver has been associated with esophageal perforation in 0.5% to 1.0% of operations but may be preventable if several measures are taken. First, close communication between the anesthetist and surgeon is mandatory. The anesthetist should alert the surgeon to the distance of the tip of the dilator from the incisors marked on the side of the dilator. In addition, we pass a small dilator (36F to 40F) first to insure that there is unobstructed passage into the stomach. While smaller dilators tend to fold back on themselves before perforating the esophagus, larger dilators more commonly will perforate if they run into an irregular stricture or diverticulum. Equally important, the inferior traction on the Penrose drain must be released and the laparoscope pulled back as the tip of the dilator passes into the stomach.

Once the dilator has been placed, a fundoplication is fashioned. The position for the first stitch is chosen by pulling the two limbs of the fundus together at the 10 to 11 o'clock position on the esophagus. There should be no circumferential tension on the fundus with this maneuver. If there is tension, another point on the stomach, closer to the greater curvature is chosen. Once the suture positions are chosen, the first stitch (0 Ticron, 9 cm long) is introduced through the 11 mm trocar, and the needle is passed first through the left limb of the fundus, then the esophagus (2.5 cm above the GE junction), and then through the right limb of the fundus. A slipknot is placed, and as the knot is sliding into place the two halves of the fundus are pulled into their final position. The "floppiness" of the fundoplication is tested by placing a closed grasper between the dilator and the fundoplication to the left of the esophagus, and distracting the fundoplication away from the esophagus. This maneuver should allow 1 cm worth of redundancy between the fundoplication and the esophagus (Fig. 16.8). If the fundoplication is too loose or too tight, the first stitch is removed and another is placed. Subsequent stitches should be placed no more than 1 cm apart, thus creating an anterior distance between the top and bottom of 2 cm. It is important to place the superior stitch first, and the inferior stitch last, just above the GE junction. We generally place all three esophageal sutures to the right of the anterior vagus nerve (Fig. 16.9). We place an additional suture posteriorly between the esophagus and the fundoplication to avoid posterior fundoplication slippage. The dilator is then removed and the fundoplication is placed in its anatomic position beneath the diaphragm.

The Penrose drain is removed and the small amount of blood that accumulates

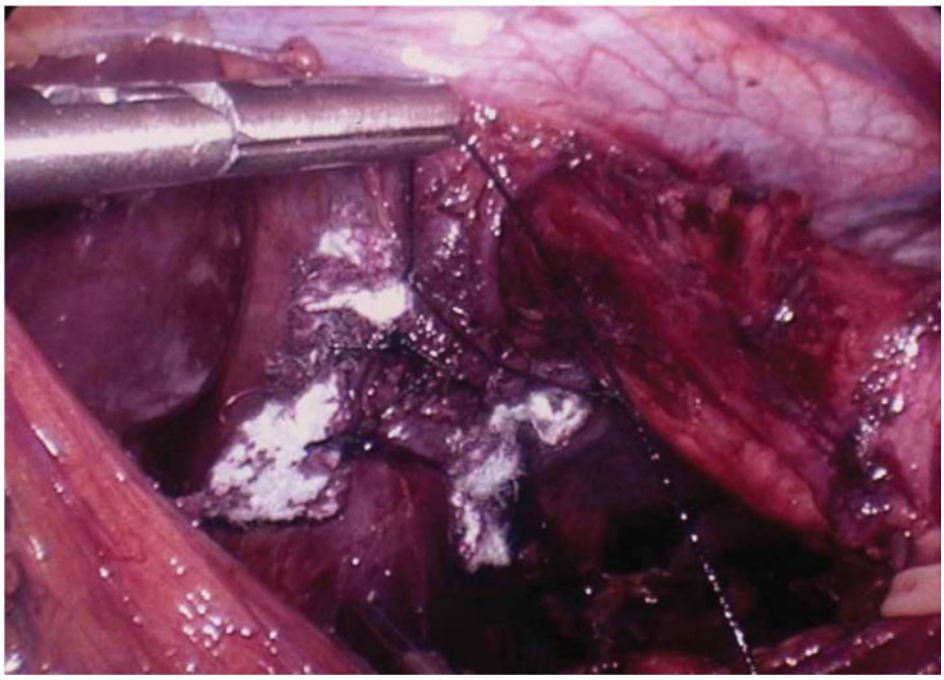


Fig. 16.6. Crural closure with pledgets.

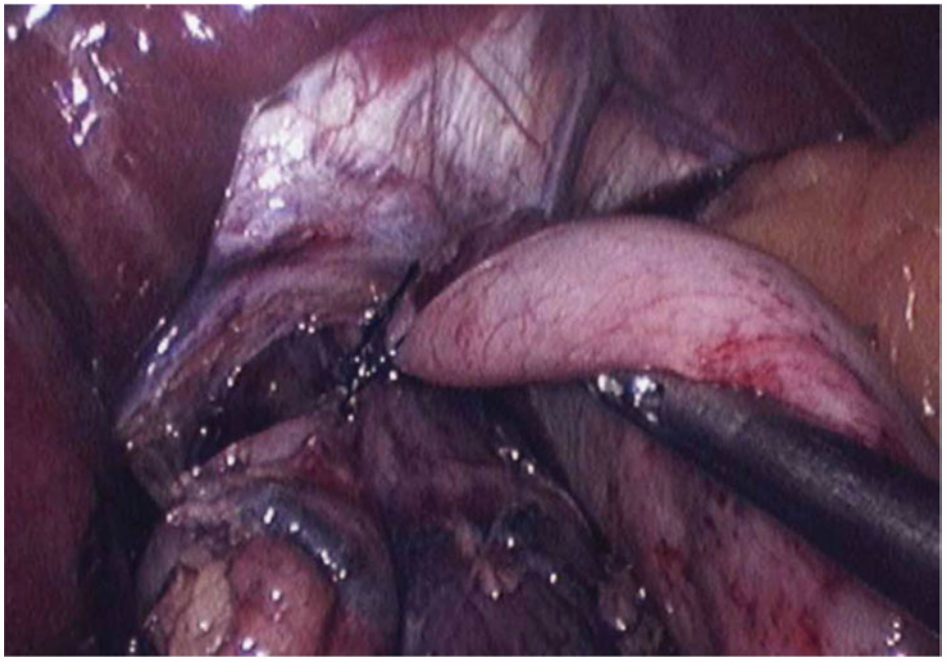


Fig. 16.8. Assessing the fundoplication for floppiness.

between the spleen and diaphragm is evacuated. The liver retractor and each of the trocars are removed under direct vision. We do not close the trocar sites in the upper abdomen that are 11 mm in diameter or less and have not seen hernia complications resulting. The skin is reapproximated with subcuticular sutures and tissue glue.

Laparoscopic Partial Fundoplication

Partial fundoplication is indicated in patients with preoperative dysphagia and poor esophageal motility. Types of partial fundoplication include a 270 degree posterior wrap (Toupet), a 180 degree anterior wrap (Dor), and a combined anterior and posterior wrap to the median arcuate ligament (Hill gastropexy). We preferentially perform a Toupet fundoplication as it is generally thought to provide superior reflux control compared with other partial fundoplications. A Dor fundoplication is used after performing a Heller myotomy due to its ability to provide coverage over an anterior myotomy. Few surgeons perform a Dor as a primary antireflux operation.

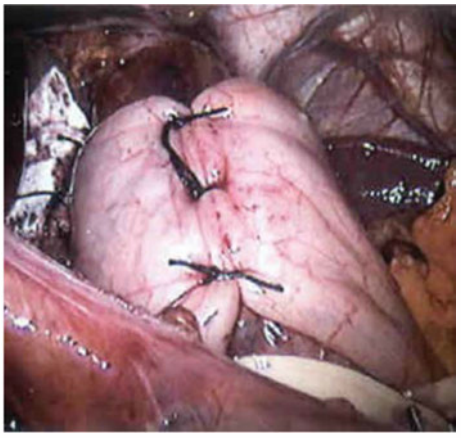


Fig. 16.9. Completed Nissen fundoplication.

The initial steps in performing a Toupet fundoplication are similar to those for LNF. After mobilization of the esophagus, division of the short gastric vessels and closure of the crura, the tip of the fundus is brought posteriorly around the esophagus as for a total fundoplication. An esophageal dilator is placed. Using three or four interrupted 0 Ticron sutures, the anterior aspect of the wrapped portion of the stomach is sutured to the right lateral wall of the esophagus just anterior to the anterior vagus nerve. The gastric fundus is sutured to the left side of the esophagus just anterior to the posterior vagus nerve.

Gastroplasty

A Collis gastroplasty was originally described in patients suffering from esophageal shortening caused by peptic stricture with severe contraction fibrosis. It was designed to create a tube of gastric cardia with a diameter similar to that of the esophagus that permits creation of an intra-abdominal, tension-free, fundoplication around this "neoesophagus."

The operation is performed laparoscopically during the course of LNF. After mobilization of the esophagus as high in the mediastinum as possible and closure of the crural defect, esophageal length is measured by means of an open Hunter grasper (2.5 cm). If 3 cm of intra-abdominal esophagus is not present, then a Collis gastroplasty is indicated.

A 48F Maloney dilator is passed and the gastroplasty performed with this in place. The neoesophagus is constructed by means of three firings of a 45 mm, purple or blue load, endo-GIA stapler (Covidien, Inc., Greenwich, CT). The stapler is inserted through a 12 mm port in the left subcostal area. The first firing is performed at a 90 degree angle to the dilator approximately 3 cm distal to the GE junction (Fig. 16.10). The second firing is performed in the same direction up to the dilator (Fig. 16.11). The final firing is done parallel to the dilator, and the wedge of stomach is removed from the abdomen (Fig. 16.12). The wrap is then performed as previously described for LNF.

Transthoracic Nissen Fundoplication

The advent of the laparoscopic approach to Nissen fundoplication has relegated this procedure to the realm of historical significance except for the occasional need for revision of a previous fundoplication where repeat laparoscopy, for a variety of reasons, is not feasible. The anesthesiologist places a double-lumen tube or bronchial blocker to permit single-lung ventilation and the patient is placed in the right lateral decubitus position. A lateral thoracotomy is performed and the chest is entered through the seventh intercostal space preserving the serratus anterior muscle. A 1 cm segment of the eighth rib may be taken posteriorly to allow greater rib spreading without causing a fracture. The distal esophagus is mobilized to the level of the inferior pulmonary vein taking care not to enter the right pleural space. The vagus nerves are preserved and are left on the esophagus. The phrenoesophageal membrane is divided near its insertion on the esophagus to enable entry into the peritoneal cavity through the esophageal hiatus. The esophagogastric junction is mobilized circumferentially. When the crura and the left lobe of the liver are exposed, the hiatal dissection is complete. The proximal stomach is pulled into the chest, the gastric fat pad is excised

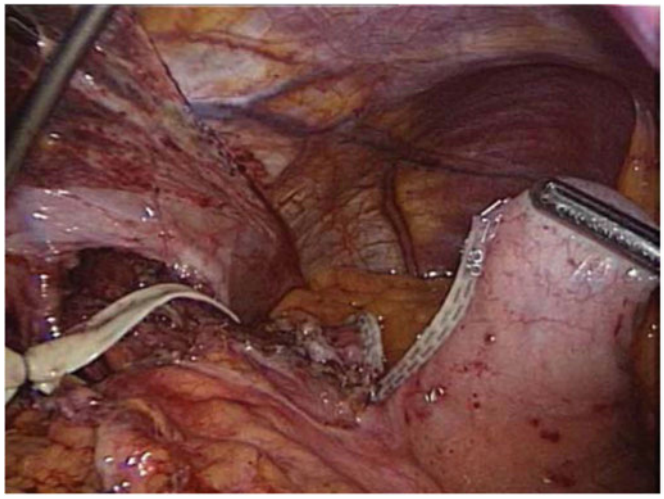


Fig. 16.10. The first firing of a Collis gastroplasty.

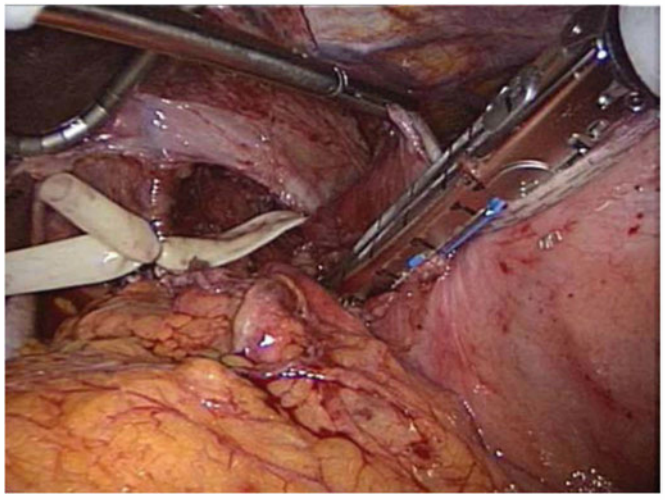


Fig. 16.11. The second firing of a Collis gastroplasty.

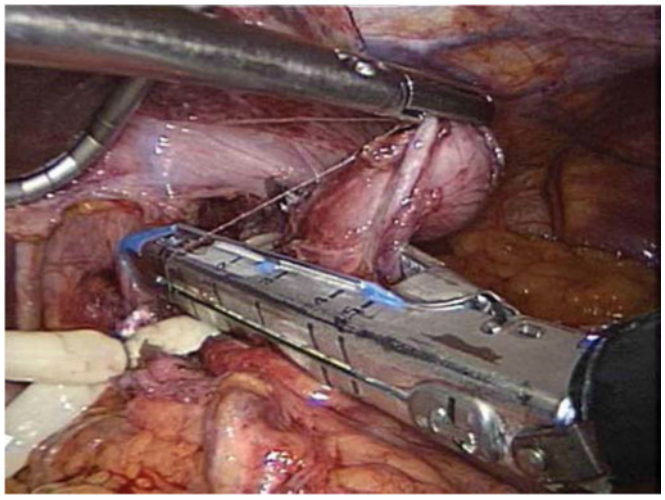


Fig. 16.12. The third firing of a Collis gastroplasty.

to permit apposition of the gastric serosa to the esophagus, and several short gastric vessels are divided to provide adequate mobilization of the stomach.

Interrupted nonabsorbable sutures are placed in the crura for future hiatal repair. The first suture is placed at the initial decussation of the crural fibers, and subsequent stitches are placed cephalad at 1 cm intervals until an adequate closure of the hiatus is possible. This usually requires three or four sutures (Fig. 16.13). The stitches are left untied until the fundoplication wrap is completed. A large (56F to 60F) dilator is placed so that it traverses the GE junction. The tip of the fundus is grasped in the surgeon's left hand and is brought posteriorly around the esophagus, where it is held with an atraumatic clamp. A portion of the stomach high on the body is pulled across the esophagus to meet the wrapped portion of the fundus. Three interrupted sutures are placed through the stomach to the left of the esophagus, through the muscularis of the esophagus, and again through the gastric fundus lying to the right of the esophagus. These are spaced at 1 cm intervals to create a 2 cm wrap (Fig. 16.14). After the sutures are tied, the dilator is removed, and the wrap is reduced below the diaphragm, where it should rest without tension. The crural sutures are tied to calibrate the esophageal hiatus, leaving enough room to allow the tip of one finger to be admitted easily alongside the esophagus. A nasogastric tube is placed and the chest is closed after an intercostal drain is placed.

Transthoracic Partial Fundoplication (Belsey Mark IV)

The initial portions of the operation are performed in a manner similar to that described for the transthoracic Nissen fundoplication, although it is usually not necessary to divide any of the short gastric vessels. Crural sutures are placed but are left untied. The partial fundoplication is begun by placing mattress sutures of interrupted nonabsorbable material through the stomach 1 cm distal to the esophagogastric junction and through the esophagus 1 cm proximal to the junction. Three sutures make up the first layer: one just anterior to the anterior vagus nerve, one just anterior to the posterior vagus nerve, and one midway between these two (Fig. 16.15). After these sutures are tied, a second row of mattress sutures is placed. These are first passed through the diaphragm, close to the margin of the central tendon. They then are placed through the stomach and esophagus in a mattress fashion, encompassing an

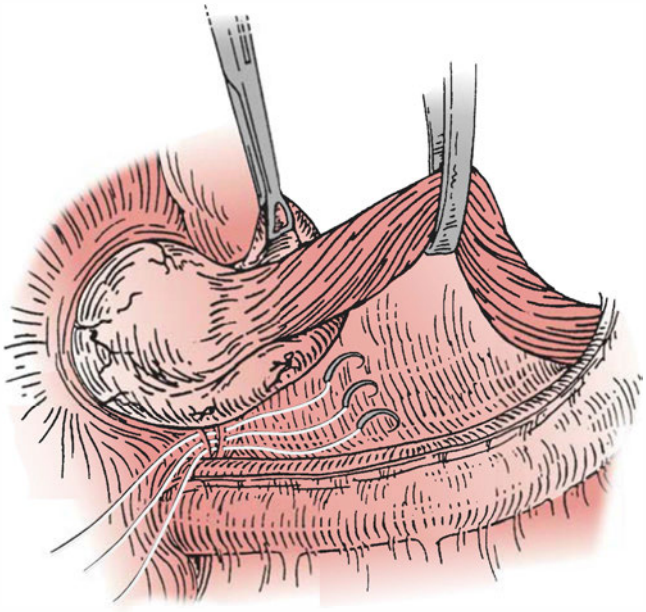


Fig. 16.13. Hiatal closure.

additional 1 cm of stomach distal to and 1 cm of esophagus proximal to the previous sutures, and finally are brought through the diaphragm again (Fig. 16.16). The wrap is reduced below the diaphragm, where it should rest without tension. The second row of sutures is then tied to create a 240 to 270 degree wrap that is anchored underneath the diaphragm. The crural sutures are tied to calibrate the esophageal hiatus, leaving sufficient room to admit the tip of one finger posteriorly alongside the esophagus (Fig. 16.17). The chest is closed after an intercostal drainage catheter is inserted.

Postoperative Care

Prior to emergence from anesthesia the patient is prophylactically treated for postoperative nausea with ondansetron and phenergan. A low dose of steroids may be added for their antiemetic effects. Ketorolac is administered to diminish postoperative pain. The patient is extubated and the Foley catheter is removed. No nasogastric tube is necessary for LNF. A clear liquid diet is offered in the evening of surgery and the patient is advanced to a full liquid diet by the first postoperative day. Antiemetic medications are scheduled and given on an as-needed basis to minimize the chance of retching with subsequent diaphragmatic stress. All medications are crushed or administered in a liquid form. Discharge occurs on the first postoperative day in patients who do not undergo thoracotomy. An esophagogram is obtained on the first postoperative day only after revisional antireflux surgery. Because of distal esophageal edema, patients are instructed to follow a full liquid diet for 5 days followed by a mechanical soft diet for 3 weeks. Specifically, they are instructed to avoid breads, meats, and raw vegetables. With strict adherence to this protocol, few patients complain of postoperative dysphagia. Without such aggressive management, food impaction, nausea, retching, and wrap disruption have been reported. Patients may resume activities of daily living immediately but avoid events involving intra-abdominal straining for 4 to 6 weeks to minimize the likelihood of acute fundoplication herniation.

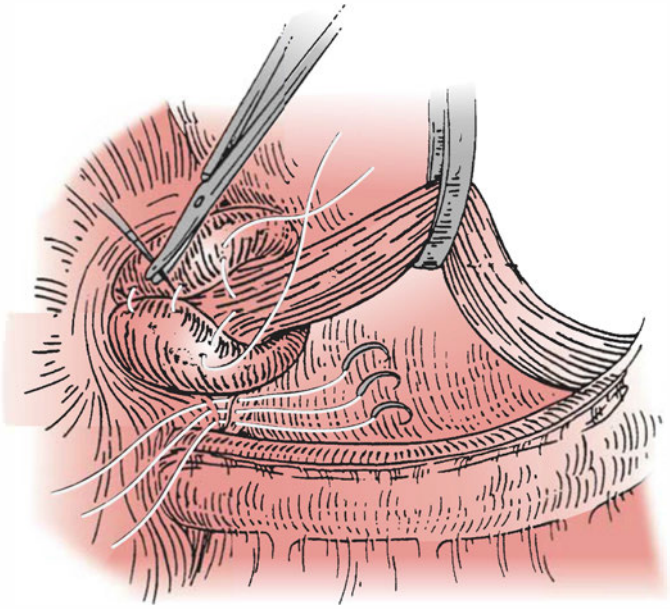


Fig. 16.14. Transthoracic Nissen fundoplication.

SURGICAL COMPLICATIONS

Serious complications after LNF are rare. A large meta-analysis demonstrated a mortality rate of 0.08%, reoperation rate of 2.7%, and splenectomy rate of 0.06%. Unrecognized gastric and esophageal injuries and acute fundoplication herniation represent potentially devastating complications. The fundamental caveat in postoperative care, therefore, is that any patient who fares poorly for any reason should be studied with a barium swallow to rule out perforation or herniation. With expeditious diagnosis and urgent reoperation, the outcome in these patients is good.

Early postoperative dysphagia can occur in up to 16% of patients after LNF, but only 2.5% of patients experience dysphagia at long-term follow-up. Patients with preoperative dysphagia, even in the face of a normal esophageal manometry study, are more likely than those without to experience postoperative dysphagia. Other postfundoplication complaints include rectal flatulence and inability to belch or vomit. These complaints tend to improve over time without specific intervention.

RESULTS OF SURGERY

Excellent long-term outcomes after LNF have been well documented in the literature. Greater than 90% of patients can expect to have reflux control at 10 years. Seventy to 90% of patients remain off PPIs and 93% of patients would undergo LNF again. Those patients with predominant

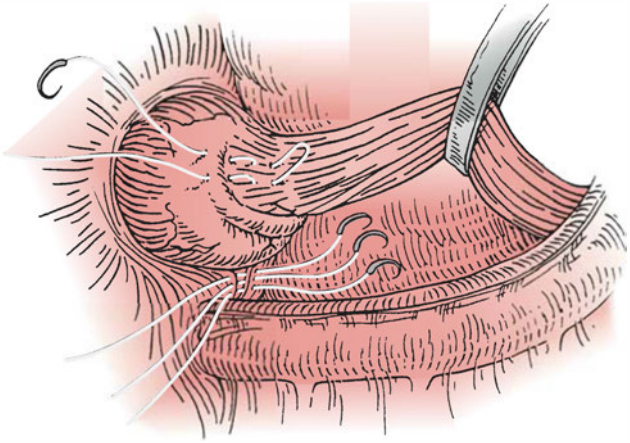


Fig. 16.15. A Belsey fundoplication is begun by mobilization of the upper stomach into the chest. The initial row of three mattress sutures incorporates 1 cm of stomach and 1 cm of esophagus and is evenly spaced between the vagus nerves.

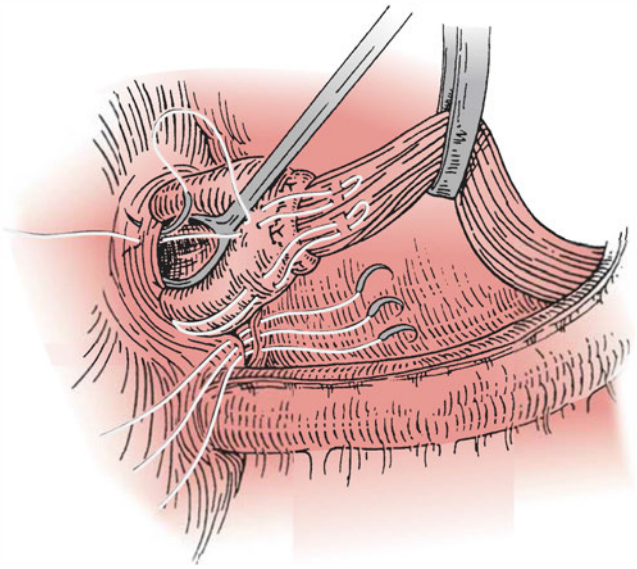


Fig. 16.16. A second row of sutures for the Belsey fundoplication is placed 1 cm from the first and incorporates the diaphragm at the beginning and end.

respiratory symptoms tend to do less well than those with typical symptoms. A substantial number of patients with Barrett's esophagus may experience regression of metaplasia and even dysplasia postoperatively. Initial success after LNF in patients with Barrett's esophagus is equivalent to those without, but there may be more anatomic failures in this group.

ROLE OF REVISIONAL ANTIREFLUX SURGERY

With the dramatic increase in LNF over the last two decades, esophageal surgeons continue to see a significant number of patients with "failed fundoplications." The definition of a failure after LNF is controversial as it can be defined in strictly anatomic terms, as resumption of PPIs, and/or may take into consideration functional outcome as determined by the patient. If failure is only defined in anatomic terms or as resumption of PPIs, then a patient with good symptom control but radiographic evidence of a small hiatal hernia would be considered a failure as would a patient with excellent reflux control with the resumption of minimal amounts of PPIs. On the other hand, if failure is defined on functional grounds then the rate of failure may increase significantly as most patients with esophageal complaints after LNF have no anatomic or physiologic abnormalities. We feel that defining failure in terms of patient symptoms is most appropriate as LNF is a at core functional procedure.

Up to 16% of patients will experience anatomic or physiologic failure and 3% to 6% of patients will require a revisional antireflux surgery. Those patients who experience recurrent esophageal symptoms should be evaluated with an upper gastrointestinal radiographic study, upper endoscopy, and/or esophageal manometry as clinically indicated. These studies will define the nature of the symptoms and define an anatomic failure. The most common mechanism of anatomic failure is fundoplication herniation into the mediastinum. Most surgeons approach revisional antireflux operations laparoscopically, especially if the first operation was performed laparoscopically. The results of revisional surgery are very good. Success rates are approximately 80% and only 16% of patients are unsatisfied with redo LNF. Success rates decrease dramatically, however, after the first revisional surgery for a variety of reasons, many of them are functional rather than anatomic. The patient who has undergone multiple antireflux operations, therefore, should be approached with caution.

SUGGESTED READINGS

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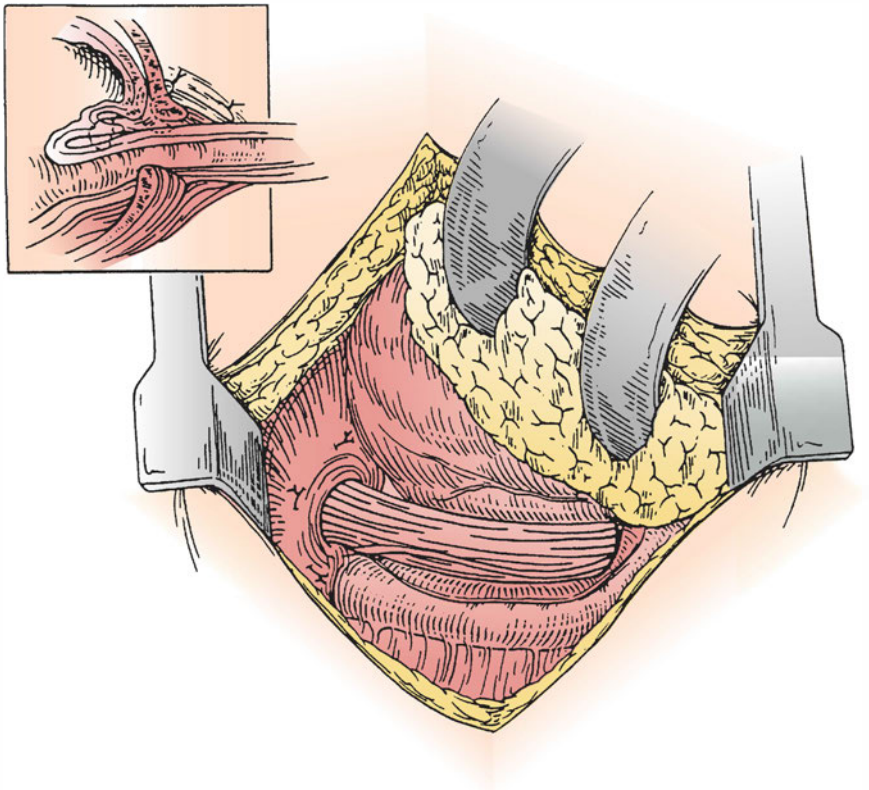


Fig. 16.17. The completed fundoplication showing the wrap anchored below the diaphragm by the second row of sutures.

EDITOR'S COMMENTS

For patients who have continued reflux symptoms despite ideal pharmacologic management, the advent of the laparoscopic Nissen fundoplication (LNF) has made the surgical alternative significantly less foreboding. Patients may be discharged on the first postoperative day with minimal pain and able to eat a full liquid diet with rapid progression to a mechanical soft diet. Both the short- and long-term results of the procedure have been excellent in terms of elimination of reflux symptoms. Interestingly, there seems to be significant variation in the number of these cases performed based on geographic areas. It seems that in certain locations the gastroenterologists seem much more willing to refer cases for operation as compared with other areas where they are extremely hesitant about referring a patient.

The key to success with LNF is patient selection just as it is with other operations designed to alleviate a functional problem. Patients should have a complete preoperative evaluation that includes a pH study, ideally over a 24-hour duration, as well as esophageal manometry. Attention to technical detail is particularly important for optimal outcome and symptom relief without causing a new problem such as dysphagia where previously it did not exist. The authors have provided the pertinent technical details that should be carefully followed. Transient dysphagia may occur in some patients but it should be short lived as the edema resolves.

Rarely is there a need for an open abdominal operation for fundoplication no matter the type of wrap chosen. The thoracic approach should be reserved for those patients in whom revision is necessary and either multiple previous operations have been attempted or previous complications have occurred. A left thoracoabdominal approach may occasionally be useful to approach the left upper quadrant that has previously been violated with multiple procedures and attendant complications. Coming from the chest allows for the esophagus to be visualized, the spleen kept out of the way, and complete mobilization to be effected. The only caveat to this approach is taking care at the time of closure to accurately reapproximate the diaphragm and the costal arch.

LRK

17 Transhiatal Esophagectomy

Mark B. Orringer

INTRODUCTION

The technique of transhiatal esophagectomy (THE) without thoracotomy and a cervical esophagogastric anastomosis (CEGA) was rediscovered in the mid-1970s. Since then, the operation has been progressively refined and has emerged as a recognized alternative to traditional transthoracic esophagectomy for both benign and malignant disease requiring resection and reconstruction. Since the earliest reported series of transthoracic esophagectomy and an intrathoracic esophagogastric anastomosis, the leading causes of postoperative morbidity and mortality have been (1) respiratory insufficiency resulting from a combined thoracoabdominal operation and (2) mediastinitis from an intrathoracic anastomotic leak. THE addressed both issues. By eliminating a thoracic incision, postoperative pulmonary complications were less. By placing the esophagogastric anastomosis in the neck, a leak meant a salivary fistula which drained externally and was not generally associated with mediastinitis. Detractors of THE have argued that the procedure is done "blindly" and has the potential for excessive bleeding, and failure to permit an extensive mediastinal lymph node dissection makes it less attractive from an oncologic standpoint. These concerns have not been substantiated in now more than 30 years of experience with THE, which the author therefore regards as the approach of choice in most patients with benign and malignant disease requiring esophageal resection and reconstruction. This chapter focuses upon the technique of THE, particularly the nuances of gastric mobilization, transhiatal mobilization of the esophagus, preparation of the gastric conduit and positioning it in the mediastinum and neck, and construction of the CEGA.

INDICATIONS AND CONTRAINDICATIONS

For most conditions for which esophageal resection and replacement are indicated, THE and a CEGA are applicable. The authors and his colleagues have reported a series of 2,007 THEs, 1,525 (76%) for carcinoma of the intrathoracic esophagus and 482 (24%) for benign disease (Table 17.1). Indications for esophagectomy in achalasia include a failed prior esophagomyotomy, a reflux stricture after a prior esophagomyotomy (often without a concomitant antireflux procedure), and a tortuous megaesophagus (>6 cm in size). Technical hazards unique to an esophagectomy for achalasia include adherence of the formerly myotomized esophagus to the adjacent descending thoracic aorta; displacement of the megaesophagus into the right chest; larger than usual aortic esophageal arteries; and a much wider than usual cervical esophagus that is more difficult to mobilize and encircle. Reflux strictures have dramatically decreased in frequency since the treatment of gastroesophageal reflux with proton pump inhibitors. Conversely, with the sharp rise in the number of laparoscopic hiatal hernia/antireflux operations being performed, multiple failed repairs or obstruction from recurrence are becoming increasingly frequent indications for THE and a CEGA. After two failed antireflux operations, the likelihood of achieving long-term reflux control and comfortable swallowing is sufficiently low that the author advocates a THE in those *requiring* further surgery. But this decision is not made lightly. Such patients must be informed about the potential functional "downside" of esophagectomy, including dysphagia from an anastomotic stricture, early satiety, posture-related regurgitation, and dumping symptoms. Occasional reflux or intermittent dysphagia from a recurrent hiatal hernia may be a better option than contending with an esophageal anastomotic stricture or chronic dumping symptoms for the rest of one's life.

In the author's experience now with more than 3,000 THEs, this operation has been possible in 98% of patients requiring an esophageal resection for both benign and malignant disease. Contraindications to THE include documented invasion of the airway by upper or middle third esophageal cancers; histologically proven distant metastatic (M1) esophageal carcinoma; and most importantly, the surgeon's assessment through the diaphragmatic hiatus at the time of the planned esophagectomy that tumor fixation to adjacent mediastinal structures precludes a safe transhiatal "blunt" dissection. In those with a history of a prior transthoracic esophagomyotomy, adherence of the exposed submucosa to the adjacent thoracic aorta often creates a technical challenge, and if the esophagus cannot be adequately dissected away from the aorta under direct vision through the hiatus, conversion to a transthoracic esophagectomy is a safer option. When performing a THE, the surgeon must always be prepared to convert to a transthoracic approach when unexpected bleeding or unusually severe fixation of the esophagus to adjacent structures is encountered.

PATIENT SELECTION AND PREOPERATIVE PREPARATION

Critical to a successful THE are careful patient selection and preoperative assessment and conditioning. Physiologic age is of far more important than chronologic age, and there is no absolute age limit to performing this operation. Baseline pulmonary function tests in those with a history of cigarette smoking and cardiology assessment of risk in those with heart disease are routine. The presence of cirrhosis and associated ascites and/or portal hypertension absolutely preclude an esophagectomy by jeopardizing venous drainage of the esophageal replacement and therefore a safe esophageal reconstruction. In those who have undergone multiple prior gastric operations that jeopardize use of the stomach as an esophageal replacement, or those with carcinoma of the cardia that may require a major gastric resection to achieve an adequate margin, the suitability of the colon as an esophageal substitute must be determined in advance. A barium enema examination to exclude severe diverticular disease and a bowel prep preoperatively are undertaken. It has not been the author's

Table 17.1 Indications for Transhiatal Esophagectomy (2007 Patients)

	Number (%)		
	Group I 1,063 Pts	**Group II 944 Pts**	**Total 2,007 Pts**
	1976–1998	**1998–2006**	**1976–2006**
Benign conditions	**278 (26%)**	**204 (22%)**	**482 (24%)**
Neuromotor dysfunction	92 (33%)	47 (23%)	139 (29%)
Achalasia	69	44	113
Spasm/dysmotility	21	3	24
Scleroderma	2	0	2
Stricture	74 (27%)	21 (10%)	95 (20%)
Gastroesophageal reflux	40	7	47
Caustic ingestion	18	6	24
Radiation	4	2	6
Other	12	6	18
Barrett mucosa with high-grade dysplasia	53 (19%)	90 (44%)	143 (30%)
Recurrent gastroesophageal reflux	21 (8%)	6 (3%)	27 (6%)
Recurrent hiatus hernia	14 (5%)	14 (7%)	28 (6%)
Acute perforation	15 (5%)	9 (5%)	24 (5%)
Acute caustic injury	5 (2%)	1 (1%)	6 (1%)
Other	4 (1%)	16 (8%)	20 (4%)
Carcinoma of the intrathoracic esophagus			
Site	**785 (74%)**	**740 (78%)**	**1,525 (76%)**
Upper third	35 (4%)	16 (2%)	51 (3%)
Middle third	164 (21%)	63 (9%)	227 (15%)
Lower third and/or cardia[a]	586 (75%)	661 (89%)	1,247 (82%)

[a]Includes pathologic gastric carcinomas involving the cardia and lower esophagus.
Source: Reproduced with permission from Orringer MB, Marshall B, Chang AC, Lee J, Pickens A, Lau CL. Two thousand transhiatal esophagectomies: Changing trends, lessons learned. Ann Surg 2007;246:363-374.

practice to utilize preoperative mesenteric angiography to evaluate the blood supply of the colon unless there is a history of mesenteric vascular disease or possible interruption of the inferior mesenteric artery in a prior abdominal aortic aneurysm repair. Abstinence from cigarette smoking for a minimum of 3 weeks before the operation is an absolute requirement of the author, even in those with cancer. Those suspected of continued smoking have a urine cotinine level determined, and if recent tobacco use is confirmed, the operation is cancelled. Patients are issued an incentive spirometer at the initial consultation and are requested to use it on a set schedule throughout the day until they are admitted for their operation. They are instructed to walk 3 miles a day to condition themselves for early postoperative ambulation. The importance of this "surgeon–patient contract" is emphasized. With severe esophageal obstruction and inability to obtain adequate fluid and caloric intake by mouth, a nasogastric feeding tube is placed into the stomach, if necessary with fluoroscopic control, and tube feedings for home use are initiated. Every effort is made to avoid a hospitalization and the need for intravenous hyperalimentation before surgery, and patients are typically admitted for the esophagectomy on the day of the scheduled procedure. A mechanical cleansing of the large bowel is undertaken in those in whom there is concern about the suitability of the stomach as an adequate esophageal replacement.



SURGICAL TECHNIQUE

The four phases of THE—abdominal, cervical, mediastinal (transhiatal), and anastomotic—are performed in a sequential and reproducible fashion from patient to patient. Placement of an epidural catheter preoperatively for both intraoperative and postoperative pain control is an important advance. Flexible esophagoscopy is performed after induction of general anesthesia to insure the availability of an adequate proximal esophageal margin as well as a healthy gastric conduit for esophageal replacement. Excessive air insufflation should be avoided. After the endoscopy, a 16F nasogastric tube is placed. A radial artery catheter for intraoperative monitoring of blood pressure during the mediastinal dissection is routine. This catheter is well-secured to avoid its dislodgment during this operation in which the arms will be at the sides, not secured to arm boards. The patient is positioned supine on the operating table with the head turned toward the right and supported on a soft head ring, and the neck extended by a small shoulder roll under the scapulae. The wrists, elbows, and arms are carefully padded with foam, placed at the sides, and secured in place with the drape sheet. This positioning allows the surgeon and his/her assistant unobstructed access to the patient's neck, chest, and abdomen. Table-mounted brackets for securing an upper-hand retractor during the abdominal phase are positioned at the nipple line. "Bumping up" one side or another for an anterolateral thoracotomy in case a transthoracic exposure is required is not done. If conversion to a transthoracic esophagectomy is required, the abdomen is quickly closed temporarily with 4 or 5 through-and-through full-thickness #2 nylon sutures, the wound covered with a plastic surgical drape, and the patient turned and repositioned in the lateral decubitus position for a true posterolateral thoracotomy. After completion of the transthoracic esophagectomy, the chest is closed and the patient turned supine once again and positioned as before for completion of the procedure. The operative field extends from the mandible to the pubis and from one midaxillary line to the other. Two suction catheters, one at the head of the table and one from below, are used routinely. This operation is best performed from the left side of the operating table, and the following operative description of THE is how the procedure would be viewed by the surgeon so positioned.

Abdominal Phase

The sequence of this phase of the operation is gastric mobilization, distal esophageal transhiatal mobilization, Kocher maneuver,

pyloromyotomy, and finally feeding jejunostomy. Through an upper midline supraumbilical incision (Fig. 17.1, inset), the abdomen is explored, the triangular ligament of the liver divided with electrocautery, and the left lobe of the liver retracted to the right with a liver blade retractor secured to the upper-hand supporting bar. A standard upper-hand abdominal wall retractor secured to the bar is used to retract the left side of the abdominal wall. The high greater curvature of the stomach is localized, and the adjacent greater omentum retracted to the left. A "clear space" between the omentum and the gastric wall is developed until the lesser peritoneal sac is entered. An index finger passed into this opening behind the greater omentum elevates the omentum away from the stomach and is used to define the high short gastric vessels and left gastroepiploic arcade. These vessels are sequentially clamped with 13-inch long right-angle clamps, divided, and tied with 2-0 silk, taking care to stay well away from the stomach to avoid ischemic necrosis. The gastric wall must be handled gently throughout this mobilization and unnecessary traction on the stomach avoided. Care must also be taken to avoid excessive downward traction on the high greater omentum that may be adherent to the spleen and result in a splenic capsular tear. Before dividing all of the high short gastric vessels, the direction of the dissection is changed, and the remaining left gastroepiploic vessels sequentially identified, clamped, divided, and ligated moving along the greater curvature toward the pylorus.

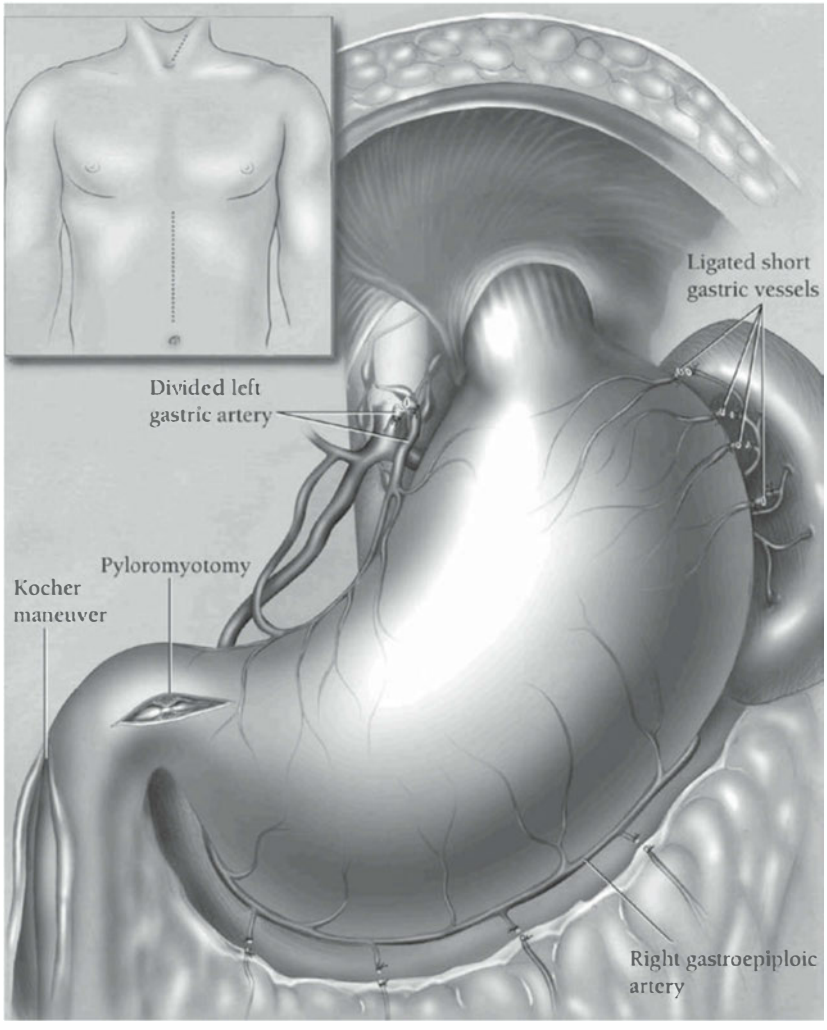


Fig. 17.1. The cervical and supraumbilical incisions used for THE (inset), and the key components of the abdominal phase: ligation of the high short gastric and left gastroepiploic vessels of the stomach; preservation of the right gastroepiploic vascular arcade, the primary blood supply of the mobilized stomach; division of the left gastric artery and vein; a generous Kocher maneuver; and a pyloromyotomy. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

The right gastroepiploic artery is identified by palpation and vision as it either terminates in the gastric wall or communicates through fine vessels with the left gastroepiploic arcade. The greater omentum is divided 1.5 to 2 cm inferior to the right gastroepiploic artery, carefully preserving it and repeatedly monitoring for the presence of a pulse before dividing and ligating each branch. The greater omentum is separated from the stomach to the level of the pylorus. Once this portion of the distal stomach has been mobilized, it can be better retracted to the right exposing the highest short gastric vessels still to be divided. The peritoneum overlying the hiatus is incised and the esophagogastric junction identified. The author no longer encircles the esophagogastric junction with a Penrose drain in order to minimize the chance of a traction injury to the upper stomach. The stomach is elevated and any adhesions between the retroperitoneum and posterior gastric wall divided with electrocautery.

Attention now shifts to the lesser curvature of the stomach. One hand is passed behind the mobilized greater curvature of the stomach into the lesser peritoneal sac until the fingers can be seen through the filmy gastrohepatic omentum on the lesser curvature side of the stomach. The gastrohepatic omentum is incised with electrocautery progressing superiorly toward the diaphragmatic hiatus, carefully palpating for a pulse in an aberrant left hepatic artery arising from the left gastric artery. This aberrant vessel is preserved if present by dividing the left gastric artery distal to its origin. By retracting the stomach to the left, the lesser curvature soft tissues are tensed, facilitating identification, mobilization, clamping, dividing, and ligating of the left gastric vein and then the left gastric artery with 2-0 silk ties. The artery is doubly ligated at its origin from the celiac trunk, and all lymph nodes in this area dissected and swept to the left attached to the lesser curvature. Similarly, all soft tissue and lymph nodes along the lesser curvature of the stomach anterior to the right crus of the diaphragmatic hiatus are swept to the left with the stomach. With mobilization of the greater curvature and high lesser curvature of the stomach completed, dissection of the esophagogastric junction and lower esophagus is begun.

The mobilized stomach is gently retracted downward as the phrenoesophageal tissue at the level of the hiatus is incised and the posterior mediastinum entered anterior to the esophagus. This space is enlarged bluntly with a sweeping

motion of the fingers so that a narrow heart-shaped retractor can be inserted and upward traction on it exerted. This exposes the distal esophagus in the posterior mediastinum. If tumor at the cardia is adherent to the diaphragm, a rim of diaphragm may need to the resected with the specimen to achieve an adequate margin. The posterior aspect of the esophagus is gently mobilized away from the spine by two, three, and then four fingers inserted into the hiatus behind the esophagus. Using a long 13-inch right-angle clamp and a long electrocautery tip, the posterior mediastinal paraesophageal soft tissues are elevated and divided. Visible paraesophageal lymph nodes are resected. The dissection alternates from one side to the other of the esophagus moving progressively toward the carina. If either lung is seen in the field, the mediastinal pleura on that side has been violated, and a chest tube will be required later. Inserting a hand into the low mediastinum and gently "rocking" the esophagus from side to side allows assessment of the degree of fixation between the esophagus and contiguous structures, particularly the spine, prevertebral fascia, and descending thoracic aorta, which could complicate transhiatal esophageal mobilization. During this low esophageal dissection, blood pressure as recorded through the radial artery catheter is carefully watched to avoid prolonged hypotension resulting from cardiac displacement by the hand inserted into the mediastinum. After mobilizing approximately 10 cm of the distal esophagus, the low posterior mediastinum is packed with a large abdominal pad to facilitate hemostasis.

The duodenum is mobilized with a generous Kocher maneuver to facilitate the eventual upward reach of the stomach through the posterior mediastinum. After an adequate Kocher maneuver, the pylorus should be able to be grasped and moved from its usual position in the right upper quadrant to the level of the xiphoid process. A pyloromyotomy is now performed. Two 3-0 silk figure-of-eight traction sutures are placed, one through the superior aspect and the other through the inferior aspect of the pylorus. The anterior pylorus is thus elevated as the pyloromyotomy is begun. Using a low cutting current from a needle-tipped electrocautery, the gastric serosa and superficial muscle layers are incised beginning approximately 1.5 to 2 cm on the gastric side of the pylorus. A fine right-angle clamp is then used to elevate the remainder of the muscle away from the submucosa as the muscle is progressively divided moving toward the pylorus. The dissection then becomes more superficial as the serosa and a few muscle fibers overlying the pylorus are cut, and the serosa and muscle of the duodenum divided for 0.5 to 1 cm. The duodenal submucosa is distinctly different from the gastric submucosa, the former being more fatty, more yellow in color, and having fine veins coursing over it, while the gastric submucosa is more pink and has relatively fewer veins on its surface. With the gastric and duodenal submucosa exposed on either side of the pylorus, the pyloric muscle is readily identified, dissected, and elevated with the right-angle clamp, and progressively divided with the needle-tipped electrocautery. Freely bulging submucosa between the two cut ends of the pylorus signifies a complete myotomy. At the conclusion of the pyloromyotomy, a silver hemoclip is placed near the pylorus at the base of either traction suture to mark the level of the pylorus for future radiographic evaluation. Should the integrity of the pyloroduodenal mucosa be violated, the pyloromyotomy is *not* converted to a pyloroplasty. Rather, the hole is closed with several interrupted 5-0 polypropylene sutures. This avoids a pyloroplasty suture line at right angles to the vertical axis of the stomach when the stomach is pulled upward through the posterior mediastinum. If repair of a pyloroduodenal mucosal tear is required, once the stomach has been brought through the mediastinum and the fundus delivered to the neck, the pyloromyotomy site, which typically comes to rest approximately 3 to 4 cm below the level of the hiatus, is buttressed with adjacent omentum or greater curvature fat loosely "tacked" in place with several interrupted 4-0 sutures. A feeding jejunostomy is performed using a 14F rubber catheter with side holes cut at its tip inserted into the bowel 18 to 20 cm distal to the ligament of Treitz. The tube is secured in place with a 4-cm long Witzel maneuver. The jejunostomy tube is not yet brought through the abdominal wall but rather covered with a towel clamped on either side to the drapes with hemostats as it emerges from the lower end of the abdominal incision to prevent its dislodgment during the remainder of the operation. Attention is now turned toward the neck.

Cervical Phase

Palpation of the cricoid cartilage identifies the level of the cricopharyngeus sphincter, the esophageal introitus. A 5 to 7 cm long oblique left cervical incision that parallels the anterior border of the sternocleidomastoid muscle is made (Fig. 17.2). The incision extends no more than 2 cm superior to the cricoid cartilage; as there is no esophagus above this point, a long cervical incision toward the angle of the mandible contributes nothing to exposure of the esophagus. The incision is deepened through the platysma muscle and then the fascia along the anterior edge of the sternocleidomastoid muscle. The sternocleidomastoid muscle is separated from the underlying cervical muscle using blunt finger dissection in the direction of the muscle. The sternocleidomastoid muscle is retracted laterally, and the underlying omohyoid muscle can be seen coursing obliquely in the opposite direction of the sternocleidomastoid muscle. The tendinous midpoint of the muscle is elevated with a fine right-angle clamp and divided with electrocautery (Fig. 17.3). Traction on the cut medial side of the omohyoid muscle tenses the omohyoid fascial layer, which is incised with electrocautery in the direction of the cervical incision. This omohyoid fascial layer leads to the underlying carotid sheath and its contents. Throughout the entire cervical portions of this operation, no metal retractors are placed against the tracheoesophageal groove in order to avoid injury

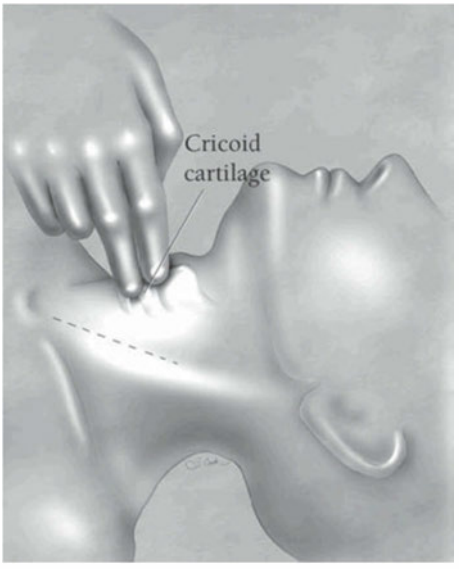


Fig. 17.2. The cricoid cartilage marks the level of the upper esophageal sphincter, the esophageal introitus. The 5 to 7 cm long incision along the anterior edge of the left sternocleidomastoid muscle should extend no more than 2 cm proximal to the level of the cartilage. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

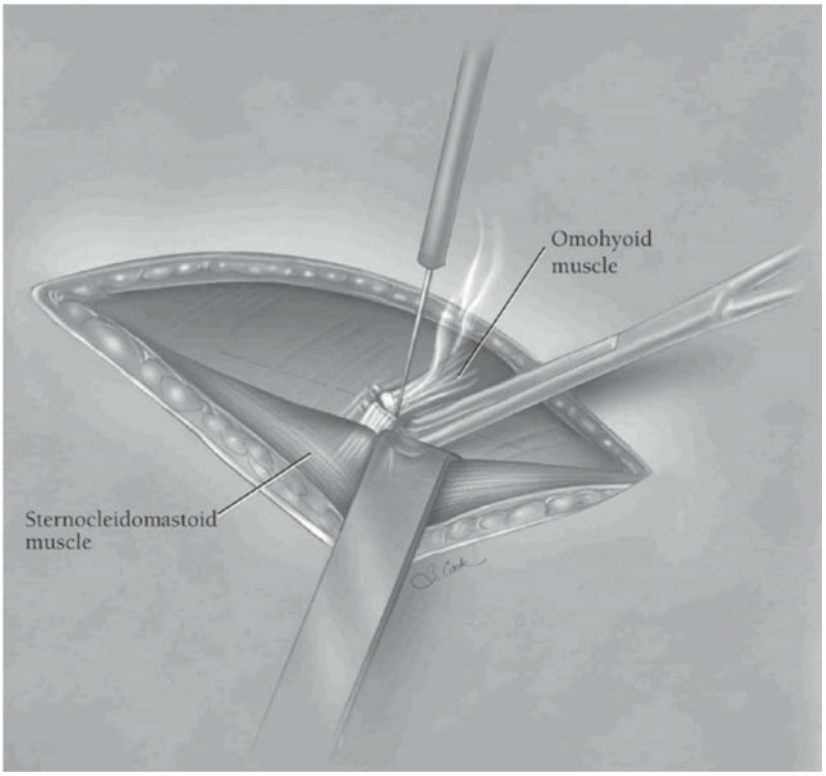


Fig. 17.3. The left sternocleidomastoid muscle is retracted laterally revealing the underlying omohyoid muscle, the central tendon of which is elevated with a right-angle clamp and divided with electrocautery. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

to the recurrent laryngeal nerve. While the sternocleidomastoid muscle and carotid sheath are retracted laterally, the thyroid and trachea are retracted medially by the index finger of the first assistant (Fig. 17.4). The middle thyroid vein arises from the medial aspect of the internal jugular vein and is clamped, divided, and ligated. A critical landmark in dissection of the cervical esophagus is the inferior thyroid artery, which consistently can be found at the level of the cricoid cartilage. This artery is clamped with fine right-angle clamps, divided, and ligated. The cricopharyngeal sphincter is found immediately at the level of the inferior thyroid artery, and all subsequent dissection of the esophagus should be inferior to this point. The deep cervical fascia anterior to the prevertebral fascia is incised in the direction of the incision, and again by blunt finger dissection in the same direction, the prevertebral fascia is cleared of tissue anterior to it and the cervical esophagus (with its contained nasogastric tube) clearly identified. By following the prevertebral fascia by blunt finger dissection into the superior mediastinum, the cervical esophagus is mobilized posteriorly. The trachea and esophagus are

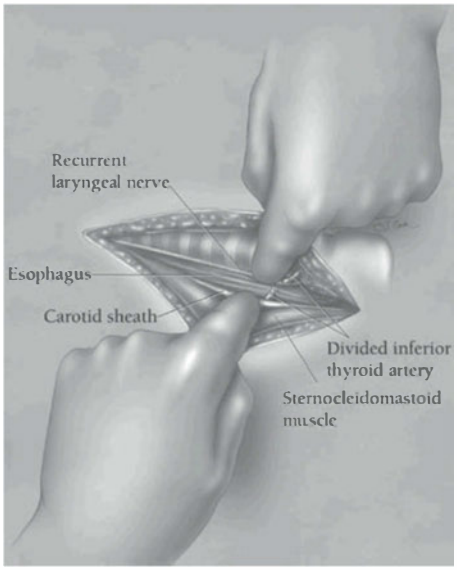


Fig. 17.4. The thyroid and trachea are retracted medially by the index finger of the assistant, never by a metal retractor, to avoid injury to the recurrent laryngeal nerve in the tracheoesophageal groove. The inferior thyroid artery is reliably found medial to the carotid sheath at the level of the cricoid cartilage and is ligated and divided. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

retracted medially, tensing the left cervical strap muscles, which are elevated with a right-angle clamp and divided with electrocautery to provide optimal exposure of the esophagus at the level of the thoracic inlet.

An important maneuver in mobilizing the cervical esophagus is upward traction on the tracheoesophageal groove applied at the level of the cricoid cartilage by the first assistant's index finger (Fig. 17.5). The additional length of cervical esophagus made accessible by this maneuver facilitates its subsequent mobilization. Sharp dissection posterior and lateral to the tracheoesophageal groove is performed, but the recurrent laryngeal nerve is not actively exposed or dissected. By finger dissection, the anterior wall of the cervical esophagus is freed the length of the cervical wound. The surgeon's index finger is then advanced medially and posteriorly around the esophagus until the prevertebral fascia on the right side is palpated. As the first assistant's index finger continues to elevate the cervical esophagus out of the superior mediastinum, and with the surgeon's left index finger around the esophagus palpating the prevertebral fascia, a long right-angle clamp is passed behind the esophagus along the front of the spine until its tip is felt by the left index finger. The right-angle clamp is spread with its tip against the left index finger completing the pathway around the esophagus so that the surgeon's left index finger can now be advanced until it appears in the wound coming from behind the esophagus. With the cervical esophagus now circumferentially mobilized, a 1-inch rubber drain is placed around the esophagus from behind, protecting the recurrent nerve the entire time with the left index finger to avoid injury to it (Fig. 17.6). The esophagus is mobilized out of the superior mediastinum by upward traction on the rubber drain as the volar aspects of the right index and middle fingers progressively sweep away the periesophageal soft tissue (Fig. 17.7). Once the cervical esophagus has been encircled and a 4 to 5 cm length of upper esophagus mobilized in the superior mediastinum, the cervical phase of the operation is completed.

Mediastinal (Transhiatal) Phase

As mentioned previously, the author now avoids an encircling Penrose drain around the esophagogastric junction (as shown in Fig. 17.7) to minimize inadvertent traction

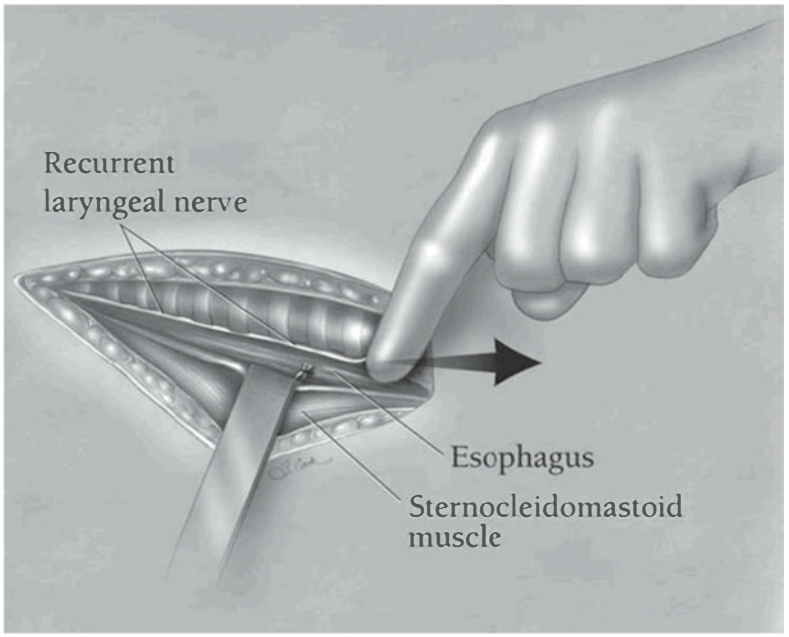


Fig. 17.5. The esophagus is elevated out of the superior mediastinum by cephalad traction on the tracheoesophageal groove by the assistant's index finger at the level of the cricoid cartilage. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

and trauma to the upper stomach. With one hand providing gentle downward traction on the mobilized stomach, the posterior mediastinal dissection is begun by advancing the opposite hand behind the esophagus along the prevertebral fascia. Resectability of the esophagus and any contained tumor, if present, is assessed by grasping the esophagus and "rocking" it to determine its mobility relative to adjacent tissues.

The drain around the cervical esophagus is elevated and retracted anteriorly, while a "half-sponge-on-a-stick" is inserted into the neck wound behind the esophagus and progressively advanced into the superior mediastinum along the prevertebral fascia (Fig. 17.8). The sponge is eventually felt by the fingers inserted through the abdomen and the hiatus. Generally, filmy posterior paraesophageal soft tissue is pushed aside and disrupted by the sponge-on-a-stick held against the finger tips. At all phases of the transhiatal mediastinal dissection, prolonged hypotension resulting from



Fig. 17.6. As the esophagus is being pulled cephalad by the assistant's index finger, the surgeon's index finger is advanced into the tracheoesophageal groove posterior to the recurrent nerve around the anterior surface of esophagus until the prevertebral fascia can be felt. A right-angle clamp passed behind the esophagus and spread against the tip of the index finger helps to develop the plane. A 1-inch flat rubber drain is passed posteriorly around the esophagus to the encircling index finger, rigorously avoiding contact between the clamp and the recurrent nerve. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

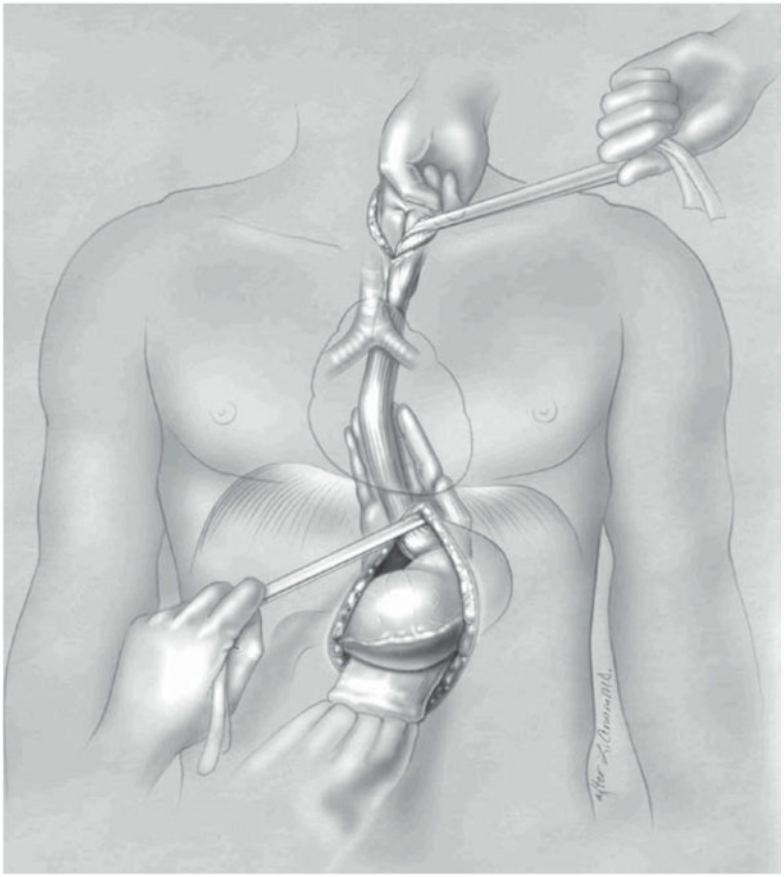


Fig. 17.7. Blunt dissection of the low cervical and upper thoracic esophagus from the superior mediastinum is performed as traction is applied to the encircling cervical drain. Through the hiatus, with gentle downward traction on the stomach (generally *not* an encircling drain around the esophagogastric junction as shown to minimize gastric trauma), the volar aspects of the fingers are kept against the posterior aspect of the lower esophagus, sweeping it away from the prevertebral fascia. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

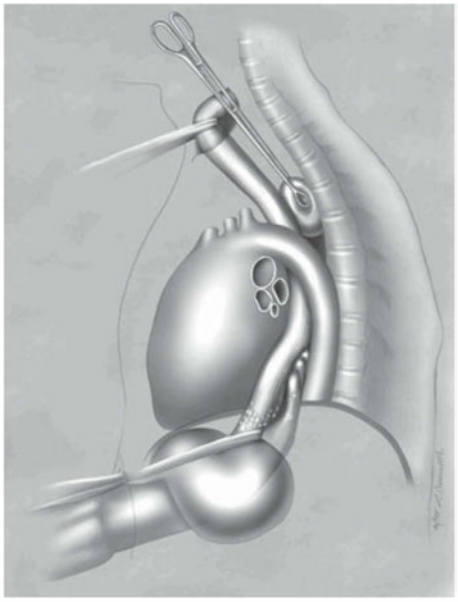


Fig. 17.8. A "half-sponge-on-a stick" is advanced downward through the cervical incision along the prevertebral fascia until it can be palpated by the fingers inserted through the hiatus behind the esophagus. Filmy attachments are swept aside and the first portion of the posterior mediastinal "tunnel" established. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

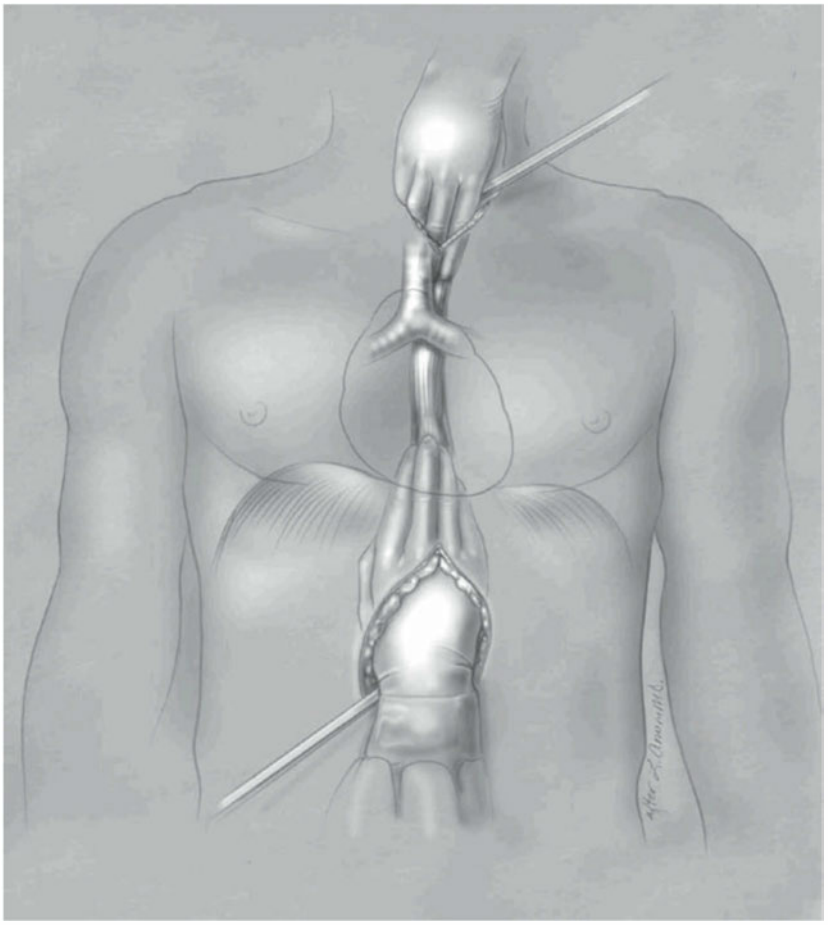


Fig. 17.9. A "mirror image" dissection along the anterior surface of the esophagus is carried out through the cervical incision and the hiatus, carefully passed behind the membranous trachea and the carina. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

cardiac displacement is avoided by visually monitoring the radial artery blood pressure that is displayed on a monitor readily seen by both the surgeon and the anesthesiologist. After the posterior mediastinal "tunnel" has been created, a 28F Argyle Saratoga sump catheter is inserted through the cervical incision and advanced into the posterior mediastinal space both to assess bleeding and evacuate blood prior to the next phase of the dissection.

The anterior esophageal dissection is now begun through the abdomen by advancing the hand through the diaphragmatic hiatus palm downward against the anterior surface of the esophagus as the other hand gently retracts the mobilized stomach downward (Fig. 17.9). The drain around the cervical esophagus is retracted superiorly and toward the patient's left shoulder as two fingers are inserted through the cervical incision, volar aspects against the anterior surface of the esophagus, and advanced into the superior mediastinum. The hand inserted through the mediastinum from below must constantly be kept as far posterior as possible to minimize cardiac displacement, and as this anterior esophageal dissection proceeds, care must be taken to avoid injury to the pericardium as well as the posterior membranous trachea as the periesophageal attachments to the trachea are gently disrupted (Fig. 17.10).

After the posterior and anterior esophageal dissections, the resulting increased mobility of the esophagus in the mediastinum allows more of it to be elevated out of the superior mediastinum through the cervical incision. While "hooking" the left index finger around the esophagus and gently lifting it, disruption of the lateral esophageal attachments is performed by the right index finger advanced downward into the superior mediastinum (Fig. 17.11). A 5 to 8 cm segment of the upper thoracic esophagus is thereby completely circumferentially mobilized and is released and allowed to retract back down into the superior mediastinum.

A hand is now inserted through the abdomen and the diaphragmatic hiatus anterior to the esophagus and advanced upward through the mediastinum to the region of upper esophageal mobilization just achieved. The circumferentially mobilized upper thoracic esophagus is "trapped" against the spine between the index and middle fingers (Fig. 17.12), and the lateral periesophageal attachments and fine vagal fibers are avulsed by a progressive downward raking motion of the fingers held against the prevertebral fascia (Fig. 17.13). As the region of the inferior pulmonary hila is reached, the fine vagal fibers often coalesce to become more substantial vagal trunks. These are either ruptured by compression between the thumb and index finger or are "hooked" by the index finger and retracted downward where they can be seen through the diaphragmatic hiatus sufficiently to be elevated with a long right-angle clamp, and divided with a long electrocautery extension (Fig. 17.13, inset).

With mobilization of the thoracic esophagus from the posterior mediastinum completed, 3 to 4 cm of the upper thoracic

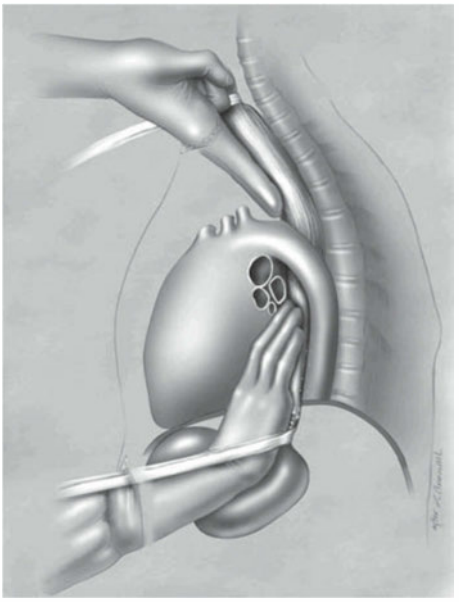


Fig. 17.10. As generally filmy attachments between the anterior wall of the esophagus and the posterior trachea are swept aside, care must be taken to avoid a posterior membranous tracheal tear. The hand inserted through the hiatus must be pushed posteriorly toward the spine to minimize cardiac displacement and hypotension. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)



Fig. 17.11. After the anterior and posterior esophageal "tunnels" have been created, the esophagus is more mobile and can be elevated somewhat out of the cervical wound by the index finger of the left hand hooked around it. The index finger of the right hand sweeps the upper lateral esophageal attachments away from the esophagus, thereby creating an entirely circumferentially mobilized portion of the upper thoracic esophagus, which drops back into the superior mediastinum when the fingers are withdrawn. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

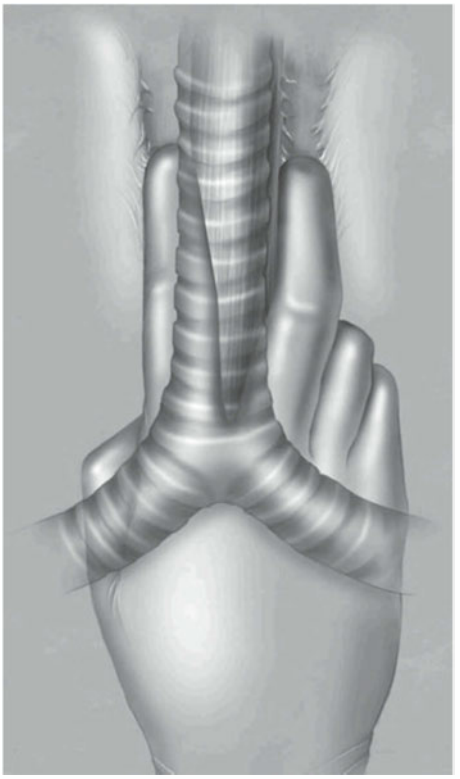


Fig. 17.12. A hand inserted through the diaphragmatic hiatus is advanced along the anterior surface of the esophagus up into the superior mediastinum. The index and middle fingers trap the circumferentially mobilized upper esophagus between them, and with a downward raking motion avulse the lateral esophageal attachments. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

esophagus is elevated out of the cervical wound, the nasogastric tube is withdrawn until its tip is at the level of the upper esophageal sphincter, and the esophagus is divided obliquely from anterior to posterior (not transversely) with a gastrointestinal anastomosis (GIA) surgical stapler so that the anterior tip is longer than the posterior tip (Fig. 17.14). Unless precluded by existing esophageal pathology such as a very high squamocolumnar epithelial junction of Barrett mucosa or a midesophageal carcinoma, the upper esophagus is intentionally divided 3 to 4 cm longer than it appears will be needed to construct the cervical esophagogastric anastomosis.

After transecting the upper esophagus in the neck, the stomach is grasped in the abdomen, and with downward traction, the thoracic esophagus is delivered out of the posterior mediastinum through the diaphragmatic hiatus, and the stomach and attached esophagus are brought out of the abdominal wound. While it is always

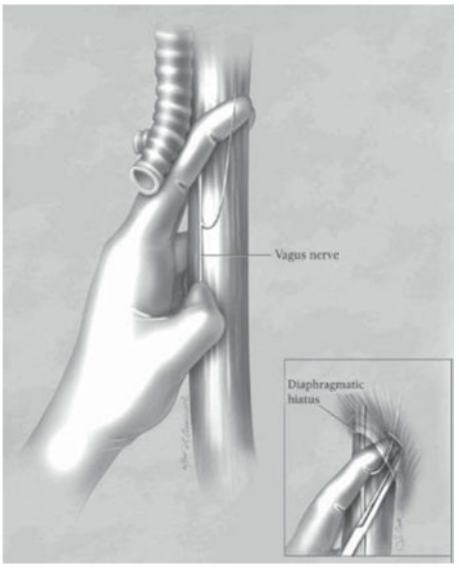


Fig. 17.13. More substantial vagal branches are "hooked" by the downward moving index finger, delivered to the level of the hiatus, elevated with a long right-angle clamp, and divided with electrocautery. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

tempting at this point to examine and palpate the extirpated esophagus, inspection of the posterior mediastinum is now the priority. Once the thoracic esophagus has been delivered out of the posterior mediastinum, the liver blade of the upper-hand



Fig. 17.14. With the entire intrathoracic esophagus now mobile, several centimeters of the upper esophagus is drawn into the cervical wound and divided obliquely (not transversely) from front to back, leaving the anterior tip longer than the posterior. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

retractor is removed and replaced with a standard body wall blade. This is because the deep retractor pulls the lesser curvature of the stomach toward the right upper quadrant, interfering with the subsequent upward reach of the stomach to the neck. The 28F Argyle Saratoga sump catheter is inserted into the posterior mediastinum through the neck wound, and with a long narrow Deaver retractor in the diaphragmatic hiatus, the mediastinum is inspected for untoward bleeding and both sides of the mediastinal pleura for entry into a pleural cavity. An occasional bleeding descending aortic esophageal artery may be encountered, clamped with a long right-angle clamp, and ligated. If either pleural cavity has been entered as indicated by a tear of the mediastinal pleura upon direct palpation of either side of the mediastinum or by seeing visible lung on one or both sides, a 28F-chest tube is placed in the appropriate side through a low interspace in the anterior axillary line, advanced to the apex, secured to the skin, and connected to underwater seal drainage. The blades of the upper-hand retractor should always be temporarily removed when chest tube placement is required, since the retraction distorts the anterior chest wall and may interfere with optimal chest tube positioning. When it has been established that there is no major mediastinal bleeding, a large abdominal pack is placed into the low posterior mediastinum through the diaphragmatic hiatus using a long packing forceps. Through the cervical incision, and protecting the tracheoesophageal groove with a finger, two narrow "thoracic packs" are placed into the upper mediastinum with a packing forceps. These packs provide pressure in the mediastinum to encourage hemostasis.

After chest tube placement and temporary packing of the mediastinum, preparation of the gastric conduit begins. Minimizing gastric trauma so that the stomach is "pink in the belly" (at the conclusion of its preparation) and "pink in the neck" (after it has been transposed through the posterior mediastinum) has become an overarching principle of the operation. The key to a successful esophagogastric anastomosis is a well-vascularized, healthy esophagus and stomach. Any technical maneuvers that can potentially harm the gastric blood supply are avoided. These include gastric traction sutures, suction devices applied to the stomach to pull it through the posterior mediastinum, aggressive "tubularization" of the stomach that excessively interrupts vital submucosal gastric collateral circulation, or suspending the mobilized stomach in the neck with sutures between the prevertebral fascia and the tip of the mobilized stomach, the point most vulnerable to ischemic necrosis.

The mobilized stomach and attached esophagus are placed upon the anterior chest, and the highest point on the greater curvature of the stomach that will reach upward to the neck identified and grasped with a moist gauze. At approximately the level of the second vascular arcade ("crow's foot") along the lesser curvature, fatty tissue and vessels are mobilized, clamped, divided, and tied to clean an area for application of the stapler. With constant gentle upward traction being applied to the tip of the stomach, and *beginning on the lesser curvature of the stomach,* the GIA 60-3.5 stapler is applied approximately 4 to 6 cm distal to the esophagogastric junction (Fig. 17.15). The stomach is progressively straightened with each successive application of the stapler, thereby gradually eliminating the natural curve of the stomach toward the patient's right side and achieving maximal upward gastric length. The emphasis is upon preserving a uniformly wide gastric conduit and resisting the tendency to angle the stapler toward the greater curvature and create a narrow gastric tip. Once the esophagus and proximal stomach have been separated from the gastric conduit, the specimen is removed from the field and inspected on the back table for adequacy of the gastric margin beyond tumor and of the proximal esophageal margin relative to the esophageal pathology. Frozen sections on the resected specimen may be obtained if there is concern about the margin; however, this is not done routinely.

While applying upward traction on the stomach to avoid "purse-stringing" the lesser curvature, the gastric staple suture line is oversewn from either end with two running 4-0 absorbable polyglycolic acid sutures interrupted in the mid portion of the lesser curvature to reduce the chance that the suture will break as the stomach is later drawn through the mediastinum and the tip into the neck. With preparation of the gastroesophageal conduit completed, placement of the stomach on the anterior chest shows that the stomach will reach above the level of the suprasternal notch; an additional 2 to 3 cm of length will be gained when the stomach is brought through

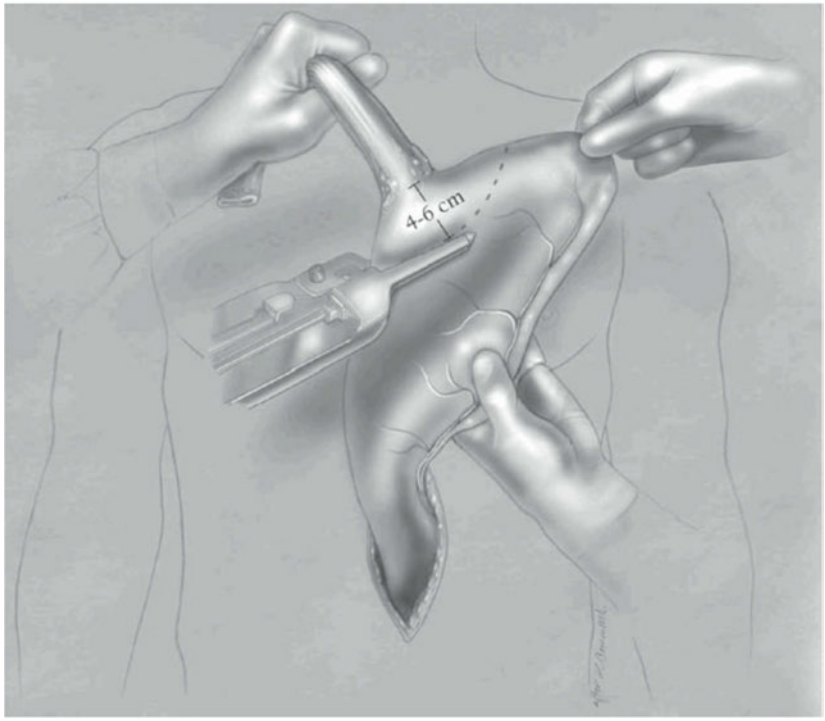


Fig. 17.15. With steady gentle cephalad traction on the tip of the greater curvature that will reach most superiorly to the neck, the GIA stapler is applied beginning on the lesser curvature at the level of the second vascular arcade ("crow's foot") 4 to 5 cm distal to the esophagogastric junction. With each progressive application of the stapler, the stomach is "straightened" to achieve maximum length. The maximum width of the stomach is preserved to insure as optimal submucosal gastric collateral circulation as is possible. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

the posterior mediastinum. The gastric staple line is toward the patient's right side (Fig. 17.16). The stomach is now lowered and brought out of the inferior end of the abdominal incision and gently wrapped in an abdominal lap pad that is moistened with warm saline.

The previously placed cervical and posterior mediastinal gauze packs are removed, and with a narrow Deaver retractor in the diaphragmatic hiatus, a final inspection for hemostasis is made. The hand and forearm are passed upward through the posterior mediastinum and advanced to the thoracic inlet to be certain that there are no major remaining adhesions, pleural bands, or vagal branches to impede the upward mobilization of the stomach. The left upper quadrant abdominal contents are retracted to the left, and the liver to the right. The wet pack around the stomach is removed and the gastric tip is "wet down" with warm saline so that it will slide more easily. As the Deaver retractor holds the hiatus open, one hand gently pushes the tip of the stomach posteriorly through the posterior mediastinum against the spine, beneath the aortic arch, and upward toward the thoracic inlet. A Babcock clamp inserted downward through the cervical incision is used to grasp the tip of the stomach held in place against the spine by the hand in the mediastinum (Fig. 17.17). To avoid unnecessary gastric tip trauma, the clamp is not ratcheted closed but rather is used to gently pull the stomach toward the neck as the hand in the mediastinum pushes the stomach upward. This continues until 4 to 5 cm of stomach comes to rest above the level of the clavicles, the stomach being pushed more from below than pulled upward from the neck wound. Proper orientation of the stomach within the posterior mediastinum should be documented by visualization in the neck of the gastric staple suture line toward the patient's right side and demonstration of the right gastroepiploic vascular arcade at the level of the diaphragmatic hiatus toward the patient's left side. Finally, insure that no twist in the stomach has occurred by gentle palpation of the anterior surface of the transposed stomach by one hand inserted through the hiatus and a finger inserted downward through the neck incision along the anterior surface of the upper stomach. Once in the neck, the tip of the stomach will generally remain in the wound without the need for traction sutures. Should it begin to slip back downward into the mediastinum, a moistened narrow thoracic pack placed gently behind the stomach at the thoracic inlet will usually stop the retraction. The author typically applies a hemostat across the stump of one of the ligated high short gastric vessels. As the abdominal portion of the procedure is completed, periodic confirmation that the clamp protruding from the neck is in place insures that the stomach remains in the neck wound. The cervical wound is covered with a saline-soaked gauze pack as attention is redirected to the abdomen.

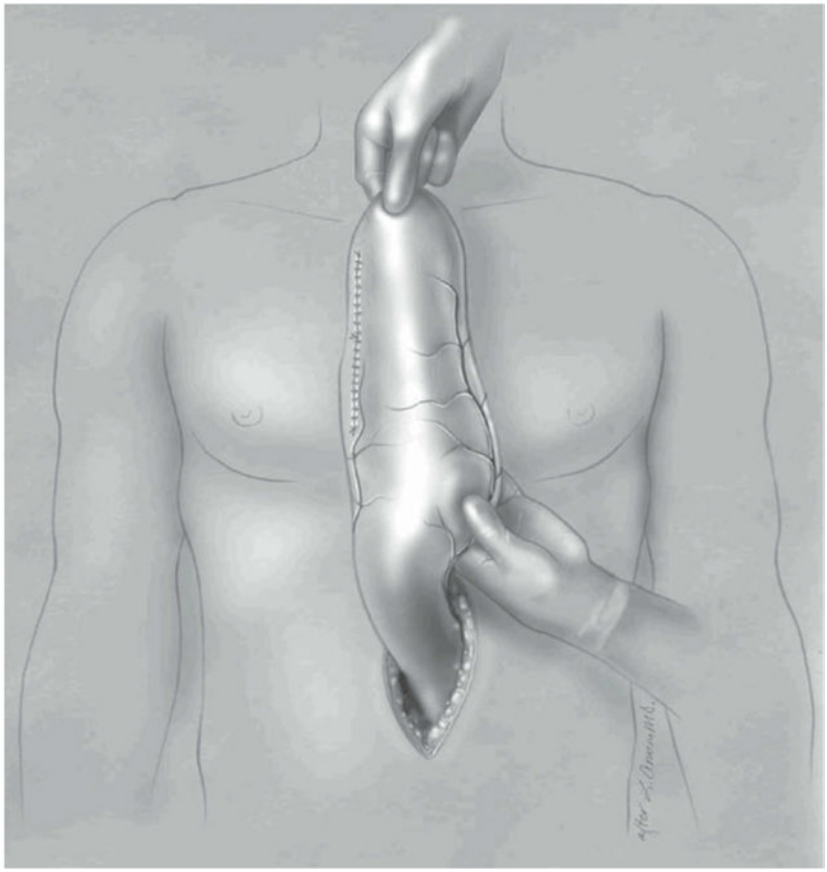


Fig. 17.16. When the mobilized stomach is placed on the anterior chest, the tip generally reaches to or above the level of the clavicles, the right gastroepiploic vascular arcade is toward the left, and the oversewn gastric staple suture line is toward the patient's right side. This orientation must be maintained as the stomach is transposed through the posterior mediastinum. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

The diaphragmatic hiatus has been enlarged, and occasionally partially torn, as the surgeon's hand and forearm were inserted through it into the posterior mediastinum during the esophageal mobilization. Herniation of intestine into the chest alongside the stomach may occur unless the hiatus is at least somewhat repaired. This is generally achieved with one or two figure-of-eight #1 silk sutures placed through the hiatus to the left of the stomach. Great care must be taken to protect the right gastroepiploic vascular arcade, the pericardium and the lung as the hiatus is being narrowed. The author typically narrows the hiatus so that it will accommodate three fingers alongside the stomach within the hiatus. It is best to err on the side of keeping the hiatus loose, as excessive narrowing may result in subsequent outlet obstruction of the intrathoracic stomach. The edge of the diaphragmatic hiatus is sutured to the anterior gastric wall at the level of the hiatus with one or two 3-0 silk sutures to further prevent herniation through the hiatus. The left lobe of the liver is returned to its normal location, and an additional 3-0 silk suture is used to tack the previously divided triangular ligament back against the left side of the hiatus. The pyloromyotomy site typically comes to rest 3 to 4 cm below the level of the hiatus, covered by the overlying left lobe of the liver. The left upper quadrant, particularly the spleen, is inspected for bleeding. The feeding jejunostomy tube is brought through the abdominal wall through a stab wound in the left upper wall. The jejunostomy tube site is secured against the anterior abdominal wall with four sutures between the intestine and the adjacent peritoneum. Prior

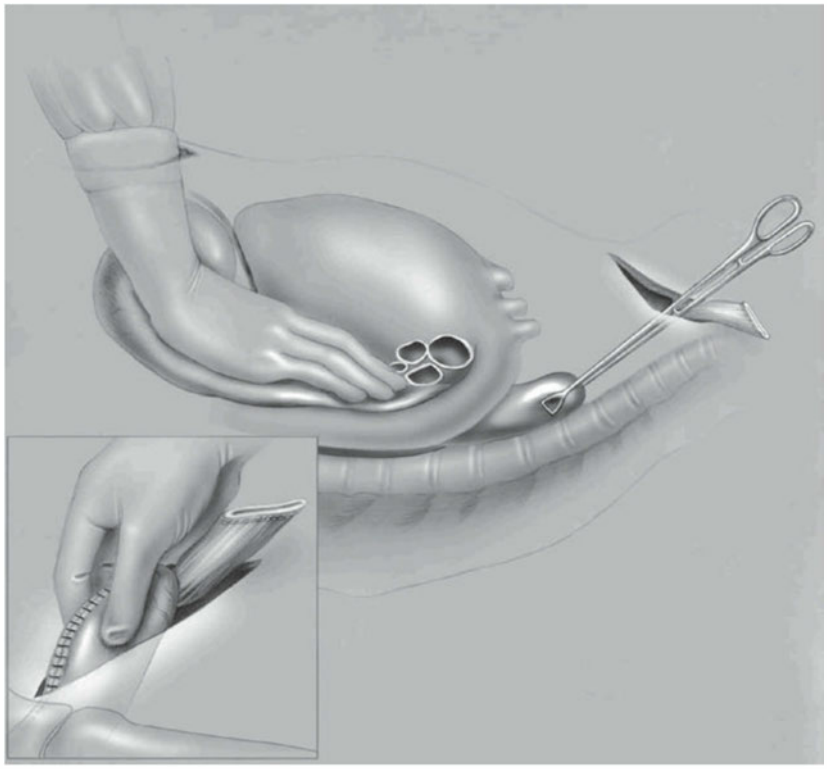


Fig. 17.17. The stomach is gently pushed upward through the hiatus and into the mediastinum between the volar aspects of the fingers in front and the spine behind and advanced beneath the carina and the aortic arch until the tip can be felt with a Babcock clamp inserted downward through the cervical incision into the superior mediastinum. The gastric tip is grasped with the Babcock clamp, which is *not* ratched closed, and is gradually drawn into the cervical wound (inset) more by pushing from below than pulling from above. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

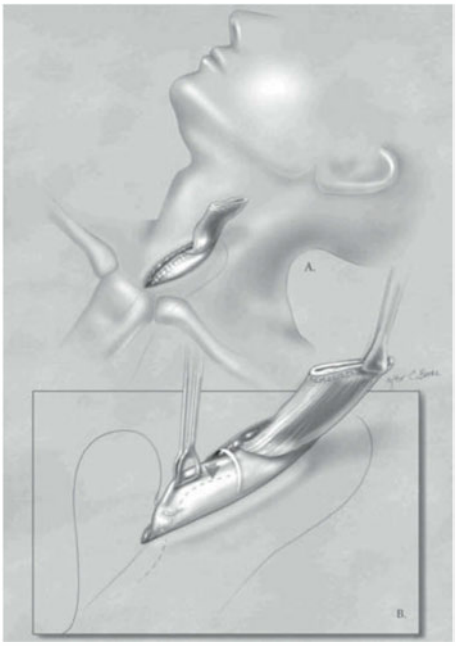


Fig. 17.18. (A) As the anastomosis begins, the gastric staple suture line is toward the patient's right side, and the divided upper esophagus is retracted superiorly. **(B)** The anterior gastric wall at the level of the suprasternal notch is gently grasped with a Babcock clamp and rotated even more medially to the right, rolling the gastric staple suture line even further away from the area on the stomach where the anastomosis will be performed. A 3-0 silk seromuscular traction suture is placed in the anterior gastric wall as far inferiorly as can be reached. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

to cutting these sutures, each is marked with a hemoclip adjacent to the knot. This provides a "target" for the interventional radiologists should percutaneous reinsertion of a feeding tube ever be needed in the future. The jejunostomy tube is sutured to the skin. As the latter final steps are being undertaken prior to closure of the abdomen, the color of the gastric tip in the cervical wound is periodically assessed to be certain that overaggressive narrowing of the hiatus has not resulted in venous congestion that would merit removal of a hiatal suture prior to closing the abdomen. The abdominal incision is then closed and covered with a sterile towel and then a separate drape to isolate the wound from contamination by oral bacteria that may occur once the esophagus is opened in the neck for construction of the anastomosis.

Cervical Esophagogastric Anastomosis

With adequate gastric mobilization, there typically is a 4 to 5 centimeter length of stomach visible above the level of the clavicles. As was established when the stomach was brought to the neck, the gastric staple suture line is oriented toward the patient's right side (Fig. 17.18A). The gastric staple suture line is displaced even further toward the right with a gently applied Babcock clamp to provide an area for construction of the anastomosis well away from the suture line. The anterior gastric wall is elevated somewhat with the Babcock clamp applied as low as possible behind the sternal notch, and a 3-0 silk seromuscular traction suture is placed inferior to the clamp (Fig. 17.18B). This is the first of three key sutures used to align the posterior wall of the divided cervical esophagus with the anterior wall of the stomach in preparation for the side-to-side stapled CEGA. The stapled end of the divided cervical esophagus is grasped with two Allis clamps and retracted toward the patient's right as the gastric traction suture is being placed.

Estimating the appropriate site for the anterior gastrotomy and the length of the cervical esophagus proximal to the anastomosis may be challenging. A 1.5 to 2 cm vertical anterior gastrotomy is created after carefully assessing where the divided end of the esophagus will come to rest once the stapled end has been transected (Fig. 17.19). This is typically done with a needle tip electrocautery on a low cutting setting. The gastrotomy must be positioned sufficiently low on the anterior gastric wall that an ENDO 45-3.5 staple cartridge eventually can be inserted to its length. The stapled end of the divided cervical esophagus is grasped from front to back with a DeBakey forceps, and the staple line is amputated sharply distal to the forceps (Fig. 17.20). This amputated end serves as the "proximal cervical esophageal margin." If there is any concern about proximity to tumor or to Barrett mucosa, the end can be submitted for frozen section analysis, but this is not done routinely. The second key suture used to set up the anastomosis is a 3-0 silk suture placed full thickness through the anterior corner of the divided cervical esophagus. The final and third stitch passes into the stomach and out 5 to 6 mm from

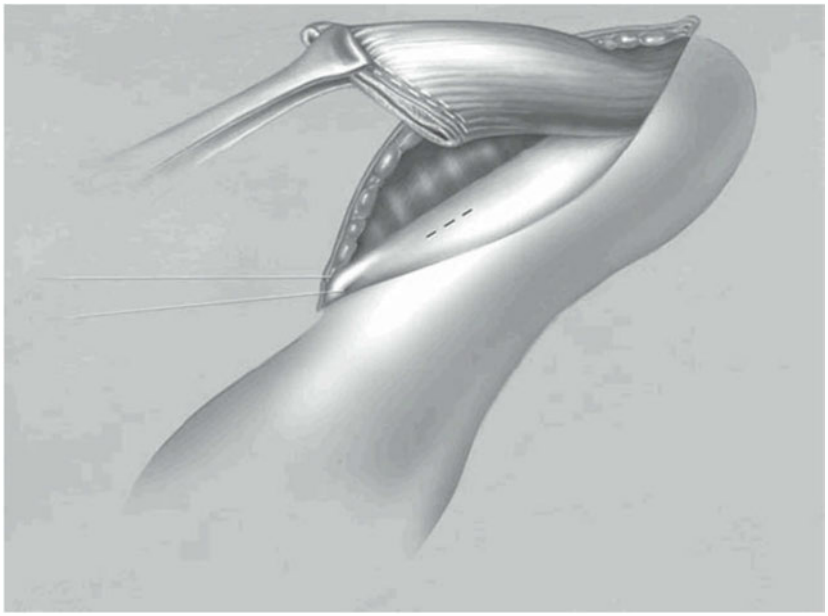


Fig. 17.19. With the gastric traction suture elevating the anterior gastric wall out of the depths of the incision to the level of the skin, the site of the anastomosis is selected by carefully estimating the length of the remaining esophagus and approximately where it will rest once the traction suture is removed and the stomach allowed to drop back into the wound. A 1.5-cm long vertical gastrotomy (*dotted line*) is made using the electrocautery cutting current. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

the upper end of the anterior gastrotomy and then through the posterior corner of the divided cervical esophagus 7 to 8 mm from the edge and emerging 2 to 3 mm from the mucosal edge (Fig. 17.21). Care must be taken to transfix the esophageal mucosa with this latter stitch. Retracting these two latter sutures inferiorly approximates the back wall of the cervical esophagus and the anterior gastric wall. As this traction is maintained, an Endo-GIA 45-3.5 stapler (Covidien, Mansfield, MA.) is inserted with the thinner anvil in the stomach and the thicker staple cartridge in the esophagus (Fig. 17.22). The stapler is gently rotated until it is pointed at the patient's right ear and the esophagus and anterior gastric wall are aligned (Fig. 17.22A, 17.22B). It is important that the gastric staple suture line continue its orientation toward the patient's right side, so that when the anastomotic stapler is fired, the gastric staple line is not transected, setting the stage for local ischemic necrosis.

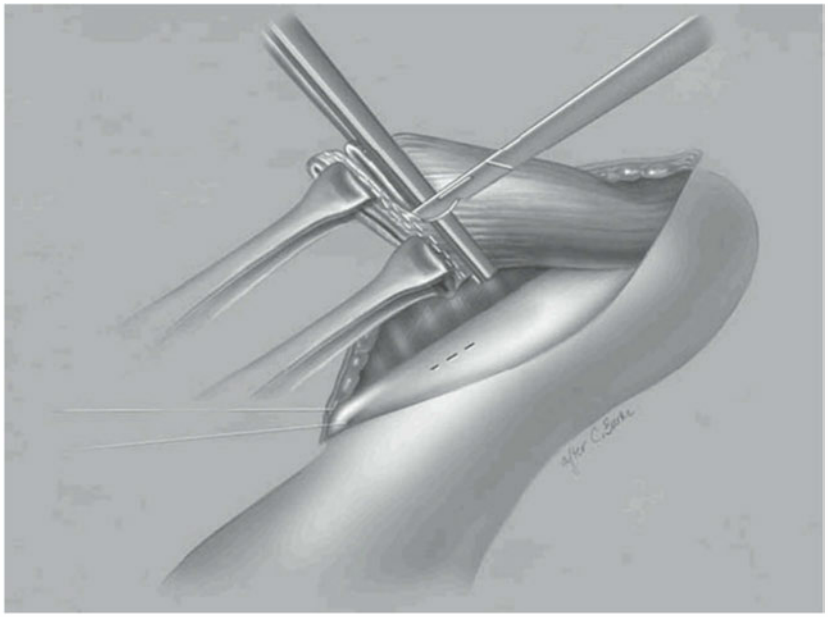


Fig. 17.20. The stapled end of the divided upper esophagus is grasped with two Allis clamps, a DeBakey forceps applied, and the stapled end amputated sharply just distal to the forceps in the same oblique plane in which the stapler was originally applied (anterior tip longer than the posterior). (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

The stapler is closed but not fired. Two "suspension sutures" of 4-0 absorbable suture, intended to minimize tension on the anastomosis, are placed on either side between the esophagus and the stomach (Fig. 17.23). The stapler is fired, thereby approximating the back wall of the esophagus and the anterior wall of the stomach and dividing the common wall between them. The stapler is removed from the field. A 16F nasogastric tube inserted by the anesthesiologist is guided by the surgeon's finger and advanced into the intrathoracic stomach. A 4-0 absorbable traction suture is placed at either corner of the opened gastrotomy and esophagotomy (Fig. 17.24). Anterior closure of the esophagotomy and gastrotomy is carried out with a running full-thickness 4-0 PDS suture on the inner layer (Fig. 17.25) and inverting interrupted 4-0 PDS sutures on the outer layer (Fig. 17.26). Either side of the anastomosis is marked with a hemoclip for future radiographic evaluation. The anterior gastric wall traction suture is removed and the stomach gently "seated" in the thoracic inlet. The cervical wound is irrigated and closed loosely over a 0.25-inch rubber drain placed near the anastomosis. No more than four or five interrupted 4-0 absorbable sutures are used to approximate the cervical muscle fascia. The skin edges are approximated with running 4-0 nylon sutures. All incisions and tube sites are dressed, and the nasogastric tube is irrigated to be certain that it is functioning properly. Prior to the patient being extubated, a portable chest radiograph obtained in the operating room verifies full expansion of the lungs, no unexpected hemothorax or pneumothorax, and proper placement of the chest tube(s).

RESULTS

The Thoracic Surgery Service at the University of Michigan Medical Center has now performed more than 3,000 THEs

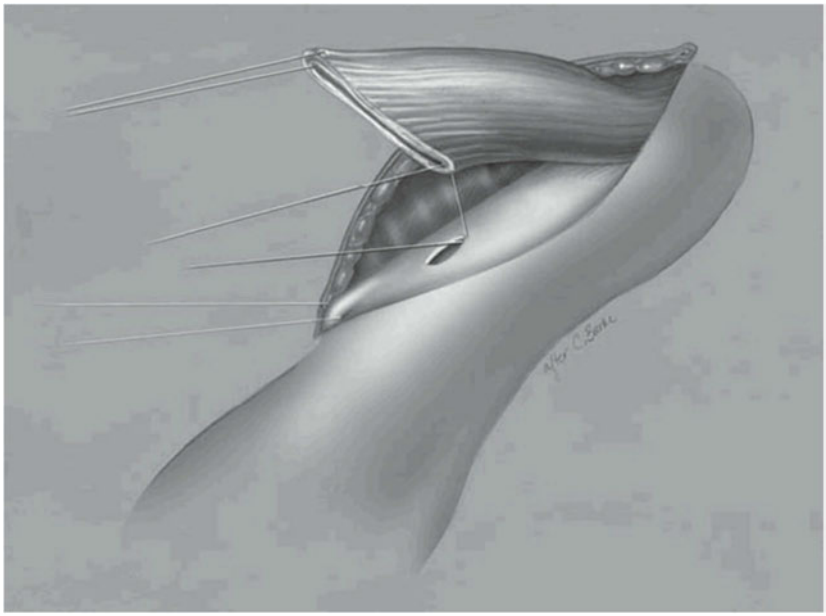


Fig. 17.21. The remaining two critical sutures needed to "set up" the anastomosis are placed. The first is a full-thickness stitch through the anterior corner of the divided esophagus. The second passes into the stomach and emerges 5 to 6 mm from the upper corner of the gastrotomy. It then passes into the posterior corner of the divided esophagus 7 to 8 mm from the divided edge and traverses the wall of the esophagus obliquely, emerging 2 to 3 mm from the mucosal edge. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

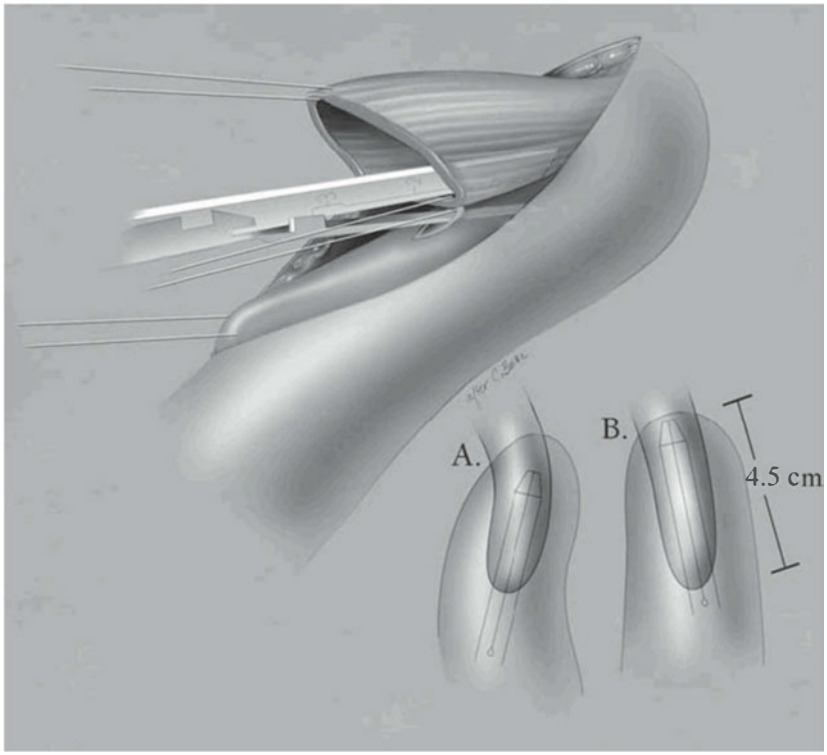


Fig. 17.22. Downward traction on the two latter sutures is maintained as an Endo-GIA 45-3.5 staple cartridge is inserted into both the esophagus and the stomach, the anvil into the stomach, and the staple-containing cartridge into the esophagus. (**A, B**) The cartridge is gently rotated outward so that its tip gradually comes to point toward the patient's right ear, aligning the posterior wall of the esophagus with the anterior wall of the stomach. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

since 1976. In our 2007 report on over 2,000 patients, approximately 75% of these operations were for carcinoma and 25% for benign disease. As the population has aged, more octogenarians and even nonagenarians have undergone THEs, 24% of our patients being 71 years of age or older, and 5% 80 years or older. The concern expressed after our early reports on the operation that the stomach would not reliably reach to the neck has proven to be unfounded. When the otherwise normal stomach is properly mobilized, the fundus will *always* reach to the neck for construction of a cervical esophagogastric anastomosis. In 97% of our THEs, esophageal replacement has been achieved using the stomach as described in this chapter. In those with a history of prior gastric ulcer disease, a gastric resection, caustic gastric injury, or tumor involvement of the stomach, a long-segment colonic interposition has been used to reestablish alimentary continuity after the THE.

There were four intraoperative deaths (<1%) from uncontrollable bleeding. Entry into one or both pleural cavities during a THE is common and occurred in 75% of our patients. It was diagnosed by inspection and palpation of the mediastinal pleura through the diaphragmatic hiatus and treated with a 28F chest tube(s) as indicated. Other intraoperative complications included splenic injury necessitating a splenectomy (3%), posterior membranous tracheal tear (<1%), and violation of pyloroduodenal integrity during performance of the pyloromyotomy (<2%). This latter complication was not managed by converting to a pyloroplasty but rather by closure with several interrupted 5-0 polypropylene sutures and buttressing the area with adjacent omentum. Early postoperative mediastinal bleeding requiring a return to the operating room for control occurred in <1%. Recurrent laryngeal nerve injury is avoidable if no metal retractor is placed against the tracheoesophageal groove during the cervical portions of the operation and occurred in 1% to 2% of our last 1,000 patients. Attesting to the merit of avoiding combined thoracic and abdominal incisions in patients undergoing an esophagectomy, and improved preoperative preparation by requiring regular use of an incentive inspirometer, walking, and cessation of cigarette smoking for several weeks before the operation, <2% have experienced sufficiently severe pneumonia or atelectasis to prolong their hospital stay. Chylothorax from thoracic duct injury occurred in <1% and was never fatal due to our very aggressive policy of early localization of the leak and thoracic duct ligation. Since adoption of the side-to-side stapled CEGA, our anastomotic

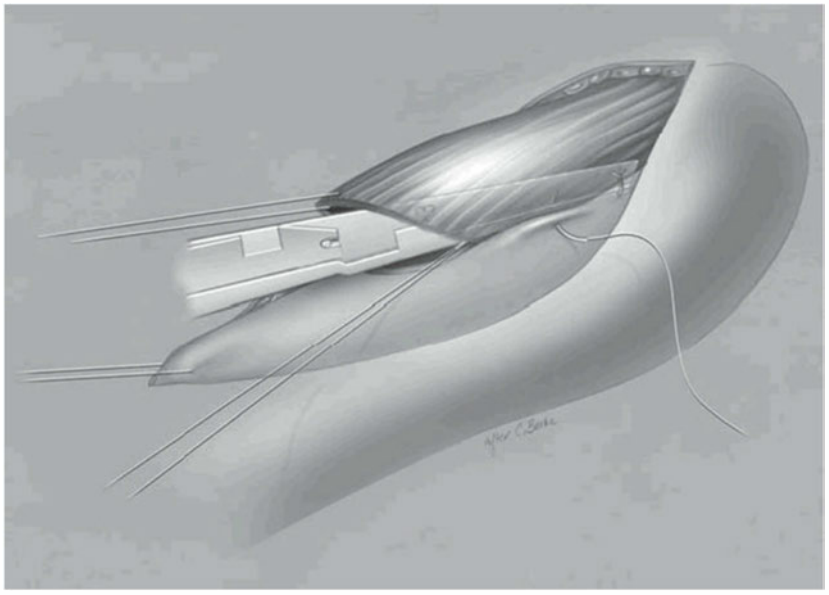


Fig. 17.23. Prior to firing the stapler, two interrupted 4-0 absorbable (Vicryl) sutures between the esophagus and the stomach are placed on either side adjacent to the stapler, essentially a "second layer" of sutures. These "suspension" sutures hold the stomach in the neck wound while taking tension off of the anastomosis. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

leak rate has fallen from 13% in our first 1,000 THEs to 9% in the second 1,000, to 3% in the author's hands. The hospital mortality for THE in our last 1,000 patients assessed was 1%, and 71% experienced no postoperative complications.

The functional results of esophageal substitution with stomach as assessed by the presence and degree of dysphagia, regurgitation, and postvagotomy dumping indicate that 76% had a good result or excellent result, 20% required an occasional anastomotic dilatation or antidiarrheal medication (fair result), and 4% required ongoing dilatations or other treatment (poor result). Overall patient satisfaction with the ability to eat has been >95%. The overall 5-year survival for our cancer patients was 29% and was significantly better for adenocarcinoma, now being diagnosed at earlier stages. In patients 75 years of age or younger with stage II and III esophageal cancer, neoadjuvant chemotherapy and radiation therapy followed by esophagectomy have become our standard treatment. Among our patients who have been complete responders (tumors rendered T0N0 after chemoradiation therapy), 5-year survival was 58% compared with 22% in all others with any residual tumor.

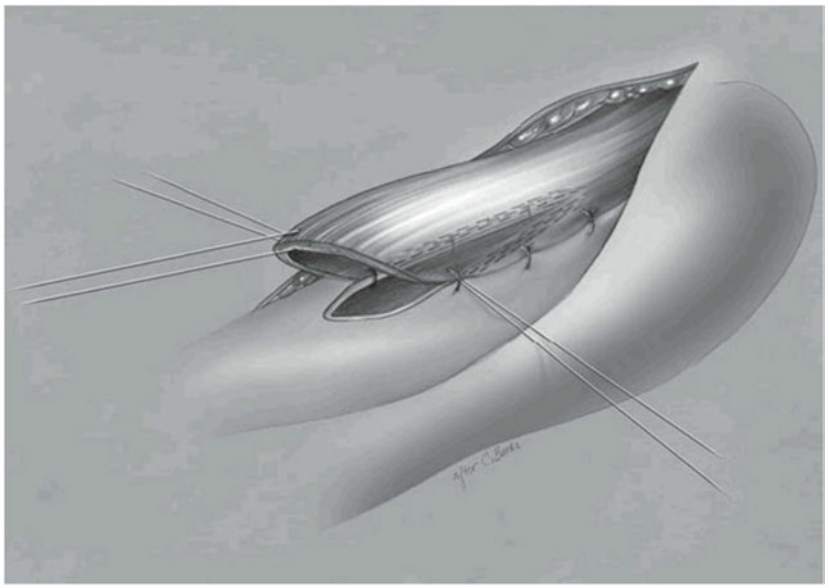


Fig. 17.24. Firing of the stapler divides the back wall of the esophagus and the anterior wall of the stomach creating a 4-cm long anastomosis. The stapler is removed, and two 4-0 absorbable (Vicryl) sutures are placed at either corner of the opened gastrotomy and esophagotomy. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

POSTOPERATIVE MANAGEMENT

With an average blood loss with THE of under 300 ml, perioperative blood transfusions are uncommon. The importance of careful preoperative assessment and physical conditioning cannot be overemphasized: complete abstinence from cigarette smoking for at least 3 weeks, regular use of an incentive inspirometer issued at the initial office consultation, and walking 3 miles a day. This regimen translates directly to fewer postoperative complications. Fewer than 4% of our THE patients ever require postoperative intensive care. They go directly to our general care thoracic surgery unit from the recovery room, start using their incentive inspirometer the afternoon of surgery, and are out of bed walking the next day. Extubation of the patient in the operating room is routine, and epidural anesthesia facilitates pulmonary hygiene and early ambulation. The nasogastric tube is typically removed on postoperative day 3, when sips of liquids are begun. Diet is progressed each day as tolerated from clear liquids, to full liquids, to a mechanical soft, and finally a soft diet. A postoperative barium swallow is obtained on day 7, and if this shows no anastomotic leak, satisfactory emptying of the intrathoracic stomach, and no obstruction to the flow of barium past the jejunostomy tube site, the patient is discharged. Tapered Maloney esophageal dilators should be part of the esophageal surgeon's armamentarium. Should a CEGA leak occur, it is managed by opening the neck wound at the bedside and initiating wound packing while providing nourishment through the jejunostomy feeding tube. 36, 40, and 46 French Maloney dilators are passed through the anastomosis within 1 week of opening the neck wound both to facilitate healing of the fistula by insuring free passage of swallowed esophageal contents into the stomach and to prevent the development of a late severe anastomotic stricture.

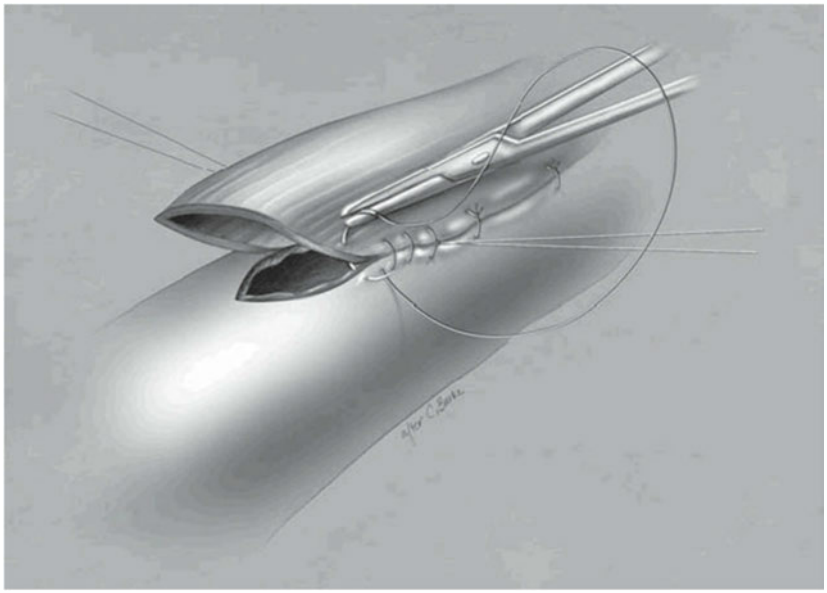


Fig. 17.25. A 16F nasogastric tube is introduced by the anesthesiologist and advanced across the anastomosis by the surgeon and positioned so that the 40 cm mark on the tube is aligned with the nostril. The first row of the closure of the gastrotomy and esophagotomy is achieved with a running full-thickness row of 4-0 PDS sutures. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

COMMENTARY

THE and a CEGA is now a well-established and efficient treatment for patients requiring an esophagectomy and esophageal reconstruction both for benign and malignant disease. In the 1970s, when the author first advocated wider use of this "less invasive" type of esophagectomy, considerable controversy was generated. Similarly, the current proponents of minimally invasive esophagectomy (MIE) must respond to those who criticize their new approach. The principles of a successful THE and CEGA, as discussed in this chapter, include (1) maximum preservation of submucosal gastric collateral circulation by preparation of as wide a gastric tube as is possible consistent with a reasonable oncologic margin, generally 4 to 5 cm distal to the esophagogastric junction; (2) progressive "straightening" and upward traction on the stomach as the stapler is applied along the curvature to achieve maximum length; (3) a generous Kocher maneuver to help achieve the maximum upward reach of the stomach that is possible; (4) a gastric drainage procedure (pyloromyotomy) to avoid postvagotomy pyloric outlet obstruction; (5) compulsive avoidance of any trauma to the gastric tip that will be used for the anastomosis; (6) direct and deliberate enlargement of the posterior mediastinal tunnel after extraction of the esophagus by the hand and forearm advanced through the hiatus to ensure that there are no remaining fibrous or neural bands to impede the transposition of the stomach through the posterior mediastinum; and (7) construction of a cervical esophagogastric anastomosis to avoid the consequences of mediastinitis associated with an intrathoracic esophagogastric anastomotic leak. In contrast, with an MIE "tubularization" of the stomach to 3 to 4 cm is advocated to facilitate passage of the gastric conduit through the posterior mediastinum that is not directly enlarged by the hand and forearm; torsion of the gastric conduit is a greater risk when the right side of the mediastinal pleura is so widely opened; a Kocher maneuver, difficult laparoscopically, is often omitted; the same applies to a gastric drainage procedure, for which a Botox injection or a "wait-and-see" posture are frequently advocated; "straightening" of the stomach during application of the stapler by the progressive application of traction to the gastric tip cannot be achieved to the same extent, and therefore there is potentially more tension on the anastomosis as a result; the tip of the gastric fundus is sutured to the end of the stapled stomach to draw the fundic end into the chest, thereby traumatizing the site of the pending anastomosis; and finally, because of the recognized increased CEGA leak rate after an MIE, proponents of the approach have gone "full circle," now advocating an intrathoracic stapled anastomosis and accepting the small but definite risk of mediastinitis associated with an anastomotic leak. Time will tell if the traditional THE and CEGA as described in this chapter is as "minimally invasive" as an esophagectomy should be.

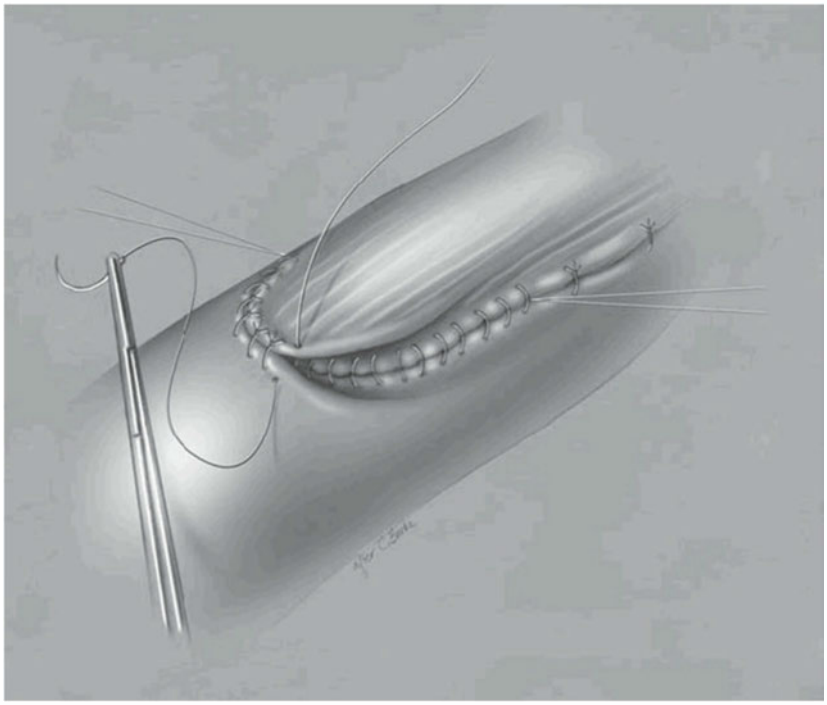


Fig. 17.26. The second layer of the closure is performed using interrupted 4-0 PDS sutures. (Reproduced from Orringer MB. Transhiatal esophagectomy without thoracotomy. Oper Tech Thorac Cardiovasc Surg 2005;10(1):63-83.)

SUGGESTED READINGS

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EDITOR'S COMMENTS

To have Mark Orringer contribute the chapter on transhiatal esophagectomy (THE) truly is a privilege and an honor since he was the one who resurrected this procedure from obscurity, championed it, and withstood the initial outcry from its detractors who refused to believe that the esophagus could or should be removed without a thoracotomy. Derisively labeled by critics as "blind esophagectomy" Orringer showed that the resection could be done under direct visualization and, while blunt, certainly was not blind. He has also demonstrated significantly lower morbidity and mortality from the operation as compared with a combined thoracic and abdominal approach and he has continued to modify and improve the operation over the years. When concern was raised about the higher incidence of leaks from the cervical anastomosis he developed the technique utilizing the linear stapler that he describes in the chapter that has significantly reduced the incidence of leak. He recognized early on an unacceptable incidence of recurrent laryngeal nerve injury and stopped the use of any metal retractor on the trachea recommending only a finger as the instrument for retraction. I am not sure there is anyone more associated with a particular procedure than Orringer is with THE. And patients, as well as surgeons, are that much the better for it.

In answer to those who tout minimally invasive esophagectomy (MIE), I have pointed out that we have been doing a MIE for years. It is called THE and it avoids a chest incision of any kind. That said, one has to marvel at those who have developed and refined the laparoscopic–thoracoscopic approach though the results have failed to demonstrate any clear superiority over THE. The recent developments in robotic esophagectomy potentially may offer some advantages based on the superior visualization offered as well as the unique range of motion of the articulating wrists that essentially mimic and exceed what can be done with the human hand. Further refinements of the robotic procedure clearly will be forthcoming.

In the meantime, it is hard to quibble with the results that can be obtained with THE and the fact that the operation is applicable to essentially all patients including the elderly, as Orringer points out. Long-term survival, yet to be definitively determined with MIE, is as good or better than that achieved with either an Ivor–Lewis approach or a three-hole procedure. THE may be the optimal approach for those patients following multiple failed antireflux procedures. As Orringer also has shown the functional results obtained following THE are superb in the majority of patients. He also has stressed optimizing patients for operation by insisting on a preoperative regimen that mandates smoking cessation for at least 3 weeks prior to operation, preoperative use and training with incentive spirometry, and exercising by walking 3 miles per day. Surgeons who insist on their patients following this regimen in addition to utilizing Orringer's technical hints, developed during the performance of over 3,000 of these operations, likely can reproduce the standard that Orringer has set for this operation. THE has more than withstood the test of time and should be a part of the armamentarium of any surgeon who wishes to venture into esophageal surgery.

LRK

18 Thoracic Approaches to Esophagectomy

Shanda H. Blackmon, Wayne Hofstetter, and Stephen G. Swisher

INTRODUCTION

Thoracic approaches to esophagectomy may be helpful for treating benign and malignant esophageal disease. Benign indications are less common but include undilatable, recurrent strictures; megaesophagus from achalasia, trauma, or ingestion of acids or alkali; and recurrent foregut surgery for benign processes (Table 18.1 and Fig. 18.1). More common indications include primary or salvage esophagectomy for localized esophageal cancer. Less common malignancies, such as sarcomatoid carcinoma, gastrointestinal stromal tumor, small-cell cancer, and locoregional involvement of other malignancies may also be addressed by transthoracic esophagectomy. Various thoracic approaches have been successfully used to address esophageal disorders (Table 18.2), the choice of which depends not only on surgeon preference but also on tumor location, body habitus, history of prior operations, patient condition, choice of esophageal substitute, and prior radiation therapy. A skilled esophageal surgeon does not limit the approach to a single modality but instead adopts the best approach to the individual patient. The primary goal of esophageal carcinoma treatment remains neoplasm resection, with relief of dysphagia. When selecting an approach, prior operations and conduit irradiation should be considered. In some situations such as in salvage esophagectomy, the use of a heavily irradiated conduit is unavoidable. In these cases, buttressing the esophageal anastomosis with a pedicled omental or muscle flap may be beneficial. Other challenges include previous extensive lung resections on the operated or contralateral side, which may require a left-sided thoracoabdominal approach instead of a traditional Ivor–Lewis right thoracic approach. High-risk esophageal resections can also be performed in a delayed or two-stage operation if there is concern regarding the performance of an anastomosis at the time of resection.

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HISTORY

The Ivor–Lewis esophagectomy is one of the only surgical procedures named after a surgeon by first and last name and was proposed in 1946 at the Royal College of Surgeons' Humanitarian Lecture. During this era, the standard approach to mid-thoracic esophageal tumors was a transhiatal blunt esophagectomy, cervical esophagostomy, gastrostomy, and skin tube reconstruction. The original operation was described as a two-stage approach that included a laparotomy and gastric mobilization (based on the right gastric and gastroepiploic arcades), followed 1 to 2 weeks later by a right thoracotomy, with esophageal resection and esophagogastric anastomosis in the chest. Exceeding the expectations of the era, the operation was successful 70% of the time. Today, this operation is usually completed in 1 day and in one stage.

RESECTION

When esophageal resection is performed for malignant disease, the entire diseased esophagus, including any dysplasia, metaplasia, or cancer, should be removed. Recurrent cancer after a low intrathoracic anastomosis can be due to incomplete resection or recurrent disease. Adenocarcinoma can occur at multiple levels, and the seventh edition of the American Joint Committee on Cancer (AJCC) staging manual (Fig. 18.2) includes proximal gastric cancer in the esophageal cancer staging system. The management of type I and II gastroesophageal junction cancers includes extending the resection to the thoracic esophagus in order to obtain an optimum the proximal margin. A gastrectomy for type III gastroesophageal junction cancers should be performed by an expert thoracic surgeon because of the potential need for simultaneous esophageal resection. Patients who are treated with definitive chemoradiation therapy and develop recurrent locoregional disease should be evaluated for a salvage esophagectomy if there is no evidence of distant disease. A transthoracic approach is favored in this situation because it allows complete resection of adjacent lymph node regions and direct placement of omental or muscle flaps to buttress these high-risk anastomoses. Palliative resection for dysphagia or bypass for tracheoesophageal fistula is seldom performed because of the availability of expandable covered stents. Patients who are at high risk for surgery can often be treated in a two-stage procedure or bypass to minimize the risks of a prolonged one-stage procedure.

PRINCIPLES OF TRANSTHORACIC ESOPHAGEAL RESECTIONS AND RECONSTRUCTION

Surgical Technique: Task List

Before the patient is brought into the operating room, the entire team should understand the operative plan. An equipment checklist will decrease intraoperative delays. All members of the team should be given a list of the planned surgical steps. The task list described below was developed to define each step and provide evidence for each decision that is made along the way.

Initial Operating Room Assessment

The patient is brought into the operating room, and after the induction of general anesthesia all appropriate lines and monitors are placed. Empiric antibiotics should be administered, along with a beta blocker, when indicated. Sequential compression devices should be applied to the lower extremities, with or without pharmacologic prophylaxis. A copy of the endoscopy reports and endoscopic ultrasound, computed tomography, and positron emission tomography images should be available in the room prior to beginning the procedure.

Table 18.1 Indications for Esophagectomy

Benign
Undilatable benign stricture
Megaesophagus from achalasia
Reoperative foregut surgery after fundoplication
Acid or alkali ingestion
Trauma
Benign tumors
Unrepairable leak or fistula
Gastric conduit necrosis
Malignant
Esophageal cancer
Primary esophagectomy
Salvage esophagectomy
Sarcomatoid cancer
Locoregional involvement of miscellaneous malignancies

Table 18.2 Transthoracic Resections

Ivor–Lewis esophagectomy = right thoracotomy + laparotomy
Hybrid esophagectomy
a. Laparoscopic abdominal approach + thoracotomy
b. Laparotomy + thoracoscopic approach
c. a or b + neck anastomosis
Minimally invasive esophagectomy
a. Laparoscopic abdominal approach + thoracoscopic approach
b. a + neck anastomosis
McKeown esophagectomy = three-field esophagectomy
Abdominal incision + thoracic incision + neck incision
Left-sided thoracoabdominal esophagectomy
Isolated segmental thoracic esophagectomy (rarely performed)



Fig. 18.1. Contrast esophagram of a sigmoid esophagus from life long achalasia.

Bronchoscopy

A bronchoscopy should be performed in all patients with tumors that occur in close proximity with the airway. The bronchoscope is advanced down to the level of the subsegmental bronchi, and the entire tracheobronchial tree is visualized for evidence of abnormalities. The absence of tracheal or left main bronchus involvement should be documented before proceeding with esophagectomy. The single-lumen endotracheal tube or laryngeal mask airway (LMA) is then removed, and a double-lumen endotracheal tube is positioned. A repeat bronchoscopy confirms that the left-sided endotracheal tube is in the left mainstem bronchus, and the tracheal opening on the right side allows for visualization of the carina and the entire right mainstem bronchus. A systems check should include monitoring the pressure in the bronchial cuff to ensure that it is not too high.

Endoscopy

Esophagoscopy should be performed to identify the proximal aspect of the tumor and any abnormal tissue near the tumor, including Barrett's metaplasia. As long as the tumor is distal to 30 cm from the incisors, an Ivor–Lewis esophagectomy is possible, with an intrathoracic anastomosis. No abnormalities including metaplasia should be visible proximal to the area of planned transaction. Once the endoscopy confirms normal squamous lining proximal to 30 cm from the incisors, the patient can be positioned supine, with the abdomen sterilely prepared and draped.

Abdominal Approach

A laparoscopic abdominal approach or open midline incisional approach can be used. The advantage of an initial laparoscopic approach is detection of potentially subclinical metastatic peritoneal or liver disease prior to performing a more invasive incision (Fig. 18.3).

Laparoscopic Approach

In the laparoscopic approach, five laparoscopic abdominal trocars are placed: a 12-mm trocar in the midline or just to the right of midline and two 5-mm trocars on either subcostal area (Fig. 18.4). The right subcostal trocar is used to place a Mediflex liver retractor to retract the liver anteriorly and laterally to the right. An energy source is typically placed through the 12-mm port. A 5- to 10-mm trocar should be placed for the camera in the medial subcostal area, approximately 4 to 5 cm inferior to the subcostal region. After all trocars have been placed safely, abdominal exploration should be performed. Checking for evidence of bowel injury or metastatic disease is necessary before proceeding with the esophagectomy. A minimally invasive approach should only be used if the exact same resection quality can be achieved as in an open approach.

Open Midline Incision

An open abdominal incisional approach can typically be performed through an upper midline laparotomy incision. Alternatively, but less commonly, a chevron or thoracoabdominal incision may be employed. The midline incision extends from the xiphoid down to just above the umbilicus. When open, a self-retaining type of retractor may be used to retract the liver and adjacent costal margin out of the way. A left thoracoabdominal incision is advantageous when the patient has undergone extensive foregut surgery and has excessive scar tissue in the left upper quadrant.

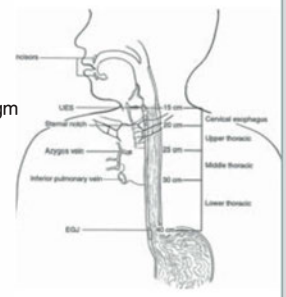
Mobilizing the Esophagus

An energy source is used to divide the pars flaccida or hepatogastric ligament. The left triangular ligament should be divided to allow the left lateral segment of the liver to be retracted. The right crus of the diaphragm should be exposed with an energy device, and the esophagus should be mobilized away from the right crus, stripping the peritoneal lining away to include all tissues. If the patient has a small hiatus, the crus can be divided approximately 1 to 2 cm to enlarge the hiatus for additional dissection. This allows for circumferential dissection around

Esophageal Cancer Staging

Primary Tumor (T)

Tx Primary tumor cannot be assessed
T0 No evidence of primary tumor
Tis High-grade dysplasia *
T1 Tumor invades lamina propria, muscularis mucosae, or submucosa
T1a Tumor invades lamina propria or muscularis mucosae
T1b Tumor invades submucosa
T2 Tumor invades muscularis propria
T3 Tumor invades adventitia
T4 Tumor invades adjacent structures
T4a Resectable tumor invading pleura, pericardium, or diaphragm
T4b Unresectable tumor invading other adjacent structures, such as aorta, vertebral body, trachea, etc.



Regional Lymph Nodes (N)

Nx Regional lymph nodes cannot be assessed
No No regional lymph node metastasis
N1 Regional lymph node metastases involving 1 to 2 nodes
N2 Regional lymph node metastases involving 3 to 6 nodes
N3 Regional lymph node metastases involving > = 7 nodes

Distant Metastasis (M)

M0 No distant metastasis (no pathologic M0; use clinical M to complete stage group)
M1 Distant metastasis

REQUIRED FOR STAGING:
Location – based on the position of the upper (proximal) edge of the tumor in the esophagus (Upper or middle—cancers above lower border of inferior pulmonary vein; Lower—below inferior pulmonary vein)
Grade

CLINICALLY SIGNIFICANT:
Distance to proximal edge of tumor from incisors
Distance to distal edge of tumor from incisors
Number of regional nodes with extracapsular tumor

Histologic Grade (G)

(AKA overall grade)

Grading system	*Grade*
2 grade system	Grade I or 1
3 grade system	Grade II or 2
4 grade system	Grade III or 3
Not available	Grade IV or 4

Additional Descriptors

Lymph-Vascular Invasion (LVI)
LVI (absent)/Not Identified
LVI Present/Identified
Not Applicable or Unknown/Indeterminate

Residual Tumor (R)
RX Presence of residual tumor cannot be assessed
R0 No residual tumor
R1 Microscopic residual tumor
R2 Macroscopic residual tumor

Adenocarcinoma, c, p

GROUP	T	N	M	Grade
0	Tis (HGD)	N0	M0	1, X
IA	T1	N0	M0	1-2, X
IB	T1	N0	M0	3
	T2	N0	M0	1-2, X
IIA	T2	N0	M0	3
IIB	T3	N0	M0	Any
	T1-2	N1	M0	Any
IIIA	T1-2	N2	M0	Any
	T3	N1	M0	Any
	T4a	N0	M0	Any
IIIB	T3	N2	M0	Any
IIIC	T4a	N1-2	M0	Any
	T4b	Any	M0	Any
	Any	N3	M0	Any
IV	Any	Any	M1	Any
Stage Unknown				

Squamous Cell Carcinoma , c, p*

GROUP	T	N	M	Grade	Tumor Location**
0	(HGD)Tis	N0	M0	1	Any
IA	T1	N0	M0	1, X	Any
IB	T1	N0	M0	2-3	Any
	T2-3	N0	M0	1, X	Lower, X
IIA	T2-3	N0	M0	1, X	Upper, middle
	T2-3	N0	M0	2-3	Lower, X
IIB	T2-3	N0	M0	2-3	Upper, middle
	T1-2	N1	M0	Any	Any
IIIA	T1-2	N2	M0	Any	Any
	T3	N1	M0	Any	Any
	T4a	N0	M0	Any	Any
IIIB	T3	N2	M0	Any	Any
IIIC	T4a	N1-2	M0	Any	Any
	T4b	Any	M0	Any	Any
	Any	N3	M0	Any	Any
IV	Any	Any	M1	Any	Any

Fig. 18.2. Seventh edition of the AJCC cancer staging system. (From Edge SB, Byrd DR, Compton CC, Fritz AG, Greene FL, Trotti A, eds. AJCC Cancer Staging Manual, 7th ed. Springer; 2009.)

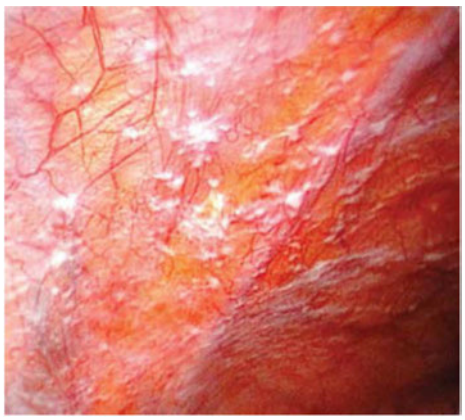


Fig. 18.3. Laparoscopic view of diffuse metastatic abdominal disease.

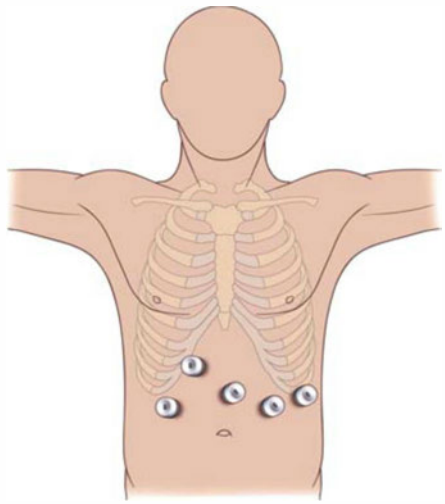


Fig. 18.4. One approach to laparoscopic trocar port placement.

the esophagus. The phrenoesophageal membrane should be taken down, and all tissue adjacent to the esophagus should be kept with the esophagus. The crural decussation should be dissected posteriorly, and the left crus should be identified from the right side of the abdomen (Fig. 18.5). The posterior esophageal window should be developed, and all tissues adjacent to the esophagus should be dissected away from the right and left crura. After dissecting in the cranial direction toward the mediastinum, the gastrohepatic ligament should be mobilized toward the lesser curve of the stomach. An appropriately sized hiatus for conduit passage is important as too large a hiatus could result in colon herniation and too tight a hiatus can result in ischemia of the conduit. It is helpful to dissect as far into the chest as is safe before closing the abdomen, as the inferior pulmonary ligament lymph nodes from both sides can be dissected from the abdominal approach. It is usually possible to reach up to the inferior pulmonary vein, depending on patient anatomy.

Mobilizing the Stomach

The right gastroepiploic artery should be identified and preserved as the gastrocolic omentum is divided and the lesser

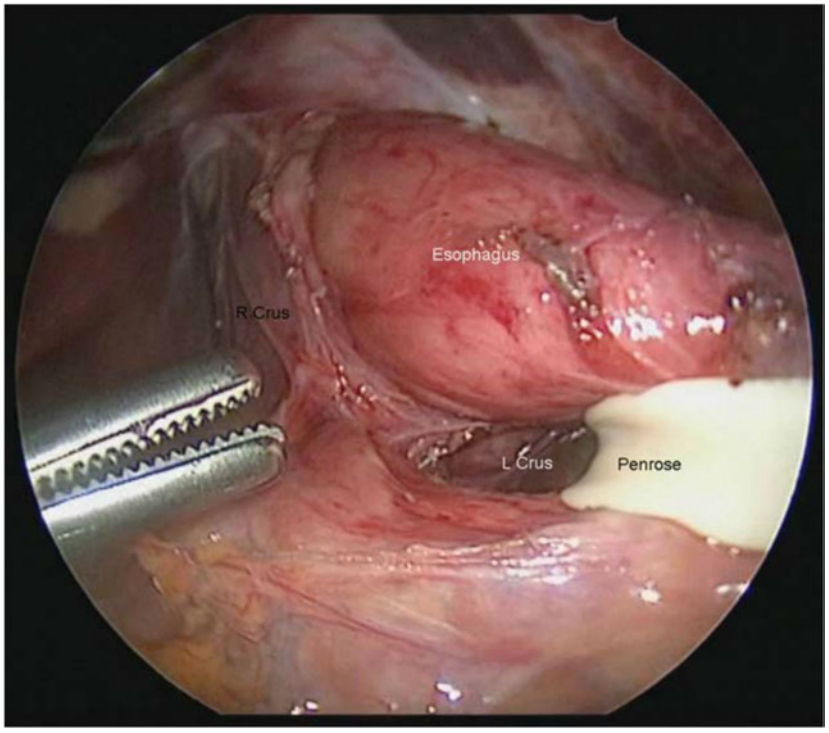


Fig. 18.5. Development of the posterior esophageal window.

sac is entered (see Fig. 18.6, "gastroepiploic artery preservation and omental dissection"). Instead of sparing a small segment of omentum, a larger omental flap can be created to be delivered into the chest to cover the anastomosis (Fig. 18.7). When manipulating the stomach, care should be taken to avoid injury when grasping and the right gastroepiploic artery should be avoided. The division should be completed along the entire greater curve of the stomach up to where the short gastric vessels are encountered. Exposure of the short gastric vessels is enhanced by grasping the posterior aspect of the stomach and rolling the tissue toward the liver and up away from the aorta. The short gastric vessels should be divided with an energy device up to the left crus of the diaphragm. The posterior attachments of the stomach to the pancreas should be divided. The cardia of the stomach should be dissected away from the left crus. The posterior esophageal window should be created, and a Penrose drain placed around the esophagogastric junction. This drain can later be recaptured and pulled into the chest to assist in mobilizing the intrathoracic esophagus. The posterior aspect of the stomach should be dissected away from the pancreas, preserving the right gastric artery. For benign disease, the modification does not typically require a complete esophagectomy or lymphadenectomy.



Fig. 18.6. Gastric mobilization with gastroepiploic artery preservation.

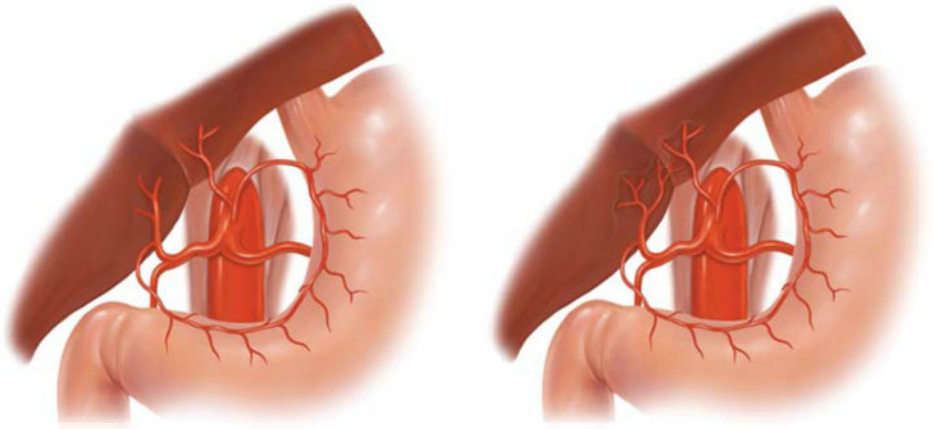


Fig. 18.7. Pedicled omental flap.

LEFT GASTRIC ARTERY DIVISION AND CELIAC LYMPH NODE DISSECTION

Once the posterior attachments of the stomach have been divided, the stomach is lifted anteriorly to allow for visualization of the left gastric artery pedicle. The left gastric artery pedicle should be divided either with individual ligation of the artery and vein or with a linear vascular stapler at the level of its origin from the celiac axis taking care to include the lymph node packet.

One should be aware for the presence of any aberrant celiac, gastric, or left hepatic artery anatomy (Fig. 18.8A, 18.8B), especially a replaced left hepatic artery coming completely from the left gastric artery. If this artery is an accessory artery and another left hepatic artery is present, it can typically be divided without consequence. However, if the entire left hepatic artery originates from a left gastric artery, a laparoscopic or open sonographic evaluation of flow dynamics can be used to determine portal flow and compensate when hepatic arterial flow has stopped. If blood flow to the liver is not diminished by more than 50%, the artery can still be divided. If the artery must be preserved, the left gastric artery should be ligated further distally, that is closer to the stomach. This will result in a lesser lymph node acquisition but will prevent hepatic necrosis. A low stapler division of this artery, as proximal as possible, allows for incorporation of all lymphatic tissue to be included with the specimen. Celiac lymph nodes can be dissected at this time and sent as separate pathologic specimens. Left gastric artery lymph nodes are then identified and dissected away from the specimen and identified for permanent specimen pathologic analysis. We recommend sending separately labeled packages to more accurately stage disease.



A B

Fig. 18.8. Aberrant celiac, left gastric, or hepatic artery anatomy. (**A**) Replaced left hepatic artery and (**B**) accessory left hepatic artery off the left gastric artery.

CREATING A TUBULARIZED STAPLED GASTRIC CONDUIT

Formation of the gastric conduit begins with dissecting the lesser omentum away from the stomach at the area of the incisura to allow for the placement of the linear stapler (see Fig. 18.9 for stapling and designing the gastric conduit). The stapler should be placed beyond the largest arcade from the right gastric artery, and one linear stapler load should be fired toward the greater curve of the stomach. Multiple firings of the linear stapler progressing toward the greater curvature are required to create the conduit. Starting the stapling at the incisura and proceeding toward the apex of the greater curvature allow for a slightly greater length of gastric conduit to be created.

A 2- to 3-cm-wide conduit is recommended as a narrower conduit is at higher risk of leakage, and a wide conduit may shorten the actual length provided. A retractor should be used from the far left subcostal area to straighten the superior aspect of the stomach, preventing twisting of the stomach or conduit while stapling. Once the staple line has been created, it can be oversewn.

The conduit can be completely separated from the specimen or left partially attached. If the conduit is completely separated, it will need to be sewn back to the specimen at the lesser curve side in two places to prevent rotation as it is delivered into the chest. A stapler can be passed into the lumen of the conduit to create a side-to-side or circular stapled anastomosis. If an omental pedicled flap is created, it should also be tacked to the edge of the conduit to facilitate delivery into the chest.

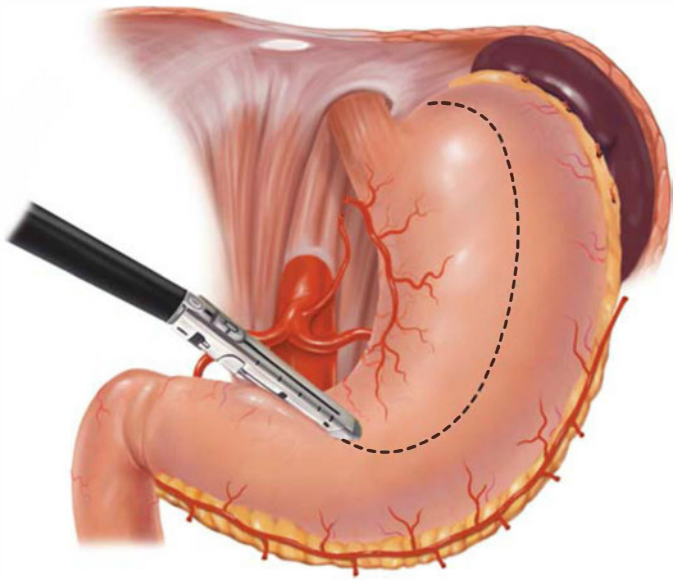


Fig. 18.9. Stapling and creating the tubularized gastric conduit.

MOBILIZATION OF THE DUODENUM

The duodenum should be mobilized with a Kocher maneuver to allow the pylorus to reach the level of the esophageal hiatus. The division of the omentum and the posterior antrum attachments should be completed to allow the conduit to be delivered easily into the chest. Any attachments to the pancreas should also be released here, without damaging the right gastric artery that comes off the proper hepatic artery, just distal to the gastroduodenal artery.

PYLORIC DRAINAGE

Three options are available for addressing the pylorus: pyloroplasty, pyloromyotomy, or paralytic injection (typically 100 to 200 IU botox). Some surgeons do not believe a pyloric drainage procedure is necessary; however, many reports in the literature indicate that delayed gastric emptying can result from inadequate drainage due to a restricting pylorus. It is impossible to identify which patients do not need a drainage procedure; because of this, we advocate performing a drainage procedure in all patients.

Laparoscopic or open pyloroplasty should be performed. The pyloroplasty is performed by placing an initial suture on the superior aspect of the pyloric muscle. Electrocautery using an L-hook allows the creation of a longitudinal incision that extends from the stomach into the duodenum, traversing and dividing the pyloric muscle along the way (Fig. 18.10A, 18.10B). It is also possible to perform a pyloromyotomy instead of a pyloroplasty; if this route is chosen, the pyloric muscle should be divided and the mucosal layer of the stomach and duodenum preserved. A long-handled electrocautery instrument can be used to achieve hemostasis during division of the pyloric muscle, and if a pyloroplasty is used, the incision should be completed all the way through the mucosa, exposing the internal lumen of the internal duodenum and stomach. Internal mucosa exposure is facilitated by using the previously placed stay suture on

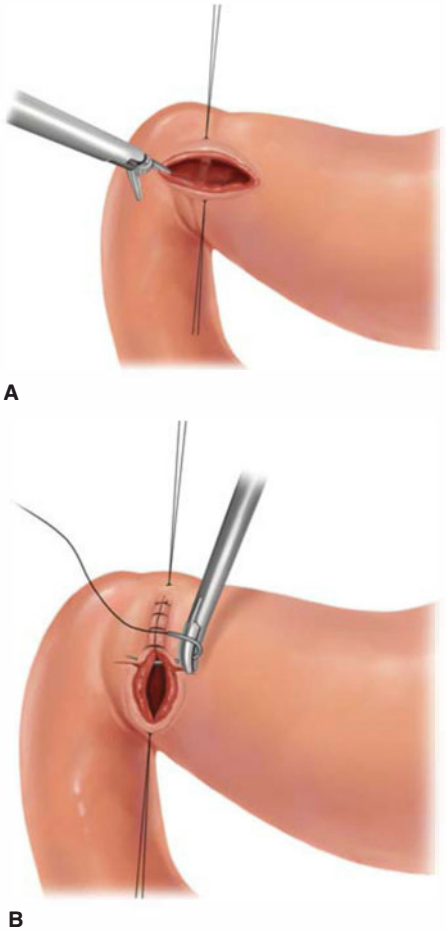


Fig. 18.10. Pyloroplasty technique. Longitudinal incision over the pyloric muscle (**A**) and transverse closure with interrupted sutures (**B**).

the superior aspect of the pyloric muscle. Interrupted sutures are used to close the longitudinal pyloroplasty transversely (in a Heineke-Mikulicz manner). Once the longitudinal pyloroplasty has been closed transversely with interrupted sutures, two large endoclips can be placed on the most superior suture to mark the area. The mobilization of the duodenum should be completed which allows the pylorus to reside at the level of the hiatus once the gastric conduit is mobilized into the chest. The pyloroplasty can be covered with a small piece of omentum.

FEEDING TUBE PLACEMENT

As an added safety measure in case of the need for postoperative enteral feeding, a tube jejunostomy should be performed. After the transverse colon has been retracted superiorly, the ligament of Treitz should be identified. Approximately 20 cm distal to this ligament, a segment of jejunum should be identified for jejunostomy tube placement. If the abdomen is open, a standard Witzel-type tube jejunostomy is performed.

Laparoscopic feeding tubes can be placed using a sutured technique, with or without an assist device, or a T-fastener technique. The bowel should be held in place against the anterior abdominal wall, and a subclavian needle should be used to traverse the left anterior abdominal wall, where the bowel meets the anterior abdominal wall, to enter the antimesenteric side of the proximal jejunum. A guide wire should be placed into the lumen of the jejunum and guided distally into the bowel. T-fasteners are placed in all four quadrants around the entry point of the guide wire. A saline injection into the bowel lumen often allows safer and easier passage of intraluminal devices and distends the bowel for easier placement of T-fasteners. As an alternative to T-fasteners, the bowel can be sutured to the anterior abdominal wall as well. The T-fasteners are tightened, and a dilator with an introducer sheath is placed over the guide wire through the left anterior abdominal wall. The dilator and guide wire are simultaneously removed, leaving the introducer sheath and the distal jejunum and allowing placement of the feeding jejunostomy tube through the introducer sheath. A grasper is used to guide the catheter into place. Ensure the back wall of the bowel is not incorporated into the sutures, as this may cause obstruction. The catheter is sewn to the anterior abdominal wall and secured on the outside. In the laparotomy approach, the feeding tube is simply sutured to the anterior abdominal wall directly.

THORACIC INCISIONS

Once the gastric conduit has been created, a thorough inspection should be made to assure hemostasis prior to closing the abdomen. The patient is then taken out of the supine position and positioned in the left lateral decubitus position. All pressure points should be padded. The arms should be placed on an arm board, with an axillary roll placed underneath the axilla. The table should be flexed to maximizing the opening of the intercostal spaces. A thoracoscopic or open approach can be used for the mobilization of the esophagus in the chest.

THORACOSCOPIC INCISIONS

In the thoracoscopic approach, four thoracoscopic trocars are typically placed (see Fig. 18.11 for chest trocar placement). A 3-cm utility port is placed in the posterior axillary line at the seventh intercostal space, just above the diaphragm, and a

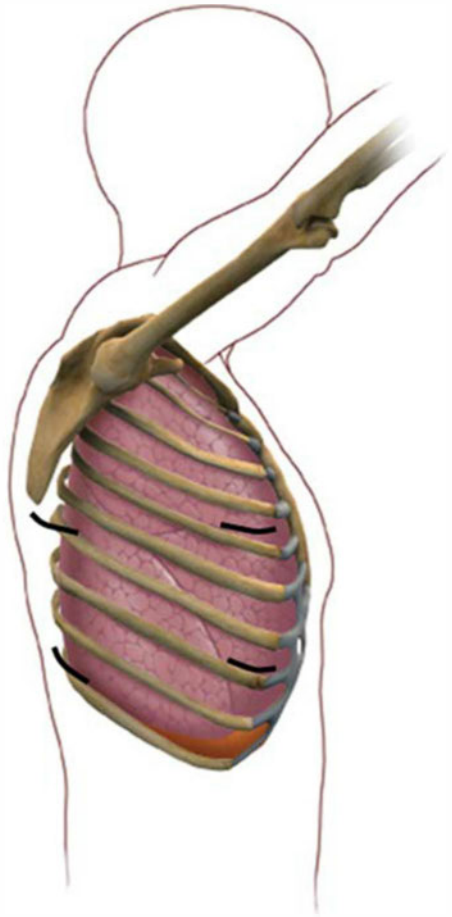


Fig. 18.11. Thoracic trocar placement, depicting four ports (two in the anterior axillary line and two in the posterior axillary line).

camera port is placed in the anterior axillary line in the eighth intercostal space. A second access port is placed in the right fifth intercostal space in the anterior axillary line. The fourth trocar maybe placed approximately 2 to 3 cm superior to the port in the posterior axillary line. A port protector may be used at the utility site to retract the soft tissue and facilitate suction, exposure, and instrument passage. One should not use suction in the chest without a vent, as this may cause negative-pressure pulmonary edema of the collapsed lung in addition to compromising visualization.

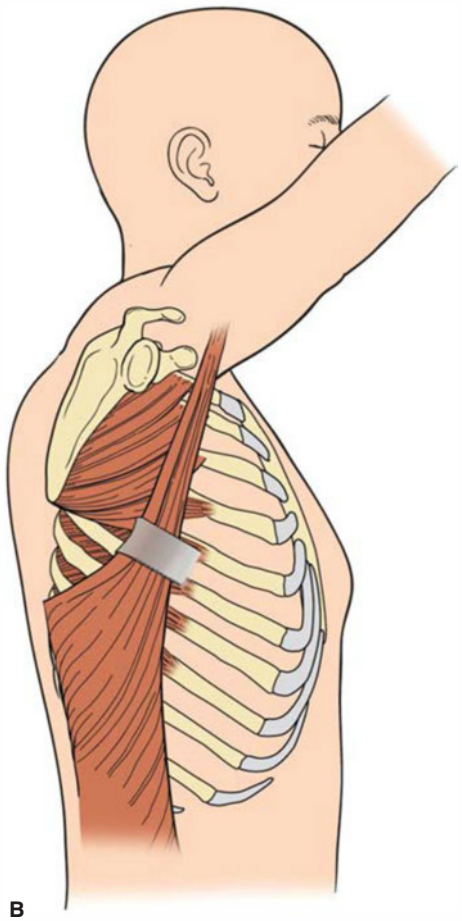
THORACOTOMY

Open thoracic incisions can be performed using a muscle-sparing or muscle-dividing approach. The advantage of a muscle-sparing approach is the preservation of the chest wall musculature that may be subsequently utilized, should a flap is required for use in the event of a postoperative intrathoracic complication (Fig. 18.12). The incision begins in the inframammary

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A



B

Fig. 18.12. Right posterolateral muscle-sparing thoracotomy incision, healed (**A**), and illustration of mobilization of the latissimus anterior as it is spared (**B**).

crease in the mid-axillary line, extending to the auscultatory triangle. This incision is typically 6 to 8 cm long in a muscle-sparing approach. If a longer incision is needed, the latissimus dorsi muscle can be divided to provide additional exposure; it is rare that the serratus anterior muscle needs to be divided. Once the ribs are exposed, the chest is entered through the fifth or sixth intercostal space. Resecting a small portion of the rib may facilitate entry into the chest, and some surgeons advocate removing a long segment of the fifth rib to provide optimal exposure. Epidural analgesia is recommended when an open incision is made, as pain control may be improved compared with a traditional patient-controlled intravenous analgesia.

LEFT THORACOABDOMINAL APPROACH

When choosing a left-sided approach for esophagectomy, surgeons must recognize the limitations of this approach. A neck anastomosis will be required if the esophageal resection is to be delivered above 25 to 30 cm. The costal arch will be divided, and if the edges of the margin are not reapproximated carefully, postoperative pain will result from mobility of the unfused ribs. A larger incision, spanning two body cavities, can result in more postoperative pain. The benefit of this approach is the superb exposure provided of the hiatal area, especially in reoperative cases, and the ability to perform a more thorough nodal dissection than that afforded by a transhiatal approach. Another advantage is the single position of the patient during the entire procedure, even when a neck incision is being used to construct the anastomosis.

THORACIC ESOPHAGEAL DISSECTION AND LYMPHADENECTOMY

The lung is retracted anteriorly to expose the mediastinum. An energy device is used to incise the pleura on either side of the esophagus, extending from the base of the diaphragm superiorly up to the level of the azygos vein. If an en bloc dissection is the goal, the azygos vein and thoracic duct are taken along with the specimen. The inferior pulmonary ligament is divided and all lymph nodes in this area are identified as an inferior pulmonary ligament lymph node package and sent for permanent histologic evaluation.

A vascular endoscopic stapler or suture ligation is used to divide the azygos vein for additional exposure of the esophagus. This stapler can be passed through a chest tube incision in the anterior axillary line in the seventh intercostal space. Creating this port, even in an open case, provides another passage for retraction instruments and better visualization or camera placement when teaching.

An energy source is used to dissect the esophagus circumferentially starting at the level of the diaphragm though much of this dissection may have been completed from the abdomen. While dissecting toward the abdomen, the esophagus is exposed inferiorly toward the diaphragm and into the abdomen from the chest to reveal the Penrose drain that was left in the abdomen encircling the distal esophagus at the gastroesophageal junction. This Penrose drain should be delivered into the chest and retracted superiorly, allowing identification of the attachments around the esophagus. By pulling up on the Penrose drain, the posterior aspect of the esophagus and all tissue adjacent to the esophagus can be safely divided with an energy device under direct visualization.

When an en bloc lymph node dissection is performed, the azygos vein is removed with the specimen and the thoracic duct is ligated (Fig. 18.13). En bloc esophagectomy includes the following structures: intrathoracic esophagus in continuity with the paraesophageal lymph nodes, thoracic duct, subcarinal (level 7) lymph nodes, low paratracheal lymph nodes, parahiatal lymph nodes, hepatic lymph nodes, celiac lymph nodes, left gastric lymph nodes, splenic artery with associated lymph nodes, spleen, and greater omentum. The resected specimen is bounded anteriorly by the pericardium and membranous trachea, laterally by the right and left pleural cavities, and posteriorly by the aorta and vertebral bodies. Some surgeons advocate routinely ligating the thoracic duct to limit the possibility of a postoperative chylothorax. When performing a modified en bloc dissection, as we prefer to do, the azygos vein, spleen, greater omentum, and thoracic duct are spared, but lymphatic tissue adjacent to the area is included. Lymph nodes should be labeled according to anatomic location to ensure that nodal counts are correct. Figure 18.14 depicts thoracic nodal staging for proper labeling. Dissecting the posterior aspect of the pericardium and the left atrium allows complete dissection around the esophagus, keeping all tissues adjacent to the esophagus with the specimen. The right inferior pulmonary ligament is incised all the way to the level of the right inferior pulmonary vein. The left inferior pulmonary ligament can be identified and lymph nodes dissected from this exposure as well. The esophagus should be mobilized to the level of the azygos vein, avoiding the posterior membranous portion of the trachea and the right and left mainstem bronchi while excising the subcarinal lymph nodes. No energy source is used during dissection of the superior aspect of the esophagus to prevent inadvertent cautery injury to the membranous portion of the trachea or mainstem bronchi.

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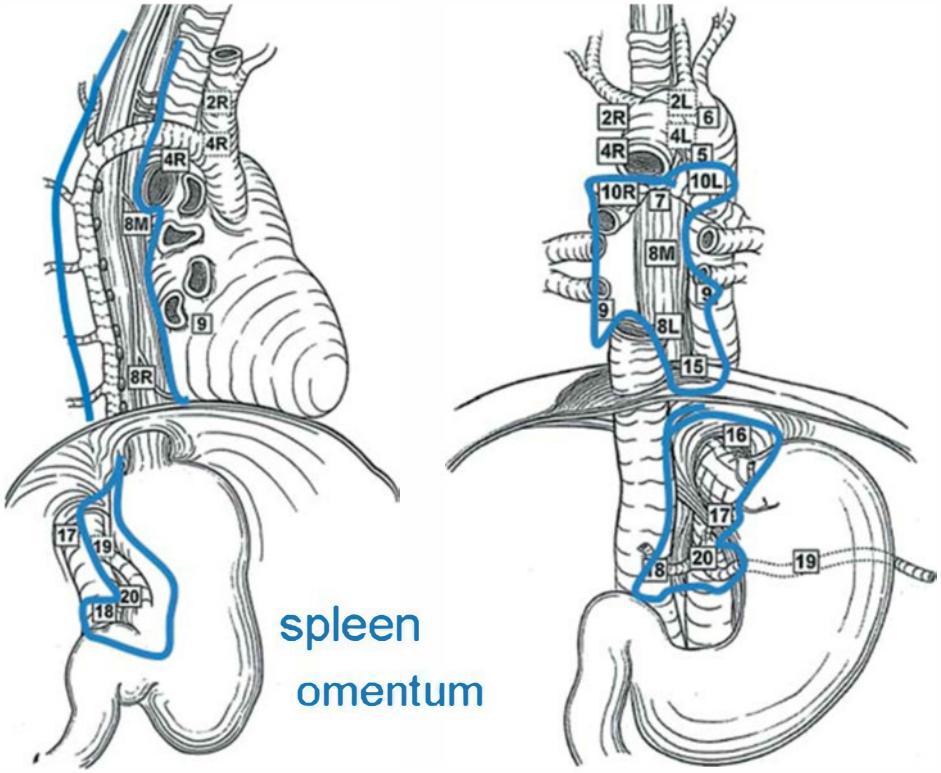


Fig. 18.13. En bloc lymphadenectomy for esophagectomy. (From Edge SB, Byrd DR, Compton CC, Fritz AG, Greene FL, Trotti A, eds. AJCC Cancer Staging Manual, 7th ed. Springer; 2009.)

Esophageal transection should be performed with an energy device or sharp instrument at or above the level of the azygos vein, depending on the tumor location, making sure to achieve a negative margin. It is possible to carry the resection to the apex of the thoracic inlet and create an anastomosis almost to the level as what would be created with a neck anastomosis, but creation of the anastomosis above the level of the azygos vein is more than adequate for distal esophageal lesions (Fig. 18.15). Proximal and distal margins should be clearly marked and frozen sections should be obtained if there is any question about either margin.

INTRATHORACIC ANASTOMOSIS

Sutures are placed on the terminal end of the esophagus to hold it without damaging or grasping the tissue that will become the anastomosis. Three types of intrathoracic anastomoses can be created to join the terminal end of the esophagus to the gastric conduit, including the more traditional hand-sewn, stapled side-to-side, and circular stapled. Each is discussed in this chapter in detail. The lack of a serosal layer and the segmental esophageal blood supply puts any anastomosis involving the esophagus at considerable risk underscoring the importance of precise technique and gentle handling of tissue. It is important to limit the dissection to preserve blood supply just proximal to the anastomosis as well as avoiding tension on sutures, which may cut through the wall. We published a propensity-matched analysis of 214 esophagectomies and found the thoracic-stapled anastomosis to be safe, effective, and less likely to cause postoperative stricture or dysphagia than

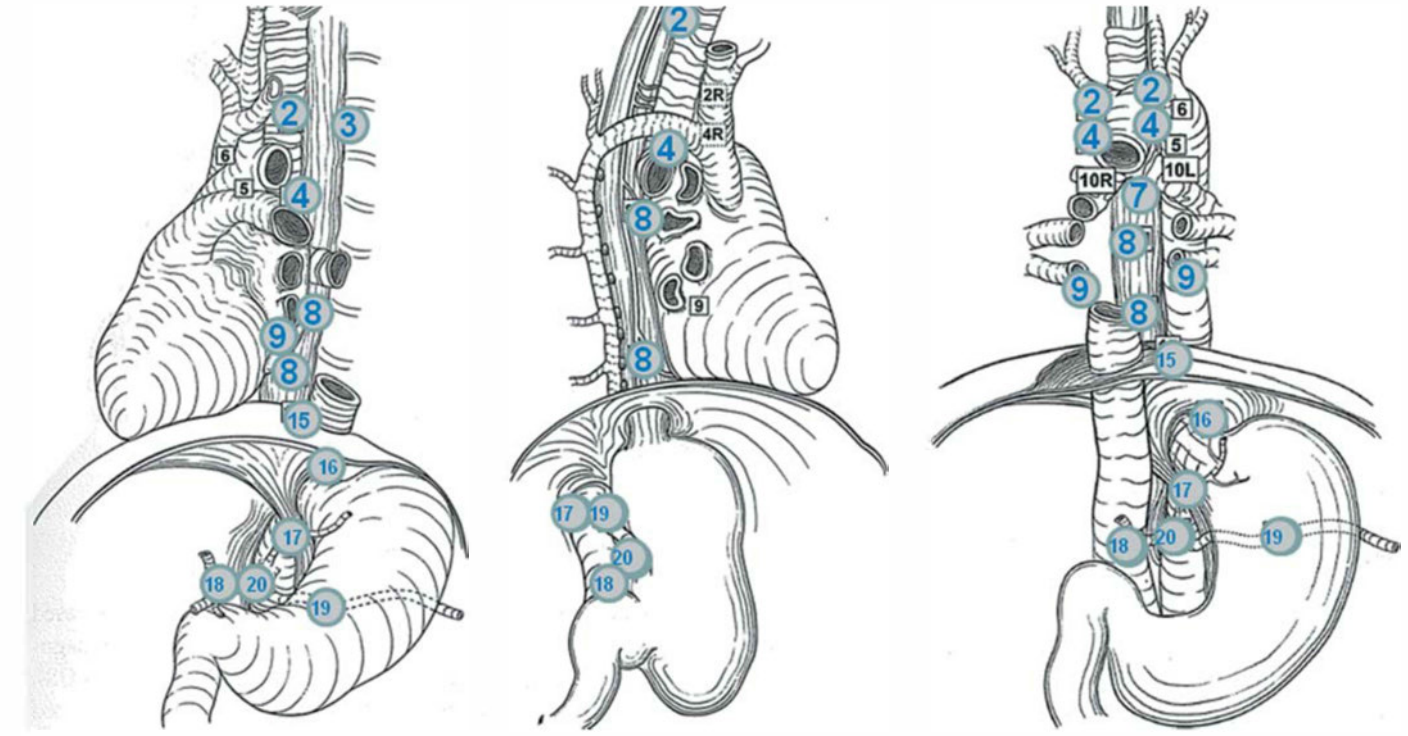
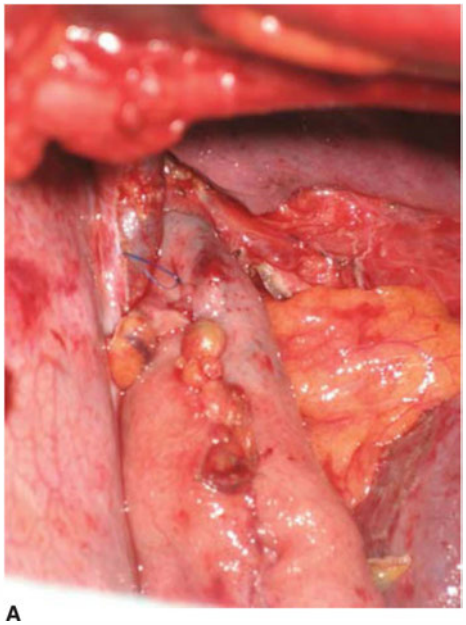
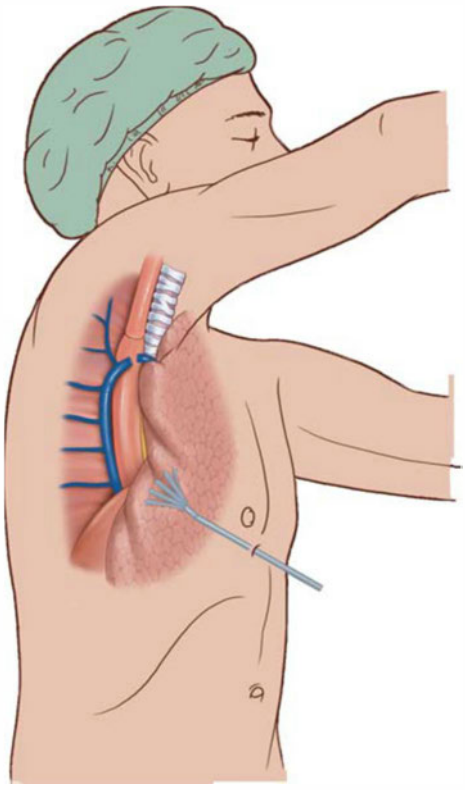


Fig. 18.14. Esophageal lymph node map. (From Edge SB, Byrd DR, Compton CC, Fritz AG, Greene FL, Trotti A, eds. AJCC Cancer Staging Manual, 7th ed. Springer; 2009.)

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A



B

Fig. 18.15. (**A, B**) Azygos vein in relation to the thoracic anastomosis.

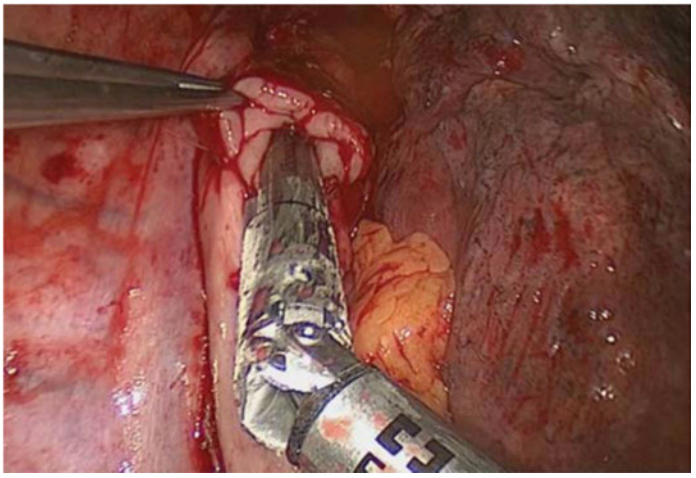


Fig. 18.16. Photograph of an intrathoracic stapled side-to-side anastomosis.

the hand-sewn anastomosis, with a similar leak rate (Blackmon et al., 2007).

The conduit should not be delivered into the chest until the surgeon is ready to create the anastomosis. If the tissue is brought into the chest too early, the conduit may be rotated or may obstruct the surgeon's view. If the conduit is sutured to the specimen, the sutures are simply cut, and the specimen is sent to confirm a negative margin. If a circular-stapled anastomosis is created, a note should be sent with the specimen noting that the final margin is the end esophageal portion of the doughnut from the stapler. It is important not to deliver too much of the gastric remnant into the chest because redundancy may cause problems later such as dysphagia and poor gastric emptying. Redundant conduit can be resected from the more ischemic tip of the conduit, or the redundant conduit can be tucked back into the abdomen and tacked into position after the anastomosis has been created (Fig. 18.16).

Circular-Stapled Anastomosis

A circular-stapled anastomosis can be created by placing a baseball or purse-string suture on the terminal end of the esophagus. Once the suture has run circumferentially around the terminal end of the esophagus passing through muscle and mucosa, the suture is tied tightly around the anvil to close the terminal end of the esophagus. A second purse-string suture is placed slightly higher up from the baseball stitch, circumferentially around the terminal end of the esophagus, leaving a more generous suture on the outside of the esophagus than on the inside; this suture should be tightened as well. If the esophagus does not appear to be tightly fixed around the anvil, an additional suture may be placed to facilitate gathering of the terminal end of the esophagus around the anvil. Any redundant tissue left around the anvil that may creep into the anastomosis will cause a leak. The stapler is placed through the open proximal end of the gastric conduit, and the spike from the EEA stapler is made to exit on a greater curve side of the gastric conduit, approximately 4 to 5 cm away from apex. The anvil is engaged in the stapler that is then fired. This allows the two structures to be joined, creating an end-to-side functional end-to-end anastomosis. Figure 18.17 shows a circular-stapled anastomosis. The gastric conduit staple line should not be any closer than 2 cm to the circular-stapled anastomosis as this will create a watershed area of poor perfusion that may result in a leak.

A significant incidence of stricture formation has been reported when using a size 25 circular-stapled anastomosis; however, some surgeons report no difference for size 25 and 28 staplers. Circular staplers smaller than size 25 should never

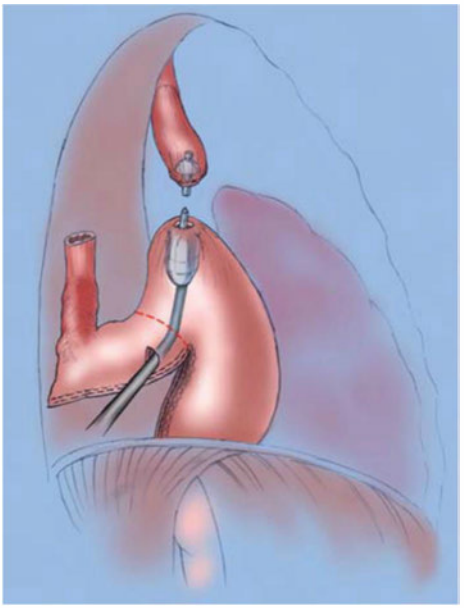


Fig. 18.17. Circular-stapled thoracic anastomosis. (From Blackmon SH, et al. Propensity-matched analysis of three techniques for intrathoracic esophagogastric anastomosis. Ann Thorac Surg 2007;83(5):1805-1813; discussion 1813.)

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be used. Tips for using the circular stapler are included at the end of this chapter.

Stapled Side-to-Side Anastomosis

Once the most proximal end of the gastric conduit has been removed and the gastric conduit is completely stapled with a linear stapler, the conduit can be positioned posterior to the terminal end of the esophagus and sutured into position to lessen tension on the anastomosis. The esophagus should be laid on top of the gastric conduit and again sutured on either side. A gastrotomy is placed on the greater curve side of the gastric conduit, approximately 4 to 5 cm inferior to the most proximal end of the gastric conduit. One fork of the linear stapler is placed in the gastrotomy and the other in the open end of the proximal esophagus and a stapled side-to-side but functional end-to-end anastomosis is created. The anterior rim (the hood) of the esophagus is hand-sewn in an interrupted manner over the opened gastrotomy (Fig.18.18).

Hand-Sewn Anastomosis

To create a hand-sewn anastomosis, the terminal end of the esophagus is placed on top of the gastric conduit, as described in Figure 18.19, for the beginning of a stapled side-to-side anastomosis. A gastrotomy is created on the greater curve side of the stomach to match the esophageal opening. Interrupted sutures are placed circumferentially, connecting the terminal end of the esophagus to the greater curve gastrotomy. A second suture layer is optional, but it may narrow the anastomosis. Interestingly, in a propensity-matched analysis, an intrathoracic hand-sewn anastomosis was associated with higher stricture and dysphagia rates but not higher leakage rates.

COMPLETING THE THORACIC PORTION

Once the anastomosis is completed and the gastric conduit tip has been removed, a nasogastric tube should be directed gently into the gastric conduit, with the tip left just above the diaphragm. Warm (cold irrigation may induce fibrillation of the heart) irrigation and insufflation of the conduit should be used to confirm that the anastomosis is completely intact. The conduit should be inspected to ensure that there is no abnormal kinking, redundancy, or malposition. The staple line should be facing the right side of the patient, and the greater curve should be facing toward the left. Figure 18.20 shows the proper orientation of the final conduit and position, with the anastomosis ideally above the level of the azygos vein. The gastric conduit may be tacked into position by suturing it to the mediastinal pleura in several areas. If an omental vascularized pedicled flap has been harvested, the flap can be positioned on top of the anastomosis or between the

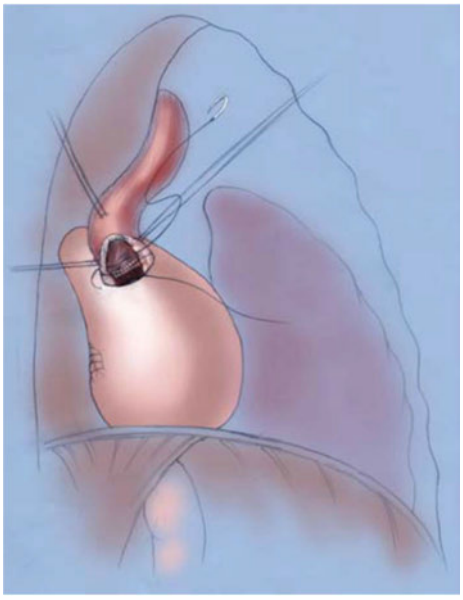


Fig. 18.19. Hand-sewn thoracic anastomosis. (From Blackmon SH, et al. Propensity-matched analysis of three techniques for intrathoracic esophagogastric anastomosis. Ann Thorac Surg 2007;83(5):1805-1813; discussion 1813.)

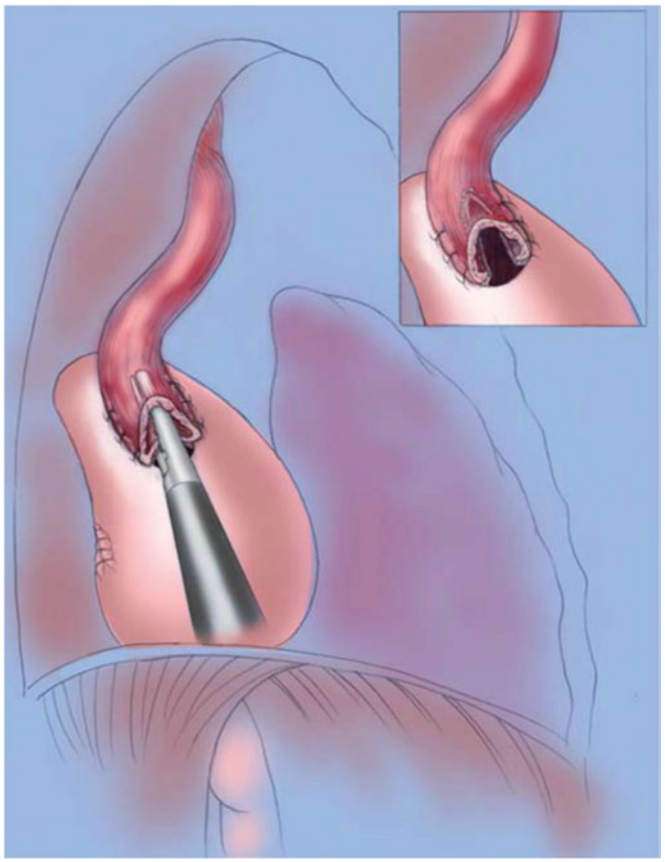


Fig. 18.18. Linear-stapled thoracic anastomosis. (From Blackmon SH, et al. Propensity-matched analysis of three techniques for intrathoracic esophagogastric anastomosis. Ann Thorac Surg 2007;83(5): 1805-1813; discussion 1813.)

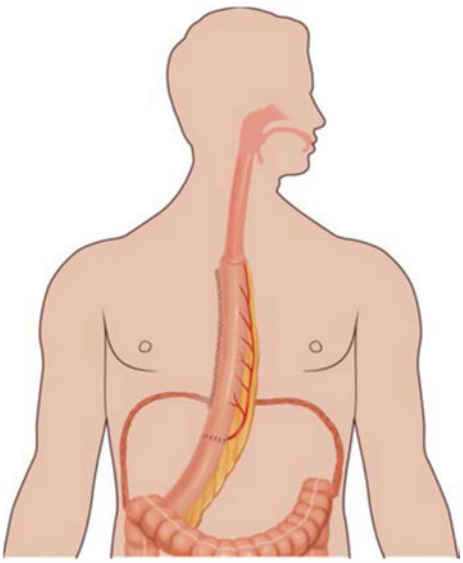


Fig. 18.20. Proper orientation of a thoracic conduit.

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trachea and the esophagus. It is important not to rotate the gastric conduit when positioning the omental flap. Any redundancy of the conduit should be reduced back into the abdomen. Once the conduit has been properly positioned, it should be tacked to the diaphragm at the level of the crura. The conduit should be straight and located directly behind the left atrium. A chest tube should be placed in the posteroapical position. We do not recommend placing drains directly adjacent to the anastomosis but in close proximity such that drainage of a leak is possible. The lung is inflated and the incisions are closed.

ADDITIONAL TIPS

1. It is important that the patient does not receive any pressors during the operation or in the immediate postoperative period because this may cause conduit ischemia and possible necrosis.
2. The nasogastric tube should be placed into position after the intrathoracic anastomosis has been completed and the terminal end of the gastric conduit has been resected to avoid accidentally including the tube in the staple line.
3. Repeat endoscopy can be performed to evaluate a questionable anastomosis.
4. The nasogastric tube should be secured into position by creating a bridal, placing two ET suction catheters through either nostrils, exiting from the mouth. Umbilical tape can be tied to the terminal end of the two ET suction catheters. Delivery of the ET suction catheters back outside the nostril allows for delivery of either end of the umbilical tape out of the nostril, with the loop passing around the septum. The umbilical tape is loosely tied away from the nostril and tightly tied around the nasogastric tube. This is not meant to hold the nasogastric tube in place but is only a reminder for the patient in the event that they pull on the nasogastric tube. Securing this too tightly may result in nasal septum necrosis.
5. When creating an esophagogastric anastomosis with the circular stapler, it is important to use a stapler with a lower staple height (3.5 mm) and as large a stapler as the esophagus can receive. Larger diameter staplers are better able to staple outside the tissue that is secured to the anvil and are less likely to develop stricture. Using too large a stapler can result in a tear in the esophagus, which can be problematic if the esophagus has been divided in the neck.
6. A laparoscopic anvil grasper will facilitate the creation of a circular-stapled anastomosis and will make it easier to grasp the anvil during manipulation.



POSTOPERATIVE MANAGEMENT

In the immediate postoperative period, the patient should be monitored in a specialized setting that is familiar with the care of esophagectomy patients. Many studies have revealed a reduction in postoperative morbidity and mortality rates when the surgery is performed in specialized centers with high resection volumes; the data indicate that survival depends on the volume of esophagectomies done in the institution as much as on the volume performed by the individual surgeon. Postoperative avoidance of intravenous pressors, generous administration of intravenous fluids, aggressive pulmonary toilet (except positive pressure oral ventilation or deep vigorous suctioning), maintaining good conduit decompression with a drainage tube, carefully securing all drainage tubes, early ambulation, and careful management of postoperative atrial fibrillation are just a few of the essential keys to good outcomes in these patients. For a list of common complications after thoracic esophagectomy, refer to Table 18.3. The mortality rate of this procedure is <0% to 4%.

POSTOPERATIVE INTRATHORACIC LEAK

The management of postoperative leaks and fistulae is the subject of another chapter, but Martin et al. clearly demonstrated the safety of early reintervention for thoracic leakage with subsequent reduction of postoperative mortality. Early reintervention and the use of muscle flaps have limited the morbidity and mortality rates that were previously associated with intrathoracic leaks. The role of off-label esophageal stenting for leaks and fistulae has yet to be established, but many centers have incorporated this newer and less invasive procedure into practice. The postoperative leakage rate from an intrathoracic anastomosis is higher in patients who have undergone neoadjuvant chemoradiotherapy. The published leak rates in these patients range from 6% to 30%. In patients who have not undergone neoadjuvant treatment, the leakage rate in high-volume centers is typically <10%. The rate is also expected to be higher in patients with a neck anastomosis rather than an intrathoracic anastomosis; this is likely due to relatively more ischemia at the tip of the gastric conduit when brought up into the neck.

Table 18.3 Thoracic Esophagectomy Complications

Leak (anastomosis, conduit, or pyloroplasty)
Bleeding
Pneumonia
Stricture
Dysphagia
Reflux
Atrial fibrillation
Dumping syndrome
Ischemic conduit

LONG-TERM MORBIDITY

Patients may develop a stricture after resection, which may require dilatation, revisional surgery, or stenting. Careful consideration of patient complaints will ensure increased satisfaction. Postoperative support groups allow surgeons and patients to learn from each other about postoperative outcomes and management. Dumping syndrome and delayed gastric emptying are well-known complications following esophagectomy. Reflux into the conduit is enhanced when the anastomosis is in a lower position in the chest. Grehlin or other appetite stimulants may lessen the characteristic postoperative weight loss. Postoperative respiratory insufficiency and atelectasis occur more commonly following thoracic esophagectomy, but the incidence of pneumonia is similar, regardless of the approach. Chylothorax, posterior membranous airway tears, and blood loss tend to occur with greater frequency following the transhiatal approach but this varies greatly depending on experience.

REVISIONAL SURGERY

Conduits that become redundant or herniate into the chest may require revisional surgery. Incomplete pyloric drainage, paraconduit herniation of abdominal contents, or constricting closure of the diaphragmatic hiatus may also require revision. Luketich et al. described revisional procedures for conduit redundancy, twisting, and incomplete conduit emptying, with excellent results; patients with less than optimal outcomes can benefit from surgical correction of the technical problems that may result from the initial surgical procedure.

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EDITOR'S COMMENTS

The surgeon who wishes to perform esophageal surgery must be facile with multiple approaches since "one size does not fit all." The emergence of minimally invasive techniques has changed the landscape but not all patients will be candidates for such an approach and every surgeon performing minimally invasive surgery should be intimately familiar with open techniques. There should never be any compromising of oncologic principles simply to carry out an operative procedure that the surgeon prefers. The operative procedure needs to be dictated by patient characteristics and specifically by the tumor. I have previously stated that the ultimate minimally invasive esophageal operation is the transhiatal approach where no thoracic incision is employed, not even small ones used for trocar placement, but this view has clearly been challenged by those who have developed significant expertise with the minimally invasive approaches. No matter the route chosen the bottom line remains the completeness of the resection and the optimal staging of the patient.

There are a number of small technical details that may have a major impact on the surgical procedure. Though an intrathoracic anastomosis is associated with a lower leak rate than a cervical anastomosis, the consequences of an intrathoracic leak can be far greater especially if the leak is inadequately drained or recognition is delayed. Most cervical anastomotic leaks may be managed simply by opening the skin to allow drainage and healing usually occurs without further complication. Stricture formation occurs with high frequency following an anastomotic leak and it probably is best to begin anastomotic dilatation expectantly. I am very concerned about the incidence of stricture following the use of the circular stapler, a device that I do not use. For years I employed a hand-sewn, single-layer anastomosis with excellent results but in recent years have employed the linear stapler technique popularized by Orringer and described by the authors in this chapter. The opened distal end of the esophageal remnant is placed adjacent to a small gastrotomy made in an appropriate location on the gastric remnant and a single suture is placed in full thickness at the midpoint of what will become the staple line. One blade of the linear stapler is placed in the gastrotomy and the other in the esophageal lumen and the stapler is fired leaving a "hood" of anterior esophagus that is sewn closed. This creates a very large anastomosis that has been associated with a lower leak rate than previously seen with other techniques. When working in the neck, care should be taken to avoid placing a retractor on the tracheoesophageal groove so as to not injure the recurrent laryngeal nerve.

When an intrathoracic anastomosis is used, a contrast swallow should be obtained on the fifth or sixth postoperative day to assure the absence of a leak. I do not routinely obtain a swallow with a cervical anastomosis but simply give the patient some grape juice and observe to see if any purple color is seen in the drain collection bulb. Clear liquids are started as soon as it is determined that no leak is present. Occasionally, a leak will present within a day or two of a seemingly negative study.

Gastric conduit necrosis fortunately is rarely seen but may be a devastating complication. When this occurs, the signs are usually present within the first 2 days following resection. The surgeon must be vigilant and maintain a high index of suspicion if the patient is febrile or hypotension occurs or if the patient simply does not "look right." There should be no hesitation to perform an endoscopy to visualize the gastric remnant if there is any concern regarding the viability of the conduit. If there is evidence of necrosis, the conduit

(*continued*)

EDITOR'S COMMENTS (*continued*)

must be taken down and resected with the patient being left with a cervical esophagostomy and facing the prospect of subsequent reconstitution of the gastrointestinal tract with a colon interposition, should they make it through the postoperative period.

There is ample evidence in the literature that supports the concept that esophagectomy should be done preferentially in high volume centers where the morbidity and mortality associated with the operation are significantly lower than that seen in centers that do few esophagectomies. This is especially true if the patient has undergone a course of neoadjuvant treatment with chemotherapy and radiation therapy. These are complex procedures where the technical performance of the operation as well as the postoperative care must be optimal.

LRK

19 Minimally Invasive Esophageal Procedures

Nikiforos Ballian, Matthew J. Schuchert, and James D. Luketich

INTRODUCTION

Minimally invasive surgical approaches are being increasingly implemented in the management of esophageal disease. The continued challenge for the minimally invasive surgeon is to maintain the fundamental principles of esophageal surgery that have been established with decades of open experience, while avoiding ill-advised shortcuts. The more complex the esophageal intervention, the more likely we are to see difficulty in reproducing the key technical steps of any given minimally invasive procedure. In most cases it is not the intent of the surgeon to perform a less than perfect operation, but one may be prodded by reports from other centers, as well as by patients and referring physicians seeking good results from small incisions. Therefore, as we evolve toward a less invasive culture and embrace new technologies, it is important that we continue to critically interpret and publish our results, while adopting a thoughtful attitude toward accomplishing the best outcomes possible.

This chapter summarizes the role of minimally invasive surgery in the treatment of esophageal cancer. Though laparoscopic fundoplication represents one of the cornerstones of minimally invasive esophageal surgery, this topic is discussed in detail in the chapter on antireflux procedures (Chapter 16). Similarly, other advanced minimally invasive esophageal techniques including esophageal staging techniques (Chapter 2), myotomy with fundoplication for esophageal motility disorders (Chapter 20), diverticulectomy (Chapter 22), palliative esophageal procedures (Chapter 21), and paraesophageal hernia repair with or without gastroplasty (Chapter 35) are covered in this text. The minimally invasive approach to esophagectomy highlighted in this chapter includes a detailed description of operative technique as well as a discussion of the published outcomes with comparison to the comparable open procedures. Complex procedures such as minimally invasive esophagectomy (MIE) are routinely being performed in only a limited number of specialized centers. Prospective studies ultimately will be required to objectively identify the benefits and limitations of these minimally invasive approaches.

BACKGROUND—ESOPHAGEAL CANCER

Esophageal cancer is the sixth leading cause of cancer-related death worldwide. In the United States, there will be an estimated 17,460 new cases of esophageal cancer diagnosed in 2012, with over 15,070 deaths (ninth leading cause of cancer-related deaths in the United States). The incidence of esophageal cancer increases steadily with age, with the median age of diagnosis being 68 years. Though squamous cell carcinoma has been demonstrated to be four to five times more common in African Americans, the incidence of adenocarcinoma has been increasing rapidly among Caucasians. In the mid-1990s, adenocarcinoma overtook squamous cell carcinoma as the most common type of esophageal cancer in the United States. Risk factors for the development of adenocarcinoma include chronic gastroesophageal reflux disease (GERD), obesity, and the presence of Barrett's esophagus. Esophageal adenocarcinoma is currently the solid malignancy with the most rapidly increasing incidence in the United States and the Western world, having increased by almost 600% since the 1970s. Overall survival rates, however, remain grim, ranging between 5% and 16%. The introduction of new molecular diagnostic tests in addition to new and emerging therapeutic techniques, and the development of more effective multimodality management strategies, provide hope for earlier identification and hopefully improved outcomes in the management of this lethal condition.

ENDOSCOPIC MANAGEMENT OF HIGH-GRADE DYSPLASIA AND SUPERFICIAL ESOPHAGEAL CANCER

High-grade dysplasia (HGD), referred to by some as carcinoma in situ, is characterized by malignant histologic findings that are confined to the esophageal epithelium and do not penetrate the basement membrane. HGD is believed to represent a late step in the dysplasia–carcinoma sequence, and can exist both as an isolated lesion and in association with invasive cancer. Although not all patients with HGD have, or will develop, cancer, malignant progression has been documented in 16% to 59% of patients followed for more than 5 years. Detection is notoriously difficult due to the lack of visible mucosal changes, and diagnosis is complicated by significant interobserver variability in the interpretation of the pathologic specimens. Esophagoscopy employing the Seattle protocol (four-quadrant jumbo biopsies taken every 1 cm) remains the standard method for evaluating segments of Barrett's esophagus. When HGD is suspected based upon pathologic evaluation, confirmation of the diagnosis by an independent pathologist is performed.

The principal management options for patients diagnosed with HGD include endoscopic surveillance, mucosal ablation, or esophagectomy. Intensive endoscopic surveillance can be performed every 3 to 6 months in the setting of HGD, with surgical or ablative therapy being employed once there is definitive evidence of adenocarcinoma. Although it is true that not all patients with HGD will go on to develop cancer, by the time biopsy specimens reveal HGD, between 40% and 60% patients will already have an invasive malignancy. Intensive surveillance strategies might avoid the risks associated with esophageal resection, but there has not been a clear demonstration that current intensive surveillance protocols are cost-effective or improve

mortality from esophageal cancer. Extensive work is underway evaluating methods to enhance the yield of surveillance protocols utilizing improved imaging modalities (e.g., fluorescence endoscopy, narrow-band imaging). However, at this time, none of these modalities has been clearly established as the standard of care in the management of HGD and early esophageal cancer.

Endoscopic ablation represents another option in the treatment of HGD or carcinoma in situ. The intent of this procedure is to eradicate the abnormal Barrett's epithelium to allow for subsequent restoration of normal squamous mucosa. Multiple techniques have been developed to selectively eradicate Barrett's mucosa and intramucosal carcinomas, including photodynamic therapy, endoscopic mucosal resection (EMR), and radiofrequency ablation (RFA).

Photodynamic Therapy—Technique

Photodynamic therapy (PDT) employs a photosensitizing drug (porfimer sodium) that is absorbed and retained at higher concentrations in neoplastic tissue when compared with normal tissue. Porfimer sodium (Axcan Scandipharm, Inc., Birmingham, AL) is injected at a dose of 2 mg/kg, and is allowed to circulate for 24 to 48 hours prior to treatment. Endoscopy is performed, and a PDT probe of appropriate length (1 cm, 2.5 cm, or 5 cm) is chosen and deployed through the endoscope under direct visual guidance. The probe is typically deployed in the stomach, pulled back to the designated site of treatment, and maintained in immediate proximity to the targeted lesion throughout the course of therapy. Focal stimulation of porfimer sodium in vivo with a laser light of appropriate wavelength (630 nm) induces a photochemical reaction that results in mucosal destruction. Total light dose ranges between 175 and 300 J/cm. After the initial application is completed, a second look is performed typically within 48 hours to permit assessment of treatment response, perform endoscopic mucosal debridement, and to apply a second dose of PDT, if needed. Patients are maintained on a liquid diet between PDT treatments advancing to a soft diet in 2 to 3 days following the completion of therapy.

Photodynamic Therapy—Results

The endoscopic ablation of Barrett's esophagus and superficial cancers with PDT has been evaluated in several prospective studies. Complete ablation of HGD or carcinoma in situ can be accomplished in 50% to 77% of cases, with complete squamous reepithelialization occurring in about half. Residual subsquamous intestinal metaplasia has been identified in 5% to 50% of patients after PDT, raising concerns regarding the possibility of continued malignant progression. The most common complication following PDT is stricture formation, occurring in one-third to half of the patients. Other complications following PDT include cutaneous photosensitivity, chest pain, nausea, pleural effusions, *Candida* esophagitis, atrial fibrillation, and odynophagia. Although more severe complications such as esophageal perforation and tracheoesophageal fistula formation have been reported, they are exceedingly rare (less than 1% in most large studies). Progression to cancer following PDT for HGD has been reported in 5% to13% of patients. Our experience at the University of Pittsburgh included 50 high-risk patients with either HGD or localized esophageal cancer. At a mean follow-up of 28.1 months, the intent-to-treat success rate was 38% in HGD and 30% in cases of focal carcinoma. Strictures occurred in 42% of patients.

Endoscopic Mucosal Resection—Technique

EMR is also a viable treatment option in patients with nodular or focal HGD or early esophageal cancer. This group of patients has a lower risk of lymph node metastasis, and thus local resection techniques can be performed with the intent to cure. Advantages of EMR also include the generation of a tissue specimen that provides information on stage and margins and lower morbidity and mortality when compared with esophageal resection.

At endoscopy, the targeted lesion must be carefully assessed to determine the extent of resection. Visual delineation of the borders of the lesion can be enhanced by the use of chromoendoscopy or narrow-band imaging. Endoscopic ultrasound (EUS) is also frequently utilized to assess lesion depth and the status of the periesophageal lymph nodes. The EMR technique involves raising the mucosal or submucosal target area by intramural saline and/or suction and then performing a snare resection (Fig. 19.1). This technique can be performed focally or circumferentially. The most common EMR technique utilized in the esophagus is the "cup and suction" or EMR-cap technique. Following injection of saline (with or without epinephrine or methylene blue) into the submucosa to elevate the targeted lesion, a plastic cap is then positioned over the targeted site and the mucosa is drawn into the cap by suction. A snare that is positioned around the cap is then closed around the base of the lesion, which is then removed by electrocautery (Fig. 19.2).

Another commonly employed EMR technique is band ligation. The targeted lesion is suctioned into a ligation cylinder (Duette System, Cook Medical, Winston-Salem, NC) and a band is deployed at the base, which results in the formation of a pseudopolyp. A snare is deployed just below the base of the pseudopolyp to excise the lesion. Other EMR techniques include the strip biopsy, which is most useful for raised or polypoid lesions. No submucosal injection is performed with this technique.

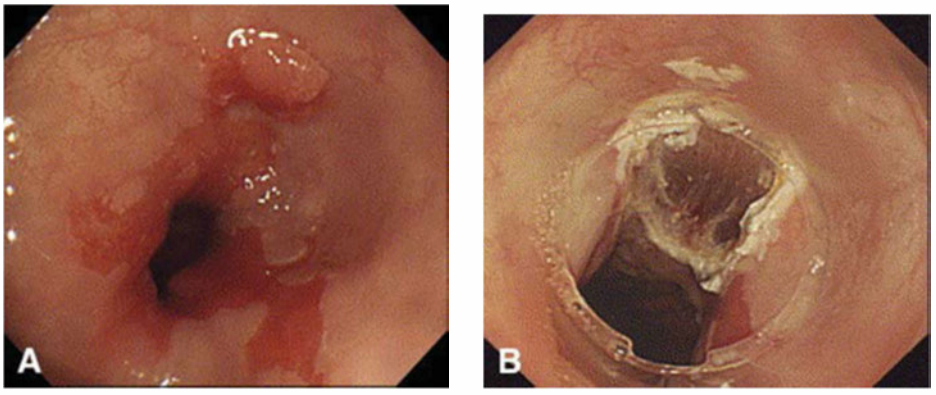


Fig. 19.1. Endoscopic mucosal resection. (**A**) Localized polypoid lesion involving the distal esophagus. (**B**) Appearance of mucosa following endoscopic mucosal resection. (Reprinted from Peters FP, et al. Endoscopic treatment of high-grade dysplasia and early stage cancer in Barrett's esophagus. Gastrointest Endosc 2005;61:506-514, Copyright (2005) with permission from Elsevier.)

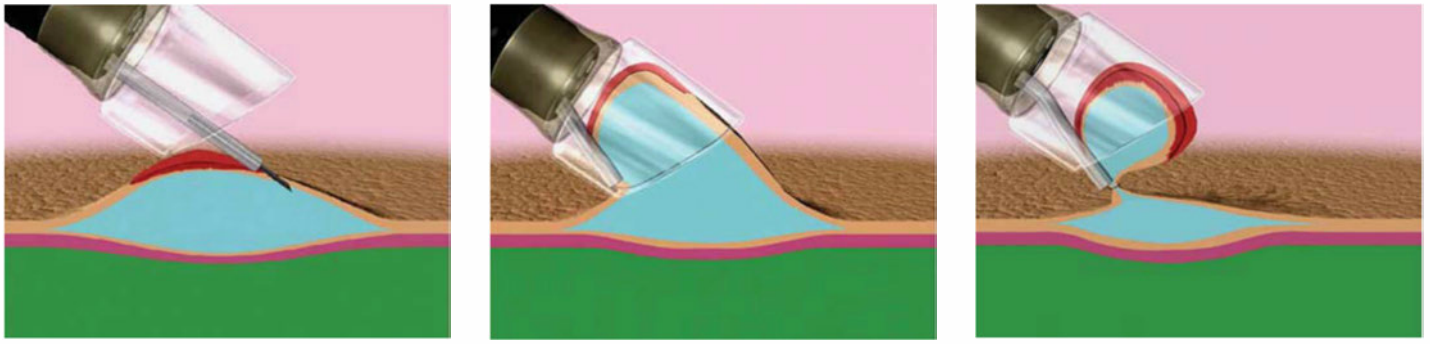


Fig. 19.2. Endoscopic mucosal resection. (**A**) Target lesion is raised by the submucosal injection of saline with epinephrine. (**B**) Suction is applied to the mucosa drawing the target lesion into the cap. (**C**) The snare is then deployed to remove the mucosal specimen. (Reprinted from Inoue H, et al. Endoscopic mucosal resection for early-stage gastrointestinal cancers. Best Pract Res Clin Gastroenterol 2005;19:871-887, Copyright (2005), with permission from Elsevier.)

A snare is introduced through the working channel and the targeted lesion is removed by electrocautery. A variation of the strip biopsy is the "inject, lift, and cut" technique, which requires a two-channel endoscope. Following submucosal injection, the lesion is lifted by forceps introduced through one channel, and removed by snare electrocautery performed via the second channel.

After resection, the specimen is oriented and submitted to pathology for assessment of pathologic T stage, surgical margins, and for the presence of lymphovascular invasion. For lesions with negative margins and no lymphovascular invasion the probability of a curative resection is >95%. For lesions where margin status remains in question, close endoscopic follow-up is warranted to rule out the possibility of local recurrence. If there is suspicion of lymphovascular invasion or nodal involvement, standard surgical resection with nodal dissection is recommended.

Endoscopic Mucosal Resection—Results

In properly selected patients, EMR can achieve complete remission in over 95% of cases, with corresponding estimated 5-year survival rates as high as 98%. When local recurrences are encountered, they can almost always be treated with by repeat EMR. EMR is now being performed with high success, both focally and circumferentially, for HGD and superficial cancers. Bleeding is the most common minor complication. Esophageal stricture is a late complication and is reported in up to 30% of cases, especially when EMR is performed circumferentially or when combined with other ablative modalities such as PDT. Risk of esophageal perforation is less than 1%.

In conclusion, EMR is a viable technique for the management of HGD and early esophageal cancer. It can be labor intensive and time consuming, especially for larger lesions. Large lesions typically require piecemeal excision, which can complicate assessment of surgical margins. Further randomized studies with long-term follow-up will be required to fully delineate the potential benefits of this approach.

Endoscopic Submucosal Dissection—Technique

Endoscopic submucosal dissection (ESD) represents an extension of the EMR technique and employs an electrocautery knife to dissect and excise larger (>2 cm) mucosal lesions in an en bloc fashion. Similar to EMR, the borders of the lesion are delineated by scoring the mucosa with electrocautery. A saline–epinephrine solution is then injected in the submucosal plane. Other injectable solutions (e.g., hyaluronic acid) can be utilized to maintain the lifting effect on the mucosa, which is then excised utilizing an endoscopic electrocautery knife. A peripheral incision is performed to initiate the dissection. Submucosal fibers are sequentially hooked and cut, and hemostasis is maintained by the use of electrocautery (Fig. 19.3). A transparent cap can be utilized on the tip of the endoscope to enhance exposure. A variety of endoscopic cutting knives has been utilized as reported in the literature, including the Needle Knife (Olympus, Tokyo, Japan), Hook Knife (Olympus, Tokyo, Japan), Flex Knife (Olympus, Tokyo, Japan), Triangle Tip Knife (Olympus, Tokyo, Japan), IT Knife 2 (Olympus, Tokyo, Japan), Duaknife (Olympus, Tokyo, Japan), Safe Knife (Pentax Corp., Tokyo, Japan), Flush Knife (Fujinon Corp, Omiya, Japan), and Hybrid Knife (Erbe Corp, Tübingen, Germany).

Endoscopic Submucosal Dissection—Results

Similar to EMR, successful en bloc resection achieving negative margins has been reported in 95% to 100% of cases with the ESD technique, with recurrence reported in <3% of cases in small series. Complications of ESD include bleeding, perforation, stricture, and pain. Bleeding is the most common complication (7% to 11%), and can be managed with electrocautery and applications of topical epinephrine. Specialized endograspers (Cograsper, Olympus, Tokyo, Japan) are particularly useful in applying electrocautery to areas of bleeding during ESD. Perforation rates, as reported, range from 0% to 1%. Stricture is most common in cases that involve resection of greater than one-half of the esophageal circumference, and is managed with periodic esophageal dilation.

A recent report from a multiinstitutional collaborative in Europe ($N = 27$ esophageal cases) demonstrated successful en bloc resection in only 77.1% of cases. Morbidity rates were 18.5%, including three cases of perforation (11.1%) and a postprocedural bleeding rate of 7.4%. These data indicate that significant additional experience must be gained with this technique to confirm the promising initial experience reported from Japan. Furthermore, long-term results and survival data are lacking for the ESD technique and this approach will need to be validated by prospective, randomized trials prior to more widely applied clinical application.

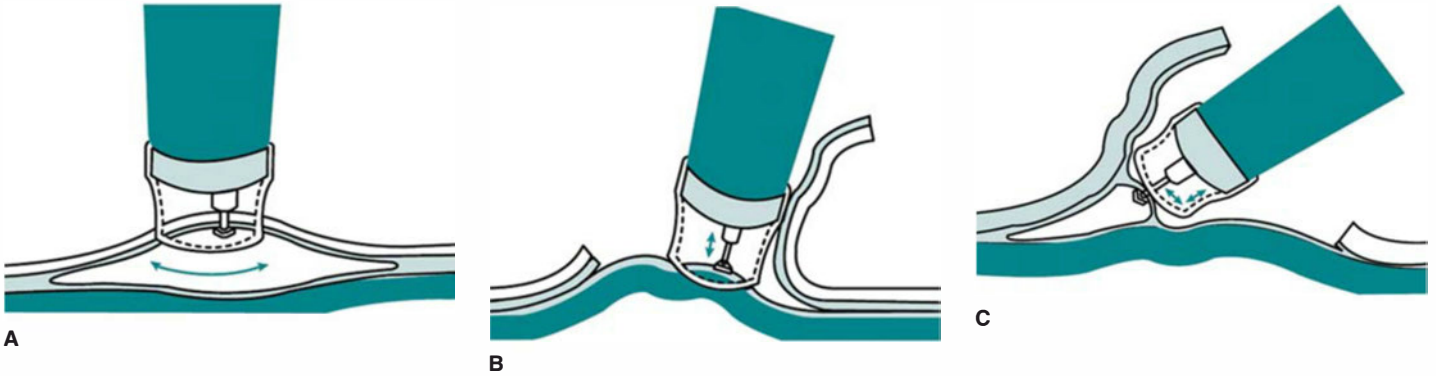


Fig. 19.3. Endoscopic submucosal dissection. (**A**) Marginal cutting utilizing a triangle tip cutting knife. (**B**) Hemostasis is maintained with spot surface coagulation. (**C**) Submucosal dissection is completed to permit complete removal of the targeted lesion. (Reprinted from Inoue H, et al. Endoscopic mucosal resection for early-stage gastrointestinal cancers. Best Pract Res Clin Gastroenterol 2005;19:871-887, Copyright (2005), with permission from Elsevier.)

Radiofrequency Ablation—Technique

The RFA system (Halo^{360} System, BARRX Medical Corporation, Inc., Sunnyvale, CA) is a balloon-based catheter ablation system that applies radiofrequency energy via a 3 cm bipolar microelectrode with 60 electrode rings. Radiofrequency energy is delivered through the balloon, generating heat which leads to coagulative necrosis and tissue destruction in the adjacent tissue. A sizing balloon is initially deployed to measure the inner diameter of the targeted segment. The appropriately sized balloon delivery system is then advanced into position over a wire and RFA is performed per a standardized protocol. The closely spaced circumferential electrodes deliver the radiofrequency energy pulse in less than 1 second, accomplishing a uniform distribution of energy density. The depth of ablation is typically limited to the muscularis mucosa. This is slightly more superficial than the EMR and ESD techniques, which excise tissue down to the level of the submucosa. As a result, RFA has been thought to produce a lower rate of stricture formation. The catheter can be repositioned to treat the entire length of Barrett's esophagus as required (Fig. 19.4). Noncircumferential probes (Halo^{90} System, BARRX Medical Corporation, Inc., Sunnyvale, CA) that can apply energy to focal islands of residual Barrett's tissue are available.

Radiofrequency Ablation—Results

In a study of 70 patients with Barrett's esophagus with no dysplasia treated with RFA (12-month follow-up), metaplasia was completely eradicated in 70% of patients, and partially eliminated in 25% of patients. Adverse events were rare and included chest discomfort and fever. No strictures or residual subsquamous metaplasia were noted. In a multicenter U.S. registry report on 142 patients with HGD treated with RFA, complete resolution was accomplished in 90.2% of patients with HGD, 80.4% with dysplasia, and 54.3% of patients with intestinal metaplasia.

In a multicenter, sham-controlled study evaluating the use of RFA ($N = 84$) for Barrett's esophagus with dysplasia, RFA produced complete eradication of dysplastic Barrett's esophagus in 77.4% of patients, as compared to 2.3% among controls ($P < 0.001$). Patients in the RFA group had a reduced rate of dysplastic progression (3.6% vs. 16.3%; $P = 0.03$) during follow-up and a lower incidence of cancer (1.2% vs. 9.3%; $P = 0.045$). Bleeding occurred in one patient and the esophageal stricture rate was 6.0%. On long-term follow-up (mean follow-up = 3 years), RFA was associated with eradication of dysplasia in >85% of patients, with no evidence of intestinal metaplasia

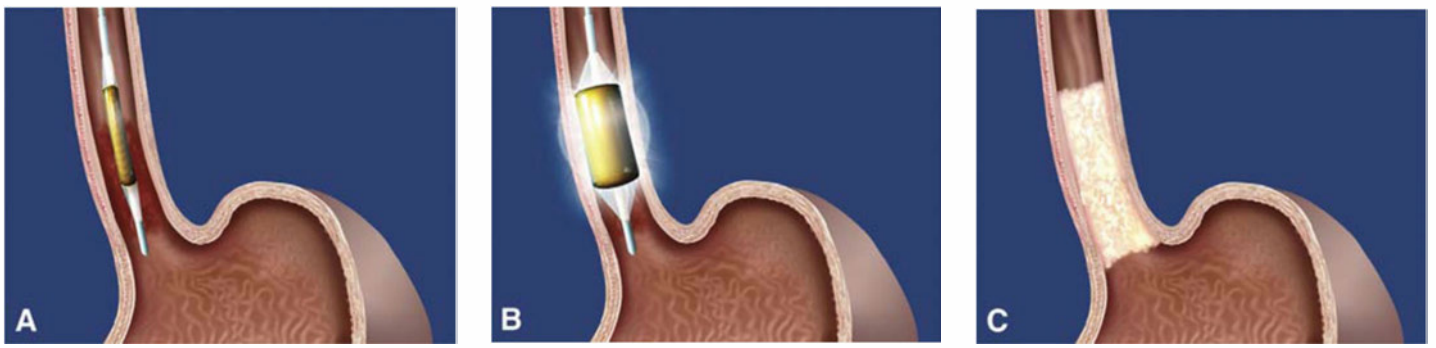


Fig. 19.4. Radiofrequency ablation of Barrett's esophagus. (**A**) Insertion of ablation probe over wire. (**B**) Balloon expansion and RFA treatment pulse. (**C**) Mucosal ablation of treated segment. (Reprinted from Sharma VK, et al. Balloon-based circumferential endoscopic frequency ablation of Barrett's esophagus: 1-year follow-up of 100 patients. Gastrointest Endosc 2007;65:506-514, Copyright (2007), with permission from Elsevier.)

in >75%. The rate of neoplastic progression was 1 in 73 patient-years, and the rate of progression to carcinoma was 1 in 181 patient-years.

There is increasing interest in the combination of modalities in the management of Barrett's esophagus with HGD and/or early cancer. The combination of EMR with RFA has been evaluated in a European multicenter study, in which 24 patients with HGD or carcinoma *in situ* were treated by EMR for visible mucosal abnormalities, followed by serial RFA to treat the underlying Barrett's esophagus. At a median follow-up of 22 months, neoplasia and intestinal metaplasia were eradicated in 95% and 88% of patients, respectively. The combination of RFA followed by antireflux surgery has also been evaluated in a series of 14 patients, and was shown to be performed safely with no significant difficulties in subsequent esophageal dissection. In this study, complete eradication of Barrett's metaplasia was accomplished in 50% of patients. No progression of Barrett's esophagus was observed.

These data suggest that RFA is a safe and durable procedure for the management of patients with Barrett's esophagus with dysplasia. RFA can be performed with low morbidity and can be combined safely with other modalities as required to augment the treatment response.

Endoscopic Ablation—Alternative Techniques

A variety of other ablative approaches has been utilized in the management of Barrett's esophagus with HGD or superficial cancer. Nd:YAG laser ablation, argon plasma coagulation (APC), multipolar electrocoagulation (MPEC), and cryotherapy are all capable of ablating neoplastic and metaplastic mucosa, followed by repopulation with normal squamous epithelium. YAG laser, APC, and MPEC can eradicate Barrett's mucosa with success rates ranging from 25% to 88% in reported series, but can be associated with a higher risk of perforation and stricture formation. Cryotherapy is a simple technique that can ablate Barrett's mucosa via a low-pressure liquid nitrogen spray. This technique has been shown to downgrade HGD or eliminate cancer in 80% of patients with intramucosal carcinoma, and 68% with HGD at 1-year follow-up. Further experience with these alternative modalities is required, which are currently best viewed as adjuncts in the multimodality approach to patients with HGD and superficial esophageal cancer.

Endoscopic Therapy—Conclusions

In a position statement by the American Gastroenterological Association, eradication of Barrett's esophagus with HGD by RFA, EMR, or PDT is recommended in lieu of intensive endoscopic surveillance. EMR (or ESD) is specifically recommended in cases where there is a visible mucosal irregularity in an effort to most accurately determine the T stage of the neoplastic lesion. Given the frequent multifocality of Barrett's dysplasia following EMR, a concomitant ablative procedure (e.g., RFA) is recommended as required to ensure complete eradication of disease.

MINIMALLY INVASIVE ESOPHAGECTOMY

MIE is a complex and technically challenging procedure that has evolved over the past two decades. Various techniques have been developed over time, including hybrid approaches that combine open surgery with either thoracoscopy or laparoscopy, as well as totally MIE techniques (transhiatal, three-hole, Ivor–Lewis).

Variants of MIE include "thoracoscopic esophagectomy," which utilizes thoracoscopy to achieve esophageal mobilization in combination with standard laparotomy and a cervical incision for the completion of the esophagectomy. Other approaches include "lap-assisted esophagectomy," where laparoscopy is utilized for mobilization and preparation of the gastric tube, and "hand-assisted laparoscopic transhiatal esophagectomy," which introduces a hand port to assist with mediastinal mobilization during transhiatal esophagectomy (THE). Also, robotic thoracoscopy/laparotomy approaches have been developed utilizing the da Vinci operating robot.

Totally MIE (total MIE) techniques have been developed utilizing thoracoscopy and/or laparoscopy exclusively. The most popular totally minimally invasive approaches to esophagectomy are the Ivor–Lewis MIE and the three-hole (McKeown) MIE. In some centers, a complete thoracoscopic/laparoscopic approach is not feasible or preferred, and the use of hand-assisted techniques in the approach to esophagectomy has been explored. While offering some potential advantage in cases where organ integrity is important, a hand inserted near the esophageal hiatus or into the mediastinum frequently obscures the view and is generally unnecessary during MIE. The optimal minimally invasive approach remains to be determined but surgeon experience and preference must be considered as major factors. Until further randomized data become available, the chosen approach should be based on tumor and patient characteristics, as well as the surgeon's personal preference and expertise.

The first totally laparoscopic esophagectomy at the University of Pittsburgh was performed in 1996. This initial approach has evolved into one combining thoracoscopy and laparoscopy for several reasons. Esophageal mobilization can be tedious and cumbersome via a completely laparoscopic approach. In addition, visualization of paraesophageal structures (such as the inferior pulmonary vein and the main stem bronchi) and the performance of mediastinal lymph node dissection can be very limited when employing an exclusively transabdominal approach. In the first 77 patients at the University of Pittsburgh undergoing MIE, a combined thoracoscopic/laparoscopic approach was utilized in the majority, achieving a median length of hospital stay of 7 days, and a stage-specific survival similar to or better than open-surgery results. The authors have now performed MIE on over 1,000 patients with HGD or cancer. Over time, our approach to MIE has evolved from a three-hole (McKeown) technique to a minimally invasive Ivor–Lewis esophagectomy. The Ivor–Lewis approach avoids the morbidity of a neck anastomosis (higher anastomotic leak and stricture rates, recurrent laryngeal nerve injury, and dysphagia). We currently reserve three-hole MIE for patients with long-segment Barrett's esophagus and proximal tumors, where there is concern about achieving adequate proximal resection margins. Reduced postoperative pain and shorter hospital stay are noted when comparing MIE with open approaches. Furthermore, MIE may be associated with reduced overall postoperative morbidity, while achieving comparable oncologic outcomes and long-term survival.

MIE should be performed by surgeons who have extensive experience in minimally invasive esophageal surgery. Patients must be deemed fit for operation, and must have resectable lesions as characterized by EUS or computed tomography (CT). During the early portion of the learning curve, surgeons should consider performing MIE in patients who have HGD, small tumors, a favorable body habitus, and minimal or no prior abdominal or thoracic surgery. As experience increases, we have found that previous abdominal or thoracic surgery and preoperative chemotherapy or radiation therapy do not represent contraindications to a minimally invasive approach for either staging or resection of esophageal cancer.

Minimally Invasive Ivor–Lewis Esophagectomy—Technique

The procedure is begun with an on-table esophagogastroduodenoscopy (EGD) to assess tumor location and the suitability of the gastric conduit for reconstruction. If the EGD, EUS, or CT scan findings suggest gastric extension, T4 local invasion, or possible metastases, we perform a staging laparoscopy, staging thoracoscopy, or both. The patient is then intubated with a double-lumen endotracheal tube to permit single-lung ventilation, and the laparoscopic portion is performed first. Five laparoscopic ports are inserted as demonstrated in Figure 19.5, starting with a 10-mm port placed via direct cut-down two-thirds down a line joining the xiphoid process and umbilicus and 3 cm to the right of the midline. The abdomen is insufflated with carbon dioxide to a pressure of 15 mmHg and the remaining ports are placed. We routinely use a 30 degree 10 mm laparoscope for visualization.

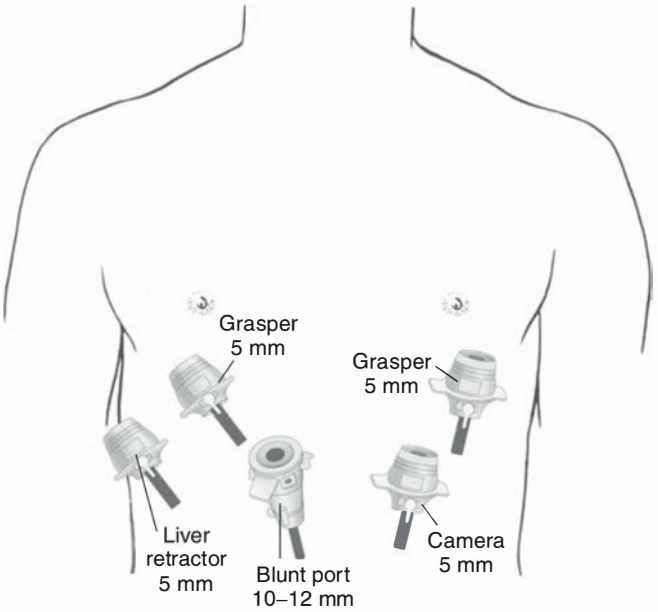


Fig. 19.5. Standard port placement for laparoscopic gastric mobilization, pyloroplasty, and J-tube insertion.

Staging laparoscopy is then performed, and if no metastatic disease is present we continue with gastric mobilization by dividing the gastrohepatic ligament. The right and left crus are dissected taking care to avoid early division of the phrenoesophageal ligament to prevent loss of pneumoperitoneum. The short gastric arteries and gastrocolic omentum are then divided with ultrasonic coagulating shears, while preserving the right gastroepiploic arcade. In some cases, we mobilize an omental pedicle at the mid- to upper-third of the greater gastric curve. This pedicle is supplied by two feeding vessels and is used later in the procedure to wrap the anastomosis. The stomach is then rotated to the patient's right and all posterior attachments to the pancreas are divided. A celiac lymph node dissection is performed along the splenic artery toward the splenic hilum. The left gastric artery and vein are then exposed and divided with an endoscopic linear cutting stapler, taking care to divide the vessels proximally to allow resection of the lesser curve lymph nodes and fat with the specimen. The pylorus and antrum are then mobilized to allow the pylorus to reach the right crus without undue tension. The right gastric vessels are preserved. Using ultrasonic shears and 2-0 braided nonabsorbable suture, a Heineke–Mikulicz pyloroplasty is performed (Fig. 19.6). A gastric conduit is then created with multiple firings of the endoscopic linear cutting stapler using 3.5 to 4.8 mm staples, starting along the lesser curve at the level of the antrum and directing the stapler toward the angle of His (Fig. 19.7). For exposure of the stomach during this part of the procedure, the assistant grasps the

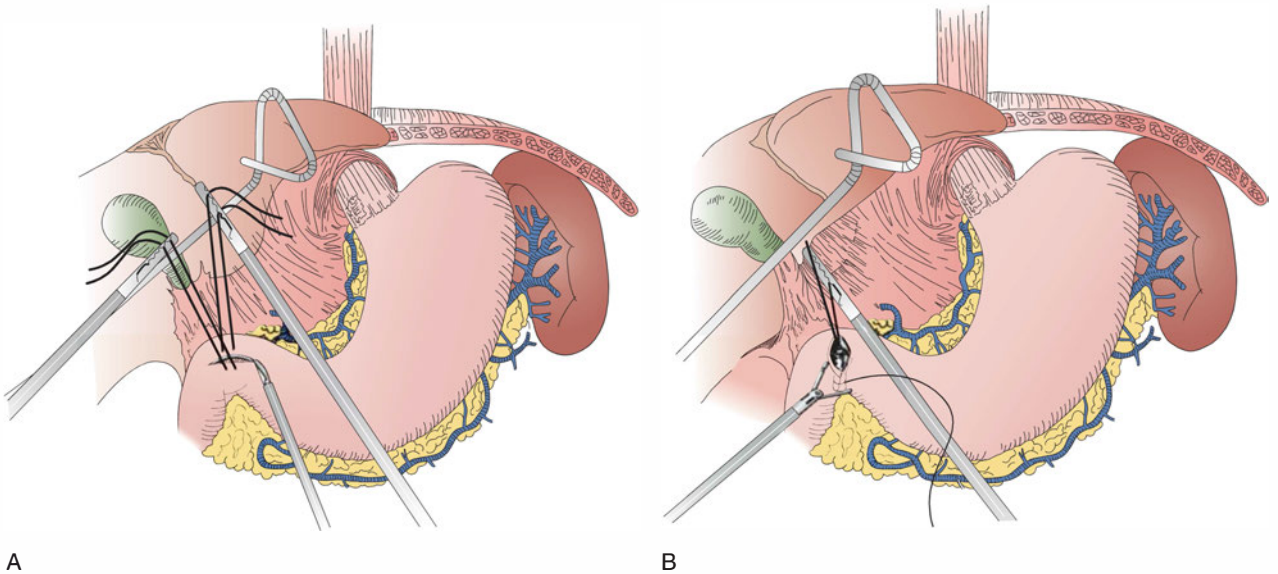


Fig. 19.6. Laparoscopic pyloroplasty. (**A**) The pylorus is divided longitudinally after placement of stay sutures. (**B**) The pylorus is then closed transversely with interrupted 2-0 Surgidac sutures. A portion of the omentum may be tacked over the closure to provide additional coverage.

gastric fundus and applies traction toward the spleen. At the same time, the antrum is retracted inferiorly and to the patient's right to maximize conduit length. We limit conduit diameter to 4 to 5 cm to optimize emptying. The conduit is then sutured to the specimen with two 2-0 Endo Stitches (U.S. Surgical, Norwalk, CT) (Fig. 19.8). Alignment of the conduit is maintained to prevent rotation, with the staple line facing the patient's right at all times. If harvested, the omental pedicle is tacked to the proximal conduit to prevent traction injury during advancement into the chest.

A feeding jejunostomy tube is then placed. With the patient flat, the transverse colon is retracted cephalad and the ligament of Treitz is identified. A 10-mm right lower quadrant port is inserted and the jejunum is grasped 30 cm distal to the ligament of Treitz and elevated toward the patient's left abdominal wall. The jejunum is tacked to the anterior abdominal wall with a single Endo Stitch and a needle jejunostomy catheter is inserted just distal to the tacking stitch under laparoscopic visualization. Intraluminal placement is confirmed by bowel distention after injecting 10 ml of air through the catheter. Three circumferential 2-0 Endo Stitches are placed at the catheter entry site to secure that part of the jejunum to the anterior abdominal wall. The jejunum distal to the catheter insertion site is then secured to the anterior abdominal wall along a distance of several centimeters with interrupted 2-0 Endo Stitches to prevent torsion of that segment of bowel. Once this is accomplished, the phrenoesophageal ligament is divided circumferentially to allow mobilization of the distal esophagus and passage of the conduit into the chest. A Penrose drain can be placed around the distal esophagus to aid with retraction. An omental patch is then used to cover the pyloroplasty and the posterior crura are approximated with 0 Endo Stitches to prevent hiatal herniation.

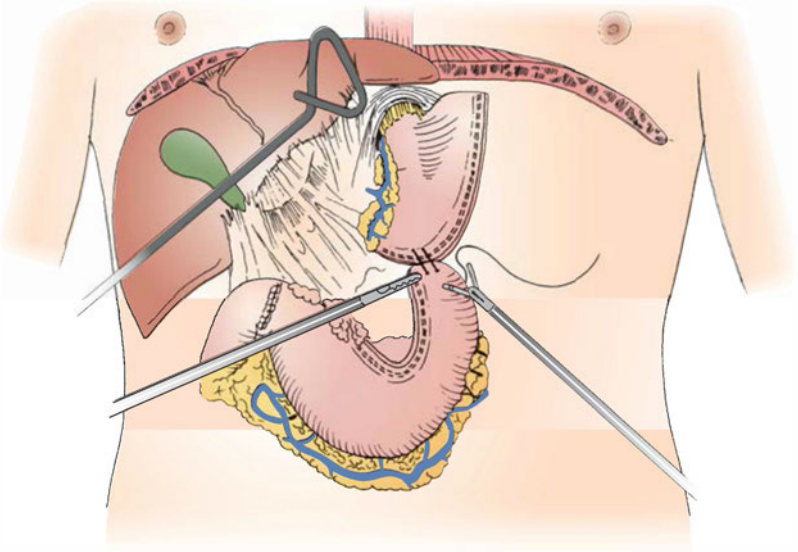


Fig. 19.8. The gastric tube is sutured to the specimen to facilitate alignment during pull-up.

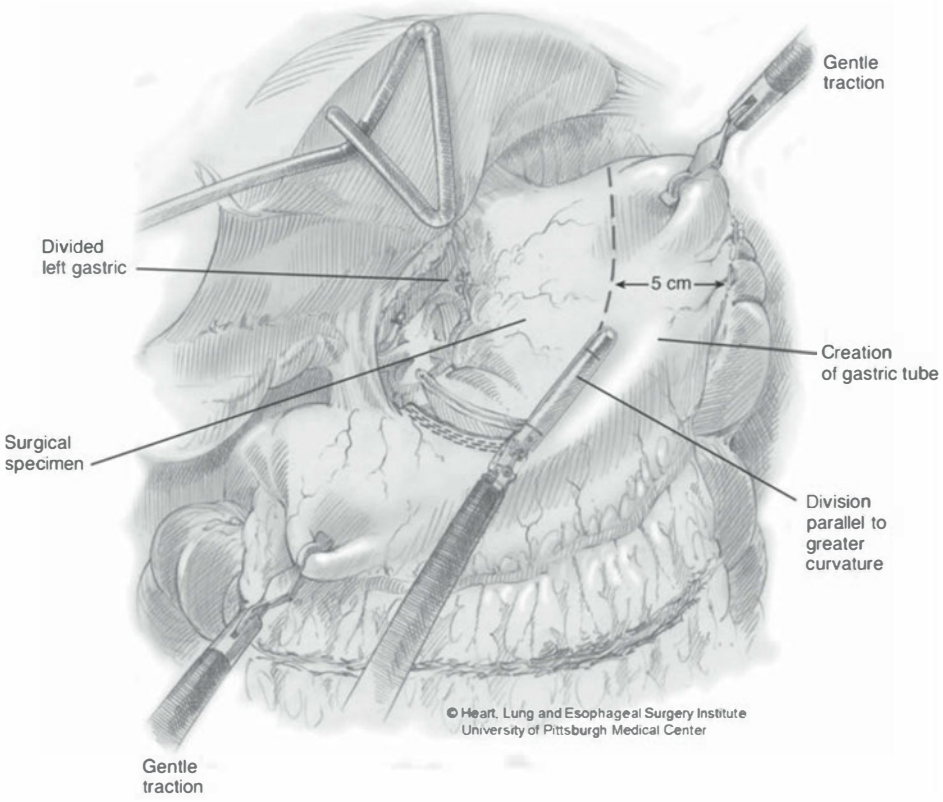


Fig. 19.7. Creation of gastric tube.

The patient is then repositioned in a left lateral decubitus position for the thoracoscopic portion of the procedure. Five ports are then inserted in the right chest as follows (Fig. 19.9): a 10-mm port is placed in the seventh intercostal space, midaxillary line for the thoracoscope; a second 10 mm port is placed in the eighth intercostal space 2 cm posterior to the posterior axillary line for the ultrasonic shears; a final 10 mm port is placed in the anterior axillary line at the level of the fourth intercostal space, and is used for placement of a fan retractor to assist with anteromedial lung retraction and exposure of the esophageal bed; a 5-mm port is placed posterior to the tip of the scapula. An additional 5 mm port can be placed anteriorly in the fifth or sixth intercostal space for suction. A retracting suture (0-Surgidac; U.S. Surgical Corp., Norwalk, CT), is placed in the central tendon of the diaphragm to improve visualization of the hiatus and is brought out of the inferior anterior chest wall through a 1-mm skin incision using the Endo Close device (U.S. Surgical Corp., Norwalk, CT). This traction suture allows downward retraction on the diaphragm without the need for manual retraction, and provides excellent exposure of the distal esophagus at the level of the diaphragm.

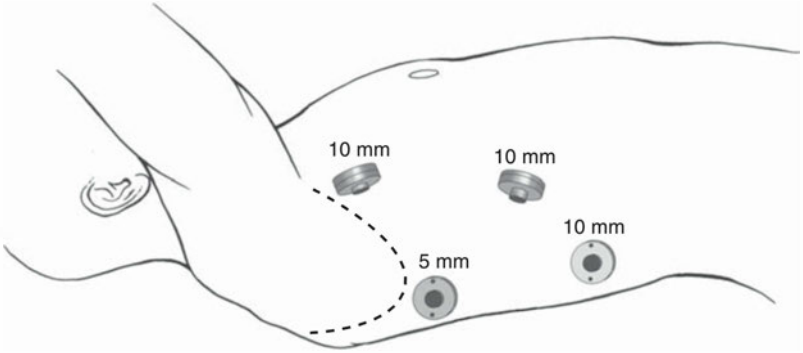


Fig. 19.9. Thoracoscopic port placement for minimally invasive esophagectomy.

Next, the inferior pulmonary ligament is divided. The mediastinal pleura overlying the esophagus is divided anteriorly and posteriorly, and the entire thoracic esophagus is exposed. The authors generally choose a plane distant to the tumor while dissecting circumferentially around the esophagus. Encircling the esophagus with a Penrose drain facilitates traction and exposure (Fig. 19.10). Circumferential mobilization of the esophagus is then performed to include the surrounding lymph nodes, periesophageal tissue, and fat from the level of the diaphragm to the thoracic inlet. The dissection is carried along the pericardium and aorta and the contralateral mediastinal pleura, up to (but not including) the thoracic duct and azygous vein laterally. Aortoesophageal vessels are sequentially ligated and divided with the Autosonic shears (U.S. Surgical Corp., Norwalk, CT). Clips are deployed liberally during this portion of the dissection (especially laterally in the vicinity of the thoracic duct) to minimize the risks of bleeding and thoracic duct leak. The azygous vein is isolated circumferentially and divided using the Endo GIA stapler with a vascular load (U.S. Surgical Corp., Norwalk, CT). Dissection is then carried proximally toward the thoracic inlet along a plane immediately adjacent to the esophagus to minimize the risk of injury to the recurrent laryngeal nerve and posterior trachea. Care is taken to preserve the pleura above the azygous vein. We believe that this pleural layer helps to maintain the gastric tube in a mediastinal location, and may also help to seal the plane around the gastric tube near the thoracic inlet, thereby minimizing the extension of a cervical leak downward into the chest. The authors do not routinely dissect out the recurrent laryngeal lymph nodes or perform a cervical lymph node dissection. The distal esophagus and conduit are then pulled gently into the chest taking care to maintain proper conduit orientation (Fig. 19.11). The proximal esophagus is then divided at or above the level of the azygous vein utilizing scissors. The sutures securing the proximal stomach to the conduit are then cut. The eighth intercostal space incision is enlarged to a length of approximately 5 cm without rib spreading, and a wound protector (Applied Medical, Rancho Santa Margarita, CA) is placed to facilitate removal of the esophagus. The specimen is sent for frozen section analysis to assess the esophageal and gastric margins. The anvil of the 28-mm end-to-end anastomosis (EEA) stapler is inserted into the esophagus and secured with two 2-0 purse string Endo Stitch sutures. The EEA stapler is then introduced into the chest through the eighth interspace incision. A gastrotomy is created near the tip of the conduit to allow insertion of the EEA stapler. The stomach is gently pulled over the tip of the EEA stapler, and is advanced cephalad to meet the anvil within the proximal esophagus (Fig. 19.12). An end-to-side anastomosis is thus created between the esophagus and conduit above the level of the azygous vein. The remaining gastrotomy defect is closed with the Endo GIA stapler (Fig. 19.13). If harvested, the omental pedicle is wrapped circumferentially around the anastomosis and secured with 2-0 Endo Stitches. The intercostal nerves are then blocked with 1 to 2 cc of 0.5% bupivacaine in dilute epinephrine for

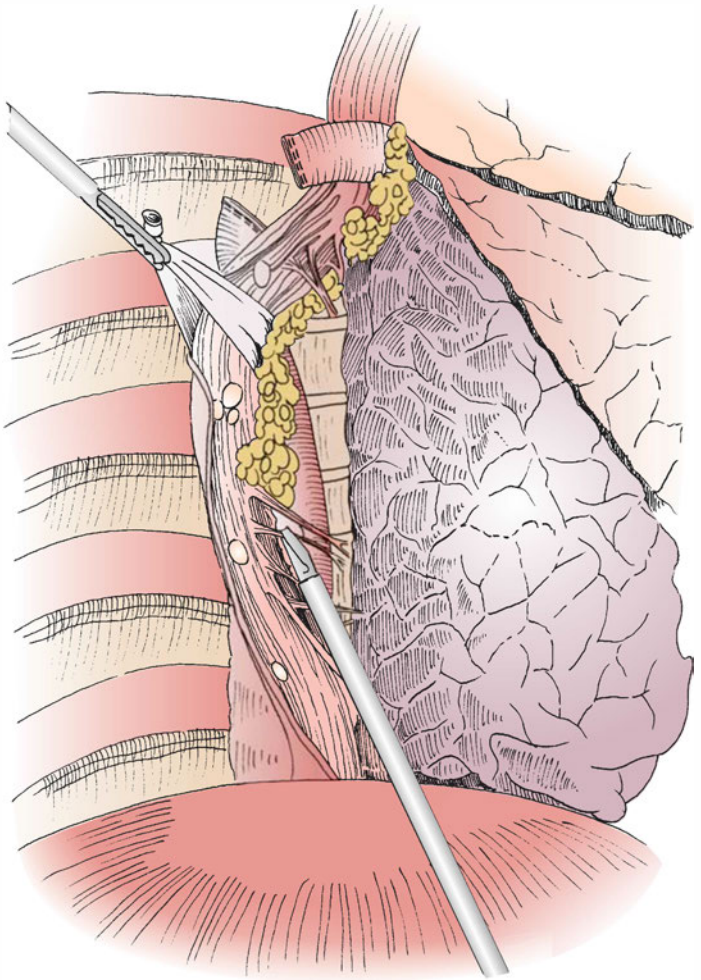


Fig. 19.10. Thoracic esophageal mobilization.

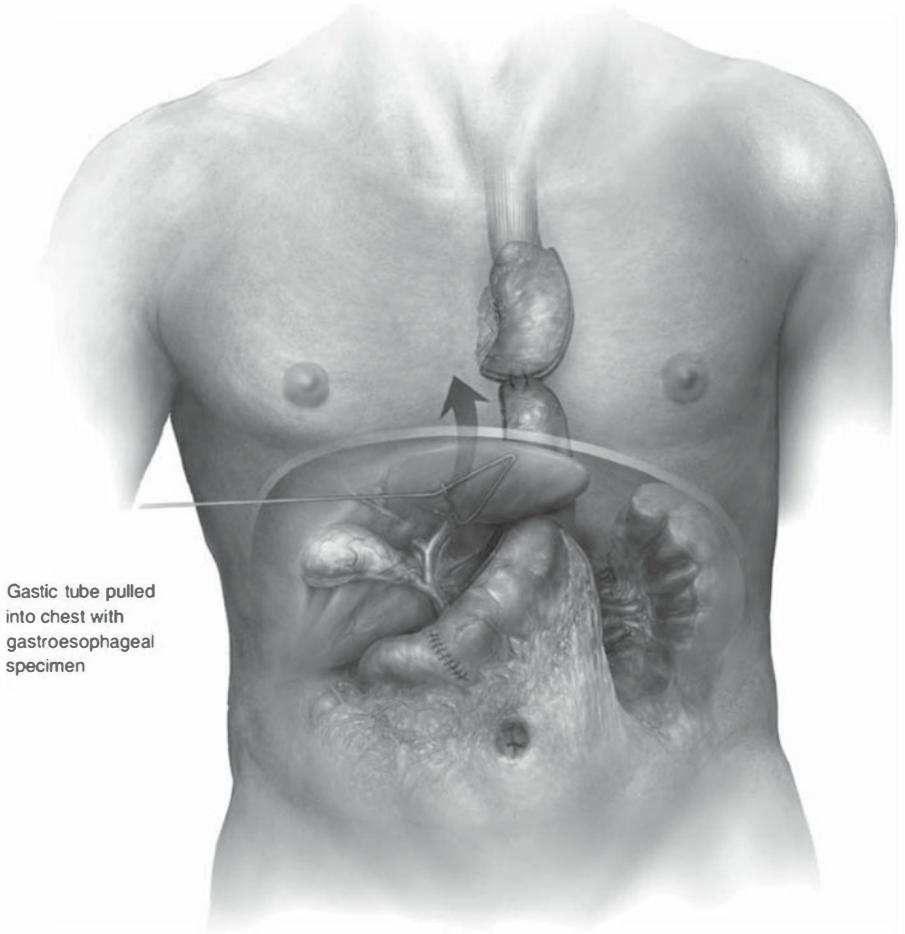


Fig. 19.11. Advancement of specimen and gastric tube into the chest. (Reprinted from Tsai WS, et al. Technique of minimally invasive ivor lewis esophagectomy. Oper Tech Thorac Cardiovasc Surg 2009;14:176-192, Copyright (2009), with permission from Elsevier.)

control of immediate postoperative pain. A nasogastric tube is inserted under direct vision with the tip positioned at the level of the hiatus, and a 28F chest tube is inserted through the camera port and directed posteriorly. The conduit is then tacked to the hiatus with several 2-0 Endo Stitches to prevent herniation. A #10 Jackson-Pratt drain is placed directly posterior to the anastomosis and brought out of the skin through a stab incision at the costophrenic angle. The lung is reinflated and the port sites are closed.

Minimally Invasive Three-Hole Esophagectomy—Technique

Previously, our preference was a three-hole (McKeown) MIE. The thoracoscopic and laparoscopic portions of the procedure are essentially identical to those described above with the difference that the thoracoscopic part is performed first. The proximal esophageal dissection is carried proximally through the thoracic inlet to the base of the neck. The patient is then positioned supine and the laparoscopic portion is completed. Division of the phrenoesophageal membrane is performed last to prevent escape of pneumoperitoneum into the chest. A 4 to 6 cm horizontal left neck incision is then made two fingerbreadths above the sternal notch. The cervical esophagus is mobilized and exposed. A finger or sponge stick is used to retract the thyroid and avoid injury to the recurrent laryngeal nerve. The dissection is carried distally, until the thoracic dissection plane is encountered. The esophagus is divided 1 to 2 cm below the cricopharyngeus, and the esophagogastric specimen is carefully pulled out of the wound while the laparoscopic assistant carefully delivers the specimen and gastric tube in proper alignment into the mediastinum (Fig. 19.14). The specimen is sent to pathology for frozen section analysis of the surgical margins. An anastomosis is performed between the esophagus and gastric tube using a hand-sewn, end–side EEA or side–side technique with an Endo GIA II stapler. When a McKeown MIE is performed, we prefer a very high anastomosis to ensure adequate resection of any tumor or Barrett's involvement, as well as to enable anastomotic leak drainage through the neck incision. A nasogastric tube is passed through the anastomosis and distally into the gastric tube for postoperative decompression.

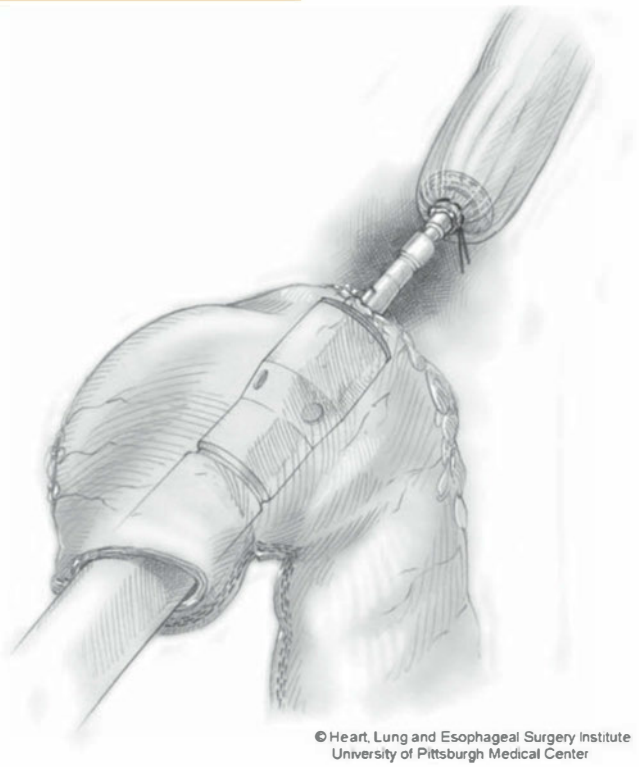


Fig. 19.12. Intrathoracic EEA anastomosis.

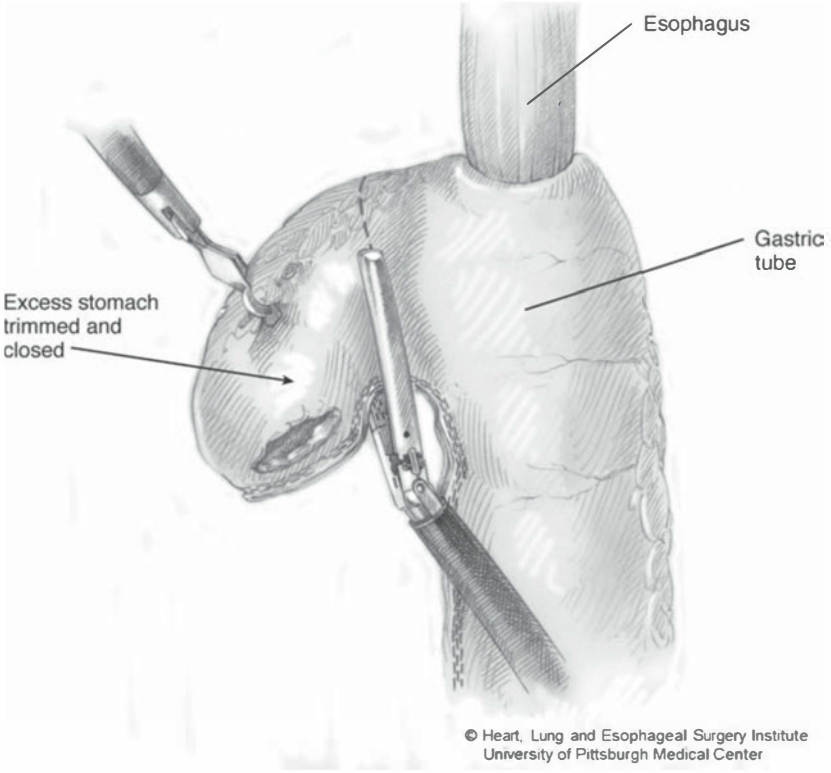


Fig. 19.13. Excision of gastric tip following completion of esophagogastric anastomosis.

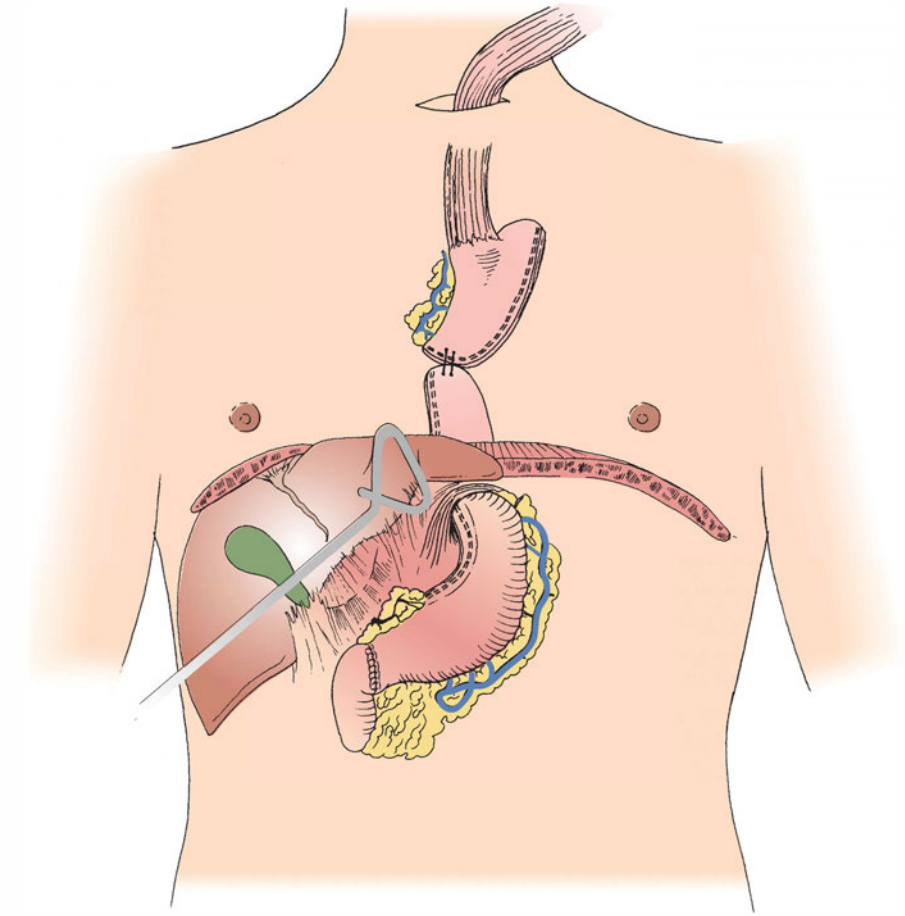


Fig. 19.14. Through a low transverse cervical incision, the esophagogastric specimen is removed and the gastric tube is advanced to the neck.

Any redundant gastric conduit is then pulled back into the abdomen under direct visualization. The gastric tube is tacked to the diaphragm to prevent herniation of abdominal contents into the chest, using the Endo Stitch. The abdominal instrumentation is withdrawn and the ports are closed. The skin of the neck is loosely approximated with staples. The completed reconstruction is shown in Figure 19.15.

Minimally Invasive Esophagectomy—Results

We reported our initial experience with 222 cases of MIE at the University of Pittsburgh in 2003. The median age was 66.5 years (range 39 to 89). The gender distribution was 186 (84%) men and 36 (26%) women. Preoperative indications for surgery included carcinoma (79%) and HGD (21%). Neoadjuvant chemotherapy was used in 35% of patients and radiation in 16%. Prior to MIE, esophageal stents were inserted in 6%, feeding tubes (G or J tubes) in 3%, and previous open abdominal surgery had been performed in 25% of the patients. The majority of cases were completed using a minimally invasive three-hole approach. Reasons for conversion to open esophagectomy included dense pleural adhesions, bleeding from esophageal and intercostal vessels, and difficulty with an intrathoracic anastomosis. Open thoracotomy is also occasionally necessary in the setting of locally advanced tumors. In the initial 222 cases performed at this institution, mini thoracotomy was liberally performed in 8 of the first 15 patients, with only four additional thoracotomies utilized in the next 207 cases. Conversion to open laparotomy was performed in four patients due to dense abdominal adhesions. Overall, MIE was successfully completed in 206 patients (93%), with a 30-day operative mortality of 1.4% ($N = 3$). The reason for this trend is not established, though lower rates of respiratory and wound-related complications are evident when compared with the reports of standard open esophagectomy, which in turn may account for the lower mortality. The most frequent minor complication was atrial fibrillation (12%), followed by pleural effusion (6%), which was treated with bedside thoracentesis or pigtail catheter drainage. Major complications occurred in 32% of patients. The most common major complication was anastomotic leak (12%). Most anastomotic leaks were localized to the neck, and were managed conservatively. Pneumonia was the second most common major complication, occurring in 8% of patients. Vocal cord palsy (4%), chylothorax (3%), and gastric tip necrosis (3%) were rare,

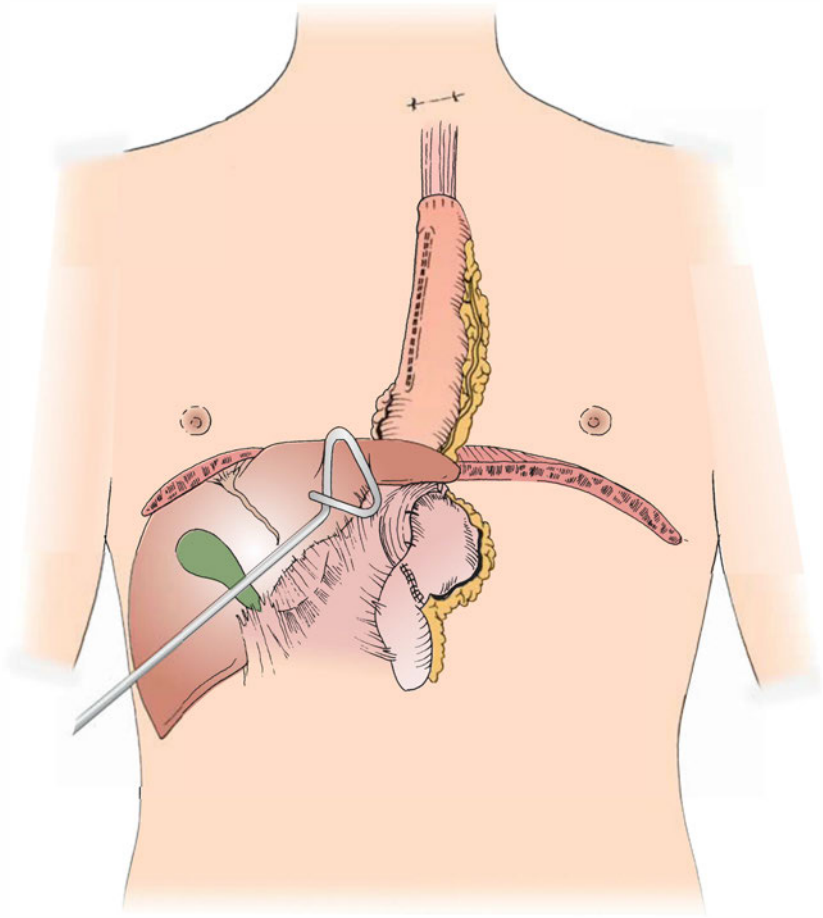


Fig. 19.15. Completed cervical anastomosis.

but serious, complications. Early Teflon or Gelfoam injection of the vocal cords in the setting of recurrent laryngeal nerve palsy improves swallowing function and lowers the risk of aspiration. Oncologic outcomes appeared comparable to published open series.

Our initial experience with minimally invasive Ivor–Lewis esophagectomy was reported in 2006 and included 50 patients. The initial 35 procedures included a planned mini thoracotomy, and the remaining 15 patients had a totally laparoscopic/thoracoscopic MIE. The anastomotic leak rate was 6% and pneumonia (10%) was only encountered in those patients who underwent mini thoracotomy. Median ICU length of stay was 1 day for the entire group. Patients who underwent mini thoracotomy had a significantly prolonged length of hospital stay (9 days) compared with those who underwent a total MIE (7 days).

We have now performed over 1,000 MIEs, with the Ivor–Lewis esophagectomy constituting our preferred minimally invasive approach. In our review of 1,011 patients undergoing MIE, 481 patients underwent three-hole MIE and 530 the Ivor–Lewis MIE approach. The operative mortality was 1.7% for the entire series (2.5% three-hole vs. 0.9% Ivor–Lewis). There were no intraoperative deaths. The frequency of conversion to open esophagectomy remained stable at 4.5%. Median ICU stay (2 days) and overall hospital length of stay (8 days) were similar between the two approaches. The median number of lymph nodes harvested was 21 (23.5 Ivor–Lewis vs. 19 three-hole; $P < 0.001$). A significant reduction in vocal cord paresis/paralysis was noted in the Ivor–Lewis group when compared with the three-hole approach (8% vs. 1%; $P < 0.001$). At a median follow-up of 20 months, the stage-specific survival was comparable to open series.

The early results of MIE compare favorably with the results of many series of open esophagectomies. Mortality rates of open esophagectomy from several of the largest open series range from 3% to 9.8%. In a highly publicized analysis of esophagectomy outcomes derived from the national Medicare claims database, published by Birkmeyer and coauthors, high-volume hospitals were found to have the lowest mortality (8.1%), with mortality exceeding 15% in medium- to low-volume centers. By comparison, 30-day operative mortality after MIE in our series was 1.7%. These data suggest that MIE can be performed safely with good results in an experienced center.

In one study comparing minimally invasive ($N = 18$) and open ($N = 16$) esophagectomy, Nguyen and coworkers found that the mean operative time (364 minutes), blood loss (297 ml), and length of intensive care unit stay (6 days) were decreased with MIE as compared with the open esophagectomy performed at the same institution. The incidence of respiratory complications (pneumonia, pulmonary embolism, respiratory failure) was similar between the MIE and open approaches. It should be emphasized that there are significant differences between the study groups in this retrospective comparison. The patients who underwent open esophagectomy had more advanced cancers. Additionally, the open operations were performed by four surgeons with variable experience, whereas the MIE procedures were performed by a single surgeon with expertise in minimally invasive esophageal surgery. The open operations were performed several years before the MIE procedures, so there may also have been differences in practice accounting for longer hospital stays.

Quality-of-life subjective assessments after MIE have been reported to be similar to preoperative values and population norms. In our series of 222 patients, the mean postoperative dysphagia score was 1.4 on a scale from 1 (no dysphagia) to 5 (severe dysphagia). Since reflux can be an issue after esophagectomy, heartburn severity was measured using the Health-Related Quality of Life Index (HRQOL). The mean heartburn score was 4.6 (on a scale from 0 to 45), which represents a normal (no reflux) score. SF36 scores were also measured and were not significantly different compared with age-matched normal values during follow-up.

In conclusion, MIE is a technically demanding procedure with a significant learning curve. Operative times typically decrease from 7 to 8 hours to 4 to 5 hours after performing 20 operations. Therapeutic outcomes compare quite favorably with most open series and are superior in many instances. Such encouraging results will serve to broaden the applicability of this technique to higher-risk patient groups such as the elderly and patients who have received prior neoadjuvant or endoscopic therapy (e.g., chemoradiation, stents). Prospective studies will be required to determine whether postoperative pain,

recovery time, and cost are improved. The Phase II Eastern Cooperative Oncology Group intergroup study (E2202) evaluating the safety profile of MIE has completed accrual, and results should be forthcoming in the near future. Until these results are available, the optimal surgical approach for each patient should be decided based on surgeon experience, tumor characteristics, and patient preference.

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EDITOR'S COMMENTS

There is not another group in the country and perhaps in the world with greater experience in minimally invasive esophagectomy (MIE) than the Luketich group at the University of Pittsburgh. In this chapter they elegantly describe their technique as well as outcomes, which compare favorably with those reported for open procedures. As they correctly point out, there is a significant learning curve associated with this operation but the technique can be mastered especially by the surgeon experienced with open esophagectomy. The Pittsburgh group clearly favors the Ivor Lewis operation done via laparoscopy and thoracoscopy over a three-hole technique where the anastomosis is placed in the neck. Leak rates are lower when the anastomosis is placed within the chest and the incidence of recurrent nerve injury is negligible when dissection in the neck is avoided.

I am not yet convinced that the morbidity or mortality results with MIE are any better than those achieved by an experienced group performing standard open esophagectomy. The length of stay does not appear to be significantly different and frankly I would not expect it to be. The length of time that it takes to heal an anastomosis and start a patient on oral intake should be the same no matter how the resection is performed and discharge really cannot occur until the anastomosis is demonstrated to be intact and the patient is eating. Whether or not there is a difference in postoperative pain still remains to be determined although intuitively one would expect those undergoing MIE to have less pain without a big abdominal incision and a rib spreading thoracotomy. That said, I have been convinced for years that I have been doing a MIE and it is called a transhiatal esophagectomy, an operation that involves no thoracic incision, not even small ones. It is likely that those completing cardiothoracic residencies today may have more experience with the MIE operation described by Luketich and colleagues than with the transhiatal approach. In addition MIE using the robot may further enhance the capabilities of the minimally invasive approach with the enhanced three-dimensional visualization and increased technical capability provided by the robotic arms. But as I have repeatedly stated the complete esophageal surgeon must have the capability to perform an array of approaches since there are so many variations in presentation that are encountered. One approach does not fit all. MIE clearly adds to the armamentarium of the esophageal surgeon and it behoves one interested in this area to master the procedure.

LRK

20 Surgery for Achalasia and Other Motility Disorders

Farzaneh Banki and Tom R. DeMeester

The diagnosis and treatment of esophageal motility disorders can be an important part of a thoracic surgeon's practice. The majority of patients with esophageal motility disorders are referred to surgeons following a primary assessment by a gastroenterologist. The patients are commonly on various medications and have undergone multiple endoscopies and dilatations. The effects of these treatments may be of some benefit but the patient's symptoms remain and a more definitive solution is desired. It is crucial for the thoracic surgeons prior to surgery to validate the accuracy of the patient's diagnosis. To do so, they must understand esophageal physiology, be able to perform a thorough esophageal assessment, and determine the best treatment option.

Since the last edition of this book, there has been a shift in the surgical approach to esophageal motility disorders. A left thoracotomy, once the standard approach to perform a Heller myotomy, has been replaced by the laparoscopic approach. Similarly, sophisticated diagnostic tools have been developed, such as high-resolution manometry, esophageal impedance, and 48-hour Bravo pH capsule. In this chapter, we will review the diagnosis and treatment of the primary and secondary esophageal motility disorders. We will focus on the diagnostic assessment and the use of endoscopic, minimally invasive, and open surgical therapy.

CLASSIFICATION OF ESOPHAGEAL MOTILITY DISORDERS

1. Primary esophageal motility disorders (absence of extra esophageal causes)
    a. Achalasia
    b. Diffuse esophageal spasm
    c. Nutcracker esophagus
    d. Hypertensive lower esophageal sphincter
    e. Ineffective esophageal motility
2. Secondary esophageal motility disorders (due to extra esophageal causes)
    a. Collagen vascular disease (scleroderma, polymyositis, dermatomyositis, systemic lupus)
    b. Neuromuscular disease (myasthenia gravis)
    c. Endocrine and metabolic disorders

ACHALASIA

Definition, Prevalence, and Natural History

Achalasia is a primary esophageal motility disorder that can affect the esophageal body, lower esophageal sphincter, and stomach. It is characterized by the absence of peristaltic contractions in the esophageal body, a hypertensive lower esophageal sphincter, complete or nearly complete loss of the lower esophageal sphincter relaxation on swallowing and, an elevation of esophageal luminal pressure above gastric baseline pressure. Some patients may also have delayed gastric emptying. The latter can occur years after the onset of the disease.

The frequency of the disease in the western world is one per 100,000 population per year with geographic pockets of higher incidences. There are two peaks in the occurrence of the disease, one between ages 20 and 40 and the other between 60 and 70. If left untreated, achalasia will cause a dilated esophagus from the accumulation of liquid within the lumen. The weight of the retained liquid column causes the loss of the normal esophageal axis and the formation of a sigmoid esophagus. Eventually, the patient loses the ability to drink water or take solid nourishment. In the past, this resulted in the death of the patient from dehydration and starvation. Patients with achalasia have higher risk for squamous cell carcinoma of the esophagus.

Vigorous Achalasia

A subgroup of patients with achalasia have isolated simultaneous segmental contractions in the esophageal body of high amplitude (>180 mmHg). These patients present with chest pain in addition to dysphagia. These patients do not do as well after surgical therapy due to the persistence of chest pain in about 50% of those operated on.

Etiology and Symptoms

The cause of achalasia is unknown. Hereditary, degenerative, autoimmune, and viral infections have been identified as possible etiologies. The most common symptom is dysphagia, which is reported in 94% of patients, followed by regurgitation in 76% and chest pain in 41%.

Up to half of the patients may complain of heartburn from esophageal distension, acidic fermentation of retained food, or delayed clearance of rare reflux episodes.

Diagnosis

A videoesophagram, an esophageal motility study, and an upper endoscopy should be obtained in patients suspected of having achalasia. Computed tomography (CT) scan and pH monitoring are selectively used.

Videoesophagram

In patients with early achalasia, the esophagus is normal in diameter and peristalsis is absent. The distal esophagus is characterized by a smooth tapering of the barium column resembling a bird's beak. As the disease progresses, the esophagus becomes more dilated and tortuous. Retained food and saliva collect within the esophageal lumen and form an air–fluid level when the patient is in the upright position (Fig. 20.1). A gastric air bubble is typically absent.

Upper endoscopy

In patients with achalasia, upper gastrointestinal endoscopy shows a dilated

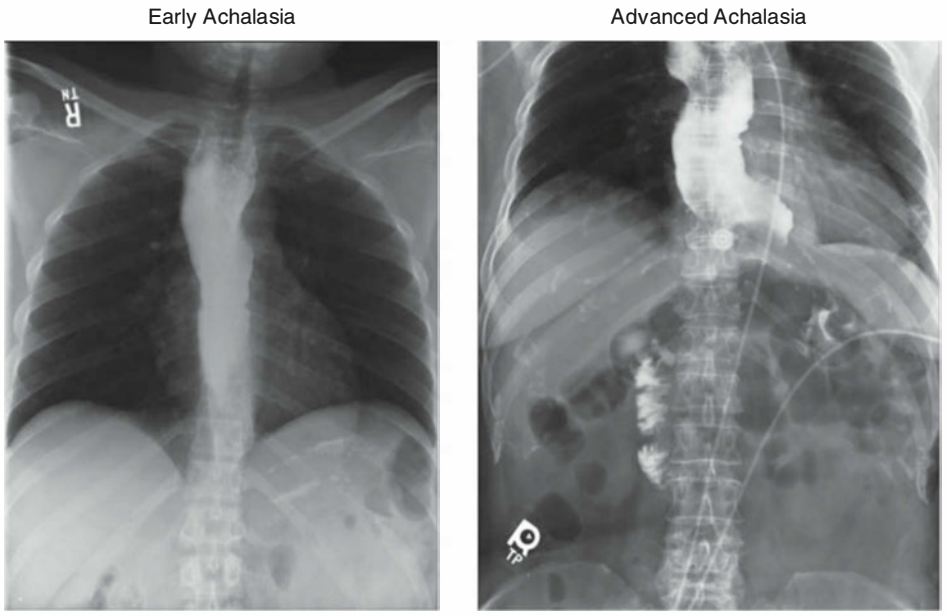


Fig. 20.1. Barium esophagram in patients with early and advanced achalasia. In early achalasia, the esophagus is dilated, the axis remains straight, peristalsis is absent, and the esophagus is narrowed at the level of the diaphragm suggesting a "bird's beak" deformity. In advanced achalasia, the esophagus is extensively dilated, the axis is deviated, usually to the right, and a "bird's beak" deformity is commonly seen at the level of the diaphragmatic hiatus.

esophagus containing foamy saliva and retained food (Fig. 20.2). The gastroesophageal junction is closed but allows a 9-mm endoscope to be pushed through the junction with minimal but definite resistance. Rarely is balloon dilation required to facilitate the passage of the scope. Pseudoachalasia can occur secondary to esophageal obstruction from a malignant lesion at the level of the gastroesophageal junction. Upper endoscopy usually can exclude this possibility, but occasionally the diagnosis can be difficult due to normal mucosa that overlies a submucosal tumor. A CT scan usually resolves the issue. The upper gastrointestinal endoscopy should be performed by the operating surgeon prior to the day of the surgery. If performed immediately prior to surgery, the upper abdominal viscera can become distended and obstruct the laparoscopic image.

Esophageal Manometry

The gold standard for the diagnosis of achalasia is the complete or nearly complete loss of the lower esophageal sphincter relaxation in response to wet swallows and the absence of peristalsis in the esophageal body on esophageal manometry (Fig. 20.3). A hypertensive lower esophageal sphincter is present in less than half of the patients. Commonly, there is an elevated esophageal baseline luminal pressure reflecting outflow resistance. Patients with vigorous achalasia have in addition isolated segmental contractions of high amplitude (> 180 mmHg; Fig. 20.4).

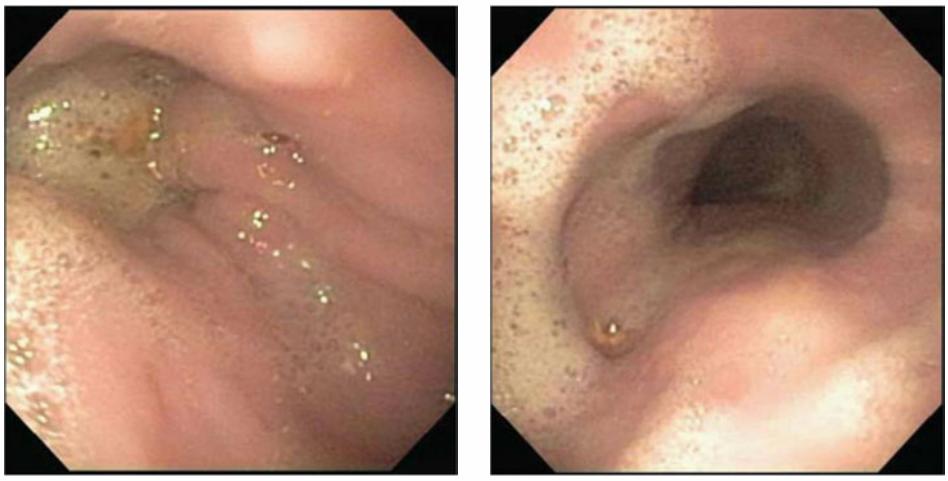


Fig. 20.2. Upper gastrointestinal endoscopy in a patient with achalasia showing a dilated esophagus, foamy saliva, and retained food.

Computed Tomography Scan

A CT scan is not necessary except when pseudoachalasia is suspected, and the gastroesophageal junction needs to be imaged to identify a mass compressing the esophagus.

pH Monitoring

pH monitoring is performed when the diagnosis of achalasia is questionable and gastroesophageal reflux disease is suspected. It is not uncommon to confuse the two entities. Achalasia patients can complain of heartburn from esophageal distension, acidic fermentation of retained food, or delayed clearance of rare reflux episodes. To exclude gastroesophageal reflux disease, pH monitoring should be performed. The pH tracing must be carefully analyzed to distinguish the episode of gastroesophageal reflux from long periods of low pH secondary to fermentation of retained food or episodes of rare reflux that clears slowly from the esophagus.

Treatment

The goal of therapy for achalasia is deceptively simple: to relieve dysphagia by reducing esophageal outflow resistance without causing unimpeded reflux of the gastric juice. To achieve this goal is more difficult than it sounds.

Nonsurgical Treatment

Botox Injection

Botulinum toxin inhibits the calcium-dependent release of acetylcholine from nerve endings involved in the relaxation of the lower esophageal sphincter. It is therapeutically effective in relieving dysphagia in about 85% of patients. Over a period of 6 months, the symptoms recur in more than 50% of the patients possibly because of regeneration of the affected receptors. Patients older than 60 are likely to have a sustained response lasting for 1 to 5 years.

A Heller myotomy after Botox injection can be difficult and less effective

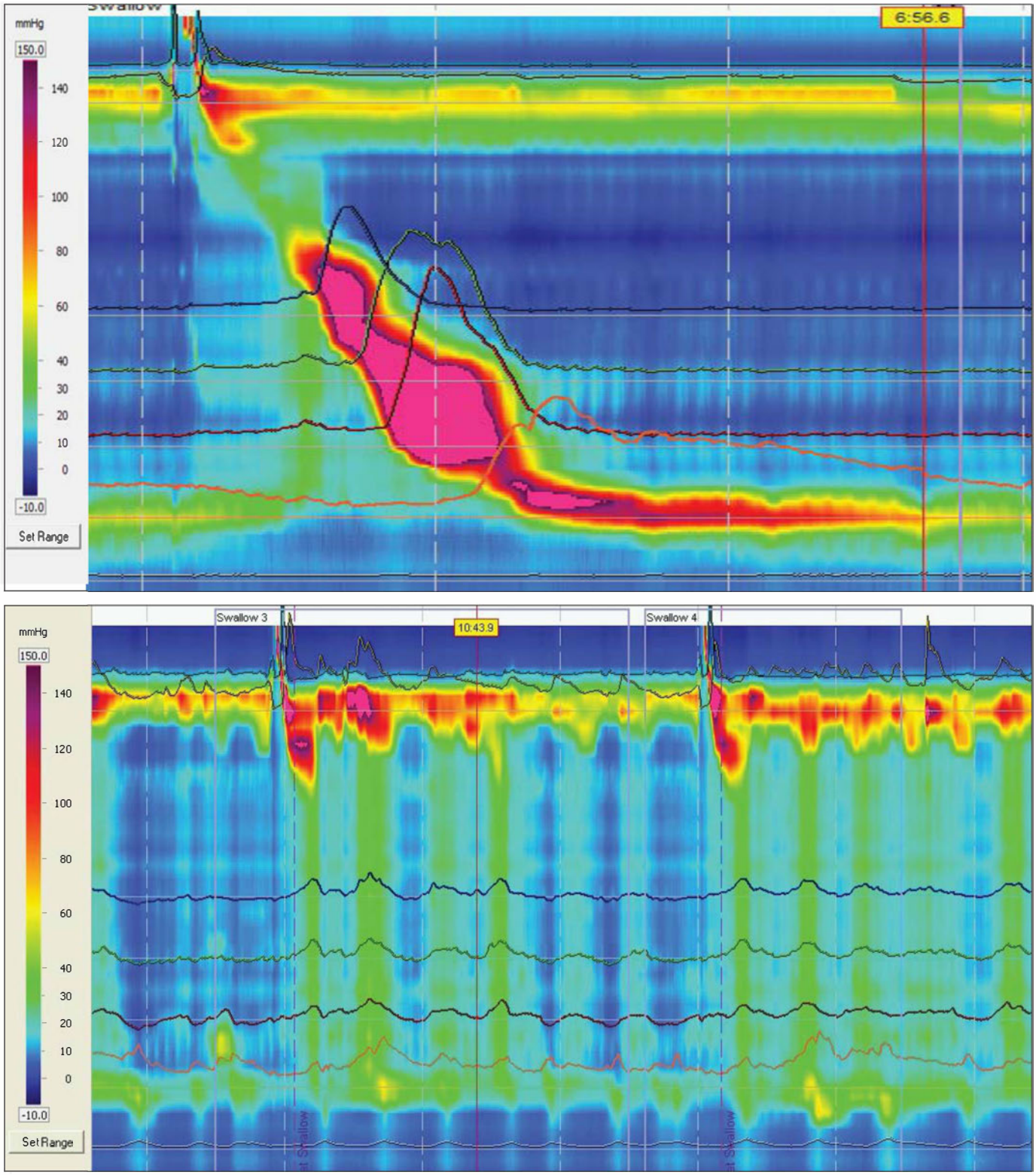


Fig. 20.3. High-resolution manometry in a normal subject (upper) and in a patient with achalasia (lower). In normal subjects, a pharyngeal swallow (in *red*) is followed by a peristaltic wave in the body of the esophagus (also in *red*). Simultaneously, with the pharyngeal swallow the lower esophageal sphincter relaxes (shown by disruption of the *green* band). In patients with achalasia, the pharyngeal swallows are not followed by a peristaltic wave and the lower esophageal sphincter does not relax. The lower esophageal sphincter pressure is elevated compared with the normal subject. The luminal pressure in the esophageal body is elevated (*green columns*) above gastric pressure (*blue color*).

because of submucosal scar formation. The fibrotic reaction can obliterate the anatomical plane between the mucosa and the muscular layer of the esophagus. This increases the risk of mucosal perforation when performing the myotomy. For this reason, Botox injections are used only in high-risk elderly patients with significant comorbidities.

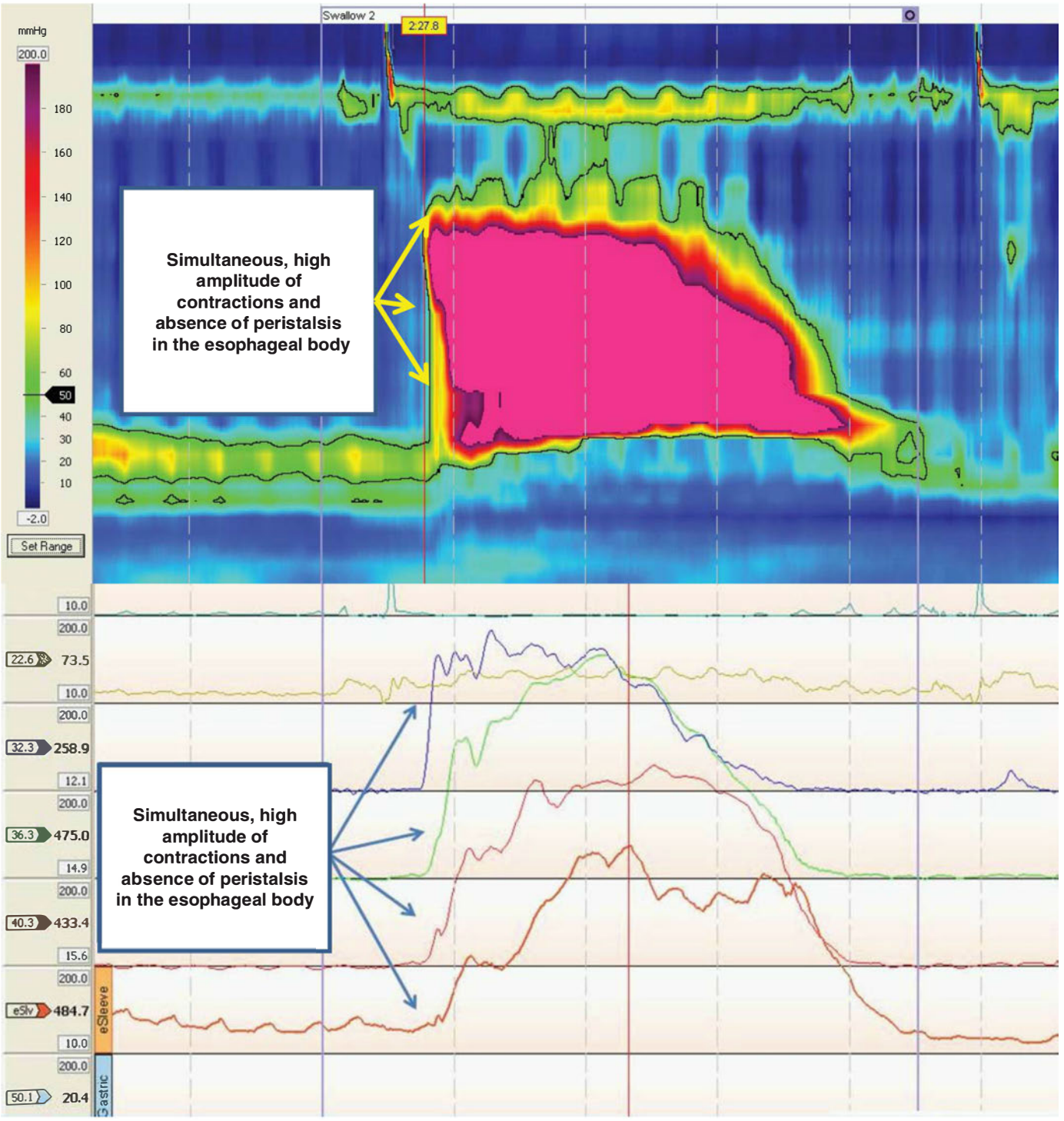


Fig. 20.4. Vigorous achalasia on high-resolution manometry (upper) and on standard manometry (lower).

Pneumatic Balloon Dilation

Pneumatic balloon dilation of the lower esophageal sphincter is the most effective nonsurgical treatment for achalasia. A special balloon, used to perform dilatation, is inflated to a diameter of 30 to 40 mm or a circumference of 90F to 120F. The procedure is monitored under fluoroscopy, and the balloon is commonly filled with liquid contrast for a better visualization during inflation. The waist of the balloon is positioned at the level of the hypertrophied lower esophageal sphincter and is inflated to a diameter of 30 mm. A follow-up procedure can be performed if dysphagia persists. This is usually done by inflating the balloon to a diameter of 40 mm. The procedure is associated with a 1.6% to 4% risk of esophageal perforation. Predictors of a poor outcome are an age $<$40 and a lower esophageal sphincter pressure $>$10 mmHg 3 months after the procedure.

Surgical Treatment

Heller Myotomy

The most effective and durable treatment for achalasia is a Heller myotomy. Initially, this procedure was performed transthoracically either via a thoracotomy or

thoracoscopy. Today, the procedure is universally performed by laparoscopy. Critical steps of the operation as done by us involve performing a left lateral myotomy in line with the greater curvature of the stomach, starting at the apex of the diaphragmatic esophageal hiatus dividing the esophageal muscle, all the oblique fibers at the angle of His and extending the myotomy 3 cm beyond the gastroesophageal junction onto the stomach. The loss of esophageal axis and a long duration of symptoms are predictors of a poor outcome.

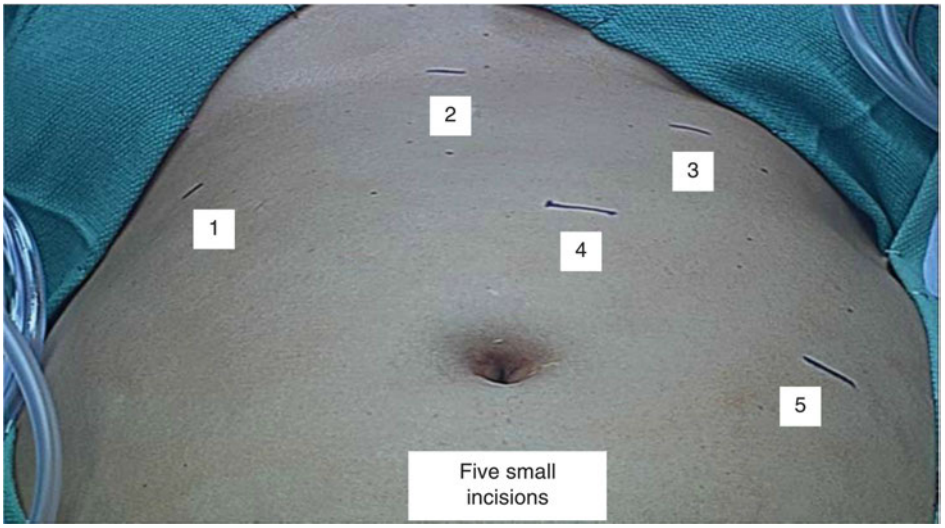


Fig. 20.5. Port locations for laparoscopic myotomy and Dor fundoplication.

Fundoplication after Heller Myotomy

After Heller myotomy, 40% to 60% of patients have gastroesophageal reflux. For this reason, it is recommended that a partial fundoplication should be added. A prospective randomized trial of myotomy alone or myotomy plus Dor fundoplication showed a reduction in postoperative reflux from 48% to 9%, respectively. In contrast, a more recent randomized trial on the degree of partial fundoplication, that is a 180-degree Dor anterior fundoplication versus a 270-degree Toupet posterior fundoplication, did not show a difference in reflux protection. The advantage of a Toupet fundoplication is the retraction of the edges of the myotomy, potentially reducing reapproximation and healing of the myotomized muscle edges together resulting in recurrent dysphagia. The disadvantages are the need to dissect posterior to the esophagus with the potential to damage the posterior vagus nerve and leaving the mucosa at the myotomy site uncovered. In contrast, the Dor fundoplication does not require posterior dissection and covers the mucosa at the myotomy site.

Surgical Technique of Heller Myotomy and Dor Fundoplication

The patient is placed on the operating table and positioned in the reverse Trendelenburg with the lower extremities spread apart in leg holders. A 1.5-cm incision is made just to the left of the midline, two-third the distance between the xiphoid and the umbilicus. An 11-mm Opti-port and a zero-degree scope are used to obtain access into the abdominal cavity under direct vision. The zero-degree scope is exchanged for a 30 degree scope. Under direct vision, a 12-mm multipurpose port is placed in the left upper quadrant, a 5-mm retraction port in the left lower quadrant, a 5-mm dissection port in the right upper quadrant, and a 5-mm port is used to create a channel into the abdomen just below the xiphoid for the insertion of the liver retractor (Fig. 20.5). The liver retractor supplied with the "Iron Intern" is passed through the channel, placed under the left lobe of the liver, elevated and retracted to the right to expose the esophageal diaphragmatic hiatus (Fig. 20.6).

The superior upper third of the gastric greater curvature is mobilized by dividing the short gastric vessels. The posterior fundus is dissected off the lateral surface of the left crus. The left lateral wall of the esophagus is separated from the left crus and the esophagus is mobilized from the crural decussation up to the apex of the diaphragmatic hiatus and into the inferior mediastinum.

The gastroesophageal fat pad is dissected off the esophagus in a left-to-right direction allowing the right side of the fat pad to remain attached. A right-angle instrument is passed through the right upper quadrant dissection port and a small window is created in the base of the mobilized gastroesophageal fat pad at the point where the stomach flares from the tubular esophagus. A vessel loop is passed through the right upper quadrant dissection port, guided through the window and is brought back out through the right upper quadrant port site. The port is removed to free the vessel loop from the lumen of the port and the port is reinserted adjacent to the vessel loop. The esophagus is rotated to the right by retraction of the vessel loop so that the greater curvature of the stomach is in an anterior position (Fig. 20.7) of the stomach.

The myotomy is performed on the left lateral surface of the esophagus in line with

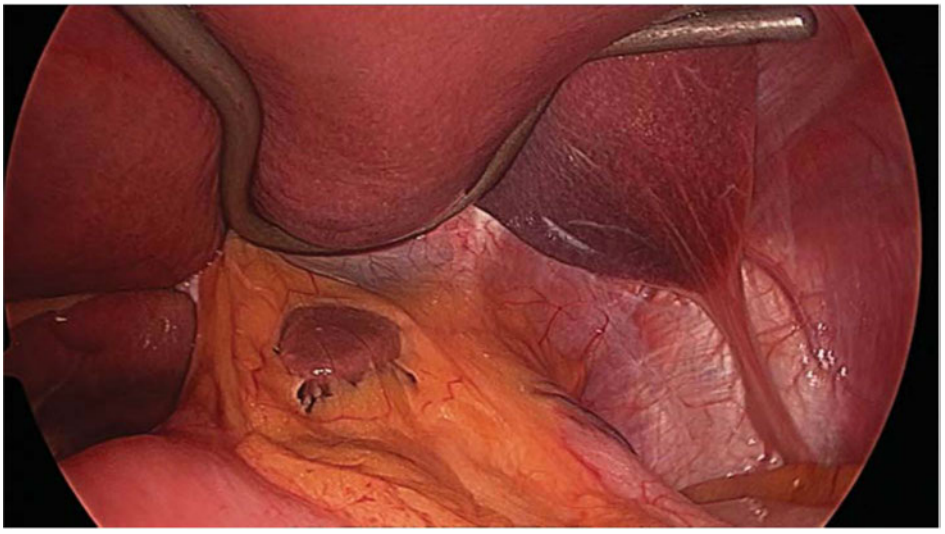


Fig. 20.6. The liver retractor is placed under the left lobe of the liver, elevated and is retracted to the right to expose the esophageal diaphragmatic hiatus.

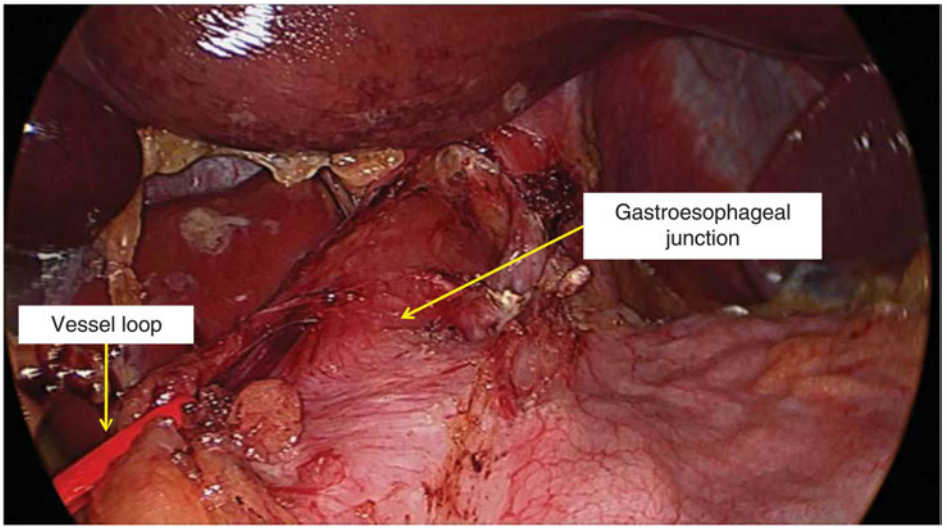


Fig. 20.7. The gastroesophageal junction identified by the point where the stomach flares from the tubular esophagus. Retraction on the vessel loop rotates the esophagus to the right so that the left lateral surface of the esophagus is exposed.

the greater curvature of the stomach. It extends from the apex of the hiatus down to the gastroesophageal junction and along the greater curvature of the stomach for 3 cm. Electrocautery is used to mark the site of proposed myotomy (Fig. 20.8). The myotomy is started by using a laparoscopic curved dissector to gently separate the longitudinal and circular muscular layers and identify the mucosa. This part of the operation must be meticulously performed. Once all the muscular fibers are separated, the mucosa bulges out. From this point, the myotomy is performed superiorly and inferiorly. In the superior direction, the esophageal muscle is divided by electrocautery using scissors up to the apex of the diaphragmatic hiatus. In the inferior direction, a laparoscopic hook is used to extend the myotomy down to the gastroesophageal junction and for 3 cm along the greater curvature of the stomach. The esophageal muscle is usually thick and white in appearance. Any remaining small septae on the mucosal surface are divided (Fig. 20.9). A Kittner dissector is used to push back the edges of the myotomized esophageal muscle to widen the myotomy and create a lip of the myotomized muscle at the edge of the myotomy. If persistent bleeding occurs from the muscle edges, it can be stopped with electrocautery; In doing so, care should be taken not to cauterize the mucosal surface.

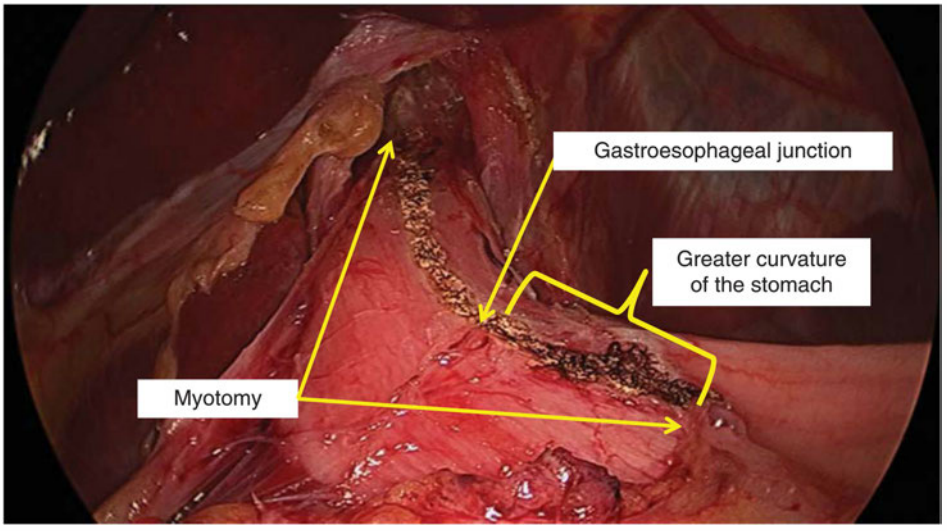


Fig. 20.8. Laparoscopic view of the location of the myotomy marked by the electrocautery down the left lateral surface of the esophagus and along the greater curvature of the stomach. The esophagus has been rotated to the right by the retraction of the vessel loop (not shown).

An upper endoscopy is performed to confirm the extent of the myotomy and to assess for a mucosal leak. During the endoscopy the stomach should readily distend with insufflation of air into the esophagus. The scope should pass easily into the stomach. The stomach is deflated in preparation for a Dor fundoplication.

A Dor fundoplication is placed over the myotomy site. In doing so, the exposed gastric mucosa in the myotomized portion of the stomach is opposed to the exposed esophageal mucosa in the lower portion of the esophageal myotomy. This mucosal to mucosal apposition constructs a thin frenulum between the stomach and the esophagus eliminating outflow resistance and functions as a flap valve to oppose regurgitation.

To construct the flap valve mentioned above, the Dor fundoplication is started inferiorly by placing an interrupted stitch through the left crus at the decussation and the myotomized muscle at the level of the gastroesophageal junction. This begins the folding of the stomach against the esophagus. The second stitch is placed through the lip of the myotomized stomach, through the left crus 1 cm cephalad to the prior stitch and through the left lip of the myotomized esophageal muscle.

Subsequent stitches are placed through the serosa of the stomach, the left crus, and the left lip of the myotomized esophageal muscle at 1 cm intervals until the apex of the myotomy at the diaphragmatic esophageal hiatus is reached. These stitches are placed so that the greater curvature rolls superiorly in line with the middle of the esophageal myotomy site (Fig. 20.10). At the apex of the diaphragmatic esophageal hiatus, a stitch is placed through the stomach, through the apex of the diaphragmatic hiatus, through the esophagus above the myotomy site, back through the apex of the diaphragmatic hiatus, 1 cm to the right, and through the apex of the rolled up stomach and tied. This anchors the superior end of the myotomy and the Dor partial fundoplication to the diaphragm and avoids the formation of the esophageal diverticulum in the apex of the myotomy. The stitches are continued down on the right side, suturing the stomach to the right lip of the esophageal myotomy. The last stitch is placed at the level of the gastroesophageal junction in line with the initial stitch placed on the left in the same position. There should be a good distance between the two stitches so that the partial fundoplication has the shape of a triangle.

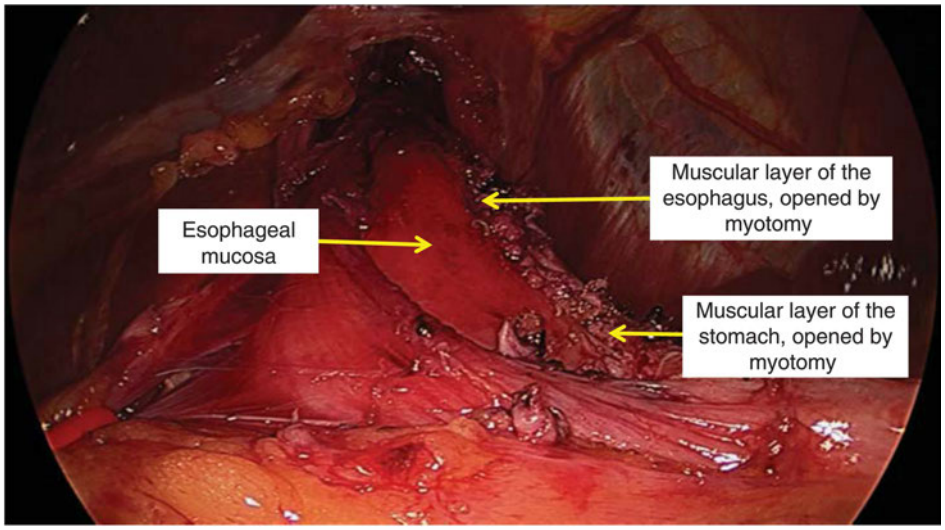


Fig. 20.9. Laparoscopic view of a complete myotomy extending form the apex of the esophageal diaphragmatic hiatus, across the gastroesophageal junction, and down 3 cm along the greater curvature of the stomach.

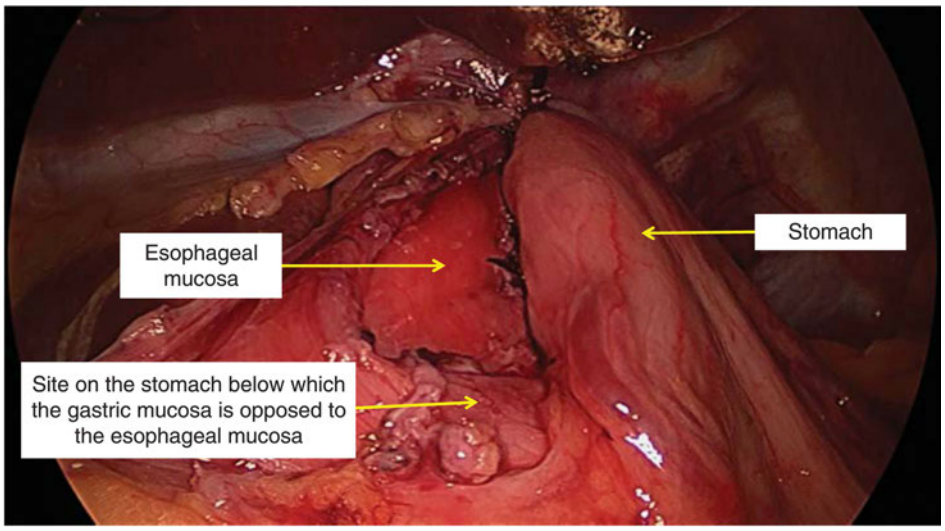


Fig. 20.10. Laparoscopic view showing the stomach attached to the left side of the myotomy form the gastroesophageal junction up to the apex of the esophageal hiatus. The mucosa of the gastric myotomy site has already been opposed to mucosa of the esophageal myotomy site.

At this point in the operation, a second endoscopy is performed to assure easy access into the stomach and to rule out any torsion of the repair. A retroflexed view of the gastroesophageal junction should show an edematous but well-formed Dor fundoplication with a frenulum formed by the apposition of the gastric and the esophageal mucosa in the lower portion of the myotomy site.

The vessel loop is removed, and the stomach and esophagus are rolled back to their normal anatomic position. The Dor fundoplication lies in the left lateral position, covers 180 degrees of the esophageal surface, and does not distort the normal plane of the stomach (Fig. 20.11). The remnants of the short gastric vessels remain in their normal anatomical position with no evidence of torsion or twist of the stomach (Fig. 20.12). A nasogastric tube is not placed in the stomach as patients with achalasia rarely swallow excessive amounts of air. The steps of the laparoscopic myotomy and Dor fundoplication are summarized in Table 20.1.

The patient is hospitalized the night of surgery. The day after surgery, the patient is given a clear liquid breakfast and a full liquid lunch. If taken well, the patient is discharged home. The patient is instructed to continue on a full liquid diet for 2 weeks. A clinical evaluation is then performed and the patient, if doing well, is advanced to a soft diet with instructions to progress to a regular diet as tolerated.

Management of a Failed Heller Myotomy

A failed Heller myotomy is suspected when a patient has persistent or recurrent dysphagia after the procedure.

Persistent dysphagia is usually due to one of the following technical problems:

1. The myotomy may have been too short or fibers of the lower esophagus were left intact. It only takes a few intact fibers to cause dysphagia.
2. The Dor fundoplication may be twisted, causing narrowing of the gastroesophageal junction.
3. An injury to the esophageal mucosa by the electrocautery may have caused a stricture.

Recurrent dysphagia occurring after a symptom-free interval can be due to the following:

1. Reapproximation and healing of the myotomized muscle.
2. Poor transit of medications causing a distal esophageal drug stricture.
3. Formation of an esophageal stricture from severe gastroesophageal reflux.
4. The presence of an esophageal malignancy.

A thorough evaluation must be performed in patients with recurrent dysphagia and includes the reports of the initial detailed history, review of the operative report and the reports of the initial barium swallow, upper endoscopy, esophageal manometry, and pH monitoring. The treatment options depend on the etiology of the dysphagia. Pneumatic dilatation can be used to widen a narrowed esophagus caused by the healing of the myotomy site or a stricture from severe reflux esophagitis. The latter is usually of temporary benefit and the former may require a redo-myotomy for relief. A cautery-induced nonreflux stricture can be managed by a mucosal plasty. Patients with severe unrelenting dysphagia from a persistent or recurrent reflux stricture usually require a vagal-sparing mucosal stripping esophagectomy (described further in this chapter).

Peroral Endoscopic Surgical Myotomy

In 2009, Haru Inoue described an endoscopic Heller myotomy that has engendered considerable enthusiasm. This new endoscopic technique is performed under general anesthesia. A small mucosal opening is made

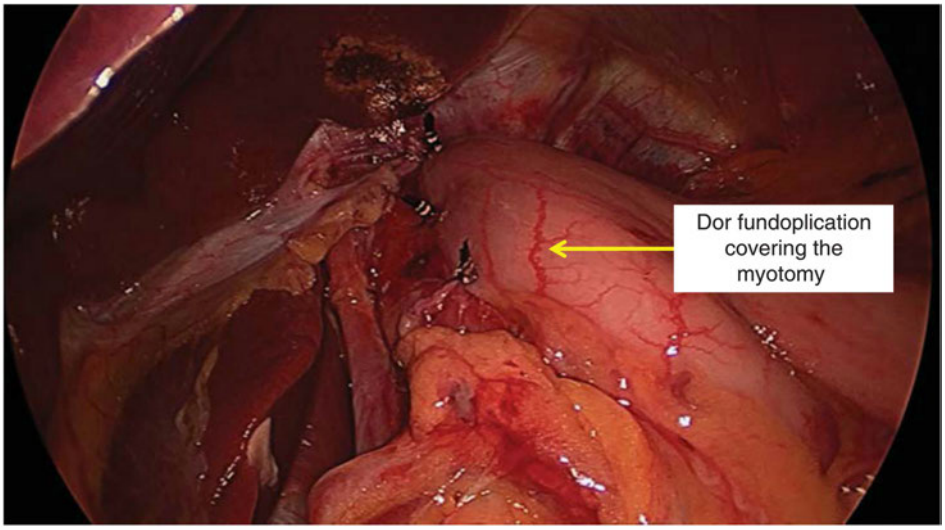


Fig. 20.11. Laparoscopic view of a Dor fundoplication with the stomach in its normal plane and the left lateral myotomy site covered by the Dor fundoplication.

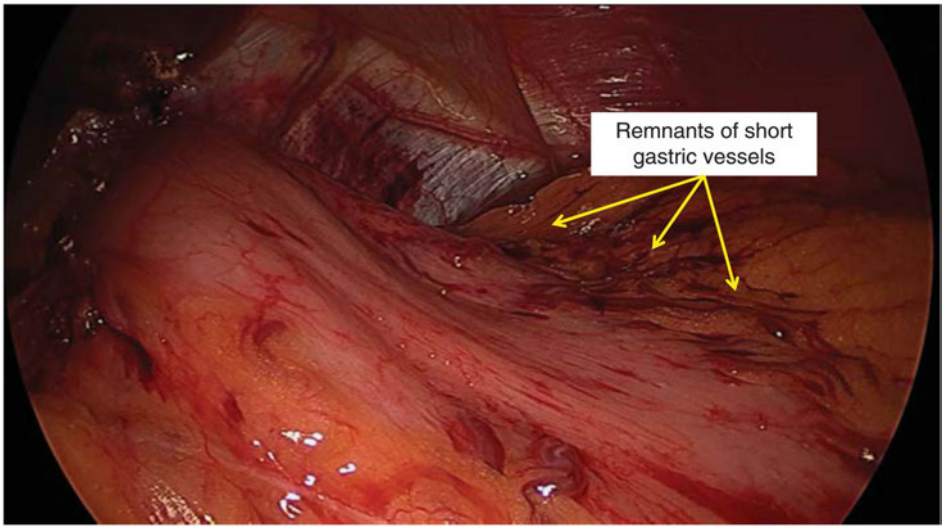


Fig. 20.12. Laparoscopic view of the short gastric vessels after the Dor fundoplication. The remnants of the short gastric vessels remain in their normal anatomical position with no evidence of torsion or twist of the stomach.

in the mid-to-distal esophagus and a submucosal tunnel is developed. The tunnel is extended down across the gastroesophageal junction and onto the stomach. A myotomy of the circular muscle fibers is performed. The length of myotomy is about 8 cm, 6 cm in distal esophagus, and 2 cm beyond to the gastroesophageal junction in the stomach. The small opening in the esophageal mucosa is closed with an endoscopic clip. A smooth passage of an endoscope through the gastroesophageal junction is confirmed at the end of the procedure. Early reports on the outcome of this procedure show a significant improvement in the complaint of dysphagia and a reduction in the resting lower esophageal sphincter pressure. To date, no serious complications of the procedure have been reported. Further studies on long-term improvement of the dysphagia, the degree of the postoperative reflux, and comparison with traditional therapies are expected before wider use of this technique is recommended.

Esophagectomy for Treatment of Achalasia

If the axis of the esophagus is straight, an esophagomyotomy will provide good relief of dysphagia and will allow the esophagus to empty by gravity. In patients with end-stage achalasia who have a sigmoid-shaped megaesophagus of greater than 10 to 11 cm in diameter (Fig. 20.13) retention of food and saliva can occur in the tortuous segment. In these patients, an esophagomyotomy is likely to be of little benefit.

In such patients, we recommend that the esophagus be removed. Our preferred technique of removal is a mucosal stripping vagal-sparing esophagectomy. This procedure was developed to avoid the need for mediastinal dissection and the morbidities associated with standard esophagectomy by preserving the vagal nerves. The procedure straightens the esophageal axis to improve emptying.

Steps of Mucosal Stripping Vagal-Sparing Esophagectomy

Mucosal stripping vagal-sparing esophagectomy removes the esophageal mucosa while preserving the esophageal muscular wall and the vagal plexus. By these means, it avoids mediastinal bleeding from the enlarged arteries associated with a megaesophagus and maintains vagal integrity to eliminate postoperative dumping and diarrhea. Gastrointestinal continuity is re-established with a gastric conduit constructed after performing a highly selective gastric vagotomy along the lesser curvature of the stomach.

The procedure is started by performing an upper midline abdominal incision. The right and left vagal nerves are identified, circled with a tape, and retracted to the right. A highly selective proximal gastric vagotomy is performed along the lesser curve of the stomach from the incisura to the gastroesophageal junction. The left gastric artery is left intact. The gastric conduit is constructed by using a GIA stapler to form a tube, 3 cm wide, along the greater curvature of the stomach.

A neck incision is made along the anterior border of the left sternocleidomastoid muscle and the strap muscles are exposed. The omohyoid muscle is divided at its pulley, and the sternohyoid and sternothyroid muscles are divided at their manubrial insertion. The left carotid sheath is retracted laterally and the thyroid and trachea medially. The left inferior thyroid artery is ligated laterally as it passes under the left common carotid artery. The left recurrent laryngeal nerve is identified and protected. The esophagus is dissected circumferentially from the left neck with the scissors pointed in the direction of the apex of the right chest, to avoid injury to the right recurrent laryngeal nerve. The esophagus is encircled with a Penrose drain.

Returning to the abdomen, the gastroesophageal junction is divided with a GIA stapler. A 1 cm opening in the mid-portion

Table 20.1 Steps of Laparoscopic Myotomy and Dor Fundoplication

Laparoscopic Heller myotomy
1. Place the patient in the reverse Trendelenburg position with the lower extremities spread apart in leg holders
2. Insert five laparoscopic ports
3. Take down of the short gastric vessels along the upper third part of the greater curvature of the stomach
4. Separate the left lateral wall of the esophagus from the left crus and mobilize the left lateral wall of the esophagus from the deccusation to the apex of the diaphragmatic esophageal hiatus
5. Dissect the gastroesophageal fat pad from the esophagus in a left-to-right direction
6. Place a vessel loop through a small window created in the base of the fat pad and retract the gastroesophageal junction to the right
7. Mark the myotomy site using electrocautery from the apex of the diaphragmatic esophageal hiatus across the gastroesophageal junction and 3 cm down the greater curvature of the stomach.
8. Perform the myotomy with electrocautery using scissors in the superior direction up to the level of the diaphragmatic hiatus and a laparoscopic hook in the inferior direction down to 3 cm beyond the gastroesophageal junction along the greater curvature of the stomach
9. Free the edges of the myotomy from the mucosa
10. Perform upper endoscopy to assess the extent of the myotomy and presence of a leak
1. The stomach should easily insufflate from the esophagus
2. The scope should pass without resistance into the stomach
Laparoscopic Dor fundoplication
1. Start inferiorly by placing an interrupted stitch through the left crus at the decussation and the myotomized muscle at the level of the gastroesophageal junction to fold the stomach against the esophagus
2. Place the second stitch through the lip of the myotomized stomach, through the left crus 1 cm cephalad to the prior stitch, and through the left lip of the myotomized esophageal muscle
3. Place the subsequent stitches through the serosa of the stomach, the left crus, and the left lip of the myotomized esophageal muscle at 1 cm intervals until the apex of the myotomy at the diaphragmatic esophageal hiatus. The greater curvature should roll up in line with the middle of the esophageal myotomy site
4. Place the apical stitch by passing the needle through the stomach, the diaphragmatic hiatus, the esophagus above the myotomy site, the diaphragmatic hiatus 1 cm to the right, the stomach, and tie
5. Continue the stitches down on the right side, suturing the stomach to the right lip of the esophageal myotomy
6. Place the last stitch at the level of the gastroesophageal junction in line with the initial stitch placed on the left in the same position. There should be a good distance between the two stitches so that the partial fundoplication has the shape of a triangle
7. Perform an upper endoscopy to assess the ease of access to the stomach and the configuration of the fundoplication

of the staple line is made to flush the esophagus with Betadine solution. The remaining portion of the staple line is opened and a vein stripper is passed up into the cervical esophagus in the neck.

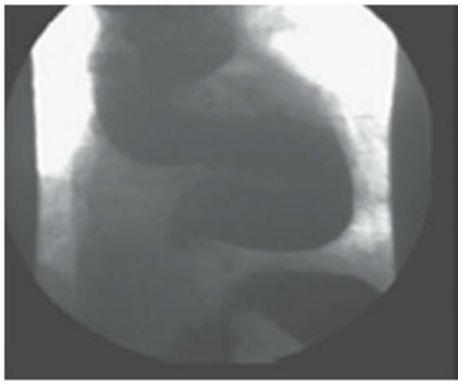


Fig. 20.13. Barium esophagram showing a sigmoid-shaped megaesophagus.

Returning to the neck, the muscle on the left lateral surface of the cervical esophagus is excised over 180 degrees of the esophageal circumference to expose the mucosa. The mucosa is circumferentially separated from the remaining intact esophageal muscle and divided transversally. The thin tip of the stripper is exchanged for a mushroom head, and the esophageal mucosa is secured tightly around the stripping cable using an endo-loop and umbilical tape. Sufficient length of one limb of the umbilical tape should remain to use as a trailer.

Returning to the abdomen, the stripper is pulled in an inferior direction to strip the esophageal mucosa from the muscular wall of the dilated esophagus. The vagus nerves remain intact along with the esophageal muscular wall. The muscular tube is inspected for bleeding and if present, a lap pad soaked with epinephrine diluted 1:10,000 is pulled through to pack the muscular tunnel to stop the bleeding. A Foley catheter containing 90 cm^3 of fluid in the balloon is used to dilate the muscular tunnel. The gastric conduit is pulled up through the muscular tunnel into the neck and anastomosed to the cervical esophagus.

The procedure avoids a thoracotomy as well as the risk of severe bleeding that can occur in these patients with a transhiatal resection. The placement of the gastric conduit within the native esophageal muscular tube straightens the esophageal axis.

DIFFUSE ESOPHAGEAL SPASM

Definition

Diffuse esophageal spasm is a primary esophageal motility disorder occurring in about 3% to 5% of patients with esophageal motility disorders. It is characterized

by >20% simultaneous contractions in the esophageal smooth muscle and symptoms of dysphagia and chest pain. It differs from achalasia in that the lower esophageal sphincter relaxes on swallowing.

Etiology and Symptoms

The cause of diffuse esophageal spasm is uncertain. It is seen in all ages but most commonly in patients >50-year old. The symptoms of dysphagia and chest pain vary in intensity from mild to severe, last for seconds to minutes, are precipitated by solids or liquids, and can occur during the fasting state.

Diagnosis

Videoesophagram

The videoesophagram in patients with diffuse esophageal spasm can show a "corkscrew" esophagus (Fig. 20.14) but due to the intermittent nature of diffuse esophageal spasm, the finding is only seen in 30% of patients.

Esophageal Manometry

The primary motility abnormality in diffuse esophageal spasm is the occurrence of simultaneous contractions following more than 20% of the wet swallows (Fig. 20.15). Contrary to achalasia, normal peristaltic waves are always seen between episodes of simultaneous contractions. The lower esophageal sphincter is usually normotensive but can be hypertensive and, in contrast to achalasia, relaxes normally on swallowing. Other less common manometric findings are long duration, multipeaked contractions, and nonswallow-induced spontaneous contractions.



Fig. 20.14. Barium esophagram showing a corkscrew esophagus in a patient with diffuse esophageal spasm.

pH Monitoring

Some patients with gastroesophageal reflux disease can masquerade as diffuse esophageal spasm. Esophageal pH monitoring is helpful to identify these patients.

Treatment

Nonsurgical Treatment

Smooth-muscle relaxants and anticholinergics can decrease high-amplitude contractions but do not consistently relieve chest pain. Pneumatic dilation or injection of Botulinum toxin helps some patients with diffuse esophageal spasm, particularly those with dysphagia, a hypertensive lower esophageal sphincter and documented delay in the esophageal emptying on barium swallow.

Surgical Treatment

Symptomatic patients with diffuse esophageal spasm who do not respond to medical therapy can be considered for surgical therapy. The key to success of surgical therapy is careful analysis of the percentage of pathologic simultaneous nonperistaltic esophageal contractions. If the patient has more than 75% pathologic simultaneous nonperistaltic esophageal contractions, a myotomy is likely to improve the dysphagia. If the patient has <75% pathologic simultaneous nonperistaltic esophageal contractions, a myotomy is less likely to improve the dysphagia and may make the dysphagia worse by destroying the existing normal peristaltic contractions. In principle, a myotomy is more likely to improve dysphagia and less likely to add to the degree of the dysphagia when the number of normal peristaltic contractions is below 25%.

The surgical treatment of diffuse esophageal spasm is similar to that of achalasia except for the length of the myotomy. In diffuse esophageal spasm, the myotomy starts above the level on the esophagus where the simultaneous nonperistaltic esophageal contractions occur on manometry and extend down the esophagus across the lower esophageal sphincter and 2 cm over the stomach. Sparing the lower esophageal sphincter from the myotomy has been reported to result in esophageal obstruction with the accumulation of liquid and food above the sphincter and the potential for esophageal rupture.

NUTCRACKER ESOPHAGUS

Definition

Nutcracker esophagus is a primary esophageal motility disorder characterized by high amplitude peristaltic esophageal contractions (>180 mmHg) of long duration (>6 seconds). These contractions are usually asymptomatic but can be associated with chest pain. Dysphagia is uncommon.

Etiology and Symptoms

The etiology of nutcracker esophagus is unknown even though it is the most common motility disorder reported. Consequently, when not associated with symptoms it may be a normal variant of esophageal contractility, that is an amplitude above the 95% threshold used to make the diagnosis.

Diagnosis

Videoesophagram

The videoesophagram in patients with nutcracker esophagus is commonly normal.

Esophageal Manometry

Esophageal manometry in patients with nutcracker esophagus is characterized by peristaltic waves of high amplitude (>180 mmHg) and prolonged duration (>6 seconds) (Fig. 20.16). Lower esophageal sphincter can be normotensive or hypertensive but shows complete relaxation with wet swallows.

pH Monitoring

Some patients with gastroesophageal reflux disease can masquerade as nutcracker esophagus. Esophageal pH monitoring is helpful to identify these patients. If gastroesophageal reflux disease is present, the motility pattern is secondary rather than primary, and therapy should be directed toward the correction of the abnormal reflux.

Treatment

Nonsurgical Treatment

Patients with nutcracker esophagus are treated only if symptoms are present.

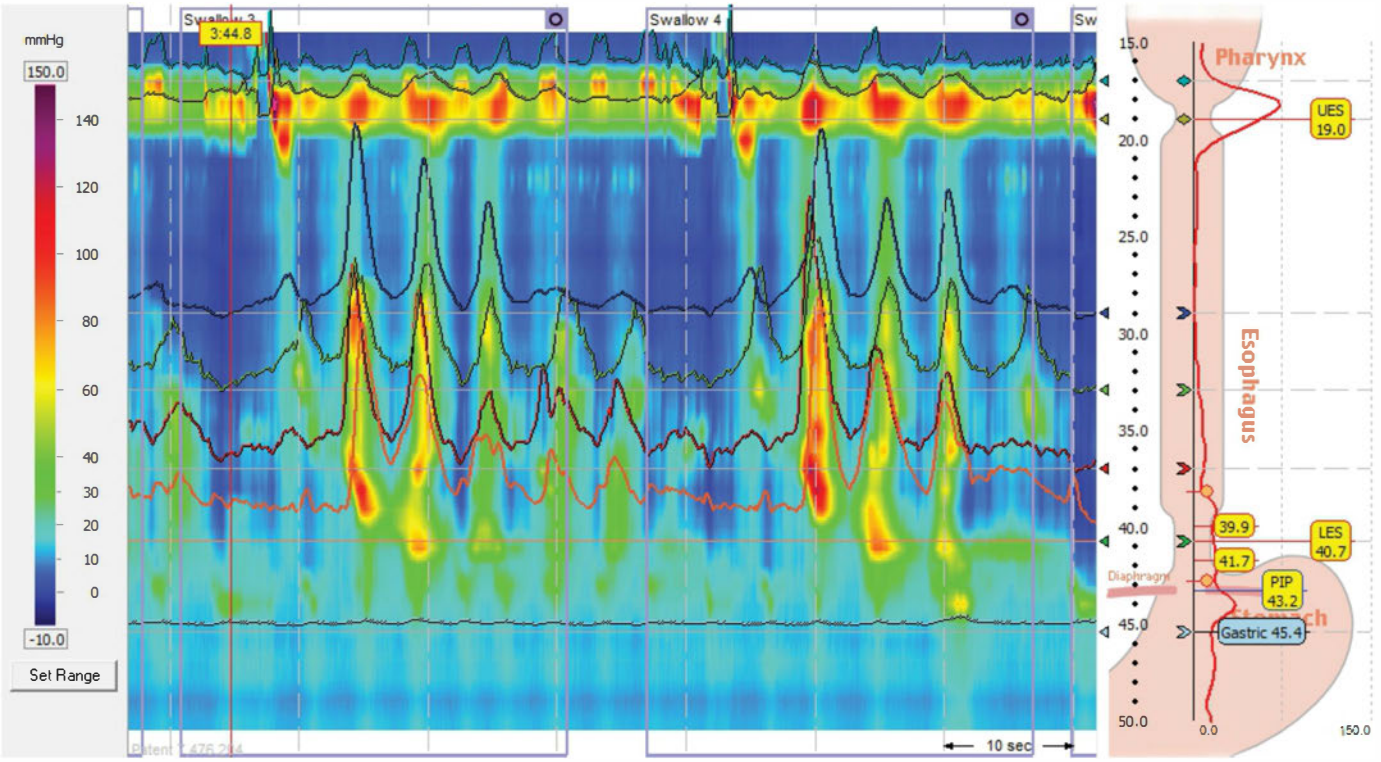


Fig. 20.15. High-resolution manometry showing episodes of simultaneous contractions in a patient with diffuse esophageal spasm.

Smooth-muscle relaxants and anticholinergics have been tried with minimal benefit. Pneumatic dilation or Botulinum toxin is of some help but the benefit is unpredictable.

Surgical Treatment

Surgery may be beneficial to a very select group of patients with nutcracker esophagus who have dysphagia and a hypertensive lower esophageal sphincter. In this situation, a myotomy of the lower esophageal sphincter may improve the dysphagia. It is unlikely that the esophageal body abnormality is contributing to the symptom of dysphagia and should be spared of any surgical intervention.

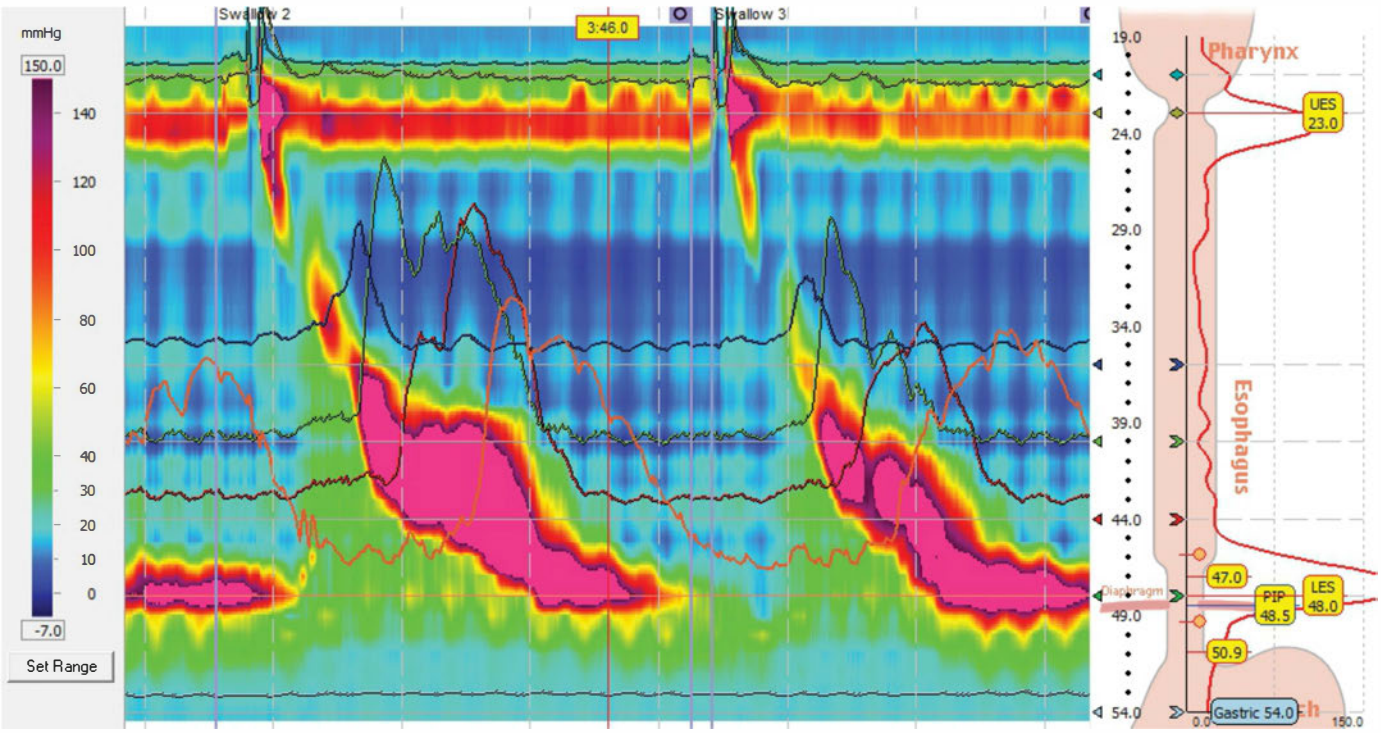


Fig. 20.16. High-resolution manometry showing peristaltic high amplitude of contractions in a patient with nutcracker esophagus.

HYPERTENSIVE LOWER ESOPHAGEAL SPHINCTER

Definition

Hypertensive lower esophageal sphincter is a primary esophageal motility disorder occurring in 7.2% of the patient population referred for motility studies. The diagnosis is made by manometry with the measurement of an abnormally high pressure in the lower esophageal sphincter that completely relaxes. Hypertensive lower esophageal sphincter can occur as an isolated condition or in association with gastroesophageal reflux disease, a paraesophageal hiatal hernia, an esophageal epiphrenic diverticulum, or nutcracker esophagus.

Etiology and Symptoms

Patients with hypertensive lower esophageal sphincter are a heterogeneous group with regard to symptoms and etiology. Symptoms may be caused by the motor abnormality in the primary form of the disease, or, in its secondary form, by gastroesophageal reflux, the altered geometry of the cardia that occurs with a paraesophageal hernia, an epiphrenic diverticulum, or a nutcracker esophagus. Dysphagia and chest pain that can masquerade as heartburn are the most common symptoms in patient with primary disease. Of those who complain of heartburn only 26% will have increased esophageal acid exposure on pH monitoring. This indicates that those symptomatic patients with normal esophageal acid exposure could be easily misdiagnosed as gastroesophageal reflux disease.

Diagnosis

Esophageal manometry is used to identify patients with hypertensive lower esophageal sphincter. The manometric definition of a hypertensive lower esophageal sphincter varies among motility laboratories and depends on the method used to read the lower esophageal sphincter pressure. If the pressure is measured at the peak of the diaphragmatic excursion, a pressure around 45 mmHg is usually the threshold, above which the sphincter is considered to be hypertensive. If the pressure is measured at the respiratory inversion point, where the effect of the diaphragmatic movement is minimal, then 26 mmHg is the threshold. Patients with primary hypertensive lower esophageal sphincter have higher sphincter pressures compared with those with secondary disease.

The criteria used in our motility laboratory to diagnose a patient with primary hypertensive lower esophageal sphincter require the following:

1. Elevated lower esophageal sphincter pressure at rest (mean pressure >26 mmHg)
2. Complete relaxation of the lower esophageal sphincter on swallowing (residual pressure <8 mmHg)
3. Peristaltic esophageal contractions of normal amplitude in the body of the esophagus.

Treatment

Nonsurgical Treatment
Medical management of the primary hypertensive lower esophageal sphincter is largely limited to drugs like nifedipine or sildenafil, and botulinum toxin to reduce lower esophageal sphincter pressure. Often, transient manometric improvement occurs with these drugs but is not associated with an equivalent clinical improvement. Dilatation has been tried with disappointing results. Acid suppression therapy relives reflux symptoms, but their effect on dysphagia and chest pain is minimal.

Surgical Treatment
The surgical approach to a symptomatic patient with hypertensive lower esophageal sphincter is tailored based on the patient's clinical findings. In symptomatic patients with primary hypertensive lower esophageal sphincter, a myotomy of the lower esophageal sphincter is done. Like in achalasia a Dor fundoplication is added to prevent reflux.

In patients with secondary disease, that is hypertensive lower esophageal sphincter associated with gastroesophageal reflux disease, or a paraesophageal hiatal hernia, a Nissen fundoplication is performed. We have shown in this situation that a Nissen fundoplication, without a myotomy of the lower esophageal sphincter, relieves the patient's heartburn, dysphagia, and chest pain. In symptomatic patients with hypertensive lower esophageal sphincter and an epiphrenic diverticulum, the diverticulum is resected and a myotomy is done from the level of the diverticulum across the lower esophageal sphincter and down to the stomach for about 2 to 3 cm and a Dor fundoplication is added. In symptomatic patients with hypertensive lower sphincter and nutcracker esophagus, a myotomy is limited to the lower esophageal sphincter.

INEFFECTIVE ESOPHAGEAL MOTILITY DISORDER

Definition

Ineffective esophageal motility is defined by a >30% frequency of low amplitude contractions <30 mmHg in the distal esophagus following wet swallows.

Etiology and Symptoms

Ineffective esophageal contractions in the esophageal body are seen in patients with gastroesophageal reflux disease and are thought to be due to inflammatory damage of the esophageal muscle. Heartburn and regurgitation are common symptoms. Dysphagia is usually mild but when severe, suggests an anatomical problem such as esophagitis or a peptic stricture.

Treatment

Nonsurgical Treatment
The nonsurgical treatment consists of acid suppression therapy for the gastroesophageal reflux disease.

Surgical Management
The most effective surgical procedure to treat gastroesophageal reflux disease is a Nissen fundoplication. A well-formed Nissen fundoplication can impose an esophageal outflow resistance up to 20 mmHg without producing dysphagia in patients with normal esophageal motility. If a patient has global peristaltic esophageal contractions <20 mmHg in the distal third of the esophagus, then a Nissen fundoplication is likely to result in dysphagia and partial fundoplication should be done. If an antireflux procedure is performed early in the disease, the inflammation in the esophageal body resolves along with improvement in the amplitude of esophageal contractions, and the dysphagia will also improve over time. Late in the disease, the muscle damage may be irreversible and the esophageal motility is unlikely to improve. In this situation, the combination of global contractions of <20 mmHg in the distal esophagus and the presence of a partial fundoplication is likely to result in dysphagia. If the dysphagia is severe, the patient is likely to have end-stage reflux disease and a vagal-sparing esophagectomy might be a better choice.

Scleroderma

Definition
Progressive systemic sclerosis, known as scleroderma, is the most common

secondary esophageal motility disorder and the most common collagen vascular disease affecting esophageal function. The disease is characterized by the fibrosis of small arteries and arterioles causing ischemic injury and fibrosis of the organ involved. There is deposition of collagen throughout the body but most commonly seen in the skin, lungs, kidneys, and gastrointestinal tract. The esophagus is the most commonly affected portion of the gastrointestinal tract. Patients with esophageal involvement usually develop gastroesophageal reflux disease manifested by heartburn or dysphagia within 2 years of their diagnosis of scleroderma. The reflux disease is often severe and complications such as persistent erosive esophagitis, long-segment strictures, and Barrett's metaplasia can develop. The mechanism of reflux in scleroderma patients is a destroyed lower esophageal sphincter that allows the reflux of the gastric juice and an aperistaltic esophagus that is unable to clear the refluxed gastric juice. Gastric dysmotility is frequent in scleroderma and can result in delayed gastric emptying which contributes to the reflux problem. Further, scleroderma patients have impaired salivary production and are unable to chemically neutralize the refluxed gastric juice by swallowing.

Etiology and Symptoms

The etiology of the arteriolar fibrosis is unknown. Histopathologic findings in the esophagus are the deposition of collagen in the connective tissue and smooth muscle atrophy. These tissue changes result is an atonic esophagus with a defective or absent lower esophageal sphincter, resulting in significant gastroesophageal reflux often with peptic stricture formation.

Diagnosis

The diagnosis of scleroderma is based on physical exam, serum antinuclear antibody levels, and histology. The possibility of esophageal involvement should be suspected in all patients with scleroderma. The involvement of the esophagus is suggested by the symptoms of reflux and dysphagia and confirmed by esophageal motility.

Videoesophagram

A videoesophagram demonstrates an atonic or "lead pipe" esophagus, often with an associated hiatal hernia.

Esophageal Manometry

Esophageal manometry demonstrates ineffective, low-amplitude esophageal contractions involving the distal two-thirds of the esophagus and a defective lower esophageal sphincter manifested by a low sphincter pressure and a short overall and/or abdominal length.

Treatment

Nonsurgical Treatment

Nonsurgical therapy for the esophageal component of scleroderma includes aggressive acid suppression therapy with proton-pump inhibitors and dilation of esophageal strictures.

Surgical Treatment

Esophageal reflux in the setting of scleroderma is difficult to treat surgically. The combination of severe esophageal dysmotility, destroyed lower esophageal sphincter, and delayed gastric emptying leads to gastroesophageal reflux that is also refractory to medical therapy. The two surgical options for such patients are as follows:

1. For patients who do not have a destroyed esophagus, a Roux-en-Y gastric preserving esophageal–jejunal bypass can be done to prevent the reflux of the gastric juice into the esophagus.
2. For patients with a destroyed esophagus from an intractable long stricture or Barrett's esophagus with high-grade dysphagia, a vagal-sparing esophagectomy can be done.

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EDITOR'S COMMENTS

When it comes to diseases of the esophagus, there probably is no greater authority than the senior author of this chapter, Tom DeMeester. His contributions to our knowledge of esophageal pathology are legendary, and the DeMeester score for quantitating acid reflux is the standard used the world over. In this chapter, Banki, a former trainee of DeMeester along with her mentor provide an outstanding summary not just of the various motility disorders but give us detailed step-by-step procedures involved in their operative treatment. This is especially true in their vivid description of the mucosal-stripping vagal-sparing esophagectomy. I must admit that this procedure seems to have some significant advantages to transhiatal esophagectomy when performed for benign disease, and I am surprised that it has not been more widely accepted and utilized.

Perhaps, the most critical issue when dealing with esophageal motility disorders is the decision when to operate or more specifically if an operation is more likely than not to fix a functional problem. When performing nonextirpative surgery the "proof is in the pudding." That is, if the patient's symptoms are not improved or alleviated then the patient has undergone an unnecessary procedure. It is clear that certain motility disorders respond better to operation than others and patients should be well informed prior to operation that the result is never "perfect." For the patient with achalasia, the well-performed myotomy simply allows the esophagus to drain by gravity without obstruction from an incompletely relaxed or hypertensive lower esophageal sphincter but the esophagus obviously remains aperistaltic. I am intrigued by the concept of the peroral endoscopic surgical myotomy and more widespread use of this procedure awaits further outcome data both short-term, specifically morbidity, and long-term functional results.

There is no question that laparoscopic Heller myotomy has, for the most part, replaced the open thoracotomy or thoracoscopic approach and has been shown to be highly efficacious in well-selected patients. More recently, robotic Heller myotomy has been used in some centers also with excellent results. The authors favor the routine use of a partial fundoplication as part of definitive treatment for achalasia since most of these patients have some element of acid reflux either preoperative or after the myotomy is performed. I agree with the authors and in experienced hands either the Dor or Toupet partial fundoplication adds little extra time to the operative procedure. The most important aspect of the operation is the completeness of the myotomy being sure to divide every last fiber of circular muscle as well as making sure the length of the myotomy is adequate. Dealing with a failed myotomy presents significantly greater challenges.

LRK

21 Esophageal Conduits and Palliative Procedures

Jenifer L. Marks and Ara A. Vaporciyan

Esophageal cancer is on the rise throughout the world and esophageal adenocarcinoma is now the most common type of esophageal malignancy requiring surgical resection in the United States. Esophageal resection for benign and malignant cases mandates conduit creation to reestablish gastrointestinal continuity. There are a number of options for conduit creation, including the stomach, colon, jejunum, and a skin tube. Selection depends on a number of patient factors as well as a surgeon's personal experience with any of the potential conduits. There are advantages and disadvantages for each conduit, and none has proven superiority in all settings. The stomach is the most widely used conduit and therefore has the most data on outcomes. The colon and jejunum are used less often, but are the first choice in some clinical settings such as when the stomach is not available. Skin tube reconstruction has also been described as an esophageal replacement, although utilized uncommonly today. Today's thoracic surgeon must be familiar with all the conduit choices and understand the technique and implications of using each method for esophageal replacement.

STOMACH

The stomach remains the most commonly used conduit for esophageal reconstruction. It is a large, durable organ with abundant native blood supply and it can be partially or completely tubularized and advanced into the chest or neck for anastomosis to the proximal esophagus. The most important aspect of preparing the stomach as an esophageal conduit is protecting the blood supply for the conduit, the right gastric and right gastroepiploic artery (Fig. 21.1). The remainder of the blood supply to the native stomach is sacrificed during mobilization of the stomach. This includes the short gastric vessels, the pancreaticogastrics, the left gastric artery, and the left gastroepiploic artery. The stomach when used as a conduit is able to survive on the blood supplied by the right gastroepiploic artery because of the stomach's rich submucosal vascular networks. Radiation can damage these networks and one must always take this into account when operating on patients who have received prior radiation. Alternative conduits should be considered if the stomach does not appear viable after mobilization.

Several aspects of protecting the right gastroepiploic and its vessel of origin, the gastroduodenal artery, deserve mention. When mobilizing the greater curvature of the stomach, care must be taken to palpate the right gastroepiploic artery and stay a safe distance away from it when dividing the gastrocolic omentum with any energy source. Also, the stomach should not be grasped by the greater curvature itself but by the body of the stomach, using gauze or the nasogastric tube to maintain tension. When dissecting the distal greater curvature at the gastrocolic ligament, one must be aware of the gastroduodenal and gastroepiploic artery at all times as this region frequently has redundant folds of omentum making it difficult to identify the artery or the wall of the colon. Careful separation of these folds may be necessary to ensure preservation of the vessel and avoid injury to the colon wall.

The right gastroepiploic artery normally terminates approximately two-thirds of the way along the greater curvature. It is rare for there to be actual continuity in this region of the stomach with the left gastroepiploic or the short gastric vessels but infrequently a collateral arcade may be found within the gastrocolic ligament. This arcade supplies the area distal to the termination of the right gastroepiploic artery and, when preserved, can enhance the blood supply to the distal portion of a conduit. Any major injury or accidental ligation of the gastroduodenal or gastroepiploic artery requires the use of an alternative conduit for esophageal replacement.

The decision to tubularize the gastric conduit is surgeon dependent. Some surgeons prefer not to perform any tubularization, advancing the stomach essentially intact into the chest. While this is an acceptable approach, most surgeons feel that such a conduit leaves the patient more prone to reflux and difficulty with gastric emptying. Therefore, most surgeons advocate some type of tubularization of the stomach.

When performing an intrathoracic anastomosis, the tubularization of the conduit can be done in a number of ways. Some prefer to completely tubularize the stomach while in the abdomen after transection of the distal margin of the specimen. A 4-cm wide conduit is prepared by sequential firings of the GIA stapler with either 4.8 or 3.5 mm loads, depending on the thickness of the organ. The staple line begins at the incisura and ends at the cardia with the proximal extent based on the length of conduit needed and the location of the tumor. The stomach is uncurled and straightened with each successive firing allowing it to lengthen. Oversewing of this staple line with interrupted Lembert stitches for hemostasis as well as to reinforce the staple line may be added. Tubularizing the entire remaining stomach in such a manner provides the greatest possible length allowing the conduit to reach easily into the neck for a cervical anastomosis. For an intrathoracic anastomosis, the proximal end of the now tubularized stomach must be attached temporarily to the distal end of the specimen so that it can be delivered into the chest during the thoracotomy.

An alternate method, when creating a conduit for a thoracic anastomosis, still begins in the abdomen. One or two firings of the stapler are begun at the incisura extending toward to cardia, to begin

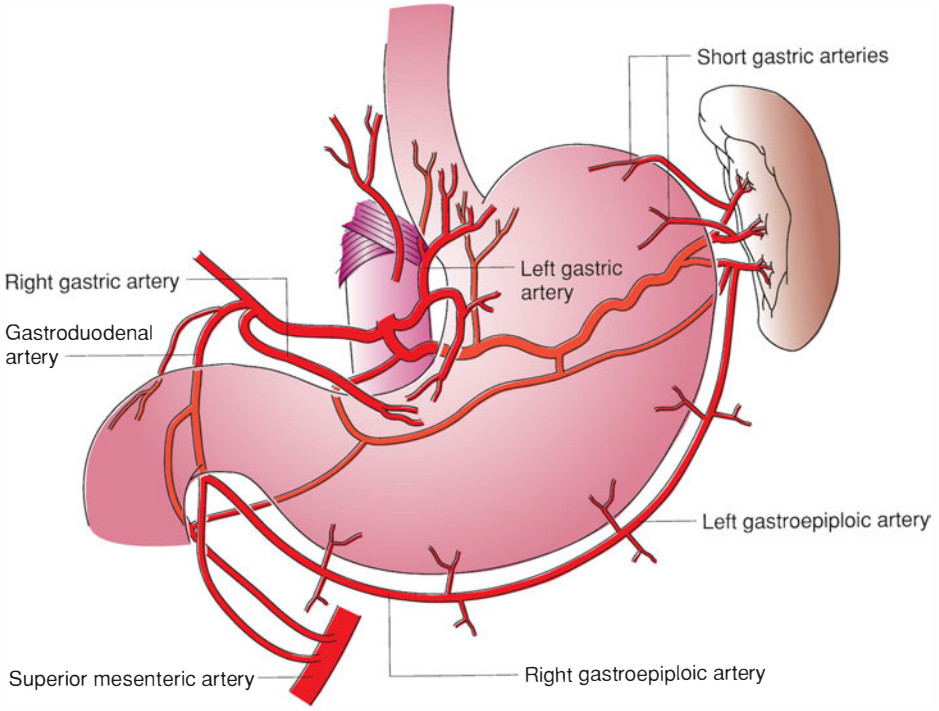


Fig. 21.1. Blood supply of stomach. (From Fiser SM. The ABSITE Review. 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008.)

the tubularization of the conduit. The stomach is again uncurled as the staplers are applied. The specimen and the conduit remain attached. After the abdomen is closed and the chest is opened, the specimen and conduit are brought through the hiatus. The anastomosis is completed at an appropriate location on the greater curvature of the conduit. The gastric staple line is then completed allowing the specimen to be removed.

Using the stomach for esophageal replacement requires only one anastomosis. As such, it is less technically demanding than any other of the conduit choices. Disadvantages to using the stomach as a conduit include dumping syndrome, early satiety, chronic reflux of gastric contents into the remaining esophageal segment, and the risk of aspiration. Dumping syndrome and early satiety tend to improve with time and lifestyle/dietary modifications. Reflux, regurgitation, and aspiration persist for the remainder of the patient's life and methods to prevent these sequelae must also be maintained.

Necrosis of the gastric conduit is a rare but very serious and sometimes fatal complication. The most proximal aspect of the conduit is most at risk for ischemia. Early postoperative acidosis or other signs of clinical deterioration should prompt esophagoscopy to determine the viability of the conduit. Focal areas of mucosal ischemia can be watched closely as they may progress. An anastomotic leak may also be an early sign of partial conduit necrosis. When gastric necrosis is suspected, the conduit and patient's status must be monitored closely and the conduit resected if there is any evidence of full thickness necrosis. We do not recommend immediate reconstruction in this situation and a diversion should be performed by stapling off what, if anything, is left of the gastric conduit and constructing a cervical esophagostomy.

COLON

The colon is another option for esophageal reconstruction. Either the left or the right colon can be used for conduit creation. Both techniques employ the use of a portion of the transverse colon. Previous abdominal operations or colon pathology may exclude the use of the colon as an esophageal replacement. To assess the suitability of the colon, the surgeon must evaluate its blood supply and the presence of any pre-existing pathology preoperatively. The middle colic vessels provide the blood supply for a right colon conduit, whereas the ascending branch of the left colic provides the supply for a left colon conduit. The transverse colon portion of a left-sided conduit is supplied by the marginal arteries or the arc of Riolan. The colon as a conduit provides more length than is possible with the stomach and can be carried up as high as the pharynx when the blood supply is adequate. Some also believe the colon to be the better choice of conduit for patients with an extended life expectancy since the storage function of the stomach can be preserved. The consequences of an anastomotic leak are often less with a colon conduit since the anastomosis is always placed in the neck. It has also been suggested that over time the colonic conduit is able to generate propulsive forces to help with motility. The colon is resistant to peptic strictures and other late complications seen with a gastric conduit. Finally, the use of the colon maintains the reservoir function of the stomach in those in whom the stomach is not involved with the primary malignancy.

Preoperative evaluation of the colon is required in any patient in which the colon may be utilized as a conduit. Colonoscopy should be performed to rule out any primary colon lesions, significant diverticular disease, or vascular lesions. Alternatively, a double-contrast barium enema may also be obtained. A CT angiogram or MRA should also be obtained to verify the patency of major vessels on which the conduit may be based. A bowel regimen is also recommended preoperatively; a liquid diet for 48 hours prior to surgery is usually sufficient in clearing the colon of large amounts of stool which could hamper the procedure. Oral antibiotics no longer are routinely used as part of the bowel preparation having been supplanted by intravenous antibiotics at the time of surgery.

The procedure is begun by mobilizing either the right or left colon, depending on which is being used. Care should be taken not to injure any vessels in the transverse mesocolon when separating the colon from the omentum. Depending on which colon is to be used, the appropriate feeding vessel is identified via transillumination. The middle colic vessel is used for a right colon graft and the ascending branch of the left colic for a left colon transposition. While the left colon can be utilized as an isoperistaltic conduit, the right colon is positioned in an antiperistaltic orientation. The right colic artery is divided and the conduit is based on the middle colic allowing the proximal colon to be rotated into the neck. After deciding which vessels will be used, the mesentery is opened on the appropriate side of the middle colic vessel. An estimate of the length needed for transposition can be obtained by measuring the distance

from 5 cm below the xiphoid to the angle of the jaw with an umbilical tape. This length is sufficient if the conduit traverses the posterior mediastinum. If the retrosternal route is to be used, the measurement should extend to the earlobe.

Once the length of the graft is determined, temporary vascular isolation is obtained by placing bulldog clamps on the vessels that will be divided. Following this temporary occlusion, it is important to observe the graft for 5 to 10 minutes to assess for any evidence of ischemia or venous congestion. If the graft appears viable, then the appropriate vessels may be ligated. For a left colon graft, the middle colic artery is ligated at its origin from the superior mesenteric artery. For an antiperistaltic right colon graft, the right colic artery is ligated near its origin from the superior mesenteric artery. For an isoperistaltic right colon graft, an uncommon selection because of limitations in length, the middle colic artery is ligated, thus basing the graft on the right colic and advancing the distal end of the colon graft into the neck. Finally, the length of the colon needed is once again confirmed and then the colon is transected 1 cm beyond the remaining marginal vessels. The graft should now be free and ready for transposition.

The graft should be placed retrogastric and can transverse the mediastinum using either a posterior, anterior (retrosternal), or subcutaneous route. Delivery of the conduit through the mediastinum is facilitated using a sterile plastic bag (such as an intraoperative ultrasound bag) into which the conduit is placed for passage. This helps minimize damage to small vessels. When using the retrosternal or the subcutaneous route the hemimanubrium, adjoining clavicular head and medial first rib on the side of the anastomosis should be removed to provide a more natural lie for the conduit and some additional room. In addition, pressure on the mesocolon or conduit at this location can compromise either the arterial inflow or venous outflow and lead to congestion and potential anastomotic problems. Strictures will also form in this location if there is a constant pressure point on the conduit from the manubrium, clavicle, and first rib.

Advantages to using the colon as a conduit relate mostly to the ability to preserve the stomach. Since the stomach is not a part of the reconstruction, it can be resected for oncologic purposes if necessary such as when dealing with a Siewert type III gastroesophageal junction tumor with extensive involvement of the stomach. In addition, gastric reservoir function of the stomach can be maintained by leaving the native distal stomach in place. This can potentially aid in the resumption of normal dietary habits. Finally, the colon is also a barrier to reflux, providing some protection to the proximal esophagus from bile or acid reflux.

Disadvantages to using the colon relate mostly to the technical aspects related to the procedure. It can be a lengthy operation that requires three anastomoses (esophagocolic, gastrocolic, and colocolic). Also, up to 30% of patients ultimately will require reoperation after a colon conduit due to redundancy or dilation of the conduit. The development of significant redundancy presents as poor emptying of food into the stomach leading to stasis and dilation within the chest. The optimal time to prevent this complication is at the time of the initial operation by limiting any redundancy of the conduit and performing the cologastric anastomosis high on the posterior wall of the stomach. The length of conduit chosen and its final positioning within the chest must be precise and without excess to avoid a redundant conduit. Any additional length should be placed below the diaphragm and the conduit secured in such a position during the initial operation.

Additional complications associated with the use of the colon include those found with a gastric conduit. These include ischemia or necrosis of the conduit, anastomotic leaks, and stricture formation. Esophagocolonic anastomotic leaks require reoperation for repair less often than an esophagogastric anastomosis perhaps due to the less erosive nature of colonic secretions and its cervical location. Small leaks will often heal without operative intervention. The risk of gastrocolic reflux and peptic colitis can be reduced by placing the cologastric anastomosis high on the posterior portion of the stomach near the greater curvature. Additionally, a short segment of the conduit (8 to 10 cm) is positioned and secured in the infradiaphragmatic position near the hiatus to act as a barrier to reflux from the abdominal to the thoracic portion of the conduit.

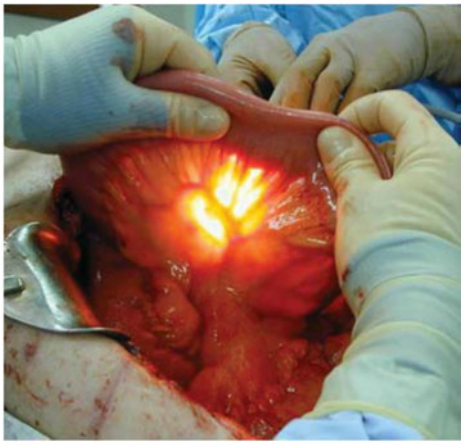
JEJUNUM

The jejunum that is naturally resistant to both acid and bile offers a third choice for esophageal reconstruction. The abundant length and mesenteric vasculature of the jejunum make it another option for both long and short segment esophageal replacement. The vascular anatomy of the jejunal mesentery allows the graft to be supercharged using intrathoracic or cervical vessels anastomosed to jejunal vessels. This improves the arterial supply and venous drainage for the graft allowing greater reach.

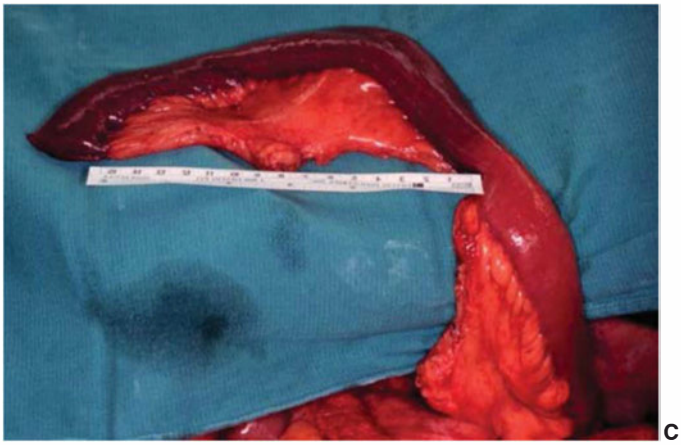
Long Segment

Unlike the colon or the stomach, the existing vascular anatomy of the jejunum limits the length of a graft based solely on any one pedicle. Therefore, the technical aspects of creating a long segment jejunal interposition graft for esophageal replacement are centered around enhancing the collateral blood supply to the segment being transposed into the chest or neck. The proximal jejunum is evaluated and the arcading pattern of its vasculature assessed via transillumination (Fig. 21.2A). To use a segment of jejunum, the vascular arcade needs to have several main branches off of the superior mesenteric artery that feed the jejunum in continuous arcades. A segment with three to four sequential branches must be identified. These three to four arcades generally provide a length of up to 50 cm for transposition. The proximal two or three branches are divided while the distal vessel is left intact (Fig. 21.2B). Opening of the mesentery between the arcades may be necessary to straighten the conduit for better function and reach. The distal arcade that is spared functions as the pedicle for the jejunal flap (similar to the left colic artery for a left colon conduit). Once the segment of jejunum is transposed to the neck, the proximal divided arcade vessel is used for a microvascular anastomosis, providing "supercharging" of the graft. The in situ vessels frequently used for this anastomosis are the internal mammary artery and vein, inferior thyroid vessels, or branches of the thyrocervical trunk. This provides inflow and drainage for the proximal third of the graft. The distal third is supplied by the sparred distal arcade vessel, whereas the middle third through submucosal collaterals from the proximal and distal third (Fig. 21.2C). The retrocolic, retrogastric route is preferred for passage of the conduit and the passage may be either via the posterior, retrosternal, or subcutaneous route.

As mentioned previously as part of the discussion of the colon conduit in the retrosternal position, removing part of the manubrium, the clavicular head, and the medial insertion of the first rib on the side of the esophagojejunostomy will provide room for the conduit and avoid points of bony compression. Similarly, using a sterile plastic bag to pass the conduit through the mediastinum will protect the thin mesentery and exposed vessels from injury.



A



C



B



D

Fig. 21.2. (**A**) Transillumination of vascular supply of proximal jejunum. (**B**) Distal vessels left intact for microvascular anastomosis, the proximal vessels have been ligated. (**C**) Ischemic proximal third of graft prior to microvascular anastomosis, distal graft perfused by native jejunal vessels. (**D**) Indicator graft after skin closure.

Once the graft is positioned in the mediastinum, the microvascular anastomosis is constructed. About 4 to 6 cm of the proximal end of the jejunum is separated from the remainder of the graft while maintaining its attachment to the recently supercharged vascular arcade. This segment of the graft will eventually be exteriorized as an indicator graft to monitor postoperatively the patency of the vascular anastomoses (Fig. 21.2D). The indicator graft is then ligated and excised on the fifth or sixth postoperative day. The esophagojejunal anastomosis is then completed as a single- or double-layer handsewn anastomosis. The nasogastric tube is passed into the mid-conduit under direct vision and palpation to avoid conduit perforation.

Two anastomoses remain for completion of the procedure. The jejunogastric anastomosis is performed on the posterior aspect of the stomach, high on the wall to avoid a saddle bag deformity and poor gastric emptying. The jejunojejunal anastomosis can be completed in a standard fashion. If the stomach was previously resected, a roux-en-Y jejunojejunostomy should be performed.

Short Segment

A short segment jejunal interposition graft may be used to replace the distal aspect of the esophagus. This procedure has been referred to as the Merendino procedure initially described to correct severe gastroesophageal reflux. Unlike the long segment interposition, a short segment interposition requires mobilization of only one vascular arcade. In addition, this one arcade remains in situ as a pedicle for the graft obviating the need for any additional revascularization. The segment of jejunum is selected on the quality of the vascular arcade as well as its ability to reach the distal end of the transected esophagus. The bowel at the proximal and distal margin of the segment is transected, the mesentery mobilized, and then the segment passed in a retrogastric position. Care is taken to ensure isoperistaltic orientation of the segment. The esophagojejunal anastomosis resides in the chest and is constructed using an ELEA stapler

or can be handsewn if the exposure is adequate. The jejunogastric anastomosis is completed using standard techniques, with care taken to avoid redundancy of the conduit in the upper abdomen. The distal jejunojejunal anastomosis is performed using standard techniques. The mesenteric defect should be closed with care taken not to put any tension on the mesentery of the interposed segment.

Advantages to utilizing a jejunal conduit include the range of lengths that can be obtained and the vascular arcade, which can be easily augmented with cervical vessels and a microvascular anastomosis. The jejunum is perhaps more resistant than the colon to bile and acid reflux that may be present at the jejunogastric anastomosis. The jejunum is a closer size match to the native esophagus than the colon and maintains its peristalsis to help with propulsion of contents through the conduit. That peristalsis can also minimize reflux at the esophagojejunal anastomosis as well. Finally, similar to the colon, the lack of reliance on the stomach allows additional stomach to be removed if required for oncologic purposes or preservation of the stomach to maintain its reservoir function.

Disadvantages are all related to the technical aspects of the operation. The reliance on a microvascular anastomosis and submucosal collateral vessels, the duration of the operation, and the additional anastomosis required to reestablish jejunal continuity after creation of the conduit all can lead to potential complications. Additionally, obesity can lead to shortened fatty mesenteries that limit the reach of the jejunum and greatly increase the technical complexity of the operation.

SKIN TUBE

In some situations, when none of the commonly utilized conduits are available for reconstruction, a skin tube may be constructed and used as an esophageal conduit. The procedure has been described in the literature and can provide acceptable results when there are no other options for reestablishing gastrointestinal continuity. The basic technique employs a myocutaneous flap that is rolled into a tube with the skin oriented internally. Myocutaneous flaps have been based on the latissimus dorsi, trapezius, and deltoid/pectoralis muscles. A free flap from the anterolateral thigh has also been described. Methods for creating the conduit vary based on the donor site but technically a skin tube is created first. The position of the tube is always subcutaneous. Once its viability is established the proximal and distal anastomoses to the esophagus and stomach or jejunum (if no stomach remains) are constructed.

This conduit is always a conduit of last resort and as such is used only in the most extreme circumstances. When compared with other conduits, it has no natural advantages other than its availability when all other conduits have been exhausted. Its disadvantages clearly derive from its many technical complexities but are also secondary to the existing comorbidities of the patient.

CONDUIT POSITION

For an intrathoracic esophageal anastomosis, the posterior mediastinal route is the only option. However, when the conduit must reach the cervical esophagus three distinct routes are available: the posterior mediastinal, the anterior mediastinal (also referred to as the retrosternal), and subcutaneous route. In general, if the posterior mediastinum is available, this is preferred as it provides the most natural position and shortest route for any esophageal conduit. An exception to this can exist when a long segment jejunal or a supercharged colon conduit is utilized since exposure and construction of the microvascular anastomoses can be easier after retrosternal positioning and resection of the manubrium. If previous resections or disease in the posterior mediastinum have created a hostile environment, the anterior mediastinum (retrosternal) is the next choice. This still provides the conduit with bony protection and is generally an easy plane to develop in the absence of a prior sternotomy. As mentioned previously, this route should be accompanied by the removal of a portion of the manubrium, the clavicular head, and the medial insertion of the first rib on the side of the esophagoconduit anastomosis to avoid compression of the conduit at the thoracic inlet. Dysphagia is more common with a retrosternal conduit due to angulation as food passes from the posterior mediastinum, the native location of the proximal esophagus, to the anterior mediastinum, where the new conduit is positioned. This angle and subsequent dysphagia can persist despite removal of the clavicular head and hemimanubrium. However, failure to remove the bony segments can increase its frequency and also lead to compression of the conduit and eventual stenosis.

The subcutaneous route is most frequently used when the posterior and anterior mediastinum are unavailable. While the subcutaneous route can still provide adequate resumption of enteral intake, the conduit is frequently visible, especially during oral intake and can be unsightly. In addition, the angulation from the posterior mediastinum for the cervical esophagus to the subcutaneous position of the conduit is at its most extreme. This can lead to an even greater degree of dysphagia than seen with the anterior mediastinal route. Finally, the length of conduit required is the greatest when using this route and care must be taken to ensure that adequate conduit length is available.

PALLIATIVE PROCEDURES

Fifty to sixty percent of patients with esophageal cancer will present with unresectable locally advanced or metastatic disease. In addition, other patients may have technically resectable disease but their diminished performance status or coexisting comorbidities preclude surgical resection. While many of these patients without metastatic disease may be offered curative chemoradiotherapy, many may eventually be referred for palliation of disabling symptoms. Pain, dysphagia, bleeding, perforation, and fistula formation are the most common symptoms requiring palliation. The palliative techniques available broadly can be grouped into endoscopic, radiation, and surgical techniques.

Of all the symptoms that may need to be addressed dysphagia is the dominant one. Palliative radiotherapy or chemoradiotherapy provides relief of dysphagia in the majority of patients presenting with advanced disease. The necessity of additional procedures will depend on individual patient responses, the severity of symptoms that remain, and the life expectancy of the patient. Bleeding, perforation, and fistulas are the next most commonly addressed sequelae that require intervention. Pain is largely addressed with medication and usually does not require invasive procedures for palliation. An exception to this is pain secondary to direct invasion of the spine, which can sometimes be addressed with external beam radiation.

Endoscopic Palliative Procedures

There are a number of endoscopic means of providing palliation for patients with advanced esophageal cancer. Dysphagia is the most common symptom requiring endoscopic therapy for palliation. Sometimes, others problems such as bleeding, perforation, and fistulas can also be addressed endoscopically. The treatment modalities range from simple mechanical

or balloon dilation with or without stent placement, and ablative therapies. Often these modalities are used in combination with one another or with systemic chemotherapy or radiotherapy to provide the most durable palliation possible.

Dilatation

Simple mechanical dilation of an esophageal stricture or localized site of disease is an effective means of relieving dysphagia. Savary dilators or balloon dilation can be used in attempts to achieve a patent lumen. Dilation should be performed under guidance, either directly via the endoscope or fluoroscopically. Blind Maloney dilation in diseased tissues increases the risk of perforation and is not recommended. Repeat dilation is almost always necessary as dilation without a stent is usually of limited durability. Dilation is often necessary as an initial step in endoscopic therapy to achieve a lumen of adequate size for the application of other treatments, such as stent placement and ablative therapies.

Stents

The most common indication for an esophageal stent is following a dilation procedure when it is used to maintain the patency of the strictured or obstructed esophagus that has been opened. Perforation and fistula formation are other common indications where the stent can limit the contamination that can result. When deployed for dysphagia, stents can be used to treat both endoluminal obstruction as well as external compression caused by mediastinal involvement. Stents should be sized so that there is at least 2 cm of length on either side of the stricture allowing the proximal and distal ends of the stent to anchor in normal tissue. This can be difficult for distal or very proximal lesions. For distal esophageal strictures, the proximal portion of the stent should be at least 2 cm above the stricture to anchor the stent. The distal portion of the stent will be free within the stomach. Stenting across the GE junction in this fashion will lead to wide open reflux and regurgitation. This must be discussed with the patient prior to stenting as these symptoms may be as troublesome as the dysphagia. Proximal lesions can be even more difficult to address with a stent. If the stent must lie at or near the cricopharyngeus patients may experience a continuous persistent globus sensation, which can limit patient tolerance. In addition, the limited length of uninvolved esophagus proximal to the obstruction prevents an adequate landing zone making stent migration a common problem.

There are a number of stents available for use. Familiarity with each stent, their delivery system, and their available lengths and widths prior to performing the procedure is vital. Two major varieties of stents exist: the self-expanding metal systems and plastic systems. Self-expanding metal stents (SEMS) are composed of a variety of metal alloys and are available either covered or uncovered. Other features that must be selected include the length and width as well as whether the stent will be deployed from the distal end or the proximal end of the delivery system. Both covered and uncovered versions provide almost immediate relief of dysphagia but each can also cause some degree of discomfort due to their radial force. Most of the SEMS available in the United States are made of nitinol and are better tolerated than stainless steel stents secondary to their gradual expansion, although they may not provide the same rapid relief of dysphagia. Concomitant mechanical dilatation after deploying the stent can speed relief of the dysphagia. The covered stents provide a barrier to tissue in-growth and also address contamination in cases of perforation or a fistula. Alternatively, solid plastic stents are also available and are designed for easier removal. The delivery systems utilized by these types of stents tend to be bulkier but they also provide rapid relief of dysphagia.

Stent removal may sometimes be necessary if patient discomfort is excessive or if stent migration occurs. The initial SEMS were not meant to be retrievable and can fracture if removal is attempted, especially after significant tissue or tumor in-growth. The more recent versions of SEMS and the plastic stents come with a loop of suture on the proximal end that is meant to be grabbed and use to collapse and remove the stent.

Stent migration is a problem with all devices. Displacement most commonly occurs secondary to normal esophageal motility. Alternatively, if the stent is positioned low on the tumor, it can propel the stent distally as it grows. It occurs more commonly with covered or plastic stents, as these resist incorporation by tissue or tumor in-growth. Regardless of which type of stent is used, all stents should be monitored periodically for migration.

When attempting to cover a perforation or fistula, a covered stent is necessary. After stent placement, all patients should be evaluated with a contrast study to confirm complete exclusion of the defect. Complete exclusion may not be necessary if a perforation can be diminished significantly, intrathoracic drainage is adequate, and the patient's life expectancy is limited. However, if complete exclusion must be obtained then a second stent may be needed. Even after complete exclusion is obtained, the patient should continue to be monitored as displacement of the stent can lead to a recurrence of the defect. Proximal defects are especially difficult to control and bridling of the stent around the nasal septum is one way to try and prevent stent migration (Fig. 21.3). One end of an umbilical

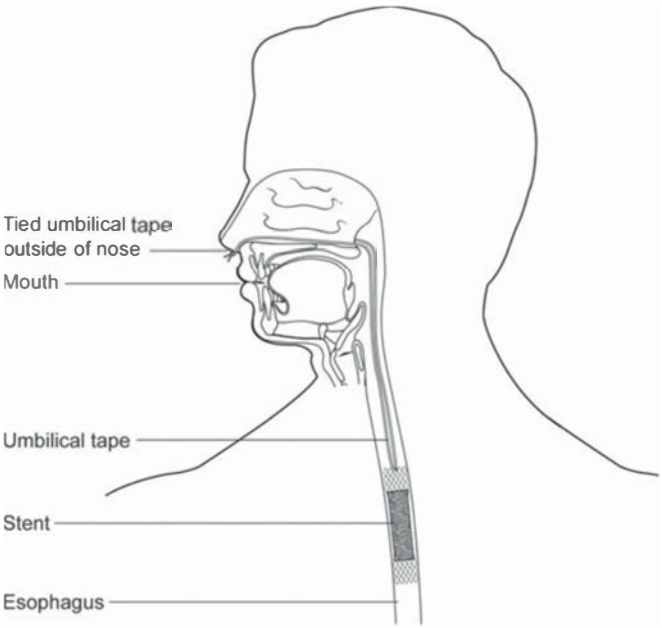


Fig. 21.3. Bridling of the esophageal stent to prevent migration. (Courtesy of Shanda Blackmon.)

tape is passed through the nostril and out the mouth, then through the stent and the stent is positioned. The other end of the umbilical tape is then brought out the other nostril and the ends secured at the nasal septum. This can prevent distal migration of the stent but still must be monitored to prevent pressure necrosis on the patient's nasal septum from the umbilical tape.

The technique of placing kissing stents, two stents in neighboring organs/vessels, can be used for fistulas between the esophagus and the trachea or rarely the esophagus and the aorta. Endovascular stenting of the thoracic aorta to treat an esophagoaortic fistula has been described and can provide a temporary solution to a life-threatening problem. For a malignant fistula in this setting, we generally do not recommend aggressive surgical intervention due to the overall poor prognosis. When treating a tracheoesophageal fistula with kissing stents, the esophagus should be stented first, followed by the trachea. This helps to limit contamination of the airway and sometimes the esophageal stent is all that is necessary to control the fistula. As always with any esophageal perforation, a collection within the chest or mediastinum should be drained with open or interventional drainage.

Ablative Therapies

Photodynamic therapy (PDT) and laser ablation are both commonly used to palliate symptoms of advanced esophageal cancer. Ablative therapies can provide rapid palliation of both bleeding and dysphagia and are generally well tolerated by most patients. Proximal and distal/GE junction tumors are best treated with this modality due to the issues with stenting. The proximity of the airway to mid-esophageal lesions can make this area prone to complications when ablative therapy is used here.

Photodynamic Therapy

PDT is performed by first administering a photosensitizing agent intravenously. Two days following the injection the region of the involved esophagus is exposed to a low power activating laser light delivered through the endoscope. It is believed that malignant cells take up the photosensitizing agent at a higher concentration than normal cells. This makes them more sensitive to the effects of the sensitizing agent after exposure to the activating wave length of light. Upon exposure to the 630 nm wavelength laser, the sensitizing agent is converted to a toxin with direct effects mediated via oxygen free radicals. Tumor cells along with their microvasculature are destroyed and the tumor becomes ischemic. Repeat endoscopy is performed 2 days after laser exposure, necrotic tumor debrided, and another exposure to the laser can be performed at this time. The amount of total light exposure determines the degree of tumor necrosis.

Increased tumor necrosis with more light exposure must be balanced against the risk of perforation, which occurs in approximately 2% of patients. Side effects such as chest pain, odynophagia, and pleural effusion occur frequently. Development of a tracheoesophageal fistula is a much more serious event that is reported in 5% to 30% of patients. This occurs more often when the mid esophagus is being treated due to its proximity in the airway. Esophageal stricture is also reported in up to 10% of patients after PDT. Patients must also be instructed to avoid prolonged exposure to sunlight and bright indoor lights after receiving the photosensitizing agent as extensive skin burns have been described due to cutaneous photosensitivity. Mild skin irritation often results from even incidental light exposure. It is recommended that patients remain completely protected from sunlight and bright indoor light in addition to eye protection from their initial injection and for at least 4 to 6 weeks after the injection. Sun block is not effective in preventing this type of burn. These restrictions must be fully discussed with the patient prior to planning their injection. After an initial PDT treatment, patients may experience tumor regrowth within 2 to 3 months and may require further therapy. PDT may be repeated or used in combination with other treatments.

Laser Ablation

Direct thermal ablation of esophageal lesions for palliation has been traditionally performed with the neodymium:yttrium-aluminum-garnet (Nd:YAG) laser. This laser is easily applied using a fiberoptic delivery catheter placed through the endoscope. The depth of penetration may be limited and associated with minimal collateral damage by carrying out precise ablation of malignant tissue. Concurrent mechanical debridement with rigid endoscopic forceps can hasten tumor removal especially when dealing with large bulky tumors. Most patients will require several treatments and even then the duration of relief provided is limited usually to 1 to 2 months. Laser therapy can be combined with other modalities for a more durable response. For example, the Nd:YAG laser can be used initially to open a severe stricture or total obstruction followed by stent placement.

Serious complications include perforation (8%), tracheoesophageal fistula (6%), and death (5%). Fire is also possible if high concentrations of oxygen are used in close proximity to the laser. This should always be kept in mind when using a laser near the airway.

External Radiation and Brachytherapy

Many patients with unresectable locally advanced esophageal cancer will receive external beam radiation therapy as part of their initial treatment. External radiation in combination with chemotherapy is associated with response rates as high as 80% achieving significant symptom relief with perhaps one-third of those patients achieving a complete response. Unfortunately, despite the initial response, up to half of these patients will recur locally and may eventually need palliation. For these patients, radiation therapy options for palliation are limited and alternative options described above will need to be considered. For those patients who require palliation and who did not previously receive radiation therapy, external beam radiation should be considered and is generally well tolerated.

Endoluminal brachytherapy is another option for local treatment of lesions causing symptoms and may be an option even in those patients who previously received external beam radiation. Both low- and high-dose endoluminal regimens are available with the low-dose regimens being better tolerated but taking a longer time to achieve symptom palliation. High-dose rate brachytherapy (HDR-ILRT) consists of 12 to 20 Gy given in one or two fractions and is the preferred method of delivery due to high success rates and the rapidity of symptom improvement. Esophageal stricture and fistula formation are potential complications of brachytherapy. The incidence of fistula formation ranges from 0% to 12%, while strictures are more common and can range from 10% to 35%.

Surgical Palliation

Open surgical procedures as palliation for esophageal cancer should be considered only after other less invasive means have failed. Symptoms such as bleeding, dysphagia, perforation, and fistulas have all been addressed surgically. Dysphagia and minimal bleeding are unlikely to require surgical intervention as these symptoms often respond to chemotherapy and radiation or endoscopic treatment. Life-threatening

bleeding due to erosion into a major vessel can be treated surgically if endoscopic techniques are unsuccessful. However, before such heroic actions are attempted, a frank conversation with the patient and their family members must take place regarding the overall prognosis. These palliative procedures are associated with significant morbidity, and if the estimated life expectancy is shorter than the time required to recover from the operation, there may not be any meaningful palliation achieved.

Operative repair often entails vessel ligation (if the vessel is not the aorta), debridement and closure of the esophagus, if possible, and soft tissue coverage of the fistula. Alternatively, a stent may be used to control the esophageal defect while the vessel is ligated surgically. An esophagoaortic fistula presenting with massive hemoptysis can be treated with both esophageal and endovascular stents; however, the overall prognosis needs to be weighed heavily against such heroic palliative interventions.

When faced with severe dysphagia, there are rare scenarios where the disease does not respond to medical or endoscopic therapy. In these cases, surgical bypass may be the only option for palliation or obstruction. Retrosternal gastric bypass can be performed with the intent of allowing the patient to recover the ability to eat. Unfortunately, such procedures have morbidity rates of up to 60% and mortality rates of 5% to 10%. They require long recovery times and may not actually improve the patients' quality of life depending on the postoperative course, lifestyle modifications necessary after undergoing such a procedure, and the natural history of their advanced disease.

Another form of surgical palliation for severe dysphagia is in the provision of enteral access. Either a jejunostomy or gastrostomy can be performed in patients with esophageal cancer. The timing of placement depends on each individual patient's disease course, their wishes, and tolerance of treatment. Some patients need feeding access upon presentation due to weight loss and disease-related malnutrition, others require it during induction therapy due to ongoing dysphagia, odynophagia, or oral intolerance, surgical candidates usually have one placed during the resection, and finally some patients have it placed toward the end of their disease course as they lose the ability to sustain themselves with oral intake alone. Regardless of when it is placed, laparoscopic, open, or endoscopic methods can be used and are well tolerated. Jejunostomy tube placement is preferred over a gastrostomy if the patient possibly is a surgical candidate due to concerns of compromising the gastroepiploic artery with gastrostomy tube placement.

When perforation of the esophagus occurs in a patient with incurable esophageal cancer, the role of surgical palliation must be carefully considered. The perforation may be primary from the tumor itself or secondary and related to endoscopic interventions. While most can be approached with placement of a stent, some are not amenable to such an approach or stent bypass may fail to achieve isolation of the leak. The first decision to be made in these cases relates to the patient's performance status and life expectancy. Patients with otherwise treatable disease should be treated aggressively with the hope of being able to complete the treatment regimen and prolong life. However, patients with limited life expectancy whose symptoms can be managed without surgical intervention should not be subjected to heroic options where only limited palliation can be achieved. In these cases, a stent sometimes accompanied by a mediastinal or intrathoracic drain can control the mediastinal spillage and avert sepsis in most situations at least temporarily. If the stent is not effective at preventing sepsis and the patient is not likely to succumb to his disease within 4 to 6 weeks, one can consider proximal diversion. It should be considered as a last choice for control of sepsis only. The overall prognosis should be considered when dealing with these difficult situations. Resection should not be considered in patients with uncontrollable local and metastatic disease.

Malignant tracheoesophageal fistula formation is another clinical situation in which some form of palliation may be indicated. This problem is largely addressed endoscopically with esophageal and/or tracheal stent placement to control pulmonary sepsis and ongoing aspiration. These patients generally present in poor condition and palliation via the least invasive means possible is the best option. Primary tracheal repair with muscle coverage and proximal diversion or resection can be considered in some rare situations. A posttreatment fistula with no evidence of disease, for example, a high squamous cell carcinoma with a complete clinical response to therapy and delayed fistula formation, might warrant consideration of surgical intervention if the patient is in good medical condition.

CONCLUSION

The modern esophageal surgeon has multiple options available for both reconstruction of the esophagus and palliation of symptoms. Blind adherence to one method is no longer an option as there clearly are clinical scenarios where alternative approaches may be beneficial to the patient. Even if one is not competent at performing each of the various options, a treating physician should at least possess an understanding of the available options and use that knowledge to guide the patient to the best treatment.

SUGGESTED READINGS

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EDITOR'S COMMENTS

As the authors point out gastric transposition, far and away, is the first choice for reconstitution of the gastrointestinal tract following esophageal resection. If for whatever reason the stomach is not available to use as a conduit, the colon remains the next best choice but introduces an additional level of complexity, in that it requires three anastomoses and is accompanied by a higher incidence of complications. A long segment jejunal graft introduces even more complexities, in that supercharging with a microvascular anastomosis is usually required.

Mobilizing the stomach for use as a conduit requires careful preservation of the right gastroepiploic artery. If a patient has had previous upper abdominal surgery, confirmation of a patent gastroduodenal artery and right gastroepiploic artery should be carried out prior to esophagectomy. Preparation of the gastric conduit should involve some level of tubularization of the stomach both to achieve adequate length and also to facilitate gastric emptying. In addition an emptying procedure, either pyloromyotomy or pyloroplasty should be performed. Ideally, the conduit should reside in the posterior mediastinum as it provides the shortest route to the neck, assuming the anastomosis is to be done in the neck.

The keys to a successful anastomosis include no tension and an adequate blood supply. Care should be taken in handling the gastric remnant to avoid submucosal hemorrhage. As mentioned, if the gastric conduit or colon is brought up via the substernal approach, the ipsilateral hemimanubrium and clavicular head should be removed in order to prevent compression of the conduit. Anastomotic leaks in the neck usually can be managed simply by opening the skin to allow drainage. An intrathoracic leak may be more devastating but usually may be managed conservatively as long as there is adequate drainage.

Palliation of esophageal carcinoma raises a number of issues. As the authors point out, there are a number of options for palliation of the major symptoms with the dysphagia being the most frequent indication for palliation in those patients who are either inoperable or unresectable. Careful consideration should be given prior to subjecting a patient a palliative procedure taking into account the predicted length of survival. The occasional patient may be a candidate for the ultimate palliative procedure, substernal gastric bypass, but as the authors point out this procedure is associated with significant morbidity. For the rare patient, who presents fairly early with a tracheoesophageal fistula but has remained in reasonable shape, substernal gastric bypass may be the procedure of choice to allow both for bypass of the fistula and protection of the lungs in addition to allowing the patient to resume oral alimentation.

LRK

22 Excision of Esophageal Diverticula

Philip A. Rascoe and W. Roy Smythe

Diverticula of the esophagus are uncommon disorders that are usually classified according to their location (cervical, thoracic, or epiphrenic), their pathogenesis (pulsion or traction), and their morphology (true or false).

The great majority of esophageal diverticula are acquired lesions that occur predominantly in elderly adults. Pulsion, or false, diverticula are the most commonly encountered type of esophageal diverticula. These localized outpouchings lack a muscular coat, and their wall is formed entirely by mucosa and submucosa. Almost all are the result of a functional obstruction to the advancing peristaltic wave, usually caused by an abnormal upper or lower esophageal sphincter. Occasionally, impedance to peristaltic progression may be the result of peptic strictures or localized motility disorders such as spasm. Pulsion diverticula thus occur most commonly at the level of the cricopharyngeus where there is a weak area of the crossing muscle fibers in Killian's triangle or the distal 10 cm of the thoracic esophagus between the inferior pulmonary vein and the diaphragm (epiphrenic location); however, they may also occur within the midthoracic esophagus.

True, or traction, diverticula are usually seen in the middle one-third of the thoracic esophagus in a peribronchial location. These diverticula are the result of paraesophageal granulomatous mediastinal lymphadenitis secondary to disorders such as tuberculosis or histoplasmosis. The ensuing desmoplastic reaction tents the full thickness of the esophageal wall, producing a conical, wide-mouthed true diverticulum. They most frequently project to the right because subcarinal lymph nodes in this area are closely associated with the right anterior wall of the esophagus. These outpouchings are rarely seen in the Western world and are usually of little or no clinical significance except in rare instances when ongoing mediastinal inflammation results in a fistulous communication with the airway or other intrathoracic structures.

ZENKER'S DIVERTICULUM

The British surgeon Abraham Ludlow is credited with the original description of a pharyngoesophageal diverticulum from an autopsy specimen that remains on display at the Royal Infirmary Pathology Museum in Glasgow, Scotland. Almost a century later, the German pathologist Zenker provided a complete clinical and pathologic description of 34 cases. The pathogenesis of this lesion was first suggested in 1926 by Jackson, who proposed that the tonically contracting upper esophageal sphincter (UES) impeded the progress of the swallowed bolus. A localized increase in intraluminal pressure forces the mucosa to herniate through the posterior midline of the inferior pharyngeal constrictor in the anatomically bare area (Killian's triangle) between the oblique fibers of the thyropharyngeus and the horizontal fibers of the cricopharyngeus. The diverticulum deviates away from the rigid vertebrae and usually presents on the left side. The exact nature of this cricopharyngeal motor dysfunction remains unclear, but most commonly an incomplete or incoordinated opening of the UES is present.

CLINICAL PRESENTATION

Zenker's diverticulum is primarily a condition of the elderly and is twice as common in men. Dysphagia for solid food and regurgitation of undigested food are the most common symptoms and are typically present. Halitosis, noisy swallowing or "gurgling" after deglutition, and globus sensation are also common. Aspiration may also result from this condition, and it may manifest as a mild nocturnal cough, morning hoarseness, or new onset adult bronchospasm caused by repeated laryngeal penetration and irritation and, on rare occasion, present as chronic lower respiratory tract infection or even lung abscess. Despite the association among hiatal hernia, gastroesophageal reflux, and Zenker's diverticulum, only a few patients present with severe heartburn and rarely do they require surgical correction of their reflux.

DIAGNOSIS

A barium esophagogram using a lateral or oblique projection usually demonstrates the diverticulum, which can be large and may protrude well into the superior mediastinum (Fig. 22.1). Esophageal manometry adds little information and should not be routinely performed. Endoscopy can be considered but often adds little to the diagnosis. Perforation of the diverticulum can result from aggressive endoscopic examination because the flexible endoscope often enters the diverticulum rather than the true esophageal lumen. Malignant change is possible but is exceedingly rare in these diverticula, with squamous cell carcinoma having been reported in no more than 0.5% of patients. If diverticulopexy is planned, the diverticulum should be carefully palpated to rule out a nodular density in the wall, and an intraoperative endoscopic examination can be performed to evaluate the interior of the sac as there will be no pathologic specimen.

TREATMENT

A small, completely asymptomatic diverticulum in an elderly patient with comorbidities that preclude general anesthesia likely can be safely observed. However, in most patients, once a diagnosis is made, treatment is suggested because these diverticula will often enlarge over time and can lead to the more bothersome complications such as aspiration. The gold standard for the surgical treatment of Zenker's diverticulum is cricopharyngeal myotomy combined with either diverticulectomy or diverticulopexy via an open transcervical approach. However, advances in instrumentation for minimally invasive surgery now allow for

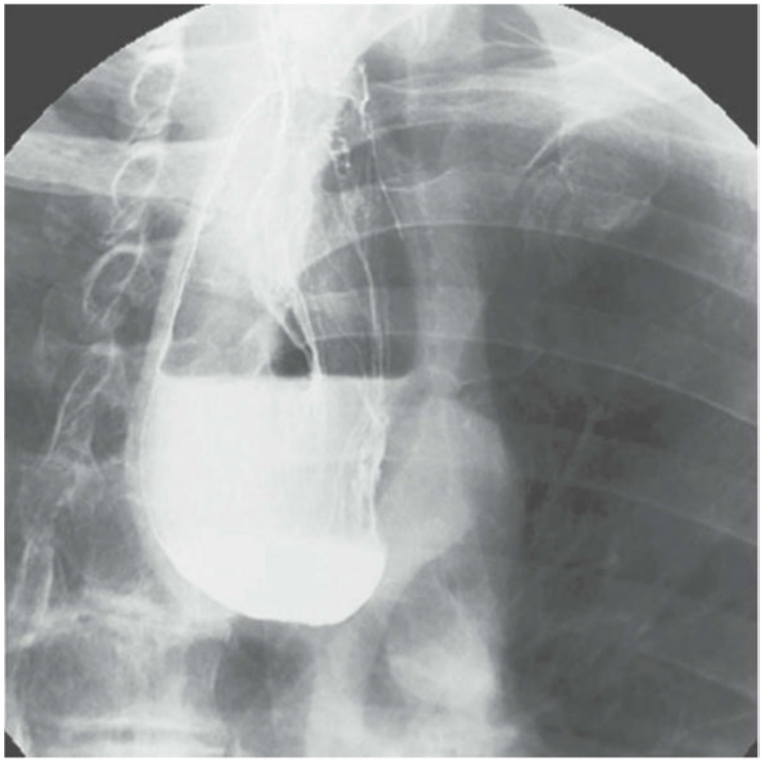


Fig. 22.1. Barium esophagram demonstrating a large Zenker's diverticulum.

the management of Zenker's diverticula via transoral endoscopic creation of a stapled esophagodiverticulostomy in many patients. This procedure utilizes a rigid diverticuloscope and an endoscopic GIA stapler to create a cricopharyngeal myotomy while bringing together the lumina of the pouch and esophagus. We believe that carefully selected patients may be offered endoscopic management with the caveat that conversion to an open procedure may be necessary should the minimally invasive approach prove technically unfeasible intraoperatively.

Endotherapy utilizing flexible endoscopes and various energy sources to divide the septum between the diverticulum and esophagus (which contains the cricopharyngeus muscle) has become more commonplace. These procedures are typically performed in an endoscopic unit by gastroenterologists. The purported benefits of this approach include the ability to perform the procedure without general anesthesia or neck extension. As only a 1.5- to 2-cm incision in the septum is recommended by most authors, diverticula >3 cm require longer incisions and, therefore, repeat procedures. A recent review of these procedures reports mediastinal emphysema from presumed microperforation in over 20% of patients and a clinical recurrence rate of approximately 20%. Given the exceptional results and minimal morbidity associated with both the traditional open repair and the transoral stapled procedure, we feel that these flexible endoscopic techniques should be reserved exclusively for poor operative candidates.

PREOPERATIVE MANAGEMENT

Endoscopic management may not be the preferred procedure in some patients due to anatomic constraints. Difficulty may be encountered in placing the diverticuloscope in patients with retrognathia, limited jaw mobility, prominent incisors, or rigid cervical kyphosis that limits neck extension. In a prospective study of factors predicting endoscopic exposure and repair of Zenker's diverticulum, the endoscopic procedure was unsuccessful in 30% of patients. Patients with short necks, high body mass index (BMI), and decreased hyomental distance were less likely to have successful endoscopic repair, and the authors advocate an open approach in such patients. In addition, the size of the diverticulum on barium esophagogram should be assessed because small diverticula (<3 cm) are generally not amenable to stapling. A small pouch limits access of the stapler anvil and prohibits the attainment of an adequate length of myotomy. Diverticula >6 cm should also be managed with an open approach because endoscopic stapling results in a large pharyngeal cavity that does not empty completely. This information is helpful in preoperative discussions with the patient concerning the likelihood of conversion to an open procedure. All patients should be prepared and give consent for an open procedure in the event that endoscopic instrumentation proves unfeasible or a complication is encountered. Patients are instructed to limit their diet to clear liquids the day before surgery.

Although many cervical operative procedures may be performed under local anesthesia, general endotracheal anesthetic is suggested for this procedure, along with the usual preincision antibiotic prophylaxis.

ENDOSCOPIC SURGICAL TECHNIQUE

After induction of general anesthesia, a shoulder roll is placed to achieve neck extension in the standard position for rigid esophagoscopy. An upper jaw dental guard is placed. Direct endoscopy is performed using a lighted suspension laryngoscope. The esophageal lumen, diverticular lumen, and their common wall are visualized. The distal blades are opened slightly to enter the esophagus anteriorly and the diverticular lumen posteriorly. The distal blades of the instrument are opened, keeping the common wall between the esophagus and diverticulum centered in the scope's aperture. The proximal scope is then widened, and the laryngoscope is attached to a suspension system to allow for bimanual instrumentation during the stapling procedure. A 0-degree bronchoscopic telescope attached to an endoscopic camera is then inserted laterally through the laryngoscope. The pouch is examined to exclude malignancy and to assess the depth of the pouch and length of the septum. A 30-mm linear GIA stapler is then introduced through the laryngoscope, and its jaws are positioned on the common wall with the longer end containing the staple cartilage within the esophageal lumen. The position of the jaws is confirmed using the telescope. If the position of the jaws is in question, the stapler is opened and the device reapplied. If necessary, traction sutures can be placed laterally in the common wall using an endosuture device and used to help pull the common wall into the jaws of the stapler. If proper positioning of the stapler cannot be confirmed before firing,

the minimally invasive approach cannot safely be performed and should be abandoned in favor of an open procedure. Once proper position is confirmed, the stapler is fired and removed. The stapler divides the common wall, including the cricopharyngeus muscle, between the diverticulum and esophagus and closes the wound edges with a triple row of staples on each side. The divided edges of the septum should retract laterally revealing an open esophageal lumen. The cut edges should be inspected to ensure hemostasis. Should a significant septum and diverticular sac still be present, a second stapler application should be performed in a similar manner. Once the septum is completely divided and hemostasis is confirmed, the procedure is terminated. Patients are offered a liquid diet the following day, and if fluids are tolerated, they are advanced to a soft diet and discharged on the second postoperative day. All patients should have a chest radiograph postoperatively to exclude perforation, which more than likely would manifest as air in the retropharyngeal space or mediastinum.

OPEN SURGICAL TECHNIQUE

After general anesthesia is induced, a shoulder roll is placed and the neck is extended with the head turned slightly to the right. An incision with its midpoint based on the cricoid cartilage is made along the anterior border of the left sternocleidomastoid muscle down. The subcutaneous tissue and platysma are subsequently divided. A superficial cervical cutaneous nerve is present in the upper one-third of the field and should be protected if possible to prevent postoperative dysesthesia of the submandibular skin. The incision is deepened through the cervical fascia medial to the sternocleidomastoid muscle. The fascia overlying the omohyoid muscle is incised, and the muscle is divided. The prethyroid muscles are retracted medially to reveal the thyroid gland, the jugular vein, and the carotid artery. The middle thyroid vein is ligated and divided, and the larynx is retracted medially. The inferior thyroid artery is ligated and divided as laterally as possible to protect the recurrent laryngeal nerve, which is located immediately beneath its branches in the tracheoesophageal groove behind the thyroid. The thyroid and cricoid cartilage are rotated medially while retracting the carotid sheath and its contents laterally. Manual retraction of the larynx with an assistant's finger is preferred to self-retaining devices or hand-held metal retractors to prevent a traction or direct compression injury to the recurrent laryngeal nerve. With the use of blunt dissection, the prevertebral fascial plane is entered posterior to the esophagus and diverticulum, and further rotation of the larynx then everts the lateral posterior pharyngoesophageal junction. The diverticulum can usually be visualized adherent within its filmy attachments to the posterior aspect of the cervical esophagus at or below the level of the cricoid cartilage. The adventitial tissues are gently dissected in the posterior midline to free up the pouch. The tip of the diverticulum is grasped with an atraumatic Babcock or similar type of clamp and is dissected away from the esophagus using a combination of sharp and blunt dissection (Fig. 22.2). The sac is elevated until its neck is clearly defined, and this may require some additional sharp dissection from the known surface of the diverticular sac toward the investing muscle fibers and scar tissue, if present. Once the neck of the diverticulum is displayed, a cricopharyngeal myotomy is created starting at the inferior border of the diverticulum on the left posterolateral or posterior aspect of the esophagus. The muscle is separated from the underlying submucosa bluntly using a small right-angled clamp. The myotomy should be extended inferiorly for approximately 3 cm (Fig. 22.3). The mucosa will bulge between

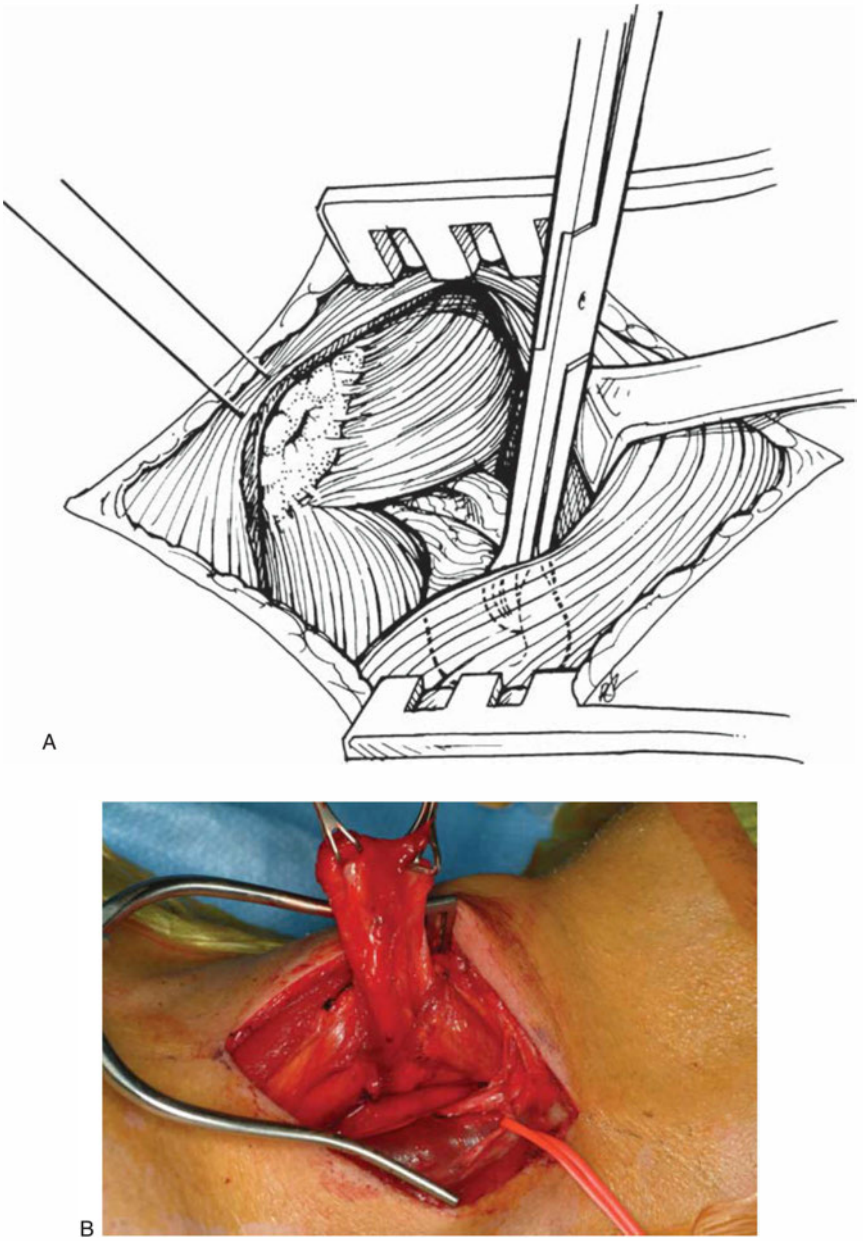


Fig. 22.2. (**A**) Exposure of Zenker's diverticulum. The sac is grasped with an atraumatic clamp and dissected away from the esophagus. (**B**) Intraoperative photograph of a Zenker's diverticulum.

the edges of the sectioned muscle. With the myotomy completed, small diverticula (<1 cm) will often disappear when released into the bulging submucosa, and thus myotomy alone is sufficient. Those that do not spontaneously resolve should be further treated with either diverticulopexy or excision. Diverticula <5 cm in length are amenable to diverticulopexy, which is a simpler and faster procedure that does not require opening of the esophageal lumen. Following the myotomy, the tip of the diverticulum may be suspended either to the posterior wall of the pharynx or to the prevertebral fascia with two to four simple 3-0 silk sutures (Fig. 22.4). Care should be taken not to transfix the sac to avoid any contamination of the surgical field. Diverticula >5 cm are too large to be suspended and should usually be resected. After myotomy, a large bougie (40F or larger, depending on patient size) is inserted into the cervical esophagus to prevent excessive mucosal resection which could narrow the esophageal lumen. If possible, a linear stapling device is applied transversely across the neck of the sac, and the sac is transected distal to the staple line. In the rare cases in which a stapler cannot be applied, the sac may be resected and the defect repaired using interrupted sutures. Care must be taken here not to remove an excessive amount of mucosa with the resection. It is better to have more than needed because tissue can be gathered into the suture line to make the lumen attain an appropriate size. Before closure, a nasogastric tube is passed into the proximal esophagus, and the mucosal integrity is tested by air insufflation by placing saline in the incision and compressing the distal esophagus below the nasogastric tube. The nasogastric tube is subsequently removed, and after hemostasis is ensured, the wound is closed in two layers without drainage. If the ends of the omohyoid are large enough, they may be reapproximated using a figure-of-eight suture technique. Patients are allowed a clear liquid diet the morning after surgery and advanced to a soft diet the following day in most cases. Most patients are discharged on the second or third postoperative day. A postoperative barium swallow is not routinely obtained before discharge. Patients should maintain a soft diet for the first week following surgery.

SURGICAL RESULTS

There have been no prospectively randomized, controlled trials comparing the different open surgical approaches, nor

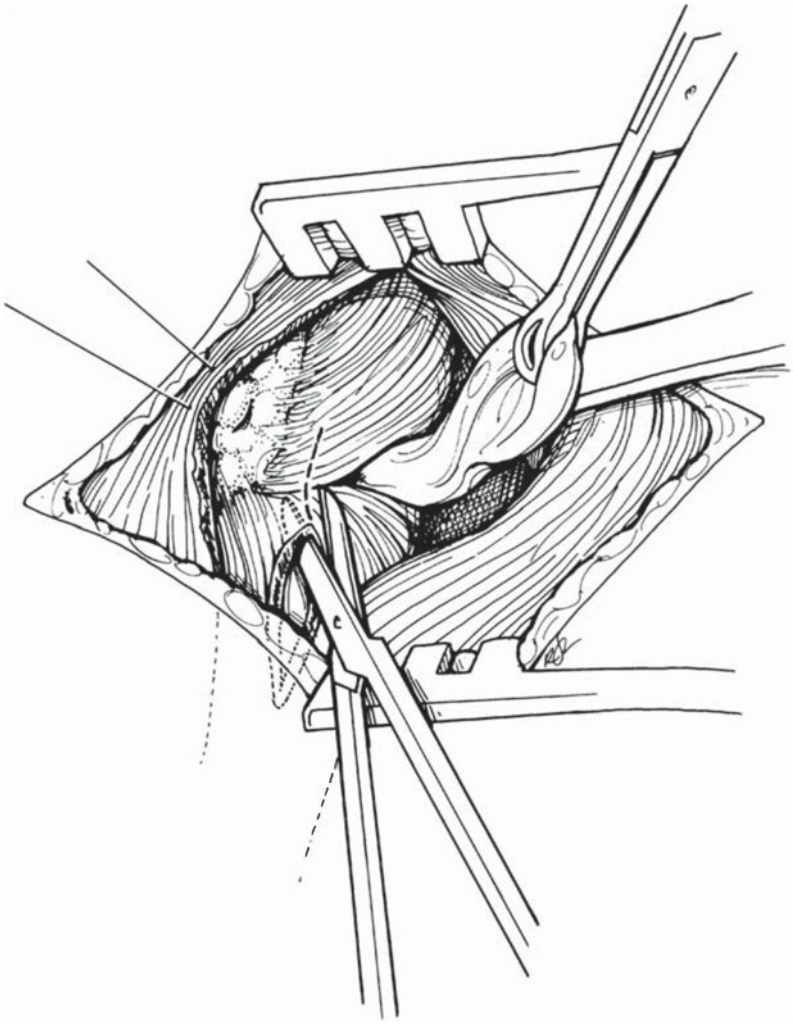


Fig. 22.3. The esophageal muscle is incised from 2 to 3 cm below the diverticulum and extended toward the cricopharyngeus.

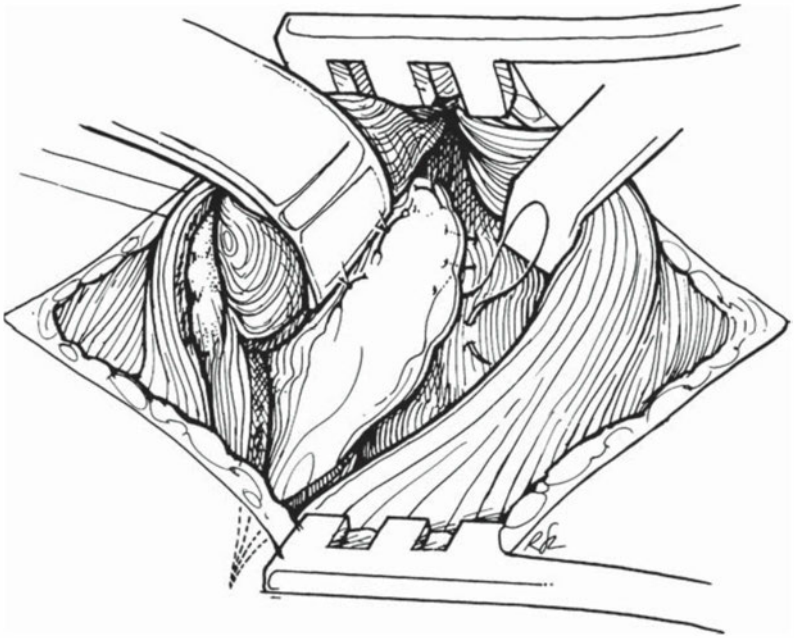


Fig. 22.4. Once the myotomy is completed, the diverticulum should be resected or sutured superiorly to the prevertebral fascia.

have there been randomized studies comparing endoscopic stapled esophagodiverticulostomy with open surgical techniques. Series of stapled esophagodiverticulostomy have reported satisfactory outcomes in approximately 90% of patients undergoing successful endoscopic repair, with both morbidity and recurrence rates <10%. These figures closely approximate those reported in series of open surgical techniques. Complications are similar after both techniques and most commonly include hemorrhage, transient vocal cord paralysis, and perforation resulting in mediastinitis. In a large retrospective Italian series of 297 patients who underwent either transoral stapled esophagodiverticulostomy or open cricopharyngeal myotomy and stapled diverticulectomy, >90% of patients were symptom-free or significantly improved after a median follow-up of >2 years regardless of the approach. Patients who underwent transoral stapling for a diverticula <3 cm were more likely to have persistent dysphagia, presumably because the common wall was too short to allow complete myotomy. Advantages of the minimally invasive approach included shorter anesthetic time, earlier resumption of normal diet, and shorter hospital stay. The endoscopic stapling technique is an effective procedure in patients with medium-sized diverticula (3 to 6 cm). However, certain patient characteristics have been demonstrated to limit successful endoscopic exposure and repair. In our opinion, conventional open repair techniques are very well tolerated and remain the gold standard therapy at present.

THORACIC DIVERTICULA

Diverticula of the thoracic esophagus are relatively uncommon and typically account for <30% of all esophageal diverticula. As stated earlier, their classification according to location or pathogenesis is probably unnecessary because midesophageal traction diverticula are now a rarity in the Western hemisphere. Almost all thoracic esophageal diverticula are associated with and perhaps caused by an esophageal motor disorder, most commonly achalasia, diffuse esophageal spasm, hypertensive lower esophageal sphincter, and nonspecific motor disorders. Using cinefluorography and manometry, Cross et al. concluded that in 150 patients with esophageal disorders, esophageal diverticula were the result of excessive intraluminal pressures. This segmental increase in pressure, caused by increased esophageal tone or a delay in sphincteric opening, acts on weakened areas of the esophagus and results in outpouching of the mucosa.

CLINICAL PRESENTATION

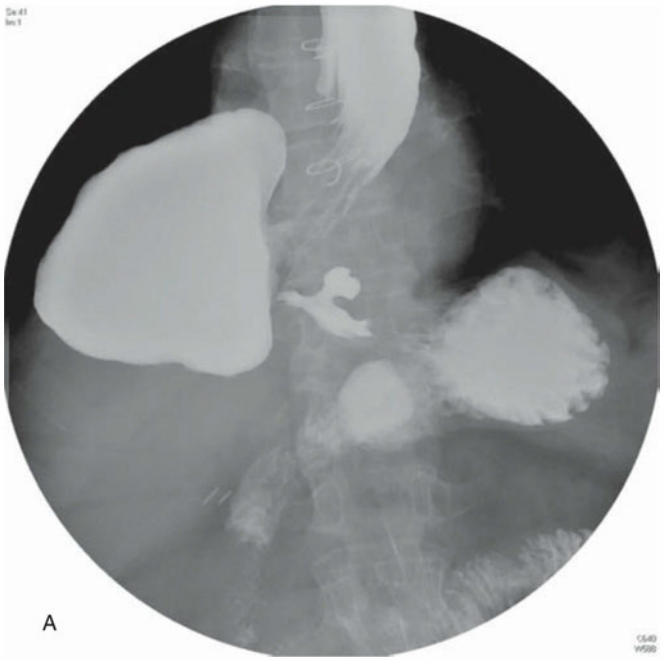
Most patients are older than 60 years of age, and many are asymptomatic or have only minimal and often vague symptoms. There is usually no correlation between diverticular size and the presence of symptoms. Many vague symptoms may be attributable to the underlying motility disorder rather than the diverticulum. Symptoms include dysphagia, postural regurgitation, belching, retrosternal pain, heartburn, and epigastric pain. As in pharyngoesophageal diverticula, pulmonary symptoms are often present but underestimated. These symptoms range from mild nocturnal cough to life-threatening massive aspiration.

DIAGNOSIS

The diagnosis is suggested by a posterior mediastinal air–fluid level on chest roentgenogram and is confirmed by a barium esophagogram. The presence of an associated hiatal hernia or carcinoma within the diverticulum (both uncommon occurrences) can be assessed by endoscopic examination, which should be performed in every case. Esophageal manometry is not essential for the diagnosis but is helpful in defining the nature and the extent of the underlying motor disorder. Manometry may require directed passage of the catheter beyond the diverticulum if difficulty is encountered.

INDICATIONS FOR SURGERY

Most would agree that surgical intervention is necessary in symptomatic patients, those with large diverticula, and those with established complications such as bronchopulmonary infection (Fig. 22.5). The indication for surgery in asymptomatic or minimally symptomatic patients is less clearly defined in the literature, and yearly surveillance for patients with an asymptomatic thoracic esophageal diverticulum is often the most prudent option.



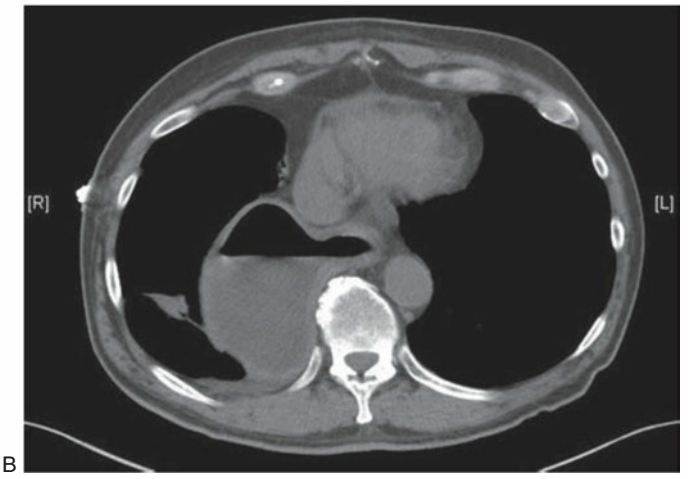


Fig. 22.5. Large epiphrenic pulsion diverticulum associated with pain and intermittent regurgitation of undigested food. **(A)** Barium esophagram. **(B)** Computed tomographic scan.

The basic elements of the operation are resection of the diverticulum and a myotomy to alleviate the underlying motor disorder. Although some authors advocate that a myotomy can be done only if a documented underlying motor disorder is present, this is a hazardous proposition because it risks the integrity of the diverticulectomy staple line in the postoperative period and a possible recurrence of the diverticulum later. Another point of controversy is the length of the myotomy. Although a limited myotomy may be satisfactory in many situations, a long myotomy from the level of the aortic arch down to the first 1 to 2 cm of the stomach adds little to the duration of the operation and essentially eliminates the need for reoperation. Naturally, once the cardia is mobilized, a nonobstructive type of antireflux repair must be added to avoid the almost certainly ensuing gastroesophageal reflux, which the aperistaltic esophagus cannot adequately clear.

SURGICAL PROCEDURE

All patients receive a preoperative dose of a second-generation cephalosporin, and this antibiotic is continued for 24 hours postoperatively. Patients with large diverticula should have the contents of the sac evacuated by a large tube preoperatively. Rapid sequence induction of general anesthesia is necessary to secure rapid control and protection of the airway against aspiration.

Notwithstanding that most diverticula project into the right side of the chest, the preferred approach for an epiphrenic diverticulum is through a left thoracotomy, which permits excellent exposure of the lower half of the thoracic esophagus, the diverticulum, and the cardia should an antireflux repair be necessary. The patient is placed in the right lateral decubitus position, and the left chest is entered through the sixth intercostal space. The use of a double-lumen endotracheal tube with deflation of the left lung greatly improves the exposure. The mediastinal pleura is incised over the esophagus from the aortic arch to the esophageal hiatus. The esophagus, including both vagus nerves, is mobilized at some distance away from the diverticulum and encircled proximally and distally with Penrose drains (Fig. 22.6). The sac is freed from the surrounding mediastinal structures with blunt and sharp dissection. Occasionally, the sac is firmly adherent to the right lung or the right mainstem bronchus (in midesophageal diverticula), and upward traction on the esophagus allows a careful dissection of the sac from those attachments.

The sac may be invested in an inflammatory rind of tissue, and this must be carefully removed to expose the wall. The neck of the diverticulum is sharply dissected from the surrounding esophageal muscle and residual scar if present, and the mucosal layer should be separated from the surrounding overlying muscle at its margins so that the true neck is obvious for later division. The vagus nerves may be very adherent to the sac and can be embedded in the inflammatory tissue surrounding it. Care must be taken not to injure the nerves during this part of the procedure.

Dissection then proceeds caudally to mobilize the gastric cardia. The phrenoesophageal membrane is sharply divided, and the hiatus is manually dilated. The mobilization of the cardia is completed by dividing the posterior attachments between the two vagi and entering the lesser sac, followed by complete excision of the fat pad in this region just superior to the cardia. The posterior attachments here may harbor an enlarged arterial branch emanating from the left gastric artery, and failure to recognize this can result in hemorrhage.

At this time, a 44F or larger bougie is placed by the anesthesiologist with

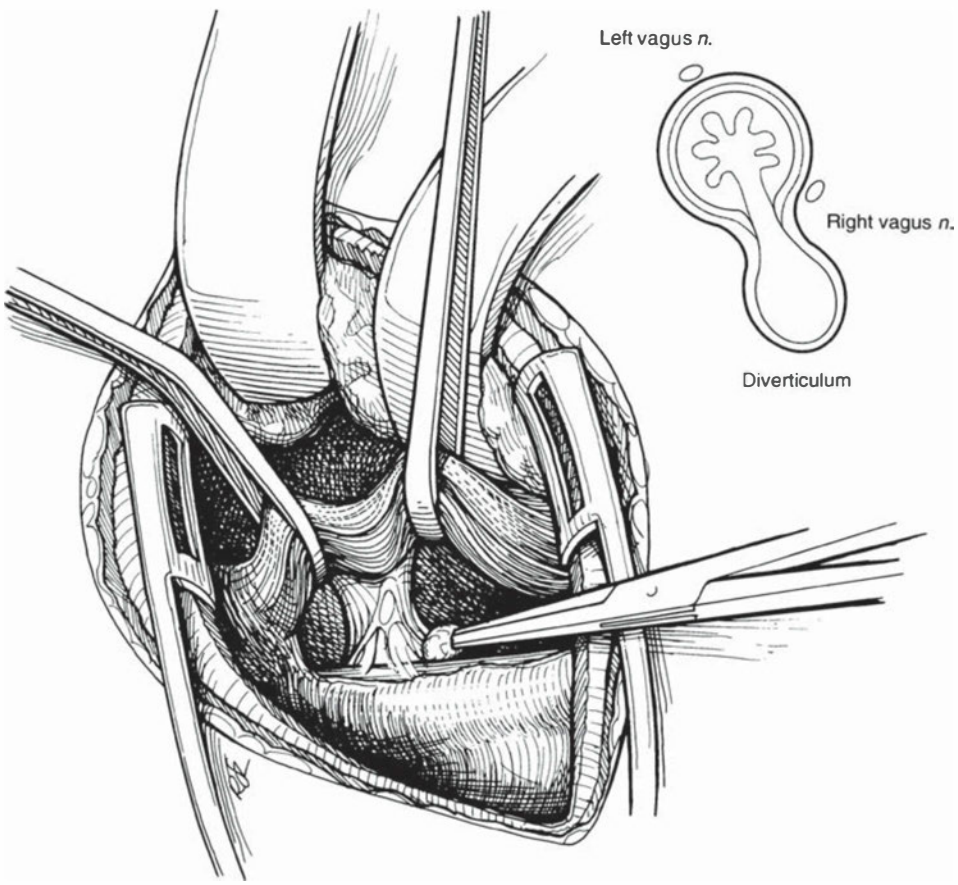


Fig. 22.6. Mobilization of the esophagus for thoracic diverticulum. Penrose drains are used to encircle the esophagus proximal and distal to the diverticulum, including vagus nerves. n., nerve.

manual guidance by the surgeon past the diverticular opening. The diverticulum is then brought into view by grasping its fundus with an atraumatic clamp and rotating the diverticulum and the esophagus anteriorly and then to the left (Fig. 22.7). If possible, a diverticulectomy is now performed by excising the sac about 5 mm from its neck using a linear stapling device. If this is not possible due to the size of the neck or necrosis at its junction at the esophageal wall, it can be sharply excised after placement of stay sutures on the cranial and caudal extents of the neck. This can then be repaired using interrupted 3-0 absorbable sutures. A nasogastric tube is passed, and the stapled closure is tested for leak by air insufflation under water with distal esophageal compression. The muscle layer is then approximated over the mucosal closure with interrupted 3-0 silk sutures (Fig. 22.8). On occasion, the muscular layer may be too edematous or attenuated for a good second-layer closure. If this is the case, an intercostal muscle flap may be mobilized and approximated to the linear repair using interrupted small silk sutures on the muscular margins. A long myotomy is then performed on the left lateral aspect of the esophagus 180 degrees from the excised diverticulum. The muscle is divided at least 2 cm proximal to the diverticular repair, and if feasible from just below the aortic arch and carried across the gastroesophageal junction onto the stomach for approximately 1 cm. The edge of the muscle is gently dissected off the underlying mucosa for several millimeters on either side to preclude premature healing of the myotomy. A nonobstructive antireflux repair should then be performed to prevent reflux sequelae. Our repair of choice is the Belsey Mark IV partial fundoplication, with sutures placed on either side of the myotomy in the muscular layer. Once the second layer of the sutures is placed, the cardia is gently repositioned into the abdomen and the second layer of the sutures is tied (Fig. 22.9). The crura are reapproximated with large nonabsorbable suture so that it will accommodate just the tip of the operator's index finger, and the nasogastric tube is advanced into the stomach.

Minimally invasive operations for thoracic esophageal diverticula have been reported. For mid esophageal diverticula, right video-assisted thoracoscopic surgery (VATS), diverticulectomy with an endo-GIA stapler, closure of the overlying muscle over the staple line, and myotomy opposite the diverticulectomy site are feasible.

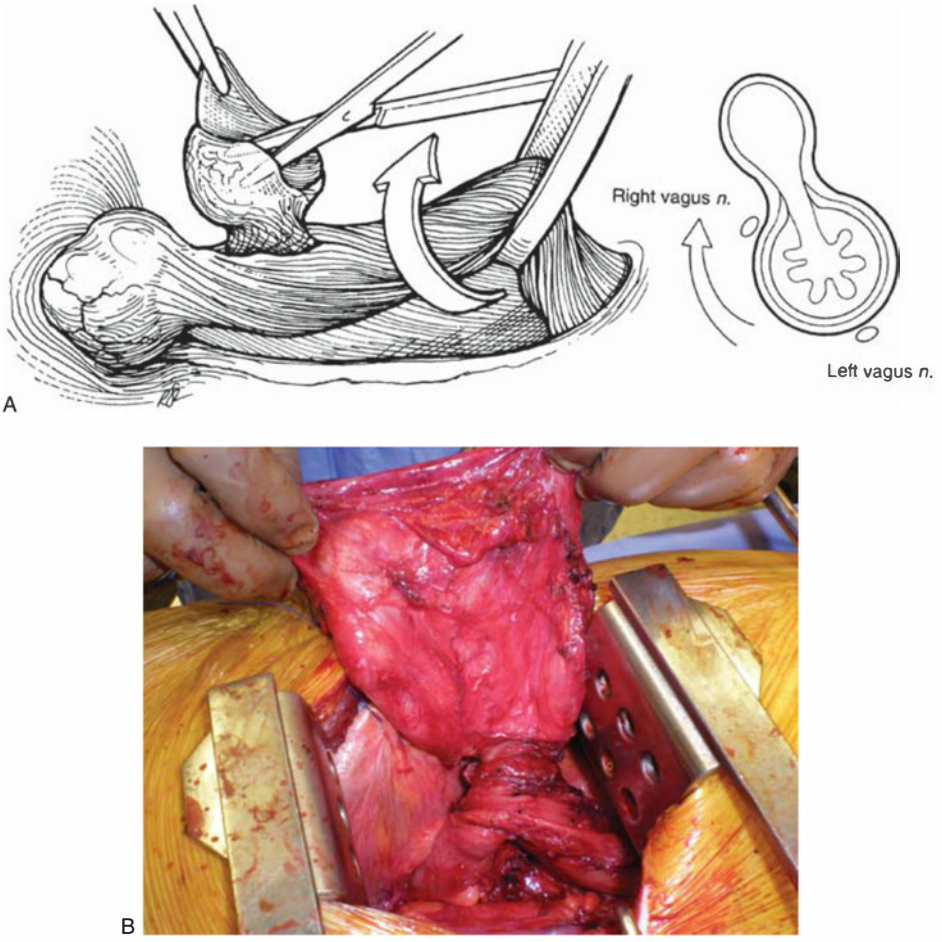


Fig. 22.7. (**A**) The esophagus is rotated to the left and anteriorly to expose the diverticulum, which is most often located on the right side. (**B**) Intraoperative photograph of a large epiphrenic diverticulum. n., nerve.

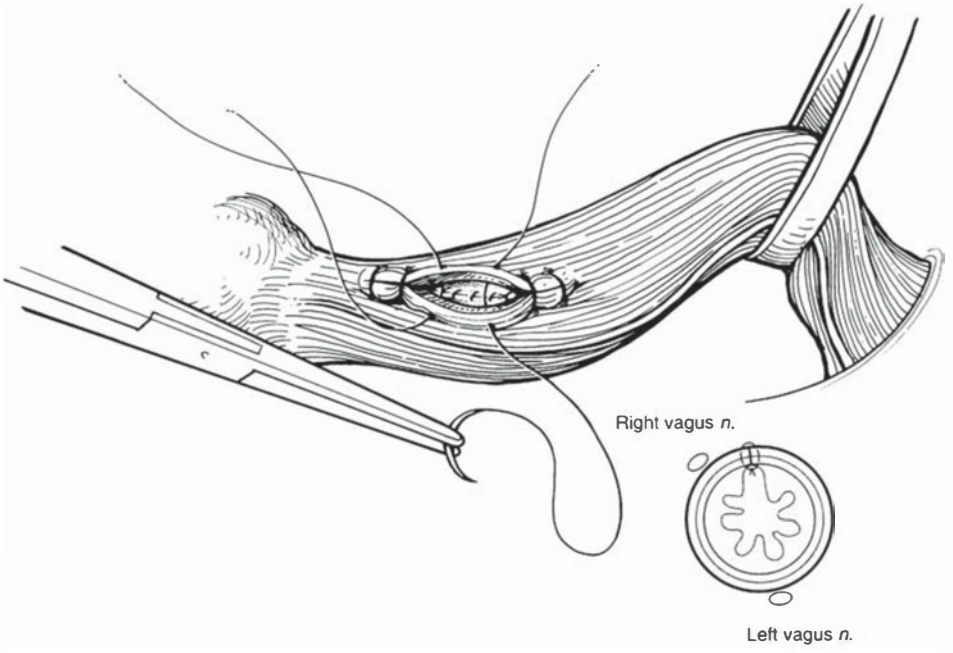


Fig. 22.8. After removal of the diverticulum, the mucosal and muscular layers are closed with interrupted sutures. The closure may be tested for leak by air insufflation and distal occlusion. n., nerve.

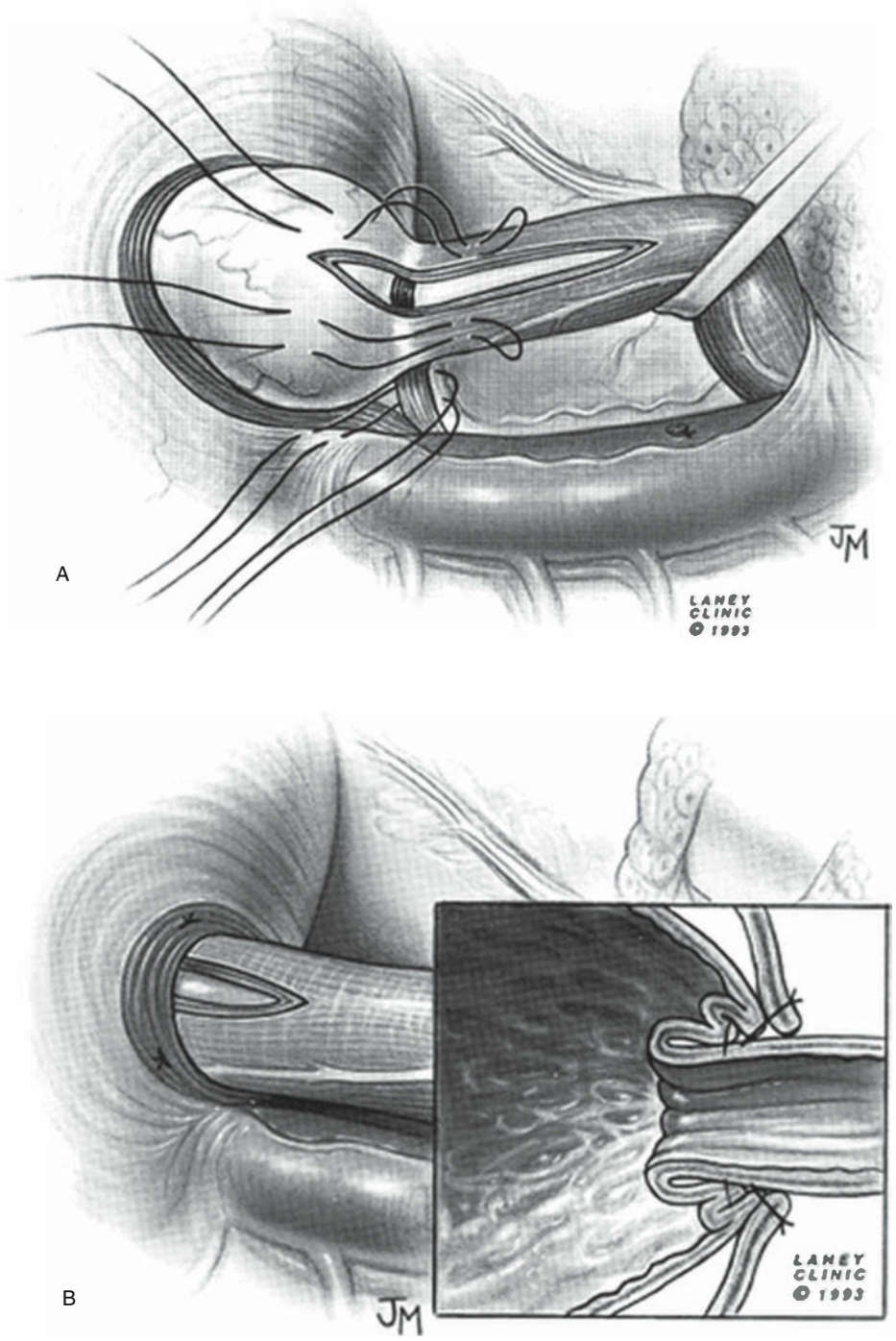


Fig. 22.9. (**A**) A Belsey Mark IV partial fundoplication, with sutures placed on either side of the myotomy in the muscular layer is performed. (**B**) Once the second layer of sutures is placed, the cardia is gently repositioned into the abdomen and the sutures are gently tied down. (Streitz JM Jr. Modified Heller esophagomyotomy. In: Shields TW, ed. *General Thoracic Surgery*. Philadelphia, PA: Lippincott Williams & Wilkins; 2005:1975–1986).

The preferred minimally invasive approach for epiphrenic diverticula involves laparoscopy, esophagogastric myotomy, stapled diverticulectomy, and partial fundoplication (Toupet or Dor). Surgical series from centers with extensive experience in minimally invasive antireflux surgery and esophagectomy have demonstrated reasonable symptomatic outcomes and morbidity and mortality, which are comparable to large open surgical series. These minimally invasive techniques are attractive. A recent series of laparoscopic repair of epiphrenic diverticula demonstrated a median length of stay of 3 days, which is certainly shorter than that reported in large series of repair via left thoracotomy. However, it bears repeating that the diverticula of the thoracic and epiphrenic esophagus are quite rare and, thus, a typical thoracic surgeon may only treat a handful of them in his entire career. It is unlikely that one can extrapolate the results obtained at centers with extensive experience in minimally invasive esophageal surgery to the thoracic surgical community at large, and open surgical repair remains the gold standard in our opinion.

SURGICAL COMPLICATIONS AND POSTOPERATIVE CARE

Retention of the nasogastric tube for 3 to 4 days is encouraged, at which time a barium esophagram is performed to rule out leak at the repair or myotomy or mechanical obstruction. If no leak is present, a clear liquid diet is begun and is rapidly advanced to a soft mechanical diet with discharge typically on the sixth or seventh postoperative day. Although success rates are very high, there has been moderate morbidity reported and a mortality of up to 9% in some series, substantiating the recommendation that small or asymptomatic diverticula should be observed. Causes of death include mediastinitis as a result of esophageal leakage and aspiration pneumonia. Morbidity and mortality are obviously negatively influenced by the advanced age of many candidates.

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EDITOR'S COMMENTS

By far the most common diverticulum of the esophagus is the pharyngoesophageal, or Zenker's, diverticulum, which, as the authors point out, is not a true diverticulum because it is a mucosal protrusion, not a true full-thickness outpouching of the esophagus. Most commonly, these patients present with difficulty swallowing, noisy swallowing, and often signs and symptoms of aspiration, especially at night. Many of these patients are elderly, and many are told that they are not operative candidates because of the risk. It is safe to say that the risk is minimal, and it is likely that there is far greater risk to the patient by allowing the diverticulum to remain, especially if the patient has signs of aspiration. If the patient is felt to be at significant risk for general anesthesia, the procedure may be carried out under local anesthesia.

It has been and remains our preference to approach a Zenker's diverticulum via the open technique though I have been pleased with outcomes when we have employed the endoscopic technique. This open procedure involves an incision along the anterior border of the left sternocleidomastoid muscle with dissection down to the prevertebral fascia. The left side is used even if the diverticulum protrudes to the right because the esophagus is easily mobilized and the left recurrent nerve is less at risk than the right. Often the diverticulum in the anesthetized patient appears smaller than what the surgeon originally thought and sometimes is difficult to identify. The myotomy is begun at the base of the diverticulum at Killian's triangle and carried well down onto the esophagus taking care to incise both the longitudinal and circular muscle fibers. For all but the largest diverticula, I prefer to perform a diverticulopexy, excluding the diverticulum from the flow of material in the esophagus but avoiding a mucosal entry. If a diverticulectomy is to be carried out, a large (50F) bougie is placed to avoid tenting of the mucosa and excessive excision that could lead to a stenosis. A staple line is placed across the diverticulum, and the patient is not fed for several days. A contrast study assures integrity of the staple line before feeding.

As described by the authors, an endoscopic approach to the treatment of Zenker's diverticulum has been described. An instrument specifically for this purpose, the Weerda diverticuloscope, should be used for proper visualization of the diverticulum and endoscopic stapler placement. One blade of the stapler must be in the esophageal lumen and the other in the lumen of the diverticulum for the procedure to be successful. This accomplishes obliteration of the "party wall" between the diverticulum and the lumen of the esophagus and accomplishes a myotomy. The long-term results of this procedure confirm that this approach offers an excellent alternative for many patients. Its utility is limited in those patients who have limited neck extension.

It is critical to recognize that any esophageal diverticulum or pseudodiverticulum represents the manifestation of a motor disorder of the esophagus, which underscores the importance of a complete myotomy. The myotomy is especially important if the diverticulum has been excised to assure the integrity of the staple line. The terms "pulsion" and "traction" diverticula are rarely used in the present-day nomenclature. Even the so-called epiphrenic diverticula are manifestations of a motor disorder, and when the operation is indicated a myotomy must also be performed. I agree with the authors that a long myotomy beginning just inferior to the aortic arch and extending down onto just distal to the gastroesophageal junction is preferred. Patients with an epiphrenic diverticulum are also subject to aspiration, but many, if not most, patients with these diverticula do not require operation. Most are asymptomatic and merely represent curiosities on plain chest radiographs or barium contrast studies. The mere presence of a large epiphrenic diverticulum is not an indication for operation, especially because these must be approached via a thoracotomy or VATS procedure.

LRK

23 Lung Transplantation

R. Duane Davis and Mathew G. Hartwig

Following the initial report of human lung transplantation by Dr. James Hardy in 1963 at the University of Mississippi, numerous subsequent attempts failed until the first successful transplant by Dr. Joel Cooper at the University of Toronto in 1983. In the succeeding years, lung transplantation has become a viable and effective treatment for patients with end-stage lung disease and has been performed in more than 32,000 recipients worldwide. Progress has been made in surgical techniques, donor lung preservation, lung reperfusion protocols, perioperative care, and long-term medical management of this challenging patient population. These refinements in care are reflected in survival improvements. One-year survival reported by the Organ Procurement and Transplantation Network (OPTN) has improved from 77% in 1997 to 82% in 2007. Likewise, the International Society of Heart and Lung Transplantation (ISHLT) Registry indicated that the median survival increased from 4.0 years if transplanted in 1994, to 5.7 years for a recipient after the turn of the millennium. In fact, the era of a particular transplant procedure remains one of the best predictors of survival in the ISHLT Registry. Similar improvements in survival have occurred in the experience of the Duke Lung Transplant Program (Fig. 23.1A). In the most recent 6-year cohort, our 30-day and 5-year survivals exceeded 97% and 65%, respectively.

Death early after transplant is commonly related to primary graft dysfunction (PGD) that is usually caused by ischemia-reperfusion injury. PGD rates have substantially decreased with better lung preservation and controlled reperfusion strategies. Although posttransplant recipient and allograft survival have significantly improved recently, long-term survival continues to be less than that for other solid organ transplants. Current 5-year survival from the ISHLT Registry approaches 55% for lungs, compared to 73% for cardiac recipients. Late deaths are most frequently related to chronic allograft dysfunction, the causes of which are related to both immunologic and nonimmunologic factors. The control of these factors with better immunosuppressive regimens and with minimizing nonimmunologic injury such as gastroesophageal reflux-related aspiration into the tracheobronchial tree appears to be improving long-term survival.

INDICATIONS FOR LUNG TRANSPLANTATION

Lung transplantation has been successfully applied as a treatment for patients with a number of different end-stage lung diseases. While specific listing criteria and indications have been widely reported, lung transplantation is typically performed in patients (1) with a limited life expectancy secondary to lung failure, estimated to be 6 to 12 months; (2) with the capability of tolerating and following the complicated posttransplant medical regimen; and (3) without comorbidities that would significantly impact posttransplant survival and quality of life, such as concurrent/recent malignancy, extrathoracic infection, or significant other organ dysfunction/failure. In select patients, concomitant other organ transplant or reparative cardiac procedures can be performed with acceptable results.

The choice to perform a single versus a bilateral, sequential lung transplant depends upon multiple factors. Clearly, recipient characteristics such as comorbidities and underlying diagnosis play an important role in directing operative selection. Likewise, the value of increasing the number of recipients transplanted by performing two single-lung transplants (SLTs) from a single donor must be balanced with the benefit of improved long-term survival that is seen in bilateral lung transplants (BLT). While the majority of nonseptic lung disease, such as chronic obstructive pulmonary disorders (COPD), idiopathic pulmonary fibrosis (IPF), and primary pulmonary hypertension, can safely undergo single-lung transplantation, patients with septic lung disease, such as bronchiectasis from either cystic fibrosis (CF) or noncystic causes, require bilateral lung transplantation to prevent infectious complications emanating from the residual native lung. Because of the numerous complications arising from the native lung such as infection, hyperinflation, malignancy, and a 25% absolute difference in survival at 7 years (Fig. 23.1B), the practice of the Duke Lung Transplant Program has been to perform bilateral, sequential lung transplants in the majority of patients. This chapter describes the steps in procuring, preparing, and transplanting lungs in a bilateral, sequential fashion. These techniques are also utilized when only one lung is being transplanted.

DONOR LUNG RETRIEVAL

The improved outcomes after lung transplantation have led to an increased number of recipients waiting for transplant. In the United States, approximately 4,000 patients are listed and waiting for lung transplant. Annually, approximately 1,100 lung transplants are performed while 450 deaths occur on the waiting list. The shortage of organs is not unique as compared to other organs, but there are a number of differences. Of organs commonly transplanted, the lungs are the most sensitive to exogenous damage. Events prior to brain injury, such as smoking, and events associated with brain injury, such as aspiration, mechanical ventilation, or neurogenic pulmonary edema, may compromise the suitability of the donor lungs for transplantation. However, there is a wide variety of opinions regarding what is a suitable lung for transplantation. Throughout the United States, there has been a gradual increase in the percentage of organ donors in which lungs were retrieved to 17%. However, this number is substantially less than the best-performing regions in the United States as well as in Australia, where the utilization is 40% to 50%. Moreover, survival rates for recipients from these high-utilization

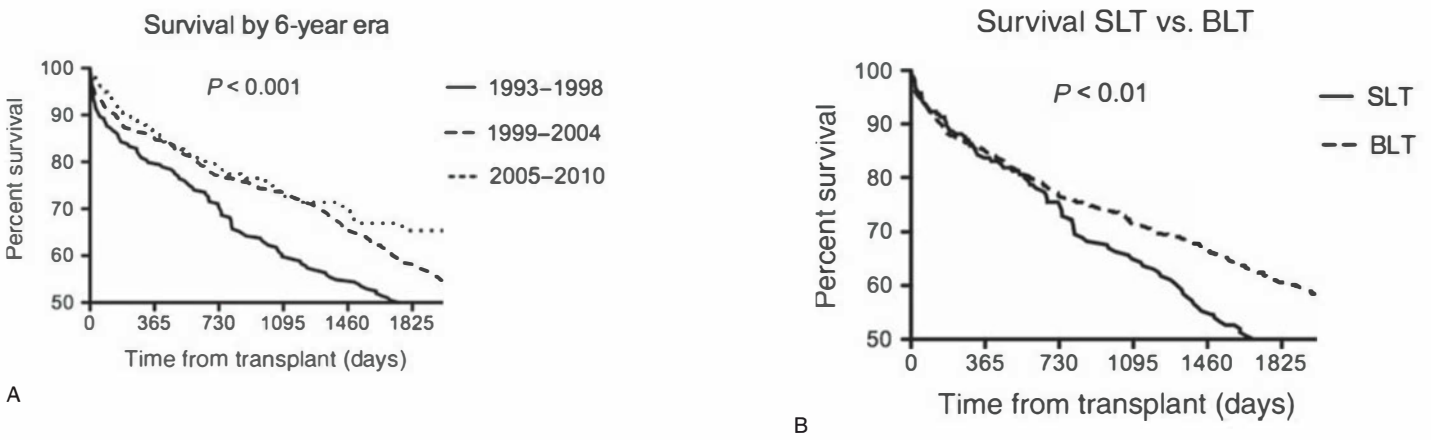


Fig. 23.1. Actuarial survival for lung transplant recipients of the Duke Lung Transplant Program for the three consecutive 6-year intervals from 1993 to 2010 (**A**) and for patients receiving single as compared to bilateral transplants (**B**). Survival has significantly improved with each subsequent time period and for patients receiving bilateral transplants.

regions exceed registry data. Aggressive donor management with hormone replacement, particularly with respect to corticosteroid administration, appropriate ventilatory settings with alveolar recruitment measures, suitable fluid resuscitation with vasoactive drug administration, aspiration prevention, and utilization of bronchoscopic suctioning have been shown to increase the yields of donor lungs. Importantly, aggressive evaluation and placement strategies by transplant centers are also factors in increasing donor lung yield.

Emerging technologies will also allow for an expanded donor pool and better utilization rates. For example, ex vivo lung perfusion (EVLP) has recently been initiated for human lung transplantation. An initial manuscript detailing 35 lungs transplanted following EVLP suggests equal outcomes in terms of PGD, BOS development, and 1-year survival when compared to 116 control recipients. Currently, this technology allows for rehabilitation of some lung allografts otherwise thought to be unusable. However, in the future it may provide a method for actually improving long-term outcomes through gene therapy or other treatments.

The donor lungs must (1) be ABO compatible with the recipient's; (2) have no unacceptable HLA antigens if the recipient has anti-HLA antibodies as determined by prospective or virtual cross matching techniques; (3) not transmit diseases such as hepatitis B or C, HIV, or malignancies; (4) be size compatible; and (5) be able to function physiologically to immediately sustain cardiopulmonary function in the recipient. With respect to disease transmission, donors who test as serologic positives for hepatitis B, hepatitis C, or HIV, or have non-CNS, nonskin malignancy (excluding melanoma) are not used except in extraordinary circumstances. One exception is that donors with positive hepatitis core antibody, but negative surface antigen serologies, can be used with very low rates of disease transmission.

Donor evaluation includes the collection of a thorough medical history for the potential donor. Specific donor criteria vary according to individual transplant centers, but in general, the focus is on the donor's blood type, size, age, smoking habits, pulmonary diseases, thoracic procedures, and mode of death. Additional information that should be provided at the time of a donor offer includes radiographic findings, arterial blood gas, ventilator settings, peak airway pressures, endotracheal tube size, and bronchoscopic findings. In an older donor with a substantial smoking history, a high-resolution chest CT can greatly assist in evaluating the lung parenchyma and allow for a better assessment of the presence of emphysema, lung nodules, and interstitial lung disease that would be contraindications to the use of the lungs. Corticosteroids (methylprednisolone 1,000 mg) should be administered to the donor and ventilatory settings should be optimized. Underventilation and inappropriate use of positive end-expiratory pressure (PEEP) is common and optimization can improve the donor arterial blood gas results. The most recent chest radiograph should be examined for pathology and can be used to help with sizing of the organs. Atelectasis does not preclude transplantation; however, clear consolidation would prevent the use of the organs, unless the area could be easily excised or the contralateral lung was to be utilized. This differentiation is rarely possible without direct visualization and palpation of the organs. Atelectasis is the most common reversible cause of low PaO2/FiO2 ratios in the donor. On-site evaluation of prospective donors greatly increases the number of lungs retrieved. In the absence of poor airway compliance (elevated peak airway pressure in the absence of a small endotracheal tube or morbid obesity) or combined abnormalities such as an older donor with extensive tobacco exposure and marginal arterial blood gases, we attempt to evaluate all donors.

Size matching of the donor and the recipient is initially based on height. Matching within 4 inches between donor and recipient is usually without consequence. However, recipients with obstructive lung disease will have larger than normal thoracic cavities, and larger donors are preferred. Recipients with restrictive lung disease, on the other hand, will have smaller than normal thoracic cavities, and smaller donors are preferred. Comparison of chest measurements, both vertical and horizontal dimensions, between donor and recipient from the chest X-ray provides additional information regarding sizing. It is important to account for size reformatting with digitized films. While significant size differences between donor lung size and recipient thoracic volume can be managed successfully in the operating room through the use of pneumoreduction techniques, appropriate size matching prior to committing to the transplant procedure is optimal.

Fiberoptic bronchoscopy is performed to evaluate for evidence of airway inflammation, intraluminal pathology, anatomic abnormalities, as well as to remove secretions from the airways. While it is common to encounter some purulent secretions,

this is not a contraindication to organ utilization unless the secretions do not clear or they recur after being suctioned clear. Donor airway cultures and gram stains are important for determining recipient antibiotic administration, but should not obviate the utility of lungs from donors.

Donor lung procurement is usually performed in the context of cardiac and hepatic organ procurement. The following donor procurement procedure can be utilized in all circumstances. Exposure of the thoracic and abdominal organs is through a median sternotomy with an extension of the incision in the midline to the pubis. The pericardium is opened in the midline with wide extensions laterally at the level of the diaphragm inferiorly. Pericardial stay sutures are placed for exposure. Both pleural spaces are opened widely, extending from the diaphragm inferiorly to the mammary pedicles superiorly. Adhesions are divided with the electrocautery. Manual examination of both lungs is performed serially to assess the general appearance, nodules, atelectasis, consolidation, edema, and compliance. Suspicious findings should be excised and sent for pathologic analysis to rule out malignancy. The elastic recoil of the lungs is then determined by inflating the lungs with bagged breaths and allowing the lungs to deflate independently by temporarily disconnecting the endotracheal tube from the ventilator. Lungs with adequate compliance should rapidly deflate following disconnection from the ventilator. During this phase of the evaluation, an aggressive alveolar recruitment is performed. Working with the anesthesiologist, the lungs are hand ventilated to expand any atelectatic areas. An arterial blood gas is obtained following these maneuvers. We use a minimum PaO_2 of 300 mmHg with the FiO_2 of 1.0 and PEEP of 5 cm H_2O as criteria for suitability. Additional information can be obtained and nonpulmonary shunt causes of impaired oxygenation can be identified by individually sampling from the four pulmonary veins. The expected PaO_2 should be greater than 400 mmHg from the veins draining appropriately functioning regions. This approach can identify areas of lung inappropriate for transplantation. Not infrequently, contused segments or areas of consolidation can be resected by wedge resection, lobectomy, or the use of only one lung. These resections are ideally done as a "back table" procedure after the procurement. Similarly, when the donor lung is oversized for the recipient chest cavity, lung resection can be performed, again, preferably as a back table procedure. In these situations, our preference is to perform a middle lobectomy on the right and a lingulectomy on the left, with a curvilinear wedge resection from the anteroapical section of either upper lobe. When even greater oversizing occurs, implanting only the lower lobe may be optimal.

Once the decision that the donor lung(s) is suitable for procurement is confirmed and this is communicated to the recipient surgical team, preparation for procurement begins by mobilizing the great vessels. Using primarily cautery dissection, the ascending aorta is dissected from the pulmonary artery (PA) and the superior vena cava (SVC). The SVC is mobilized by cautery dissection from the innominate vein to the right atrial juncture. This dissection is continued on to the heart to develop Waterston (interatrial) groove. By fully mobilizing the SVC and adequately developing the interatrial groove, the two most common sites of procurement injury can be minimized: (1) injury to the right PA posterior to the SVC often at the level of the right PA trunk bifurcation, and (2) an inadequate anterior right-sided left atrial cuff. The inferior vena cava (IVC) can usually be mobilized quickly with finger dissection. Commonly, temporary hemodynamic instability occurs, but can be lessened by placing the donor in Trendelenburg position prior to these maneuvers. The IVC should be well exposed below and above the diaphragm to ensure adequate tissue for abdominal and cardiac transplant teams.

The trachea is located by incising the posterior pericardium after gently retracting the SVC laterally and the aorta medially. An umbilical tape can be used to encircle the trachea after the plane is developed manually. Once all procuring teams are ready to cannulate, the donor is fully heparinized (250 units/kg). The ascending aorta is cannulated for standard antegrade cardioplegia. The cannula is secured using a 4-0 Prolene mattress or purse-string suture and a Rummel tourniquet. Placement of the PA cannula occurs in a similar fashion as the aortic cannula with placement comfortably distal to the pulmonary valve, but adequately proximal to assure equal perfusion through both pulmonary arteries. Care should be taken to ensure that the bent tip of the curved cannula is directed toward the bifurcation of the PA. After placement of the cannulae, a 500-μg bolus dose of prostaglandin E_1 (prostacyclin) is administered directly into the pulmonary trunk, near the pulmonary cannula. Immediately following infusion of the prostacyclin, the SVC is ligated doubly or occluded using a vascular clamp and the IVC is divided allowing decompression of the right heart. The IVC should be divided above the pericardial reflection to provide adequate cuff for both the liver and heart implantations. The aorta is cross-clamped and the cardioplegia solution is infused through the aortic cannula. Even if the heart is not to be procured, aortic cross-clamping should be performed to prevent bronchial artery flow. Left heart decompression is achieved by performing a large left atrial appendage incision. Once this incision is created, perfusion of the lungs through the PA catheter is initiated with either Perfadex® or Celsior® preservation solutions (extracellular-based solutions). The height of the solution bag above the PA cannula should not be more than 30 inches. Approximately 3 L of perfusate (35 to 50 ml/kg) are flushed through the PA (antegrade flushing of the lungs). Upon commencing perfusate flow, the thoracic cavity should be bathed in iced saline solution. The procuring surgeon then assesses biventricular filling by directly palpating the heart. Exsanguination and subsequent flow of clear fluid from the left atrial appendage should be observed, as it usually indicates successful antegrade flushing of the preservation solution throughout the pulmonary vasculature. Ventilation of the donor lungs should continue during preservation solution administration. To optimize visibility for the subsequent organ removal, the abdominal IVC should be vented inferiorly and not into the chest.

Following adequate infusion of the appropriate preservation solutions into the aorta and the PA, removal of the heart is performed. It is paramount that adequate tissue for implanting be achieved for both the heart and lung allografts. The heart is retracted to the right and a left atriotomy is initiated midway between the coronary sinus and the left pulmonary veins. The interatrial groove on the right is further developed to ensure adequate atrial cuff size. The initial atriotomy is extended inferiorly and superiorly using scissors, while being mindful of the left pulmonary veins. The surgeon on the left side of the table typically has the optimal view of the right pulmonary vein orifices and should finish creating the atrial cuff while directly visualizing these orifices. Enough cuff width should be available for the left atrial anastomosis. The SVC is divided between the silk ligatures, or by using a vascular clamp one can maximize the length of SVC taken by repositioning the clamp distally at this time. The main PA is transected at the bifurcation and the aorta is divided proximal to the cross-clamp. As with the SVC,

the vascular clamp may be moved distally in order to maximize the length of the aorta. The heart is removed from the field.

After the heart is passed off the table, retrograde flushing of the lungs is performed through each large pulmonary vein orifice. Adequate retrograde perfusion can be determined when the perfusate eluting from the PA becomes clear. Usually this requires approximately 1 to 2 L of the preservation solution (250 to 500 ml of solution for each of the four pulmonary veins). Removal of the remaining thoracic contents ensues by dividing the inferior pulmonary ligaments and posterior attachments. The trachea is mobilized by blunt dissection to at least three cartilaginous rings proximal to the carina. Prior to stapling the trachea, the anesthesia team ventilates the lungs with several bagged breaths. The endotracheal tube is pulled proximally, the lungs are allowed to deflate to approximately two-thirds of vital capacity, and the trachea is stapled with the TA-30 stapler two to three rings above the carina. A second row of staples is immediately placed proximally and the trachea is divided between the two staple lines with a scalpel or scissors. Overexpansion of the lungs is avoided to prevent alveolar stretch injury and subsequent allograft failure.

Heavy scissors can now be used to swiftly divide the remaining posterior mediastinal tissue inferiorly and superiorly in the plane anterior to the esophagus, completely freeing the lungs for removal. Alternatively, the esophagus can be divided after removal of the nasogastric tube proximally and distally with a GIA stapler. The remaining dissection then occurs between the esophagus and the spine with subsequent removal of the lungs en bloc with the esophagus. At this point, they can be bagged together for travel; if the right and the left lungs are going to separate institutions, they require division at the procurement hospital. Our preference is to perform back table dissection at the implanting hospital when possible because of better lighting and equipment.

The lungs are divided by incising the posterior pericardium, the left atrium midway between the two sets of veins, and the main PA at its bifurcation (Fig. 23.2). When lungs are traveling to separate institutions, the left bronchus can be transected between staples just distal to the carina to maintain its inflation pressure for transport. Upon arrival at the transplanting institution, the grafts are prepared for implantation. This primarily involves dividing the main bronchus for each lung either one or two rings proximal to the upper lobe takeoff. Because of the reliance of the bronchus and airways on collateral perfusion from the PA, the length of the bronchus is kept to a minimum. Care should be taken to minimize dissection along the length of the bronchus. The pulmonary arteries are inspected for the presence of pulmonary emboli. If present, they are removed with forceps and retrograde flushing through the pulmonary veins is repeated until the perfusate clears. All structures are inspected for injuries. Reconstruction of vascular injuries or inadequate cuff lengths is best achieved on the back table before initiating implantation. Direct repair is usually possible, but autologous tissue from the explanted lungs, allograft tissue from the donor, and pericardium from the recipient can be used to repair injuries. The arteries and atrial cuff should also be evaluated for residual pericardial attachments, and these should be removed at this time. If retained they may lead to kinking and blood flow occlusion after the anastomosis is completed. Samples of donor bronchus for microbiologic testing can also be taken at this time.

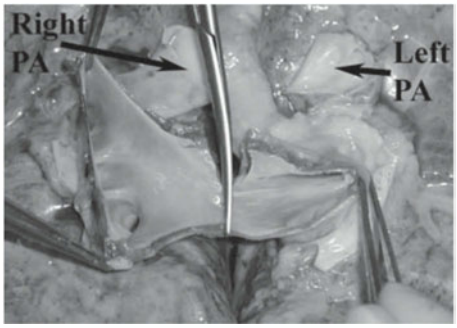


Fig. 23.2. Preparation for implantation of the lungs for separate single lung or sequential bilateral lung transplantation requires division of the donor lungs by incising the posterior pericardium, the left atrium between the 2 sets of veins, and the main pulmonary artery at its bifurcation. It is important to remove pericardial attachments from the vascular cuffs to prevent later anastomotic complications.

RECIPIENT PROCEDURE

Anesthetic Considerations

Suitable large-bore intravenous access is required for potential large volume fluid administration. All recipients have a radial artery catheter and a PA catheter placed via an internal jugular vein. We also routinely place a femoral arterial catheter because of the frequent poor functioning of the radial arterial line, particularly with clamshell incisions. Patients are intubated with a double-lumen endotracheal tube with the tip placed into the left bronchus. Confirmation of tube placement is made by fiberoptic bronchoscopic visualization. In small recipients in which a 37F or greater double-lumen tube cannot be placed, single-lumen intubation is done with the plan to perform the majority of the procedure on cardiopulmonary bypass (CPB). While others have used single-lumen tubes using bronchial blockers to isolate the left lung and advancement of the tube into the left bronchus to isolate the right, we prefer using CPB to avoid airway complications such as tube occlusion from inspissated mucous. When performing SLT, it is preferable to transplant the side that has a normal pleural space and the worst function as determined by quantitative perfusion. All transplants are done with the availability of CPB for the transplant procedure and extracorporeal membrane oxygenation (ECMO) for posttransplant support if necessary. During the procedure, minimal fluids—particularly crystalloid solutions—are administered while inotropic agents (epinephrine) are used liberally to maintain appropriate hemodynamics. Communication by the surgeons to the anesthesiologists prior to retracting on cardiovascular structures that will cause hemodynamic compromise greatly aids in the appropriate administration of boluses of vasoactive agents to prevent hemodynamic compromise. Maintenance of an appropriate red blood cell volume and clotting factors throughout the procedure is paramount. This is especially important in patients with substantial bleeding and those with pulmonary hypertension and passive hepatic congestion. We routinely use transesophageal echocardiography (TEE) to monitor cardiac function and filling, as well as to interrogate the left atrial anastomoses.

Positioning of the Patient and Skin Incision

For left SLTs, the recipient is placed in a right lateral decubitus position, with access to the left groin for possible femoral vein cannulation. The procedure is performed through a standard fifth intercostal space (ICS) posterolateral thoracotomy. Muscle-sparing thoracotomy can be utilized, particularly for patients with COPD. Right SLTs can either be performed through a standard fifth ICS posterolateral thoracotomy or preferably, through a fourth ICS anterolateral thoracotomy. Access for CPB cannulation can be achieved most easily in the chest and the groins are not exposed. For patients undergoing a bilateral lung transplant, we use a bilateral fourth ICS antero-transsternal thoracotomy (clamshell) incision (Fig. 23.3).

The patient is placed in a supine position, and both arms are lifted anteriorly and abducted. The forearms are rested on cushioned support as the arms are flexed slightly at the level of the elbow. A warming blanket or device is routinely used and is positioned from the umbilicus down for right and bilateral transplants and from the thigh down for left lung transplants. It is imperative that the patient's chest and upper abdomen are prepped widely. For a female patient, the skin incision is made at or below the inframammary crease, and the chest is opened through the fourth ICS after a breast flap is developed and retracted superiorly on each side. After ligating the internal mammary pedicles, we routinely divide the sternum transversely. While we have had a low incidence of sternal complications, other groups have reported more sternal wound problems and preferentially perform the procedure without division of the sternum through separate bilateral anterolateral thoracotomies. At this point, it is important to divide the mediastinal pleura superior to the level of the mammary vein and inferior to the level of the pericardium with cautery. The pericardium remains intact at this point, but may be opened if the use of CPB is anticipated. Prior to widely opening the retractors, the intercostal muscles in the fourth ICS are divided in the lateral and posterior direction in order to maximize the overall exposure with the clamshell incision. The overlying muscles (e.g., latissimus dorsi and serratus anterior) are relatively spared laterally. Adhesions are routinely encountered and are particularly prevalent in patients with CF, previous pneumothoraces, or previous pulmonary resections. The adhesions located along the chest wall, diaphragm, and to the mediastinum well anterior to the phrenic nerve should be divided using electrocautery. Dissection of mediastinal adhesions in the area of the phrenic nerve should be done sharply and with great caution. Following lung reduction surgery or previous lobectomies, adhesions to the mediastinum may be such that the phrenic nerve cannot be identified. In these circumstances, it is often prudent to divide the overlying lung tissue using a GIA stapler and to leave a small amount of residual lung to prevent phrenic nerve injury. This is usually accomplished after all other aspects of the pneumonectomy have been performed. Frequently, this is the most time-consuming portion of the operation.

In recipients with small chest cavities such as most patients with IPF, a figure-of-eight traction suture (0-silk) is placed into the fibrous dome of the diaphragm, and this suture is brought out of the chest using a crochet hook passed through a 14-gauge angiocatheter placed in the most inferolateral aspect of the pleural cavity. The diaphragm is retracted inferiorly while pulling down with the suture, and the suture is secured external to the chest wall with a small clamp. To aid in the exposure of the left hilum, a heavy silk retraction stitch is placed inferiorly on the pericardium, posterior to the phrenic nerve, and anterior to the inferior pulmonary vein. The silk suture is passed through a heavy-duty Rummel tourniquet, fashioned from a red rubber catheter, thus allowing the heart to be safely retracted upward and to the right to provide improved exposure during further dissection of the hilar structures and implantation of the donor lung. For bilateral lung transplants, we routinely mobilize (but not divide) both hilar structures completely before initiating the recipient pneumonectomy. Which pneumonectomy is done first is dependent on several factors listed in the relative order of importance: (1) normal chest cavity size and configuration; (2) donor lung quality (a donor lung with contusion, consolidation, or procurement injury is done second); (3) worst native lung function (this side should preferentially be done first); and (4) degree of technical difficulty for each side (more technically difficult side, usually the left, should be done first, which shortens the period in which only one transplanted lung is being perfused).



Fig. 23.3. Positioning and incision utilized for the clamshell incision. The patient is placed in a supine position with the arms lifted anteriorly and abducted. The forearms are rested on cushioned support as the arms are flexed slightly at the level of the elbow. A warming blanket is placed to cover the lower body below the level of the umbilicus. For a male patient the incision is performed at the level of the fourth intercostal space.

The recipient pneumonectomy differs from conventional pneumonectomies in that intrapericardial dissection is always done, division of the PA is done at or beyond the takeoff of the branch vessels, and the bronchus is divided immediately proximal to the upper lobe orifice. The use of the Endo GIA stapler facilitates the ligation and division of the pulmonary arteries and the pulmonary veins. After the lung is removed, the pericardium around the hilar structures is circumferentially incised. Development of Waterston groove on the right side, division of attachments from the roof of the left atrium to the PA and the posterior left atrial wall and pericardium facilitate subsequent left atrial clamp placement (Fig. 23.4). The pulmonary arteries are mobilized proximally. On the right side, this requires division of attachments anteriorly to the SVC. On the left side, attention to avoiding recurrent laryngeal nerve injury is paramount. Particularly in the anterosuperior portion of the left PA, the use of electrocautery is avoided. It is important to establish hemostasis, especially in the posterior mediastinum prior to initiating lung implantation. Particularly in patients

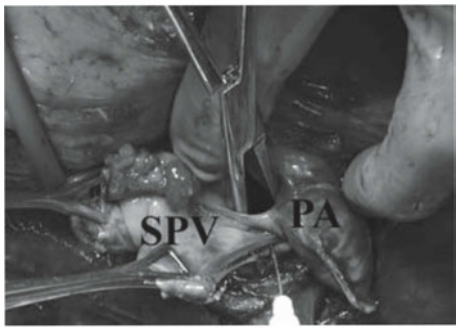


Fig. 23.4. Division of the attachments between the left pulmonary artery and superior pulmonary vein and left atrium is initiated laterally and continued medially. Completing the mobilization of the roof of the left atrium from the right side greatly assists in the placement of vascular clamps on the left atrium and subsequent anastomoses.

with CF and sarcoidosis, bleeding from lymph nodes can be problematic. Wide extirpation of these lymph node groups with clip ligation of the bronchial arteries and other source vessels provides excellent hemostasis, improves exposure, and can assist in preventing vascular anastomotic complications. The vagus nerve located posterior and lateral to the right-sided lymph node group should be identified and protected.

In patients undergoing bilateral lung transplantation, once hemostasis and adequate lengths of PA, left atrium, and bronchus are achieved, two sets of figure-of-eight #1 Maxon pericostal sutures are placed in the posterior and lateral aspect of the thoracotomy wound in preparation for closure of the chest at this point of the operation because of the excellent exposure obtained while the lung is out of the pleural cavity. Similarly, posterior pleural drainage tubes are placed in the costovertebral gutter. We use a 36F right-angle chest tube and a 24F flexible Blake drain. A smaller caliber suction catheter placed into this chest tube and connected to suction facilitates the drainage of blood and fluid throughout the remainder of the case. A flexible drainage tube can also be placed at this time in the axillary space, between the rib cage and the latissimus muscle. In female patients, the tip of this catheter can later be guided into the submammary space near the end of the operation.

Cardiopulmonary Bypass

Most lung transplants can be performed safely and efficiently without the use of CPB. However, CPB should be used whenever recipient hemodynamics, poor systemic perfusion, or technical factors dictate. We use CPB on a planned basis for patients (1) with severe pulmonary hypertension, although occasionally some patients have been done without bypass with usage of nitric oxide and inotropic support; (2) requiring intracardiac procedures such as ASD/VSD closure, valve repair; (3) with a small recipient airway; (4) who are large relative to the donor lung to avoid full cardiac output through a small vascular bed; and (5) with fragile left atrial tissue or inadequate donor left atrial cuff, to allow the anastomoses to be performed without clamps using either cardioplegic arrest or fibrillatory arrest. Particularly for patients with pulmonary hypertension and presumed passive hepatic congestion, the bypass circuit is primed with fresh frozen plasma to mitigate peritransplant coagulopathy. For right and bilateral lung transplants, cannulation is performed in the chest using the right atrial appendage for placement of a 120-degree double-stage cannula for venous return and ascending aorta cannulation for arterial return. For left lung transplants, femoral venous cannulation is done with a long Bio-Medicus venous (15 to 23F) catheter for venous return with descending aorta cannulation for arterial return. Frequently, the femoral venous cannulation is difficult in the decubitus position. In urgent situations venous return can be achieved by cannulating the left PA that allows more time to achieve either femoral venous access. Alternatively, we have been able to access the right atrial appendage following initiation of CPB. While CPB can be performed at any point in the operation as needed, our preference is to perform as much of the dissection including a pneumonectomy prior to initiating bypass. When CPB is utilized, we perform both pneumonectomies and full mobilization of all bronchi, pulmonary arteries, and left atrial cuffs before initiating any portion of the implant procedure.

Lung Implantation

There is little difference between the implantation of the left or right lung. Usually, the left lung implantation is slightly more difficult when performing a bilateral lung transplant because the heart and left atrial appendage impede the exposure for the left atrial anastomosis. Heparin is administered through a central vein (100 U/kg) with a goal ACT of approximately 300 seconds.

The recipient main stem bronchus is cut with a scalpel just proximal to the takeoff of the upper lobe bronchus. After the cartilaginous portion of the bronchus is divided with a blade, a retraction stitch is placed anteriorly to secure control over the bronchus. The remainder of the bronchial wall (posterior, membranous portion) is then divided with a pair of sharp scissors (Fig. 23.5). The recipient bronchus is now ready for anastomosis, and any secretions within the lumen are aspirated. Confirmation of endotracheal tube location is performed. The pleural space, bronchus, and endotracheal lumen are irrigated with an antibiotic solution. After aspirating the antibiotic solution, iced laparotomy sponges are placed posteriorly in the pleural cavity, and the donor lung is orthotopically positioned in the pleural cavity. The recipient bronchus and donor bronchus are aligned, and the bronchial anastomosis is performed with a 4-0 PDS suture in a running fashion. The posterior membranous portion of the bronchial anastomosis is performed first, starting from one corner of the membranocartilaginous junctions (Fig. 23.6). The anterior cartilaginous portion of the bronchus is anastomosed similarly using the running technique. Usually one bronchus is clearly larger, most commonly the recipient. By placing a transition stitch from the outside of the smaller bronchus into the inside of the larger bronchus, an intussusception is created. The orientation of the anatomy is preserved with membrane-to-membrane and cartilage-to-cartilage apposition with approximately a one-ring intussusception. After completing the bronchial anastomosis, it is evaluated for an air leak under water while the anesthesiologist manually inflates the donor lung with room air.

After completing the bronchial anastomosis, the already stapled recipient PA is occluded proximally with a Satinsky vascular clamp, and the staple line is trimmed away. The donor PA is similarly

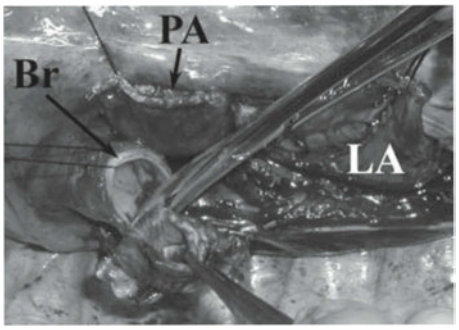


Fig. 23.5. The bronchus is divided anteriorly with a scalpel and a retraction suture is placed anteriorly to aid in the positioning of the bronchus for the subsequent anastomosis. Shown is division of the membranous bronchus. The excised recipient bronchus is sent for culture studies.

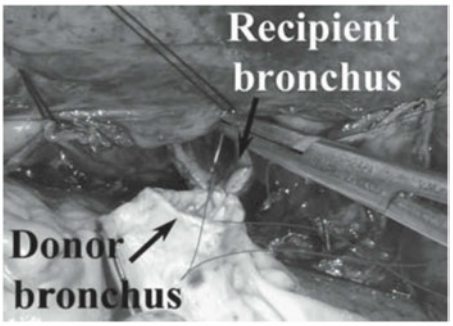


Fig. 23.6. The bronchial anastomosis is created using a running suture of 4-0 absorbable monofilament starting from the membrano-cartilagenous junction. The membranous portion is performed in an end-to-end manner.

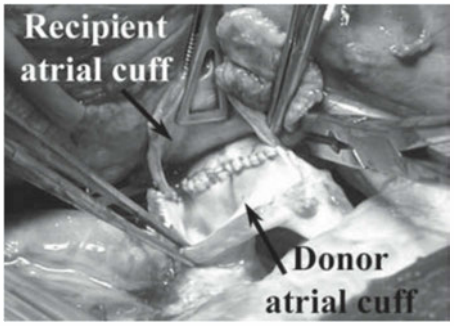


Fig. 23.8. The left atrial anastomosis is created using a running 5-0 polypropylene suture. Retraction of the anterior LA cuff by the assisting surgeon greatly assists exposure, particularly on the left side. Emphasis is place on creating an intima-to-intima approximation without incorporating muscular tissue.

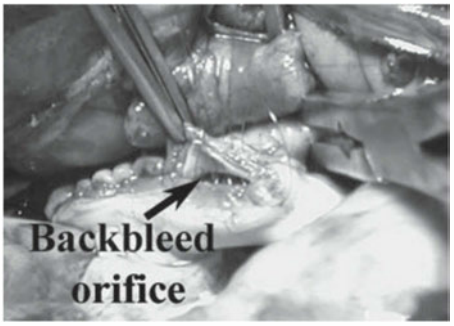


Fig. 23.9. The lungs are de-aired through the anterior most aspect of the left atrial anastomosis. Following partial removal of the PA clamp and full removal of the LA clamp, the anastomosis is secured and tied.

trimmed to an appropriate length. Care must be taken not to leave the donor or recipient PA too long to avoid kinking after the anastomosis is made. The recipient PA and the donor PA are aligned in preparation for the anastomosis, which is begun in one corner with a 6-0 Prolene in a running fashion (Fig. 23.7). On the left side, identifying the apical posterior segmental branches of the donor facilitates proper alignment. Assessment of the PA anastomosis occurs by placing a second vascular clamp (e.g., Harken II) distal to the anastomosis and subsequently removing the previously placed Satinsky clamp from the recipient PA. This method tests the PA anastomosis without reperfusion of the donor lung prematurely.

Retracting the stapled superior and inferior pulmonary vein stumps laterally with Pennington clamps assists in placement of the left atrial clamp. A large Satinsky vascular clamp is placed on the body of the left atrium. The staple lines are excised and the orifices of the superior pulmonary vein and inferior pulmonary vein are connected creating a large, recipient left atrial cuff. By placing a Pennington clamp on the anterior portion of the atrial cuff and retracting superiorly and medially, exposure is greatly improved, particularly on the left side (Fig. 23.8). Exposure for the left-sided anastomosis often requires substantial retraction of the heart. This can be facilitated by retraction of the previously placed pericardial stitch and by manual retraction. During this period, placement of the patient into Trendelenburg position and bolus administration of inotropic agents may be necessary. Occasionally, widely opening the pericardium to allow the heart to herniate anteriorly and to the right may be necessary to provide exposure and suitable hemodynamics. An endothelial to endothelial anastomosis is created using a running suture of 5-0 Prolene with a concerted effort to include the intimal layer and exclude the muscular tissue. Once the posterior portion of the anastomosis is completed, an intravenous bolus of methylprednisolone (500 mg) and mannitol (25 gm) is given.

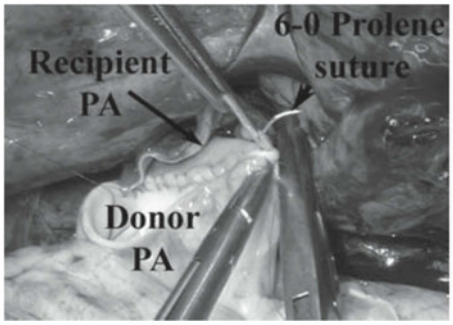


Fig. 23.7. The pulmonary artery anastomosis is performed in an end-to-end manner using a running 6-0 polypropylene suture. Frequently, a substantial size discrepancy exists that can be addressed by elongating the distensible smaller PA and taking numerous bites.

Reperfusion

With the last few throws of the 5-0 Prolene suture left loose anteriorly (Fig. 23.9), the vascular clamp on the PA is partially released to allow blood to flow into the newly implanted lung through the PA anastomosis in order to remove air from within the pulmonary vasculature. Immediately prior to tying the 5-0 Prolene suture, the Satinsky clamp on the recipient left atrium is released to force out any residual air. Reperfusion is carefully controlled to minimize the risk of PGD. Controlled, low-pressure reperfusion of the lung is achieved by gradually releasing the PA clamp over 10 to 15 minutes. Ventilation with room air is initiated by hand and then by mechanical ventilation. Pressure control mode of ventilation is preferred with 5 to 8 cm of H_2O of PEEP, a distending pressure of 16 to 22 cm of H_2O, and minimal FiO_2, preferably less than 30%. During this reperfusion period, hemostasis of the donor lung tissue is achieved. At the time of full release of the PA clamp, the PA pressures should be normal. Use of nitric oxide and loop diuretics are initiated if PA pressures are elevated or if poor systemic oxygenation is present. An additional anterior 28F pleural tube is placed on each side.

When implantation occurs on CPB, as aforementioned, bilateral pneumonectomies are performed, posterior thoracotomy sutures and pleural drains are placed, and both lungs are implanted prior to reperfusion. The first lung that is implanted is packed in ice slush during implantation of the second lung. Methylprednisolone (1 g) and mannitol (50 g) are administrated prior to reperfusion. While the lungs are ventilated with room air, gradual reperfusion is initiated starting at 5 to 10 mmHg mean PA pressure and increasing by 5 mmHg every 5 minutes until normal systemic pressures are achieved. Reperfusion of the lungs with oxygenated hypocarbic blood may be especially advantageous to maximize pulmonary vascular recruitment and parenchymal recovery.

In patients undergoing double-lung transplants without CPB, once appropriate hemodynamics and oxygenation are confirmed after reperfusion of the first transplanted lung, the remaining native lung is removed. Implantation proceeds in a fashion similar to that of the first lung. The performance of bilateral dissection and mobilization

initially greatly reduces the duration in which the entire systemic blood flow passes through only one transplanted lung, which also appears to minimize the development of PGD.

For SLTs, closure of the posterolateral thoracotomy is conventional with large-bore chest tubes placed in the anterior and posterior pleural space. For bilateral transplants performed using a clamshell incision, the anterior aspect of the clamshell opening is approximated with three sets of #5 wires, one simple set in the midline of the sternum, and one set of figure-of-eight on each side of the midline with the lateral aspect placed lateral to the mammary pedicle. The remainder of the clamshell opening is reapproximated with a series of #1 Maxon sutures in a figure-of-eight fashion. As described earlier, the Maxon sutures on the lateral most aspect of the clamshell opening have been placed while the native lungs were either deflated or removed. If not already done, a Blake drain into each of the axillary space is placed. The pectoral fascial layer, the subcutaneous layer, the subdermal layer, and the skin are then approximated.

Chest tubes are placed to suction except in circumstances in which the recipient pleural space is substantially larger than the donor lung size. In these situations, chest tubes are left to water seal to prevent overdistension of the lung allograft, stretch injury, and severe PGD. This situation can be identified by marked differences in tidal volumes between when the pleural tubes are on or off suction.

After dressings are applied, the double-lumen endotracheal tube is removed and a large, single-lumen endotracheal tube is placed. Fiberoptic bronchoscopy is performed to (1) assess the adequacy of the anastomosis, (2) clear secretions, (3) assess for the development of PGD as manifested by severe pulmonary edema, and (4) evaluate for torsion or malrotation of the allograft manifest by abnormal, crescent-shaped distal airways.

Primary Graft Dysfunction

Through the use of extracellular-based pulmonary perfusion solutions, the addition of retrograde pulmonary perfusion to the preservation regimen, minimal oxygen exposure at the time of reperfusion, control of the initial pressure and flow through the lung allograft, and the use of oxygen free-radical scavengers, the incidence of severe PGD has decreased from 15% to 25% to as little as 5%. However, development of severe allograft dysfunction as demonstrated by marked hypoxia, pulmonary edema, elevated PA pressures, and poor compliance requires immediate investigation for reversible causes. Most importantly anastomotic or mechanical causes, especially venous outflow problems, need to be identified and corrected immediately. With respect to venous outflow obstruction, if correction is delayed for more than 4 to 6 hours it is unlikely that any reasonable allograft recovery will be achieved. Interrogation of the anastomoses includes visual inspection to assess for torsion or kinking, TEE to assess for turbulent or absent pulmonary vein flow as well as left atrial anastomotic quality, the presence of intraluminal clot, and direct measurement of pressures across the anastomosis. Cardiac etiologies usually can be identified by TEE including left-sided valvular abnormalities, left ventricular failure, intracardiac shunts, and tamponade. We routinely obtain a quantitative perfusion scan within the first few hours after the transplant with lobar or greater perfusion defects necessitating further assessment, usually by operative exploration. Asymmetric pulmonary edema or crescent-shaped airways on bronchoscopic evaluation may be indicative of mechanical problems. PGD can be caused by humoral lung injury from circulating antidonor antibodies. These antibodies are identified by donor–recipient crossmatching. Treatment of donor-specific antibodies is multimodal, including plasmapheresis, column absorption, intravenous immunoglobulin, and anti-B cell therapies such as anti-CD20 (rituximab).

Severe PGD causes significantly contribute to the majority of early post-transplant deaths. Unless reversible causes could be identified, treatment has been supportive with optimization of ventilator parameters, inotropic support, and nitric oxide. We have shifted to a strategy of early institution of venovenous (V-V) ECMO when recipients develop severe pulmonary edema or require $FiO_2 > 60\%$. V–V cannulation is typically instituted via the right femoral vein with a venous catheter (Bio-Medicus) and the left internal jugular vein with a pediatric arterial cannula (Medtronic, Minneapolis, MN). Cannulae are placed percutaneously using a modified Seldinger technique over a guidewire following serial dilatations. The circuit consists of a hyaluron-based heparin-coated 3/8 inch tubing with a Quadrox D polymethylpentene (PMP) oxygenator and Jostra Rotaflow pump (MAQUET Cardiopulmonary AG, Hirrlingen, Germany). The optimal placement of circuit inflow and outflow ports are determined by the level of recirculation noted in the system. ECMO flows are approximately 2.5 to 3.5 L/minute with a sweep gas flow adjusted to maintain the pCO_2 close to 30 mmHg so as to maximize pulmonary vasodilation. During V-V ECMO support, a protective ventilatory strategy is used including low oxygen and low pressure ventilation. Weaning from V-V ECMO involves discontinuing membrane gas flow and increasing ventilatory parameters as needed. No increase in anticoagulation is required for V-V ECMO weaning. Using this strategy, patients are weaned from ECMO, usually after 3 to 5 days of support, because pulmonary vascular resistance decreases following institution of ECMO and pulmonary capillary leak usually resolves more quickly (often within 24 hours). The 30-day survival in patients requiring V-V ECMO at our institution is approximately 90%.

Principles of Postoperative Care

While an extensive discussion of postoperative care is beyond the scope of this text, a few principles are important. Optimal pain control, especially for bilateral transplant recipients, is achieved with epidural analgesia. Most patients with good allograft function should be extubated within 24 hours of the transplant and can be discharged from the hospital within 10 to 14 days of the transplant. While the optimal immunosuppressant regimen is not known, most programs use a triple-drug regimen including a calcineurin inhibitor (cyclosporin or tacrolimus), an antiproliferative agent (Imuran or mycophenolate mofetil), and corticosteroids. Induction therapy using either an anti-CD25 agent (basiliximab or daclizumab), or an immune cell depleting strategy such as polyclonal anti-T cell agents (thymoglobulin, ATGAM), or most recently anti-CD52 (Campath) are used in approximately 50% of lung transplant recipients. While acute rejection occurs commonly after lung transplant with 40% to 60% of patients having at least one episode of rejection in the first 6 months, it is an uncommon cause of early mortality. Mortality early after lung transplant is most commonly secondary to poor allograft function, infection, and neurologic and cardiac events. The immunosuppression and infection prophylaxis protocols used by the Duke Lung Transplant Program are outlined in Tables 23.1 and 23.2.

A factor that appears to be related to lung allograft injury that has been underestimated in its prevalence is aspiration injury. In addition to the infrequent, classic aspiration pneumonitis, repetitive injury

Table 23.1 Duke University Immunosuppression Protocol

Preoperative

FK506: 0.04 mg/kg PO on admission

Azathioprine: 2 mg/kg IV on induction of anesthesia

Intraoperative

Solu-Medrol: 500 mg IV prior to reperfusion of each transplant lung if bilateral; 500 mg IV prior to reperfusion of single transplanted lung.

Basiliximab (Simulect): 20 mg IV following induction of anesthesia

Postoperative

FK506: 0.04 mg/kg given sublingually every 12 h. *For patients who will also receive voriconazole or itraconazole postoperatively, reduce dose to 0.02 mg/kg.* Adjust to achieve a trough level of 10–15 ng/ml. Switch to PO when GI motility is restored.

*If creatinine >1.5, target FK506 level 8–12.

Azathioprine: 2 mg/kg IV or PO daily to maintain WBC > 4,000.

Steroids: Solu-Medrol 125 mg IV q12h × 48 h, then prednisone 20 mg PO daily. **Basiliximab (Simulect):** 20 mg IV on postoperative day 4.

Maintenance

FK506: Every 12-h dosing adjusted to maintain trough FK506 levels—mo (0–6), 10–15 ng/ml; > 6 mo, 8–12 ng/ml. A lower level may be required if patient develops significant renal insufficiency.

Azathioprine: 2 mg/kg/d PO adjusted to maintain WBC > 4,000.

Prednisone: Mo (0–3): 20 mg/d; Mo (4–6): 15 mg/d; Mo (>6): 10 mg/d.

Treatment of Rejection

Minimal or mild rejection episodes (ISHLT Grade 1 or 2): Solu-Medrol 500 mg IV daily × 3 doses followed by oral prednisone taper starting at 60 mg and decreasing by 5 mg/d until original dose is reached.

Moderate (ISHLT Grade 3) or steroid-resistant rejection: RATG (thymoglobulin) 1.5 mg/kg IV for three doses. Premedicate 30 min prior to each dose with Solu-Medrol 40 mg IV, diphenhydramine 50 mg IV, and acetaminophen 650 mg PO.

secondary to microaspiration appears to occur commonly. Factors that increase the risk of aspiration include the high prevalence of gastroesophageal reflux in patients with end-stage lung disease, particularly in those patients with IPF and CF. Gastroesophageal reflux is even more common and of greater severity following lung transplant because of issues related to both the medication regimen and vagal nerve injury. Vagal nerve injury also contributes to gastroparesis that can exacerbate the amount of esophageal reflux. Additionally, abnormal oropharyngeal swallowing appears to occur in many patients often as a result of left recurrent laryngeal nerve injury. We routinely assess all patients for gastroesophageal reflux both before and soon after lung transplant and we perform an endoscopic swallow evaluation after transplant prior to initiating oral intake. In patients with severe gastroesophageal reflux, surgical fundoplication is performed early after the lung transplant, often during the posttransplant stay. Patients who fail their swallow evaluation receive nutritional support via enteric tube placed distal to the ligament of Treitz until their oropharyngeal function normalizes. In patients with vocal cord paralysis or paraparesis, vocal cord medialization is performed.

Bronchoscopic examination is performed frequently in the early posttransplant period. It is carried out both to observe the airway anastomoses and to obtain transbronchial biopsies to investigate new infiltrates, fevers, or allograft dysfunction as manifested by worsening oxygenation, hypercapnia, and decreased spirometric values. While pulmonary embolism is uncommon after lung transplant, the potentially devastating consequence of pulmonary infarction in the face of the known lack of bronchial artery perfusion that characterizes a newly implanted lung allograft requires a high degree of vigilance with respect to diagnosis and treatment. Surgical embolectomy of large pulmonary emboli occasionally may be indicated.

CONCLUSIONS

Tremendous progress has occurred in lung transplantation over the last two decades. One-year patient and allograft survival now compares favorably to those of liver and cardiac transplantation. As with all solid-organ transplantations, the shortage of available donor organs and the development of chronic allograft injury are the primary limitations to a greater applicability to the treatment of end-stage lung diseases. While the potential development of expanded organ sources through xenotransplantation, lung organogenesis, and ex vivo perfusion systems will greatly impact the future numbers of lung transplants performed, there appears to be a substantial number of suitable organs, especially lungs, that could become available if the majority of donors were evaluated appropriately. With a better understanding of the causes of lung allograft injury and ongoing work toward developing immunologic tolerance, improved long-term outcome is achievable.

SUGGESTED READINGS

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Table 23.2 Duke University Infection Prophylaxis

Bacterial

Standard Regimen[a]

Ceftazidime: 2 g IV preoperatively on induction per anesthesia, then 1 g IV every 8 h for 7–10 d or until invasive lines are out. (Adjust doses for renal insufficiency.)

Vancomycin: 1 g IV preoperatively on induction per anesthesia, then 1 g IV every 12 h for 7–10 d or until invasive lines are out. (Adjust doses for renal insufficiency.)

CMV[b]**/HSV**[c]**/EBV**[d]

Donor negative/recipient negative

Leukoreduced red blood cell transfusions only. If HSV negative, no prophylaxis; if HSV positive, acyclovir 200 mg PO every 8 h for 12 wk.

Donor positive/recipient negative

Ganciclovir 5 mg/kg IV every 12 h for 4 wk, followed by ganciclovir 5 mg/kg IV daily for 10 wk, followed by Valcyte 450 mg PO daily, indefinitely.

Donor negative/recipient positive OR donor positive/recipient positive

Ganciclovir 5 mg/kg IV every 12 h for 2 wk followed by ganciclovir 5 mg/kg IV daily for 2 wk.

Pneumocystis carinii

Septra DS 1 tablet PO every Monday, Wednesday, and Friday starting 1 wk postoperatively and continuing indefinitely.

If sulfa allergy, Dapsone 50 mg PO daily *or* aerosolized pentamidine 300 mg every mo continuing indefinitely.

Fungal

Nystatin suspension 5 ml swish and swallow 4 times/d for oral candida prophylaxis. Continue for 6 mo.

Inhaled amphotericin B 50 mg daily for 4 d, then weekly while hospitalized immediately post-transplant (dose to be reduced to 25 mg once patient is extubated).

Toxoplasmosis gondii (use only for donor positive/recipient negative)

Septra DS 1 tablet daily starting 1 wk postoperatively (also covers PCP prophylaxis).

If sulfa allergy, pyrimethamine (Daraprim) 50 mg PO daily and folinic acid (leucovorin) 10 mg PO daily for 6 mo (use in addition to pentamidine for PCP prophylaxis).[e]

[a]The standard regimen should be amended as indicated to include (1) coverage for any other known preoperative pathogens in the recipient. This is particularly indicated for recipients with cystic fibrosis, bronchiectasis, and other septic lung diseases; (2) coverage for any additional organisms identified from donor bronchial washings. If OR and first BAL cultures are negative at 72 h postoperatively, change IV antibiotics to Levaquin or Moxifloxacin 500 mg PO daily for 7 d.
[b]Use Valcyte 450 mg PO BID for the duration of the above protocols as long as the patient is tolerating oral intake, normal GI function, WBC >5,000, and creatinine <1.5.
[c]For all EBV-negative recipients: Ganciclovir 5 mg/kg IV every 12 h for 4 wk, followed by Valcyte 450 mg PO daily, indefinitely.
[d]Patients receiving RATG (thymoglobulin) therapy treated with Ganciclovir at a dose of 5 mg/kg IV every 12 h (or appropriate treatment dose based on renal function) for 3 wk for CMV prophylaxis.
[e]If patient intolerant of daily pyrimethamine, use Dapsone 50 mg daily with pyrimethamine 50 mg weekly and folinic acid 10 mg weekly.

EDITOR'S COMMENTS

Lung transplantation is now a well-accepted therapeutic option for many patients with end-stage lung disease, yet the 5-year survival is less than 50%. The major issue relating to long-term survival continues to be the problem of obliterative bronchiolitis. Efforts have been made to improve overall survival by changing donor lung allocation methodology from simply time on the waiting list to one where patients are given a lung allocation score (LAS) that directs donor lungs to those patients who are most likely to derive the maximum benefit. The LAS is calculated from a series of formulas that take into account the statistical probability of a patient's survival in the next year without a transplant, how long that survival would be, the probability of survival following a transplant, and the projected length of survival posttransplant. In addition to this revised allocation methodology, marginal donor lungs are now being more frequently utilized. The development of ex vivo lung perfusion systems that allow for improving some of these marginal lungs is also likely to add to the donor pool. The lungs are the first organ to deteriorate in the brain-dead patient and thus are usable for transplant far less than any of the other organs.

The issue of single lung versus bilateral sequential lung transplants remains somewhat controversial though the long-term survival when both lungs are transplanted is superior. Many centers now use single-lung transplants very sparingly despite the fact that overall less recipients benefit.

I was particularly intrigued by the Duke group's low threshold for using venovenous ECMO for patients who are having oxygenation problems and specifically the outstanding success that they have experienced in this challenging group. In addition, the recognition of the role that so-called microaspiration plays in allograft dysfunction has been woefully underestimated and was initially brought to our attention by Dr. Davis and his colleagues at Duke. It has long been thought that there was an element of swallowing dysfunction following pneumonectomy; so it is not surprising that many patients post lung transplant have significant swallowing issues as well as reflux.

The issue of primary graft dysfunction continues to plague lung transplantation and has implications for long-term survival. Better lung preservation regimens have reduced the incidence but certainly not eliminated it.

LRK

24 Thoracic Outlet Procedures

Robert W. Thompson and Valerie B. Emery

Thoracic outlet syndrome (TOS) is a group of conditions caused by compression of the neurovascular structures that serve the upper extremity as they pass through the lower part of the neck, behind the clavicle, and over the first rib (Fig. 24.1). The clinical presentation of TOS depends on the specific structures compressed, giving rise to three distinct conditions: (1) *Neurogenic TOS* (NTOS), caused by compression and irritation of the brachial plexus nerves; (2) *Venous TOS* (VTOS), caused by compression of the subclavian vein leading to venous thrombosis; and (3) *Arterial TOS* (ATOS), caused by compression of the subclavian artery, leading to degenerative changes and thromboembolism. Although all three types of TOS are considered to be relatively uncommon, clinical recognition and appropriate treatment of these conditions is crucial to prevent disability in young active individuals.

NEUROGENIC THORACIC OUTLET SYNDROME

NTOS is the most frequent form of thoracic outlet compression, occurring in 85% to 90% of patients. It is due to compression and irritation of the brachial plexus nerve roots within the scalene triangle or underneath the pectoralis minor muscle tendon in the subcoracoid space. NTOS is considered to be caused by a combination of variations in anatomy (such as anomalous scalene musculature, aberrant fibrofascial bands, or cervical ribs) and previous neck or upper extremity injury that has resulted in scalene or pectoralis minor muscle spasm, fibrosis, and other pathologic changes. These muscular alterations, in turn, lead to compression and irritation of the adjacent brachial plexus nerves.

Brachial plexus nerve root compression can result in diverse symptoms that usually include pain, numbness, and tingling (paresthesia) in the neck, shoulder, arm, and hand. The intensity of these symptoms can vary, depending on levels of upper extremity activity, and are typically exacerbated with position, especially arm abduction and elevation. Although many patients with NTOS have relatively mild symptoms, with a slow gradual progression interspersed by occasional exacerbations, some exhibit a steady progression in the severity of symptoms leading to increasing and significant disability.

The diagnosis of NTOS is based on clinical evaluation, with supplemental testing procedures to exclude alternative or coexisting conditions (Table 24.1). On physical examination there is usually well-localized tenderness to palpation over the supraclavicular scalene triangle and/or the infraclavicular subcoracoid space, usually associated with reproduction of upper extremity symptoms. Most patients with NTOS report a rapid reproduction of upper extremity symptoms with provocative positional maneuvers, such as the upper limb tension test (ULTT) or the 3-minute elevated arm stress test (3-min EAST). Physical examination of patients suspected to have NTOS should also exclude any evidence of arterial or venous compromise to the upper extremity. Plain anteroposterior chest radiographs are useful to determine the presence or absence of a cervical rib, but other imaging studies of the brachial plexus are not specifically helpful in diagnosis. Conventional electrophysiologic testing (electromyography and nerve conduction studies) is often performed to exclude peripheral nerve compression disorders or cervical radiculopathy, but these tests are usually negative or nonspecific in NTOS. Image-guided anterior scalene muscle and/or pectoralis minor muscle blocks with a short-acting local anesthetic can provide support for the clinical diagnosis of NTOS, as well as help predict the reversibility of symptoms with treatment.

Initial treatment for NTOS is based on physical therapy to relieve muscle spasm, improve postural disturbances, enhance functional limb mobility, strengthen associated shoulder girdle musculature, and diminish repetitive strain exposure in the workplace. Incorrect approaches to physical therapy can result in worsening of symptoms and premature failure of conservative management. In most patients with mild NTOS or symptoms of short duration, significant improvement is observed within the initial 4 to 6 weeks and therapy is then continued on an "as needed" basis. Because NTOS is often a chronic condition subject to occasional "flare-ups" of acute symptoms (often related to overuse activities or new injury), the patient should continue regular physical therapy exercises during long-term follow-up.

Surgical treatment is recommended for patients with NTOS when the clinical diagnosis is sound, the patient has substantial disability (symptoms interfere with daily activities and/or work), and there has been an insufficient response to an appropriate course of physical therapy. Surgical treatment may also be recommended in selected patients with persistent or recurrent symptoms of NTOS following a previous operation, when there has been no response to appropriate conservative measures. Thoracic outlet decompression for NTOS may be accomplished by transaxillary operations focused on first rib resection, or by supraclavicular operations that include scalenectomy, first rib resection, and brachial plexus neurolysis. For patients with symptoms of NTOS referable to the subcoracoid space, surgical decompression may include pectoralis minor tenotomy as an addition to transaxillary or supraclavicular thoracic outlet decompression, or as an isolated procedure when this site is the dominant location of nerve compression symptoms.

VENOUS TOS

VTOS is characterized by subclavian vein compression between the clavicle, subclavius muscle, costoclavicular ligament, and first rib. The pathogenesis of VTOS involves repetitive extrinsic compression of the subclavian vein during activities involving arm

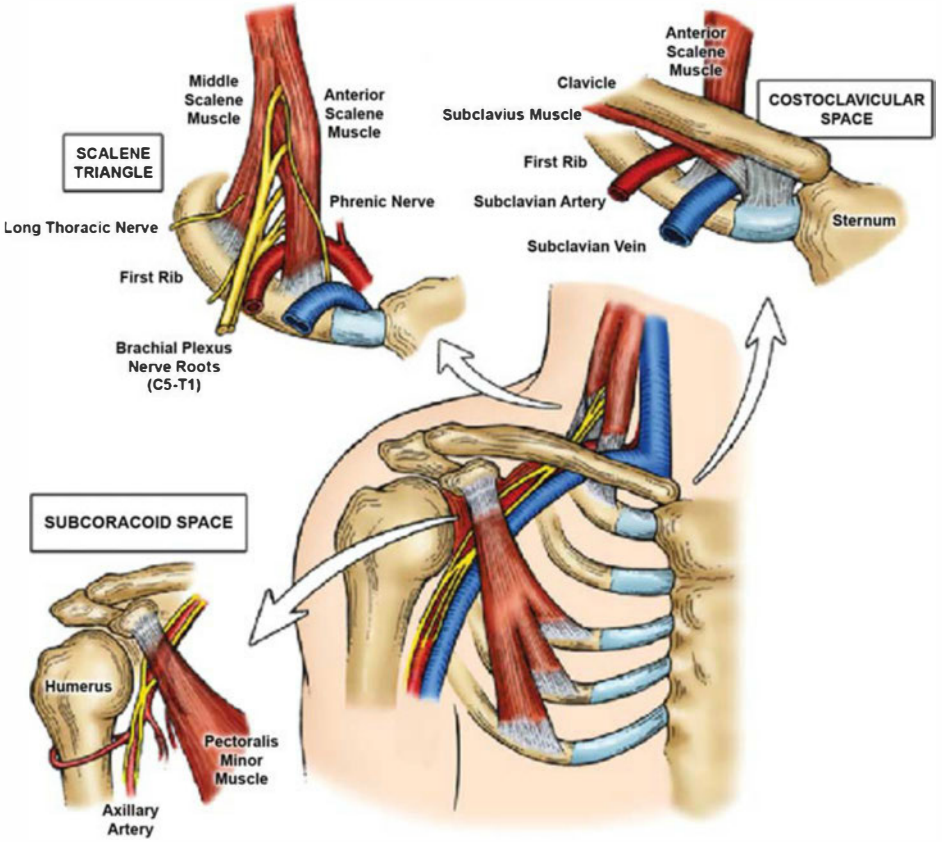


Fig. 24.1. Anatomy of the thoracic outlet. Overview of the anatomy of the thoracic outlet focusing on the scalene triangle, the costoclavicular space, and the subcoracoid space. With permission from Fischer JE et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

elevation or exertion, leading in time to chronic focal venous injury and progressive fibrous stenosis due to scar tissue formation and contraction around the outside of the vein, as well as fibrosis and wall thickening within the wall of the vein itself.

The initial phase of VTOS is usually asymptomatic, due to simultaneous expansion of collateral veins passing around the narrowed subclavian vein, but stagnant blood within the damaged segment of the subclavian vein eventually leads to thrombotic occlusion. Growth and peripheral extension of this clot into the axillary vein can further obstruct critical collateral veins, resulting in the acute clinical presentation characterized as axillary-subclavian vein "effort thrombosis" (Paget-Schroetter syndrome). Patients with subclavian vein effort thrombosis typically present with abrupt spontaneous swelling of the entire arm, often with cyanotic (bluish) discoloration, heaviness, and aching pain. Unlike more common forms of deep venous thrombosis (DVT), VTOS tends to occur in young, active, and otherwise healthy individuals, with no underlying blood clotting disorder. Pulmonary embolism from clot within the proximal subclavian vein may also occur, particularly with motion of the arm, but this is infrequent compared to DVT in the lower extremities.

The stereotypical clinical presentation of axillary-subclavian vein effort thrombosis is usually sufficient to establish the diagnosis of VTOS. Although venous duplex studies of the upper extremity may be used to confirm the presence of DVT, duplex imaging of the subclavian vein is inaccurate and cannot be used to exclude the diagnosis. In contrast, imaging studies such as magnetic resonance angiography or catheter-based venography provide more definitive information on the location and extent of axillary-subclavian vein thrombosis, and can be performed with the arms in elevation when there is incomplete subclavian vein obstruction. The distinct advantage of using catheter-based venography as the initial diagnostic step is that this can be followed immediately by thrombolytic therapy, which is often considered the first step in treatment. It is important to note that while blood coagulation testing is often performed in patients with upper extremity DVT, these tests are usually negative in those with VTOS and add little to the initial diagnosis or management.

Table 24.1 Diagnostic Features of Neurogenic Thoracic Outlet Syndrome

Unilateral or bilateral upper extremity symptoms present for at least 12 wk, meeting at least one criterion in each of the following three categories and not satisfactorily explained by another condition:

1. Presenting Symptoms
   - Pain in the neck, anterolateral chest, medial upper back, shoulder, arm and/or hand
   - Complaint of numbness or paresthesias in the hand, especially in digits 4 and 5
   - Complaint of weakness in the arm or hand
   - Paresthesias radiate from the supraclavicular or infraclavicular space to the arm and/or hand
2. Clinical History
   - Symptoms began after head, neck, or upper extremity injury (occupational or recreational)
   - Symptoms exacerbated by overhead or work-related activities, including repetitive strain
   - Presence of a cervical rib or previous fracture of the clavicle or first rib
3. Physical Examination
   - Local tenderness on palpation over scalene triangle and/or subcoracoid space
   - Reproduction of hand or digit paresthesias on palpation over scalene triangle and/or subcoracoid space
   - Weak handgrip, intrinsic muscles, or digit 5, or thenar/hypothenar atrophy
   - Positive upper limb tension test or 3-min elevated arm stress test (EAST)

Exclusion of other conditions typically includes nonspecific or negative findings on physical examination (Spurling's test, axial compression test, Tinel's sign over the carpal tunnel or cubital tunnel, and Phalen's test), imaging studies (MRI of cervical spine and shoulder), and conventional electrophysiologic tests (upper extremity electromyography and nerve conduction studies).

Adapted from preliminary consensus diagnostic criteria developed by the Consortium for Research and Education on Thoracic Outlet Syndrome (CORE-TOS). With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

Following the diagnosis of subclavian vein effort thrombosis by contrast venography, catheter-based thrombolytic therapy is recommended to reduce the amount of thrombus within the axillary and subclavian veins. In recent years this has been facilitated by pharmacomechanical approaches, where thrombolysis can usually be accomplished in a single setting. In most cases this results in a rapid reduction in the burden and extent of thrombus, and reveals a residual high-grade focal stenosis or occlusion of the subclavian vein at the level of the first rib. Balloon angioplasty may be used to reduce the degree of residual stenosis, but this is often unsuccessful at overcoming extrinsic bony compression or scar tissue within the wall of the vein. Over the past decade it has also become increasingly clear that placement of stents within the subclavian vein frequently leads to poor outcomes and should therefore be avoided. Following catheter-based venography and thrombolysis, patients with VTOS are maintained on therapeutic anticoagulation while considering further treatment options.

One option for treatment of VTOS consists of conservative treatment with chronic anticoagulation and long-term restrictions in arm activity, in the hope that recurrent venous thrombosis will not occur and that increased collateral development will eventually compensate for any residual axillary-subclavian vein obstruction. Even with satisfactory anticoagulation this approach is associated with recurrent thrombosis or chronic venous congestion in 50% to 70% of patients, and there is no information available on the optimal duration of anticoagulation for this condition. Lifelong anticoagulation and aggravation of symptoms during active use of the arm may also require considerable limitations, which are usually unacceptable for young active patients.

Surgical treatment is recommended within several weeks for almost all patients with VTOS, both to avoid long-term disability from venous obstruction and the need for long-term anticoagulation, and to permit a prompt return to normal activities. Surgical treatment is also recommended in patients with previous axillary-subclavian vein thrombosis that have remained symptomatic despite anticoagulation and restricted activity, as well as for asymptomatic individuals in whom long-term anticoagulation and restrictions on upper extremity activity are undesirable.

ARTERIAL TOS

ATOS is caused by subclavian artery compression within the scalene triangle, leading to the development of fixed subclavian artery stenosis, occlusion, or poststenotic subclavian artery aneurysms. ATOS almost always occurs in association with a congenital cervical rib or other bony anomaly. The development of subclavian artery surface ulceration or aneurysmal dilatation is often accompanied by mural thrombus formation, which frequently leads to distal thromboembolism with hand and/or digital ischemia. A second form of ATOS is observed in overhead throwing athletes, associated with occlusive or aneurysmal lesions of the distal axillary artery. These lesions are caused by repetitive trauma from hyperextension during the throwing motion, in which the axillary artery is compressed by forward motion of the head of the humerus. The thromboembolic complications of these lesions are similar to those of ATOS caused by subclavian artery lesions at the level of the first rib.

Patients with ATOS often present with acute thromboembolism in the upper extremity, characterized by the sudden onset of hand pain and weakness, numbness and tingling, and cold and pale fingers. Patients with more longstanding ischemia may present with chronic arm fatigue or claudication, nonhealing wounds, or ulcerations in the fingers. Subclavian artery occlusions or aneurysms may also be asymptomatic, with occlusions identified by a significant blood pressure differential between arms and aneurysms presenting as a nontender pulsatile mass in the lateral neck.

ATOS is usually readily suspected by clinical findings. The diagnosis may be confirmed by noninvasive vascular laboratory studies, such as duplex imaging and segmental arterial waveform analysis. Plain radiographs are important to determine if a cervical rib or first rib anomaly is present. Positional angiography (with either contrast-enhanced magnetic resonance imaging, computed tomography, or catheter-based arteriography) is typically performed to determine the presence or absence of a subclavian artery aneurysm. Similar imaging studies are performed in patients that have presented with upper extremity arterial thromboembolism to determine if a proximal source of embolism exists in the subclavian or axillary artery.

Immediate anticoagulation and urgent surgical treatment is recommended in patients with ATOS and acute upper extremity ischemia due to thromboembolism. Initial treatment may involve brachial artery exposure and thromboembolectomy to improve the distal circulation in the hand, with or without intraarterial infusion of thrombolytic and vasodilator agents. Treatment of the distal circulation should be immediately followed by thorough arteriographic assessment of the axillary and subclavian arteries, to establish a proximal source of thromboembolism, followed by thoracic outlet decompression and arterial reconstruction. For patients with stable distal circulation found to have ATOS, direct elective surgical treatment is recommended. Therapeutic anticoagulation and antiplatelet therapy are continued in all patients from the time of diagnosis until definitive surgical treatment.

SURGICAL PROCEDURES

Transaxillary Decompression

The patient is positioned supine or in lateral position under general anesthesia, with the back of the table raised about 30 degrees. The affected side is elevated with a small towel pack placed behind the shoulder. The arm is circumferentially prepped and wrapped in stockinette, with a sterile field that includes the neck, upper chest, and posterior shoulder to the scapula. Some surgeons prefer to place the arm on a table or fixed to a crossbar, but the same positioning can be accomplished by a reliable, flexible, and sturdy assistant.

A transverse skin incision is made at the lower border of the axillary hairline and carried through the subcutaneous tissues directly to the chest wall. A tissue plane extending to the apex of the axilla is established by blunt dissection near the chest wall and the long thoracic, thoracodorsal, and second intercostal brachial nerves are identified to avoid direct injury. Specific attention is directed to avoid excessive elevation of the arm, to prevent stretch injury to the intercostobrachial cutaneous nerve (which can result in troublesome postoperative pain and numbness along the medial aspect of the arm). The first rib is identified by palpation along the chest wall, and using a Deaver retractor to gently lift away the subcutaneous tissues and axillary contents, the rib is exposed in the upper aspect of the wound. The arm should be carefully elevated to facilitate this exposure and the operating surgeon should use a directed fiberoptic headlight to illuminate the operative field. Some surgeons have advocated the use of video-assisted exposure, using an endoscopic camera, as an

adjunct to visualization. Throughout the transaxillary procedure the operating surgeon must be continually aware of the position of the assistants and retractors with respect to the brachial plexus nerve roots and subclavian blood vessels, and attention must be given to avoid traction on the long thoracic nerve that exits the plane between the middle and posterior scalene muscles before coursing over the first rib to the serratus anterior muscle. A staged approach with periodic reinspection of the retractors and relief for the assistants is recommended.

Following exposure of the first rib, the subclavian vein and artery are identified along with the intervening anterior scalene muscle. These structures are carefully dissected to the point that the anterior scalene tendon can be encircled with a right-angle clamp, just above its insertion on the first rib (typically palpable as a slight bony prominence). While avoiding the phrenic nerve to expose several centimeters of the anterior scalene muscle superior to the first rib, the muscle is divided with a scissors. Because reattachment of the divided anterior scalene muscle is one of the principal causes of recurrent NTOS following transaxillary first rib resection, it is important to resect a portion of the scalene muscle rather than simply detach it from the bone.

Soft tissue attachments to the inferior and medial borders of the first rib are progressively divided by scissor dissection, starting medial to the subclavian vein (the subclavius muscle tendon and costoclavicular ligament). A periosteal elevator is then used along the inferior border of the rib to divide the intercostal muscle, and the parietal pleura is pushed away from the deep aspect of the rib by blunt dissection. At a level posterior to the brachial plexus nerve roots, the middle scalene muscle is similarly detached from the superior surface of the rib. Injury to the long thoracic nerve is avoided by keeping the periosteal elevator directly upon the rib during scalene muscle detachment. Once the posterior aspect of the first rib is exposed and the T1 nerve root is in full view and protected from injury, a C-shaped rib shears is carefully placed upon the neck of the rib and applied. The lateral portion of the divided rib is pulled downward and its anterior aspect is divided in a similar manner at the costochondral junction, immediately medial to the subclavian vein, and the rib is fully detached and removed. A Kerrison bone rongeur is then used to smooth the remaining ends of the bone beyond the level of the neurovascular structures. Once the first rib has been removed, the surgeon should identify any additional soft tissue bands that might cross the brachial plexus nerve roots and carefully divide these structures. After hemostasis is achieved, the wound is irrigated and a Valsalva maneuver is initiated to detect any rents in the pleura. A small closed-suction drain is placed through a separate wound (to be maintained on suction for 24 to 48 hours) and the incision is closed in two layers.

Supraclavicular Decompression

The patient is positioned supine under general anesthesia with the head of the bed elevated 30 degrees. The neck is turned to the opposite side and a small inflatable pillow is placed between the shoulders. The neck, upper chest, and affected upper extremity are prepped into the field, with the arm wrapped in stockinette and held comfortably across the abdomen (Fig. 24.2). A transverse incision is made parallel to and just above the clavicle, beginning at the lateral border of the sternocleidomastoid muscle and extending to the anterior border of the trapezius. The platysma layer is divided and the sternocleidomastoid muscle is retracted medially. Beginning at the lateral edge of the internal jugular vein, the scalene fat pad is detached and mobilized with ligation of small blood vessels and lymphatic tissues. A short segment of the omohyoid muscle is resected and the scalene fat pad is further mobilized from its inferior and superior attachments.

Following lateral mobilization of the scalene fat pad by gentle blunt dissection, the underlying anterior scalene muscle and phrenic nerve are exposed, along with the brachial plexus. The middle scalene muscle is exposed where it attaches to the upper surface of the first rib behind the brachial plexus, and the long thoracic nerve is identified emerging from the body of the middle scalene muscle to pass across the lateral first rib. The scalene fat pad is then held in position with several retraction sutures and the exposure is maintained with Henley self-retaining retractor (using the third arm to hold the edge of the sternocleidomastoid muscle). The resulting exposure represents the first critical view to be obtained during supraclavicular decompression (Fig. 24.3).



Fig. 24.2. Positioning and incision for supraclavicular thoracic outlet decompression. Patient position and planned incision for supraclavicular thoracic outlet decompression. The skin incision is made just above and parallel to the clavicle, extending from the lateral border of the sternocleidomastoid muscle to the anterior border of the trapezius muscle. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

To resect the anterior scalene muscle, the brachial plexus and subclavian artery are separated from the lower lateral edge of the muscle until a fingertip can be easily passed behind the muscle just above the first rib. The neurovascular structures are thereby displaced posteriorly, and blunt finger dissection is continued behind the anterior scalene to its medial border. Once the insertion of the anterior scalene muscle onto the first rib has been isolated under direct vision, it is sharply divided with scissors (Fig. 24.4).

The lower end of the divided anterior scalene muscle is elevated and its attachments to the subclavian artery and the thickened extrapleural fascia are sharply divided. The muscle is lifted further after passing it underneath to the medial side of the phrenic nerve, and its posterior attachments are divided with direct visualization and protection of the upper brachial plexus nerve roots. The anterior scalene muscle is detached from its origin on the transverse process of the cervical spine and the entire muscle is removed (Fig. 24.4). Any scalene minimus muscle fibers identified during resection of the anterior scalene muscle are resected, as are any anomalous fibrofascial bands, to ensure thorough decompression and full mobility of the brachial plexus nerve roots.

The brachial plexus nerve roots are next separated from the front edge of the middle scalene muscle to the level of the first rib and a small malleable retractor is placed between the muscle and the nerves. With medial retraction of the brachial plexus, each nerve root from C5 to T1 is sequentially identified. A second malleable retractor is placed lateral to the middle scalene muscle and first rib to permit the long thoracic nerve to be displaced posteriorly, and the attachment of the middle scalene muscle along the top of the lateral first rib is exposed and divided from the rib using the electrocautery (Fig. 24.5). The middle scalene muscle is further detached along the top of the posterior first rib using

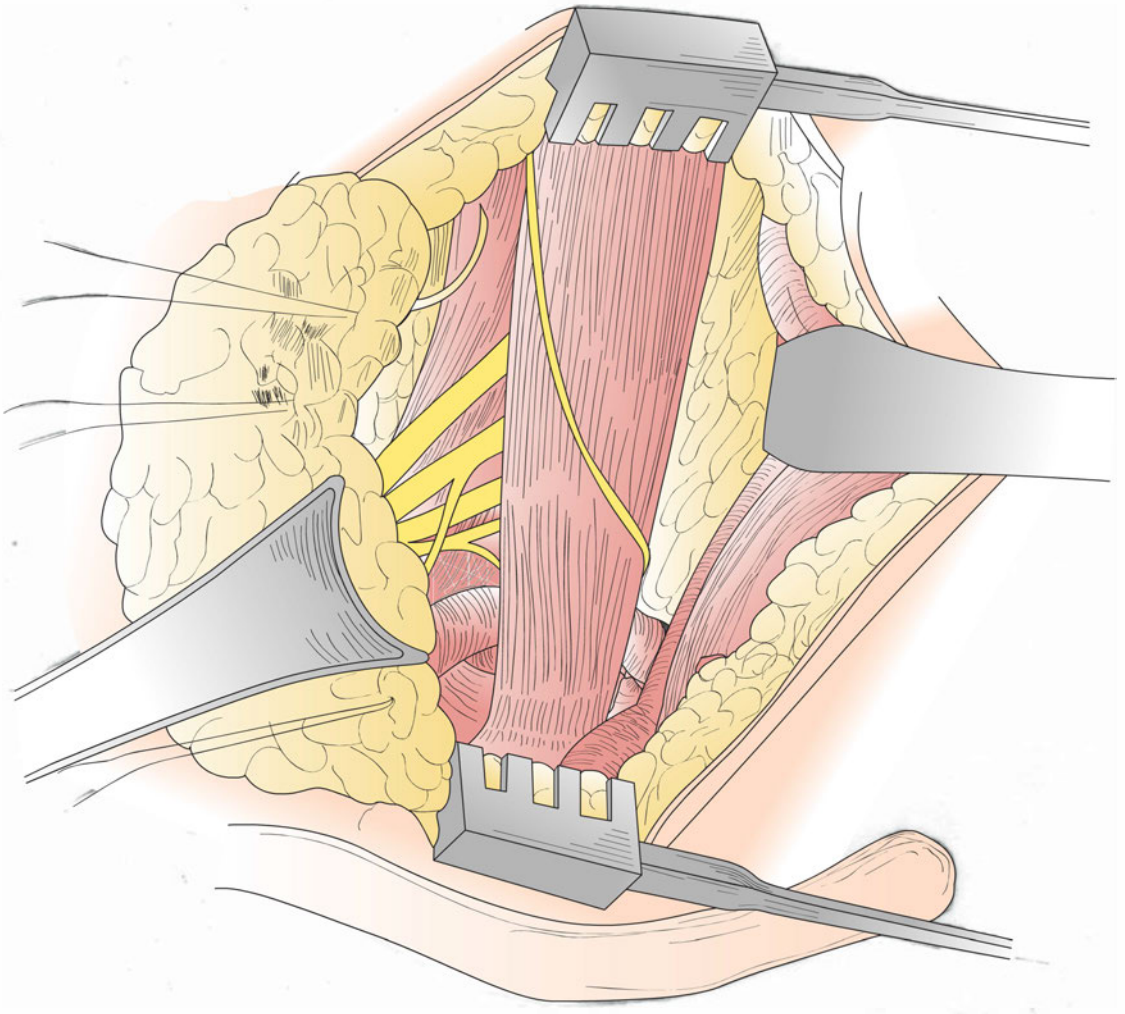


Fig. 24.3. Supraclavicular exposure. Following mobilization and lateral reflection of the scalene fat pad, direct visualization is obtained of the anterior scalene muscle, phrenic nerve, brachial plexus, middle scalene muscle, and long thoracic nerve. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

a periosteal elevator, extending to a point parallel with the T1 nerve root, and the muscle tissue anterior to the long thoracic nerve is divided to complete the resection of the middle scalene muscle.

Once the scalenectomy has been completed, the intercostal muscle attachments to the lateral first rib are divided with the cautery and blunt finger dissection is used to separate the extrapleural tissues from the undersurface of the first rib. The first rib is exposed posteriorly, where the T1 nerve root emerges from underneath the bone to join the C8 nerve root in forming the lower trunk of the brachial plexus, and a C-shaped first rib shears is inserted around the neck of the first rib. After verifying mobility of the brachial plexus and protection of the C8 and T1 nerve roots, the bone is sharply divided, the end is made smooth with a Kerrison bone rongeur, and the end of the bone is sealed with bone wax (Fig. 24.6).

The remaining intercostal muscle and fascial attachments to the first rib are divided anteriorly to the level of the scalene tubercle. While elevating the clavicle with a small Richardson retractor, the rib is displaced inferiorly to open the anterior costoclavicular space and the rib shears are placed around the anterior first rib immediately medial to the scalene tubercle. The proximal first rib is then divided and the specimen removed from the operative field (Fig. 24.7). The remaining anterior end of the first rib is remodeled to a smooth surface with a Kerrison rongeur and sealed with bone wax.

When there is a cervical rib present, these anomalous structures are found to arise within the tissue plane of the middle scalene muscle, posterior to the brachial plexus and subclavian artery. Incomplete cervical ribs typically have a ligamentous extension to the first rib, whereas complete cervical ribs attach to the lateral first rib in the form of a true joint. The posterior portion of a cervical rib is thereby readily encountered during dissection of the middle scalene muscle and divided in a manner similar that of the posterior first rib. The anterior attachment of the cervical rib is then divided and the bone is removed. When there is a true joint formed between a complete cervical rib and the first rib, the anterior portion of the cervical rib is left attached while the first rib resection is completed, and the two are removed together as a single specimen.

The last step of supraclavicular decompression is to fully mobilize each of the nerve roots of the brachial plexus and to remove any adherent fibrous scar tissue that might impair mobility (external neurolysis). Each of the contributing nerve roots (C5-T1) is meticulously dissected free

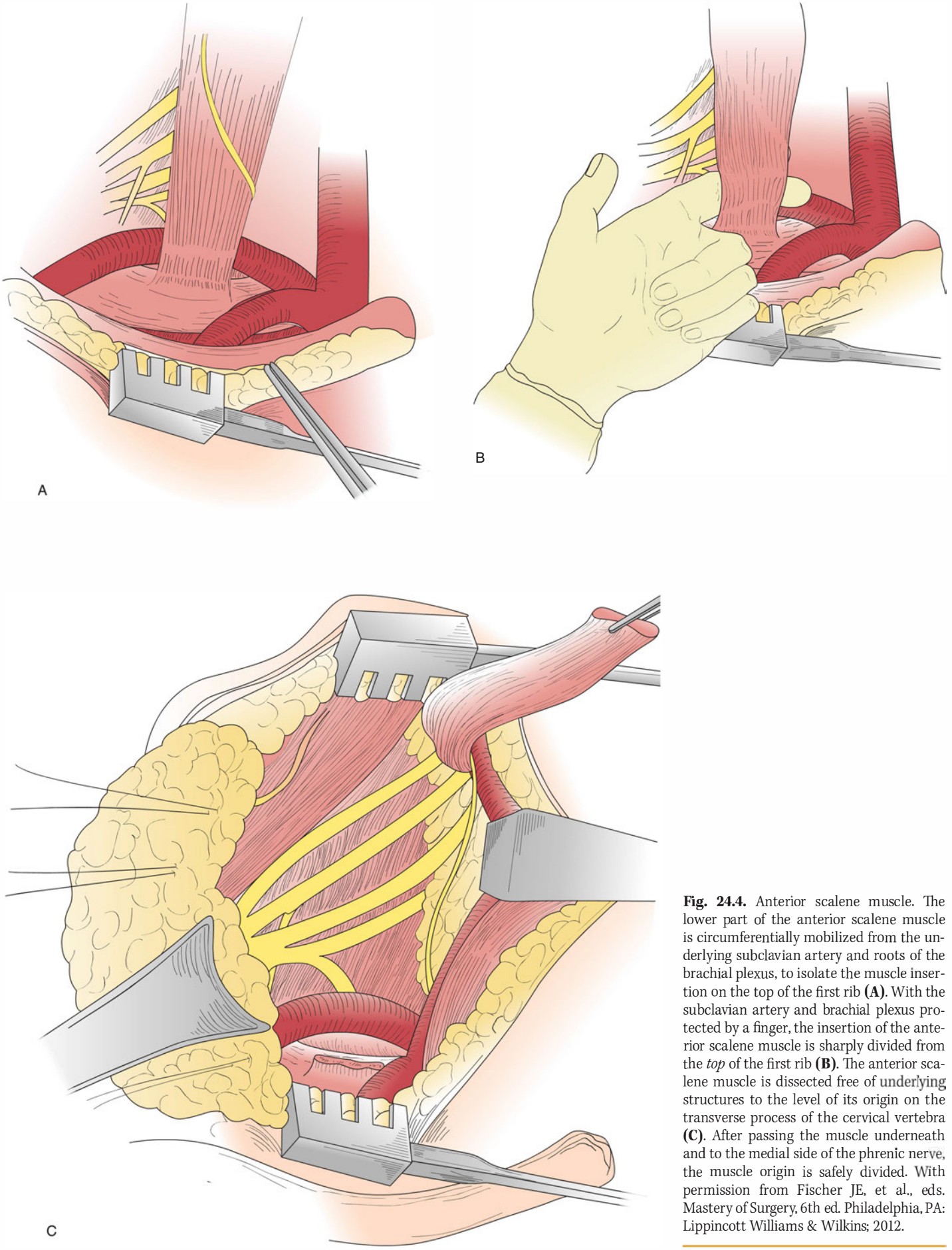


Fig. 24.4. Anterior scalene muscle. The lower part of the anterior scalene muscle is circumferentially mobilized from the underlying subclavian artery and roots of the brachial plexus, to isolate the muscle insertion on the top of the first rib **(A)**. With the subclavian artery and brachial plexus protected by a finger, the insertion of the anterior scalene muscle is sharply divided from the *top* of the first rib **(B)**. The anterior scalene muscle is dissected free of underlying structures to the level of its origin on the transverse process of the cervical vertebra **(C)**. After passing the muscle underneath and to the medial side of the phrenic nerve, the muscle origin is safely divided. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

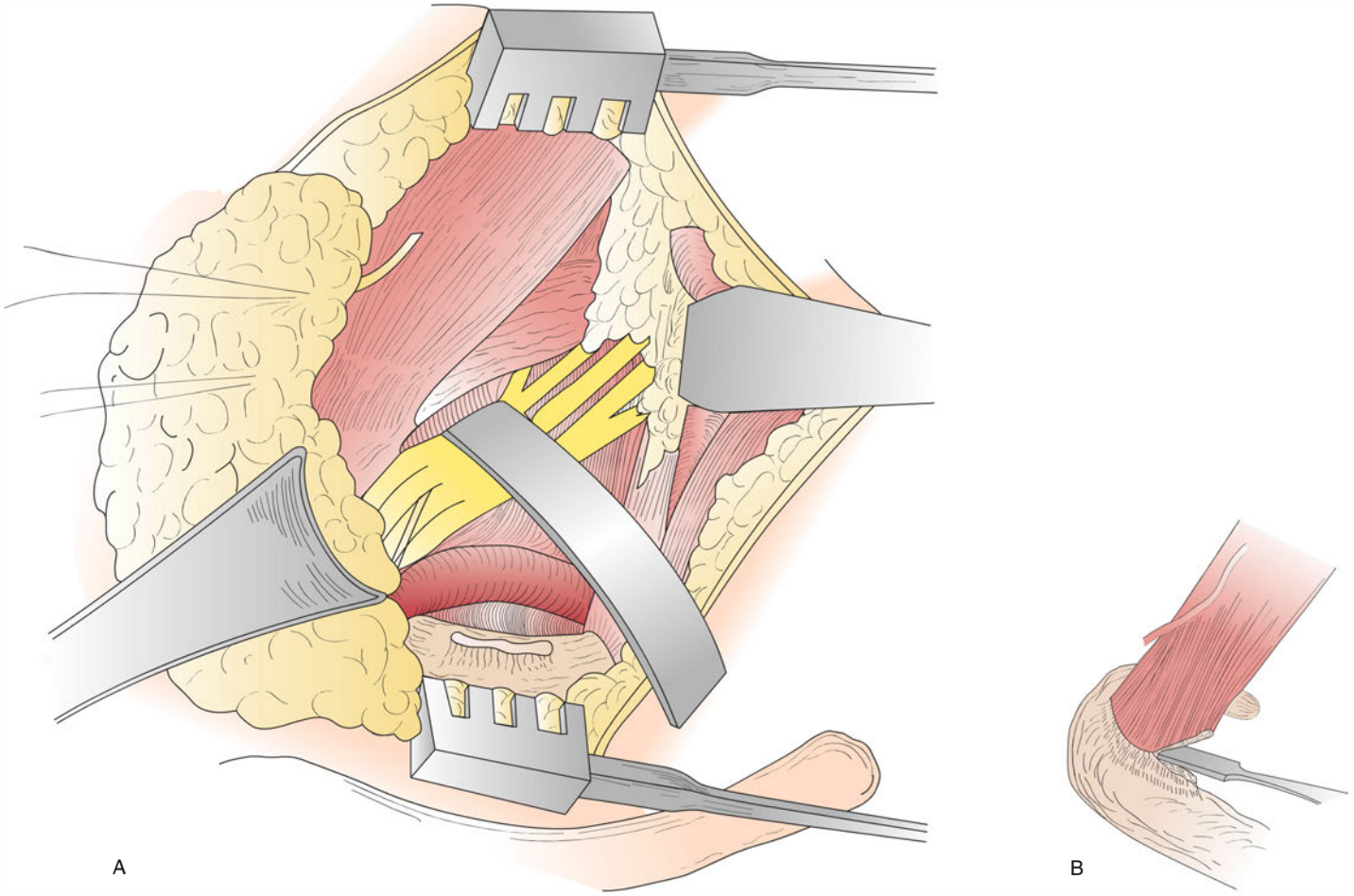


Fig. 24.5. Middle scalene muscle insertion. The insertion of the middle scalene muscle on the first rib is exposed by gentle medial retraction of the brachial plexus and posterolateral retraction of the long thoracic nerve (which is found emerging from the body of the middle scalene muscle) **(A)**. With this exposure the insertion of the middle scalene muscle can be safely divided from the *top* of the first rib with a periosteal elevator or cautery, and resected along the bone to reach the posterior portion of the first rib **(B)**. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

of any perineural fibrous tissue. This aspect of the operation is not considered to be complete until each nerve root has been completely cleared throughout its course within the operative field. Upon completion of supraclavicular decompression, the apex of the pleural membrane is opened and a closed-suction drain is placed into the supraclavicular field, where it is positioned behind the brachial plexus with its tip lying in the upper posterior pleural space. Two multihole catheters for postoperative infusion of local anesthetic are placed within the wound, adjacent to the brachial plexus and within the bed of the resected first rib. A bioresorbable (polylactide) film is wrapped around the brachial plexus to suppress development of postoperative perineural fibrosis, and the scalene fat pad is reattached to lie over the brachial plexus. The edges of the platysma muscle are reapproximated and the skin is closed.

Pectoralis Minor Tenotomy

With the patient positioned supine under general anesthesia, a short vertical infraclavicular incision is made just below the coracoid process, extending to the deltopectoral groove. The plane of dissection is kept medial to the cephalic vein as the deltoid and pectoralis major muscles are separated. The lateral edge of the pectoralis major muscle is lifted, the underlying fascia is exposed, and the space between the pectoralis major and minor muscles is developed by blunt dissection. The pectoralis minor muscle tendon is exposed and encircled near its insertion on the coracoid process, taking care to protect the underlying neurovascular bundle. The tendon is then divided with electrocautery under direct vision, within 1 to 2 centimeters of the coracoid process, allowing the divided muscle to retract inferiorly, and the edge of the divided muscle is oversewn to ensure hemostasis. The remaining clavipectoral fascia is opened to the level of the clavicle, but no further dissection of the brachial plexus or axillary vessels is performed.

Management of Venous Thoracic Outlet Syndrome (Paraclavicular Decompression)

In the surgical treatment of VTOS, we prefer to utilize an anterior approach that permits complete resection of the first rib along with direct axillary-subclavian vein reconstruction in the same setting. This begins with supraclavicular decompression as described above for NTOS, with the exception that the anterior portion of the first rib is not yet divided. Following the supraclavicular portion of the procedure, a second transverse skin incision is made several centimeters below the medial clavicle. By dissecting between the

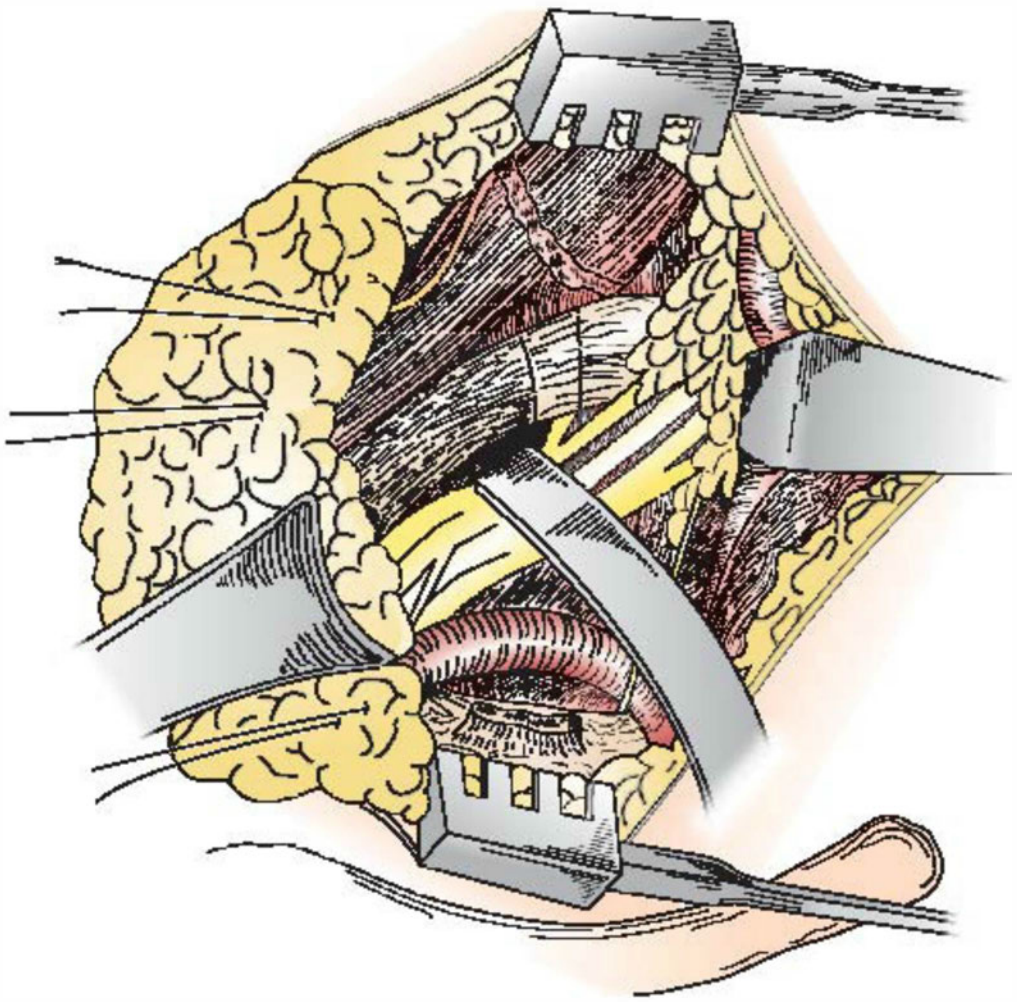


Fig. 24.6. First rib resection, posterior. The posterior portion of the first rib is exposed following removal of the middle scalene muscle, with full visualization of the brachial plexus. The C8 and T1 nerve roots are visualized immediately above and below the posterior first rib, respectively. A modified Giertz-Stille rib shears is placed around the posterior portion of the first rib in preparation for division of the bone (*dashed double line*). Following application of the rib shears, the remaining posterior first rib will be further remodeled with a Kerrison rongeur to obtain a smooth edge immediately medial to the T1 nerve root. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

upper and middle portions of the pectoralis major muscle, the cartilaginous portion of the anterior first rib is identified. This exposure is facilitated by applying downward fingertip pressure on the divided posterior segment of the first rib through the supraclavicular incision, which places the attachments between the medial first rib and clavicle under tension. Soft tissue attachments to the superior edge of the first rib are then dissected from exposure through the infraclavicular incision, and the subclavius muscle tendon, the costoclavicular ligament, and the muscles of the first intercostal space are all divided under direct vision. The anterior portion of the first rib is divided adjacent to the sternum and the entire first rib is then removed as a single specimen.

Through the lateral portion of the infraclavicular incision, the axillary vein is identified underneath the clavicle. The vein is traced and carefully separated from the subclavius muscle, and the subclavius muscle and its tendon are completely resected. Through the supraclavicular incision, the subclavian vein is exposed medially toward its junction with the internal jugular and innominate veins, and cleared of any external fibrous tissue (circumferential external venolysis). The internal jugular vein is circumferentially exposed for several centimeters above its junction with the subclavian vein, and the innominate vein is exposed for several centimeters into the upper mediastinum.

In many patients with VTOS, external venolysis is sufficient to allow reexpansion of the subclavian vein to a normal caliber. If the underlying vein is also soft to palpation and easily compressible, it is likely that no further venous reconstruction will be necessary. Additional subclavian vein reconstruction is performed when there is residual vein wall thickening despite external venolysis or residual subclavian vein stenosis by intraoperative venography. Following infusion of low molecular weight Dextran and administration of intravenous heparin, the axillary-subclavian vein, internal jugular vein, and the upper portion of the innominate vein are all controlled with clamps. The subclavian vein is opened longitudinally and the lumen is thoroughly inspected. If the luminal surface is smooth and thrombus-free, subclavian vein reconstruction is performed with a patch angioplasty using a segment of greater saphenous vein or a cryopreserved femoral vein allograft. During this type of reconstruction it is important to extend the patch angioplasty along the entire length of the affected vessel, both proximal and

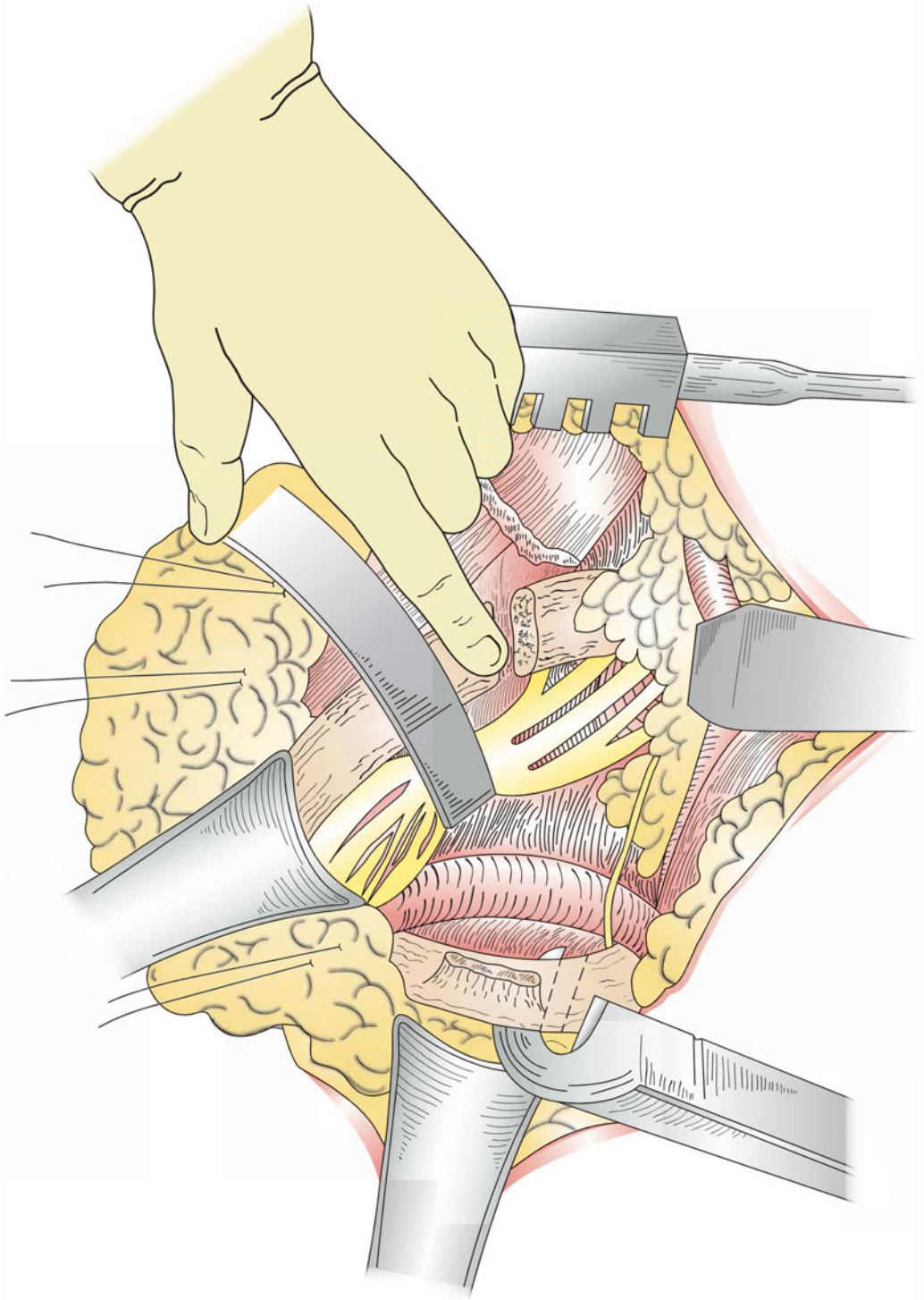


Fig. 24.7. First rib resection, anterior. Following division of the posterior first rib, the anterior portion of the rib is exposed underneath the clavicle and subclavian vein, where it will be divided immediately medial to the scalene tubercle (*dashed double line*). Fingertip pressure on the posterior stump of the rib facilitates downward displacement of the bone anteriorly, to open the costoclavicular space. A Giertz-Stille rib shears is placed around the anterior portion of the first rib in preparation for division of the bone. Following application of the rib shears, the first rib specimen is removed and the remaining anterior first rib is remodeled with a rongeur to obtain a smooth edge. With permission from Fischer JE, et al., eds. Mastery of Surgery, 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2012.

distal to the stenotic segment, including the anterior aspect of the innominate vein.

When the subclavian vein is unsuitable for patch angioplasty, due to dense fibrosis within the vein wall, ulceration of the luminal surface, or mural thrombus, the affected segment of the subclavian vein is excised and replaced. In this setting an interposition bypass graft is constructed using a widely beveled end-to-end distal anastomosis to the unaffected axillary-subclavian vein, and an end-to-side

proximal anastomosis extending into the anteromedial innominate vein. Use of a cryopreserved femoral vein allograft is usually preferred for subclavian vein bypass graft reconstruction, since the relatively small size of the saphenous vein requires creation of a panel graft to match the caliber of the subclavian vein, whereas cryopreserved femoral vein allografts of more appropriate diameter are readily available.

An intraoperative upper extremity venogram is always performed at the completion of the procedure, typically performed through the cephalic vein in the distal forearm, to confirm satisfactory reconstruction of the subclavian vein. In many patients a temporary radiocephalic arteriovenous (AV) fistula is constructed at the wrist, as a temporary (12 weeks) adjunct to increase upper extremity venous blood flow after operation. In this event the AV fistula is ligated under local anesthesia and a final follow-up venogram is performed during the same procedure.

Management of Arterial Thoracic Outlet Syndrome

Optimal surgical treatment of ATOS is accomplished through supraclavicular decompression, as described above for NTOS, including removal of the cervical and first ribs. Following decompression, attention is directed toward assessment of the pathologic changes in the subclavian artery in preparation for interposition graft repair. For subclavian artery aneurysms, depending on the length of the vessel exhibiting visible dilatation, satisfactory distal control of the subclavian artery can usually be achieved through the supraclavicular incision alone. When the lesion is more extensive, extending underneath the clavicle, exposure of the axillary artery is accomplished by adding a transverse infraclavicular incision with division of the pectoralis minor muscle tendon. After anticoagulation, clamps are placed on the proximal subclavian artery immediately distal to the vertebral artery and on the distal subclavian artery immediately beyond the aneurysmal segment. The affected segment of subclavian artery is excised and replaced with a direct end-to-end interposition bypass graft. Although reversed saphenous vein grafts may be suitable for subclavian artery replacement they are often too small in caliber, and reconstruction can be readily performed with Dacron or PTFE prosthetic grafts, cryopreserved femoral artery allografts, or other autologous (deep femoral vein or iliac artery) grafts. Following subclavian artery reconstruction a completion arteriogram is performed to evaluate the bypass graft in different positions of the arm, as well as to reassess the distal circulation.

CONCLUSION

There are few conditions in medicine and surgery that elicit more controversy than the management of TOS. In this chapter we preset a brief overview of the characteristics of each of the three types of TOS based on the principal structures affected and their varied clinical presentations, the general approaches used in diagnosis and management, and an outline of the surgical procedures most commonly used in operative treatment. While physicians specializing in the management of TOS vary in their individual approaches to diagnosis and treatment, there remains a significant proportion of patients for whom surgical treated is recommended. Similarly, different surgeons will often prefer one surgical approach over another when considering operative treatment for each form of TOS, and excellent results and outcomes can be obtained by those with abundant experience in this highly specialized area. While it appears likely that the various controversies surrounding optimal treatment for TOS will persist into the near future, there is hope that by accumulating more rigorous clinical evidence we will gradually emerge with a greater consensus upon which to base clinical decisions in the future.

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EDITOR'S COMMENTS

The majority of patients with presumed thoracic outlet syndrome when seen by either the thoracic surgeon or the vascular surgeon present with the neurogenic type for which specific diagnostic studies do not exist. As Dr. Thompson points out the diagnosis of neurogenic thoracic outlet syndrome relies almost exclusively on the clinical presentation. A chest radiograph to rule out the presence of a cervical rib probably represents the only imaging study of significant value. Most patients with this upper extremity pain syndrome can derive significant benefit from a specific program of physical therapy that must be continued at least periodically on a chronic basis. For those patients whose symptoms fail to improve with a regimen of prescribed exercises a surgical approach should be considered but patient selection likely represents the most significant predictor of a successful outcome, specifically relief of symptoms.

Most thoracic surgeons prefer the transaxillary as opposed to the supraclavicular approach for resection of the first rib as it may be accomplished without significant manipulation of the neurovascular structures. The anterior and middle scalene muscles are detached from the first rib as the rib is resected in a subperiosteal fashion. Using dedicated first rib instruments, specifically periosteal elevators both grooved and nongrooved, the rib is able to be completely mobilized prior to being cut with first rib cutters. Occasionally resection of the rib is facilitated by dividing the rib at its midpoint and then using downward traction on each segment to more easily expose the anterior and posterior aspects for division.

The supraclavicular approach probably should only be done by those with significant experience in dealing with the thoracic outlet since the potential for injury to the brachial plexus is significant because of the manipulation and specifically the retraction required. Whether or not this approach, with its greater potential for morbidity, results in a higher likelihood of success remains to be determined. Specifically whether the neurolysis advocated by Dr. Thompson adds to the probability of a successful outcome remains unknown but it certainly increases the chance for postoperative neurologic sequelae.

The vascular thoracic outlet syndromes are more straightforward specifically because of the presence of anatomic correlates that confirm the diagnosis. Imaging studies take on an important role in managing these variants of the syndrome. In our experience most of the cases of effort thrombosis of the subclavian vein may be managed by thrombolysis followed closely by transaxillary first rib resection and without any venous reconstruction. Patients are maintained on anticoagulation only for a limited duration following thoracic outlet decompression. The use of venous stents, as Dr. Thompson points out, should be condemned. Management of the arterial variant is guided specifically by the arterial abnormality with decompression of the outlet essentially an adjunct to the arterial repair, albeit an important one.

LRK

25 Chest Wall Resections

John R. Frederick and John C. Kucharczuk

INTRODUCTION

Indications for chest wall resection are broad and varied. They include resection of both primary and secondary chest wall lesions. Primary chest wall lesions are those that arise within the normal constituents of the chest wall, including skin, connective tissue, muscle, bone, and cartilage. Whether benign or malignant, primary chest wall lesions are relatively uncommon, and resection is often warranted for diagnosis and treatment. Secondary chest wall masses are caused by invasion from a process originating in contiguous organs such as the lung or breast. Lung cancer with chest wall invasion is the most common indication for chest wall resection. In these cases, chest wall resection is performed in stage-appropriate candidates as part of an en bloc resection for attempt at cure.

There are three basic tenets of chest wall resection: (1) resection of all diseases with wide margins, (2) provision of healthy soft tissue coverage, and (3) preservation of respiratory mechanics. The intent of this chapter is to review the indication and techniques of chest wall resection with reconstruction from the simple to the complex. The discussion focuses on the proper selection of reconstructive techniques from "off the shelf" synthetic material to complex soft tissue transfers.

PREOPERATIVE PLANNING

Patients being considered for chest wall resection undergo a complete medical evaluation. Special attention is focused on any past medical or surgical history that will affect the approach for resection and the choices for reconstruction. These factors include previous chest procedures, a history of radiation, evidence of active infection, and immunosuppression. All patients have radiographic imaging, including a chest radiograph and computed tomography (CT) scan of the chest. For patients with a primary chest wall lesion, magnetic resonance imaging may be helpful in delineating the local extent of disease, but as a general rule it cannot distinguish between benign and malignant masses. Patients with underlying lung cancer and contiguous chest wall invasion undergo a complete extent of disease workup to rule out metastatic disease, and if negative, they are considered for resection. If partial vertebral body resection is entertained or there is concern regarding involvement at the level of the neural foramen, preoperative neurosurgical consultation is obtained. Similarly, the need for transfer of large volumes of soft tissue for coverage or unfamiliarity with the available techniques for complex tissue transfer should prompt consultation with an experienced reconstructive surgeon. A tissue diagnosis should be obtained in most cases to differentiate between benign and malignant tumors and to identify malignant neoplasms that may be amenable to medical therapy. This consists of fine needle aspiration, core needle biopsy, or incisional biopsy of lesions larger than 5 cm. Lesions smaller than 5 cm are usually suitable for excisional biopsy.

OPERATIVE PLANNING: OPTIMIZING THE APPROACH

In selecting the appropriate incision, it is imperative that the surgeon be thoughtful, flexible, and experienced. The optimal surgical approach allows for the assessment of the extent of disease without violation of the lesion. Lateral chest wall lesions are generally approached through posterolateral incisions. If there is a planned need for soft tissue transfer, the latissimus dorsi and serratus anterior muscles are mobilized but not divided before entering the pleural cavity. If there is no need for tissue transfer, we generally divide the latissimus dorsi muscle but spare the serratus anterior in case it should be needed in the future. The pleural cavity is entered either at an interspace below the lesion or at a site anterior to the lesion. The lesion is palpated to determine the extent of resection required. Primary chest wall lesions rarely invade the lung, and pulmonary resection is usually not required. In cases of lung cancer with contiguous chest wall involvement, the chest wall resection is performed as shown in an en bloc manner. Attempts to develop an extrapleural plane or strip the tumor off the chest wall should be avoided because this risks violation of the tumor with contamination of the pleural cavity and consequently a high likelihood of recurrence. Once the chest wall resection is complete, the chest wall bloc remains attached to the underlying pulmonary parenchyma while the pulmonary resection is performed. No attempt is made to separate the chest wall bloc from the underlying lung.

Lesions involving the apex of the chest can be approached via the traditional Shaw–Paulson technique. This approach uses a long posterolateral incision carried up to the C7 vertebral body, elevation of the scapula off the chest wall after division of the trapezius and the rhomboids, and chest wall resection from a posterior approach. Recently, it has been recognized that these apical lesions may be more effectively managed through the anterior cervicothoracic approach described by Dartevelle and subsequently modified by others. Anterior chest wall lesions are often best approached with the patient supine and an anterior incision made over the location of the lesion.

Regardless of the surgical approach chosen, the basic tenets of chest wall resection must be fulfilled. These include a wide margin of excision, en bloc anatomic resection with the attached lung parenchyma, and appropriate reconstruction of the chest wall defect when required.

TECHNIQUE OF CHEST WALL RESECTION

Chest wall involvement is often heralded by the clinical finding of pain associated with

a peripheral lesion seen abutting the chest wall on CT scan. In general, it is not possible to reliably differentiate chest wall invasion from simple abutment merely by looking at the CT scan. A posterolateral incision is made, and the latissimus dorsi muscle and serratus anterior muscles are either divided or mobilized if they are needed for pedicled muscle flaps. The chest is typically entered via the fifth intercostal space and exploration carried out. If the lesion involves the fifth rib, entry into the hemithorax should be either anterior or posterior to the presumed area of involvement. Before initiating the chest wall resection, palpation of the hilum and mediastinum should be carried out to ensure resectability. It serves little purpose to take down the chest wall block only to find locally advanced disease or diffuse pleural disease that precludes resection. The combination of mediastinal lymph node positivity and chest wall involvement portends a poor outcome. If there is any suspicion of nodal involvement, mediastinoscopy must be carried out before chest exploration. A positive mediastinum usually precludes chest wall resection outside of a protocol setting, although some surgeons proceed with resection after neoadjuvant chemotherapy. Most studies have shown essentially no 5-year survivors with chest wall involvement and positive mediastinal lymph nodes.

Once it has been determined that the tumor is resectable, the chest wall resection is begun. Depending on the location, the scapula may need to be mobilized off the chest wall, which requires division of the trapezius and rhomboid muscles. At least a portion of one rib above and one rib below the lesion should be resected. The extent of resection is determined by the intrathoracic exploration. The anterior portion of the resection is done first by determining the appropriate rib and incising the periosteum several centimeters anterior to the lesion. A wide excision should be performed to ensure negative margins. We prefer to take a 1-cm piece of rib and submit this separately at each level as the anterior resection margin (Fig. 25.1A). This move creates a small amount of space for ligation of the intercostal bundle at each level. Starting at the inferior extent of the resection and working superiorly, the anterior margin is taken at each level with the pleura excised as the resection proceeds. The intercostal muscles at the inferior and superior extent of the resection are divided along with the pleura, and the posterior portions of the

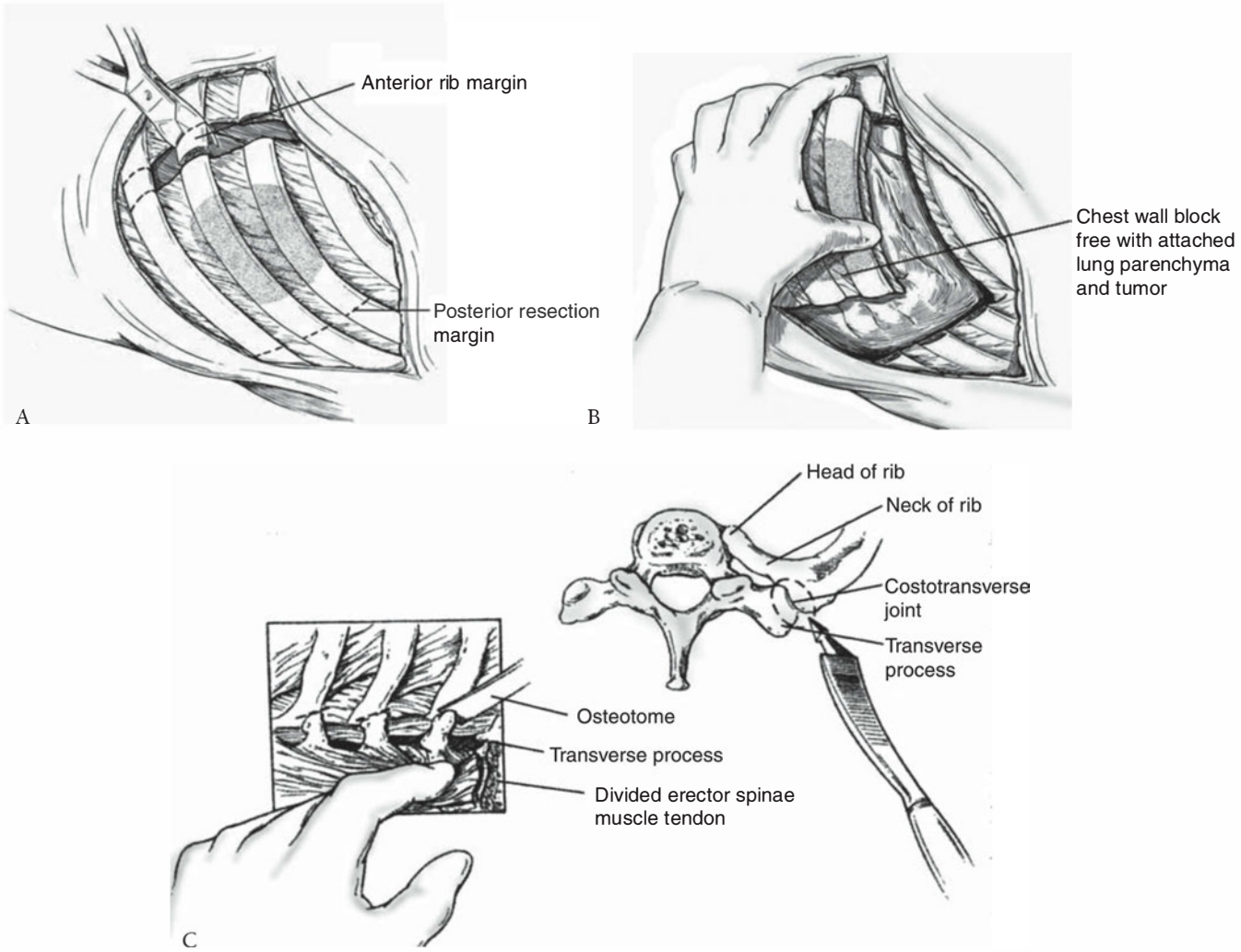


Fig. 25.1. (**A**) En bloc chest lateral chest wall resection in a patient with a primary lung cancer. The anterior line of resection is far away from the primary site of invasion, and small, 1-cm portions of the anterior ribs are removed and labeled as margins. (**B**) The completed chest wall resection; at this juncture, the entire chest wall bloc is allowed to fall into the pleural cavity and the pulmonary resection is performed through the chest wall defect. (**C**) Disarticulation of the posterior rib from the transverse process with an osteotome. (Reprinted with permission from Kaiser LR. Atlas of General Thoracic Surgery. St. Louis, MO: Mosby; 1997.)

involved ribs are divided. There is no need to take a separate posterior margin at each level as is done anteriorly; this margin will be marked on the en bloc resection specimen. Depending on the location of the lesion, the posterior rib division may be either through the rib as demonstrated in Figure 25.1B or may require disarticulation of the rib from the respective transverse process of the spine as shown in Figure 25.1C. If there is any doubt, the rib should be disarticulated and, at times, even the transverse process may need to be resected. As always the intent is to have a negative resection margin; anything less (i.e., a positive margin) is associated with poor long-term survival.

If required, division of the posterior portion of the ribs is performed in a subperiosteal manner with ligation of the intercostal bundle. The bundle at each level has already been ligated and divided anteriorly, and thus only a single ligature at each level is necessary. If disarticulation from the transverse process and vertebral body is required, the intercostal bundle must be ligated at the level of the neural foramen. To disarticulate the ribs from the transverse process, the paraspinous ligament must be reflected away from the spine down to the level of the transverse process with the use of electrocautery because there are numerous perforating vessels that must be controlled. Once down to the transverse process, the rib is retracted anteriorly as the cautery incises the cartilaginous junction between the neck of the rib and the transverse process of the spine. The correct location has been identified when the cartilage appears to "melt" with the application of the cautery and the rib neck begins to separate slightly. Once the cartilaginous symphysis is divided, a curved osteotome is inserted between the neck of the rib (Fig. 25.1C, inset) and the transverse process and, with the transverse process as a fulcrum, is rocked forward, thereby disarticulating the neck of the rib from the transverse process and the head of the rib from the vertebral body. This is facilitated by having incised the overlying pleura from within the chest. The osteotome is directed anteriorly toward the vertebral body, and thus away from the spinal canal, so there is no chance of injuring the spinal cord. The force is directed upward away from the vertebral body so as to lift the head of the rib. The rib should be gently disarticulated so as to not avulse the intercostal nerve as the dural sheath may be torn with a resultant cerebrospinal fluid leak. Once the rib has been disarticulated enough to see the intercostal nerve exiting the neural foramen, the nerve and intercostal vessels should be ligated and divided. It is not uncommon to experience some bleeding from the foramen because there are multiple small venous channels that may be opened. Only bipolar cautery should be used at the foraminal level to control any bleeding so as to prevent any thermal injury to the spinal cord. The foramen should *not* be packed with Surgicel or gel foam to control bleeding; these materials can expand within the foramen leading to spinal cord compression with catastrophic neurologic consequences.

Once the chest wall bloc has been entirely separated, it is left attached to the underlying lung parenchyma in cases of a primary lung tumor invading the chest wall. The appropriate anatomic pulmonary resection (lobectomy or, at times, segmentectomy) is then carried out. A complete mediastinal lymph node dissection should be part of the procedure even if a mediastinoscopy has been done so that the most accurate staging information is obtained. If the lesion is a primary chest wall tumor, the specimen is removed and the reconstruction is begun.

Removal of the first rib requires some special techniques and expertise. One must be intimately familiar with the relationship of the brachial plexus and subclavian vessels to the first rib to avoid injuring any of these structures. Special first-rib instruments are necessary for safe first-rib resection. These include special angled first-rib periosteal elevators either with a groove or not, rongeurs, and angled rib cutters. If first-rib resection is required, the scalene muscles that insert on the rib must be detached. The posterior scalene inserts on the second rib, so this will have already been taken. The middle scalene inserts on the first rib between the subclavian vein and artery and should be reflected with the periosteal elevator following cautery incision of the periosteum. The anterior scalene should be incised and reflected off the rib. The inferior aspect of the rib is freed with the use of a grooved elevator. A Matson elevator is placed under the rib to expose the medial aspect so as to protect the nerves and vessels. Once the medial aspect of the rib is cleared, a first-rib cutter is used to cut the anterior aspect of the rib. If the first rib needs to be disarticulated from the transverse process, care must be taken to identify the C8 and T1 nerve roots that combine to form the lower cord of the brachial plexus. These nerve roots may be identified by visualizing the head of the first rib, which is located immediately between these two structures. If the roots are not identified as they exit their respective foramina, the operator may mistakenly divide the lower cord of the brachial plexus leaving the patient with a useless ipsilateral hand. The T1 root may be taken as part of the resection if it is involved, but division of the C8 root leaves a nonfunctional hand. The degree of disability that occurs after division of the T1 nerve root usually is minimal to moderate at most.

Recent advances in chest wall resection include the use of thoracoscopy, particularly in cases where thoracoscopic lobectomy with en bloc chest wall resection is anatomically feasible. Some surgeons advocate this technique based on the theoretical benefit of alleviating rib spreading, thereby reducing postoperative pain and recovery time. More extensive studies must be performed to validate the use of this technique.

SYNTHETIC MATERIAL CHOICES

The synthetic materials available for chest wall reconstruction are polypropylene mesh and polytetrafluoroethylene patches. Both have been used in large series of patients and appear to provide equivalent outcomes. Polypropylene is significantly less expensive and can be reinforced with methylmethacrylate to create a rigid prosthesis that can be contoured to the chest wall. This may provide a better cosmetic result and a significantly better functional result especially for anterior lesions. Figure 25.2 shows the creation of a mesh/methylmethacrylate "sandwich" for chest wall reconstruction. Regardless of the material selected for reconstruction, it is inserted into the defect with interrupted, nonabsorbable suture. We use No. 1 polypropylene sutures placed through the cut edges of the lateral ribs and around the uncut superior and inferior ribs as shown in Figure 25.3. A small drill facilitates placement of suture through the cut ribs. If no superior rib remains for fixation, the prosthesis is secured on the remaining three sides with room at the apex to avoid compression of the structures of the thoracic inlet. The synthetic material is covered with healthy soft tissue; usually, this simply requires closure of the cut muscle edges over the prosthesis followed by soft tissue closure. In cases where there has been significant soft tissue or muscle resection, complex soft tissue transfer is required.

Reconstruction of the chest wall defect is not required if a posterior resection has been done and the entire defect is covered by the scapula. If the fifth rib has been taken, however, the tip of the scapula has

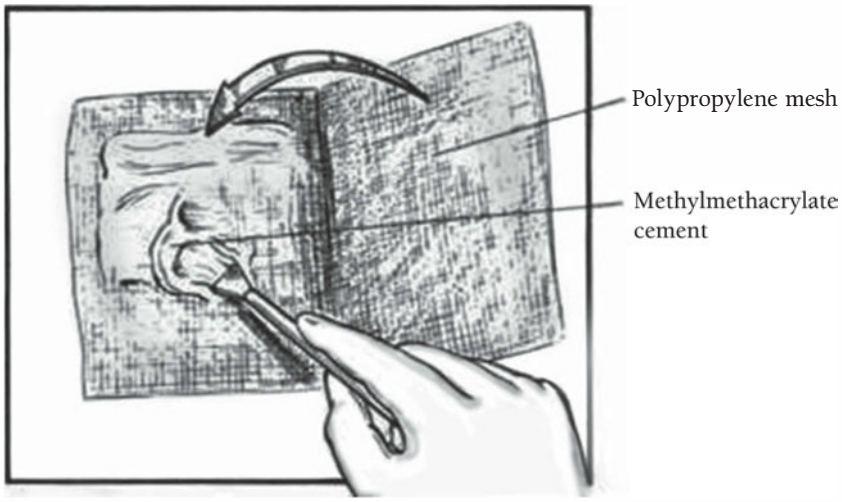


Fig. 25.2. Creation of a polypropylene mesh/methylmethacrylate "sandwich" to be used as an inset prosthesis for lateral chest wall reconstruction. (Reprinted with permission from Kaiser LR. Atlas of General Thoracic Surgery. St. Louis, MO: Mosby; 1997.)

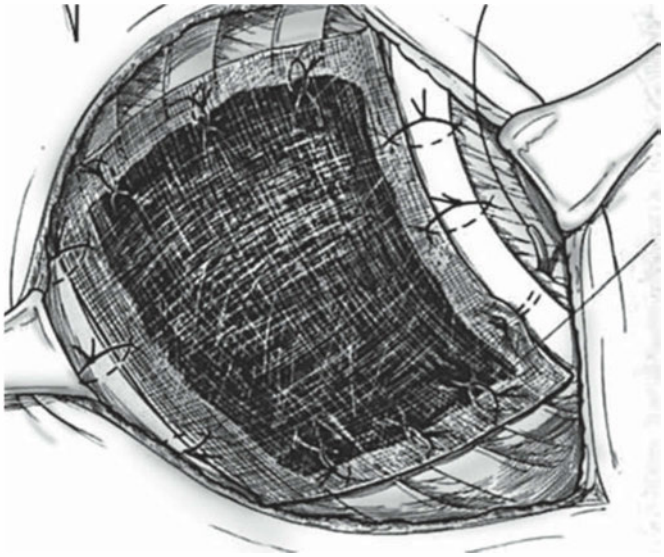


Fig. 25.3. Chest wall prosthesis inset with interrupted nonabsorbable sutures. (Reprinted with permission from Kaiser LR. Atlas of General Thoracic Surgery. St. Louis, MO: Mosby; 1997.)

Table 25.1 Pedicled Flaps Available as Either Simple Muscle or Composite Tissue Transfers for Chest Wall Reconstruction[a]

Muscle	Arterial supply	Use in chest wall reconstruction
Latissimus dorsi	Thoracodorsal	Anterior and lateral chest wall
Pectoralis major	Thoracoacromial	Anterior and midline chest wall
Serratus anterior	Lateral thoracic	Lateral chest wall
Rectus abdominis	Superior epigastric	Anterior and midline chest wall
Omentum	Gastroepiploic	Midline chest wall

[a]The neurovascular bundle that must be preserved in each flap is listed. These transfer flaps as well as a variety of others can also be used as "free flaps" with the use of microsurgical techniques to establish arterial inflow and venous drainage in complex cases.

a tendency to become trapped underneath the sixth rib, a situation that is very uncomfortable for the patient. Thus, if the fifth rib has been taken posteriorly, chest wall reconstruction with a patch of polypropylene mesh should be undertaken to prevent entrapment of the scapular tip. Rigid fixation is not required in this location. Other posterior or posterolateral chest wall defects should be reconstructed either with polypropylene mesh alone or with mesh and methylmethacrylate. Anterior chest wall defects usually require a rigid prosthesis to prevent respiratory embarrassment due to altered chest wall dynamics in the early postoperative period. This is especially important in the patient with borderline pulmonary function, where the "flail chest" physiology created by the chest wall defect is often enough to mandate mechanical ventilation without rigid fixation of the chest wall.

Several commercially available osteosynthesis systems are now being utilized for complex chest wall reconstruction. These consist of either steel or titanium rib prostheses fixed in place with modular connectors or silicone molds into which methylmethacrylate can be injected. Although more expensive, these systems may provide a structural advantage in the preservation of chest wall mechanics. It is important to note, however, that implantable materials always carry the risk of infection or displacement. Additionally, most authors still advocate coverage with mesh or tissue flaps.

TISSUE TRANSFER OPTIONS

The major muscles of the chest wall provide a good source of autologous tissue for soft tissue coverage of large chest wall defects. They may be utilized to cover prosthetic material in cases where significant soft tissue has been removed or alone without underlying prosthetic reconstruction in cases of infection. The rectus abdominis and omentum can also be transferred from the abdomen to the chest to provide coverage. Table 25.1 lists the tissue available to transfer to the chest along with the blood supply of each flap. Preservation of the neurovascular bundle of a pedicled flap is critical to its success and is an absolute requirement.

Figure 25.4 illustrates the anatomic features of each pedicled flap used in the chest. Anterior chest wall coverage is best provided by pectoralis major advancement flaps. A pedicled rectus abdominis flap or omental flap with skin graft may also be

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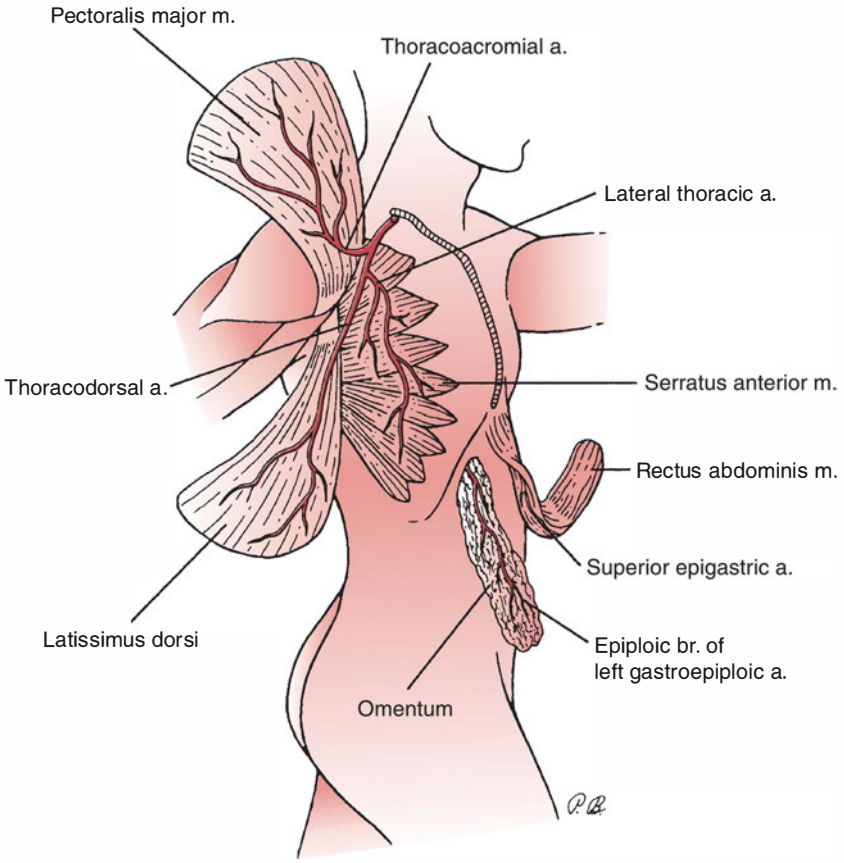


Fig. 25.4. Pedicle muscle flaps available for reconstruction of chest wall and sternal defects. a, artery; m, muscle.

utilized. Lateral defects are best addressed with pedicled serratus and/or latissimus flaps. If a composite flap (skin, soft tissue, and muscle) cannot be used, muscle alone can be transferred and a split-thickness skin graft can be applied.

STERNAL RESECTIONS

Primary sternal tumors are rare, and when they occur they almost always are chondrosarcomas arising from the body of the sternum. These tend to be low-grade lesions, and the best opportunity for cure is total sternectomy to achieve negative margins and immediate reconstruction with prosthetic material. The likelihood of cure and propensity for recurrence with sarcomas are related to the tumor's histologic grade.

Defects created by partial sternectomy may be small and reconstructed with simple pectoralis advancement flaps without the need for prosthetic material. Total sternectomy results in significant soft tissue and structural loss and usually requires complex reconstruction. This is especially true if the sternum is being resected for recurrent breast cancer after excision and radiation therapy.

TECHNIQUE FOR STERNAL RESECTION

A vertical midline incision is made over the sternum, and skin flaps may need to be raised, depending on the size of the lesion. The pectoralis muscles are reflected laterally unless muscle is involved, in which case they are left with the sternum to be resected en bloc. The sternal notch is dissected and the retrosternal plane is developed. Inferiorly, the xiphoid process is excised and the retrosternal plane entered from this aspect as well. The pleural reflections are swept laterally. The costal cartilage at each level is divided after removal of the perichondrium. The posterior perichondrium is incised to separate it from the sternum. This is facilitated by the use of a bone hook to elevate the sternum.

Often the manubrium can be preserved with total removal of the body of the sternum. If this is the case, the body of the sternum is disarticulated from the manubrium. If resection of the manubrium is required to achieve a complete resection, the clavicular heads are disarticulated and the first costal cartilage on each side incised. This allows complete removal of the sternum. Reconstruction is carried out with a polypropylene mesh/methylmethacrylate "sandwich," which is fitted into the defect and contoured appropriately. The prosthesis is inserted with monofilament nonabsorbable suture placed through the cut rib edges and clavicular heads.

Figure 25.5 illustrates an intended total sternectomy for a primary sternal lesion. The reconstruction is completed as shown in Figure 25.6A. The omentum has been transferred to cover the mediastinum; a mesh/methylmethacrylate prosthesis is covered by bilateral pectoralis major turnovers. If the skin is not removed as part of the resection, it is closed over drains to complete the reconstruction. If significant skin and soft tissue has been resected, the prosthesis can be completely covered with the addition of a rectus abdominis flap (Fig. 25.6B) and a split-thickness skin graft applied.

CONCLUSION

Primary and secondary lesions of the chest wall and sternum can be resected in appropriate patients with good clinical outcomes. Taking on these cases requires proper preoperative planning and a working knowledge of a variety of reconstructive techniques.

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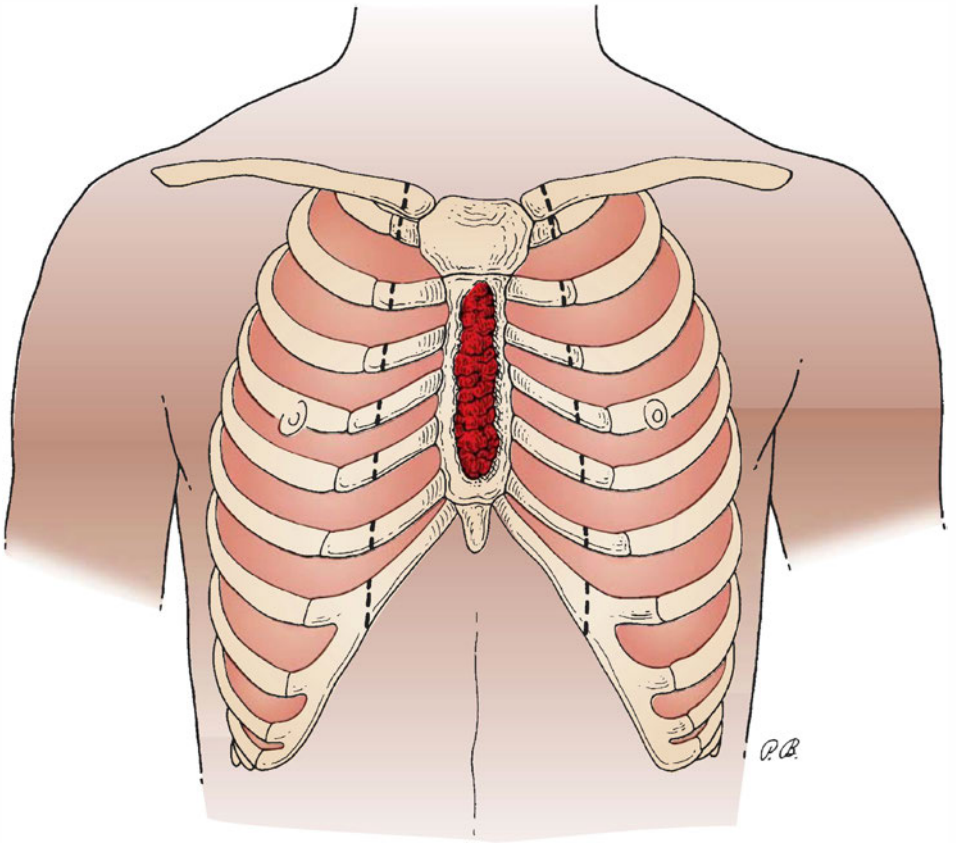


Fig. 25.5. Primary sternal tumor with intended resection lines.

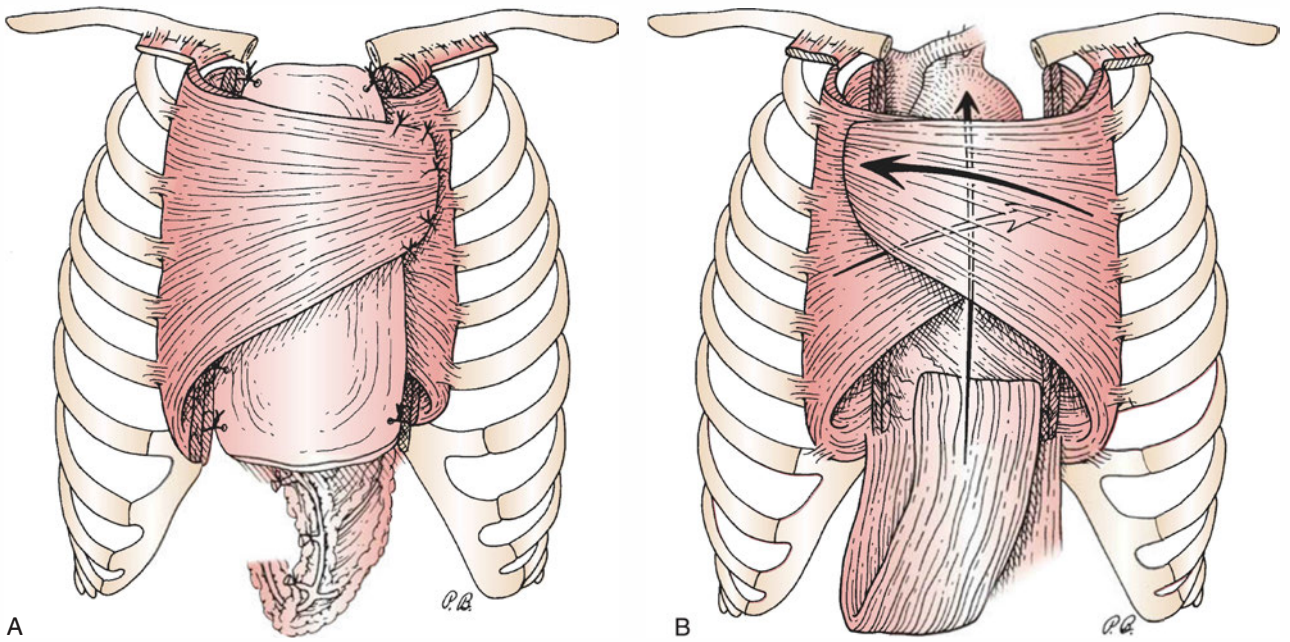


Fig. 25.6. (**A**) Reconstruction of sternal resection with omental transfer to cover the mediastinum followed by bilateral pectoralis muscle turnover flaps to cover a polypropylene mesh methylmethacrylate prosthesis. If soft tissue and skin coverage is also needed, the prosthesis can be completely covered by a rectus flap as shown (**B**) and the reconstruction completed with a skin graft.

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SUGGESTED READINGS

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EDITOR'S COMMENTS

Primary lung tumors involving the chest wall are unusual but the surgeon must be knowledgeable regarding resection techniques since chest wall involvement usually cannot be determined with certainty preoperatively. In order not to compromise the extent of the resection, the surgeon must also be intimately familiar with the techniques of chest wall reconstruction. The importance of achieving a negative resection margin must be again emphasized since this along with node negativity are the major determinants of long-term survival. There should be no hesitation about making a "big hole" if that is what is required to achieve a negative margin. In order to guide the pathologist, at least two of the margins of the resected chest wall specimen should be marked with sutures and indicated on the pathology requisition. Frozen section confirmation of bone margins is not possible so the lesion should be resected widely.

As the authors point out the combination of chest wall involvement accompanied by positive mediastinal lymph nodes portends a very poor outcome and thus a careful assessment of the mediastinum should be carried out if there is any question of node positivity. Certainly if the PET scan shows activity in the mediastinum, histologic confirmation should be obtained either with endoscopic ultrasound-guided bronchoscopic biopsy or mediastinoscopy prior to considering resection.

Apical chest wall involvement creates a unique set of issues that require special expertise as the authors point out. Because of the complex anatomy and myriad structures within the thoracic inlet, great care must be taken during resection and the surgeon must be prepared for a variety of situations. If the T1 nerve root is involved it may be taken with little if any neurologic sequelae in the majority of cases. However, care must be taken to unequivocally establish that one has identified the T1 root and not mistake it for the lower cord of the brachial plexus. This is only possible if the C8 and T1 nerve roots are identified at the foraminal level where the head of the first rib is seen between the roots and the confluence of the two roots is seen to form the lower cord. Then, and only then, should the large T1 root be divided. If the tumor extends further superior and is seen to involve the lower cord of the brachial plexus or the C8 nerve root resection should not be performed as this will leave the patient with a useless, clawed hand. Additionally, the subclavian artery may be involved by tumor and the surgeon should be prepared for this possibility that involves resection of the involved segment of artery with prosthetic graft replacement. The possibility of neurovascular involvement further underscores the importance of the surgeon being familiar with the anterior approach to apical lung tumors as opposed to the classic Shaw–Paulson posterior approach. The anterior approach is far less morbid and provides significantly better exposure to the neurovascular structures without compromising the ability to achieve an adequate resection of chest wall especially the first rib. One would think that an anterior approach for a posterior lesion is counterintuitive but one must keep in mind the very short anterior–posterior length at the chest apex. In addition, the upper lobectomy is easily accomplished through the anterior cervicothoracic incision with no need for any additional incision.

Primary chest wall tumors are exceedingly rare, as the authors correctly point out and definitive histology needs to be established prior to proceeding with resection because many of these turn out to be "medical tumors" best treated with chemotherapy and potentially radiation therapy. I am specifically referring to Ewing's tumors and chest wall lymphomas, among others. Small lesions may be completely excised for "biopsy," while larger lesions should have an incisional biopsy for the establishment of definitive histology on permanent section. Frozen section diagnosis should not be relied upon when dealing with these lesions. The biopsy procedure should be a separate procedure followed by the definitive procedure at a second session once the histology has been established and resection is indicated.

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26 The Diaphragm

Mara B. Antonoff and Jonathan D'Cunha

INTRODUCTION

Separating the thoracic and abdominal cavities, the diaphragm is a dome-shaped muscular structure that serves its key function as the principal muscle of respiration. Through contraction and relaxation, the diaphragm cyclically increases and decreases the volume of the thorax, altering intrathoracic pressure and, ultimately, permitting air entry into the lungs via the establishment of negative pressure. Sustained contraction of the diaphragm serves to increase intraabdominal pressure, useful in generating a Valsalva effect. When functioning well, the diaphragm rarely is the subject of significant clinical attention; however, dysfunction of the diaphragm poses significant and challenging dilemmas to the patient and the surgeon, emphasizing the substantial importance of this musculoaponeurotic structure.

EMBRYOLOGY AND ANATOMY

The embryologic development of the diaphragm occurs between the 7th and 10th weeks of gestation, with the definitive musculotendinous diaphragm incorporating elements of four embryonic precursors: (1) the septum transversum, (2) the pleuroperitoneal membranes, (3) the paraxial mesoderm of the body wall, and (4) the esophageal mesenchyme. From the septum transversum, myoblasts migrate into the right and left pleuroperitoneal membranes, bringing phrenic nerve branches along the way. The majority of the septum transversum gives rise to the nonmuscular diaphragmatic central tendon. The posterolateral portion of the diaphragm is formed by fusion of the dorsal mesentery with the pleuroperitoneal membranes. As these membranous structures are encountered by muscle fibers migrating caudally from the cervical myotomes, the muscular bulk of the diaphragm is formed. Migration of myoblasts into the dorsal mesentery results in formation of the bilateral crura, which originate on the vertebral column and insert into the dorsomedial diaphragm.

While the diaphragm is a fairly consistent organ, with no normal variations in anatomic structure, there are several common abnormalities that result from failed intrauterine development. As the diaphragm originates from several embryonic structures, and normal development requires complex and coordinated execution of a number of vital steps, congenital abnormalities of the diaphragm are well described. Such errors in embryologic development can result in future surgical problems, including congenital diaphragmatic hernias and eventration. Presence or absence of a sac depends on the time point at which the defect occurs, either before or after membranous fusion.

To safely operate on or near the diaphragm, one must have a clear understanding of the anatomic features of the muscular organ, as well as appreciation for the vital structures in close proximity. With its dominantly convex superior surface facing the thoracic cavity and the concave inferior surface facing the abdomen, the diaphragm consists of a mobile component centrally and a fixed component peripherally, which attaches to the inferior margin of the thoracic cage and the superior lumbar vertebrae. The peripheral muscular fibers converge radially at the central tendon. This fibrous, centrally located aponeurosis is composed of the right, left, and middle leaflets, resembling somewhat of a cloverleaf with incomplete divisions. The major muscular elements of the peripheral diaphragm include the sternal, costal, and lumbar groups (Fig. 26.1). The sternal portion is comprised of two muscular attachments to the posterior xiphoid. The costal portion comprises the right and left domes of the diaphragm, attaching to the inferior six ribs and adjacent costal cartilages. The lumbar portion arises from the medial and lateral arcuate ligaments (thickened extensions of the psoas and quadratus laborum fascias, respectively) and attaches to the three most superior lumbar vertebrae.

The left and right crura of the diaphragm arise from the anterior aspects of the lumbar vertebrae. The right crus is longer than the left, and may arise from the first three to four lumbar vertebrae, while the left crus may arise from just the first two to three vertebrae. As the muscle fibers ascend, the medial fibers of both crura decussate just anterior to the aorta. After encircling the esophagus, the muscle fibers insert superiorly into the central tendon of the diaphragm. The central tendon is fused to the inferior aspect of the pericardium via the phrenicopericardial ligaments.

Significant structures pass through three openings: the caval foramen, at T8; the esophageal hiatus, at T10; and the aortic hiatus, at T12 (Fig. 26.2). The caval opening, lying within the central tendon, allows the passage of the inferior vena cava (IVC) as well as terminal branches of the right phrenic nerve and some lymphatic channels. The esophageal hiatus is to the left of midline but is dominantly a formation of the right crus. In addition to the esophagus, this aperture also provides a path for the anterior and posterior vagus nerves, esophageal branches of the left gastric vein and artery, and lymphatics. The aortic hiatus is formed by the crura and the median arcuate ligament, and allows passage of the aorta as well as the thoracic duct.

The major blood supply to the inferior aspect of the diaphragm comes from the left and right phrenic arteries, which come directly from the abdominal aorta in the vicinity of the aortic hiatus (Fig. 26.3). These paired arteries each bifurcate, giving off a large anterior branch that courses along the anterior and superior aspects of the muscle, and merging with the pericardiophrenic artery. The posterior branches of the phrenic artery course laterally and posteriorly and ultimately merge with the intercostal vessels. The pericardiophrenic and intercostal vessels originate from the internal mammary artery. Dominant venous drainage occurs via the

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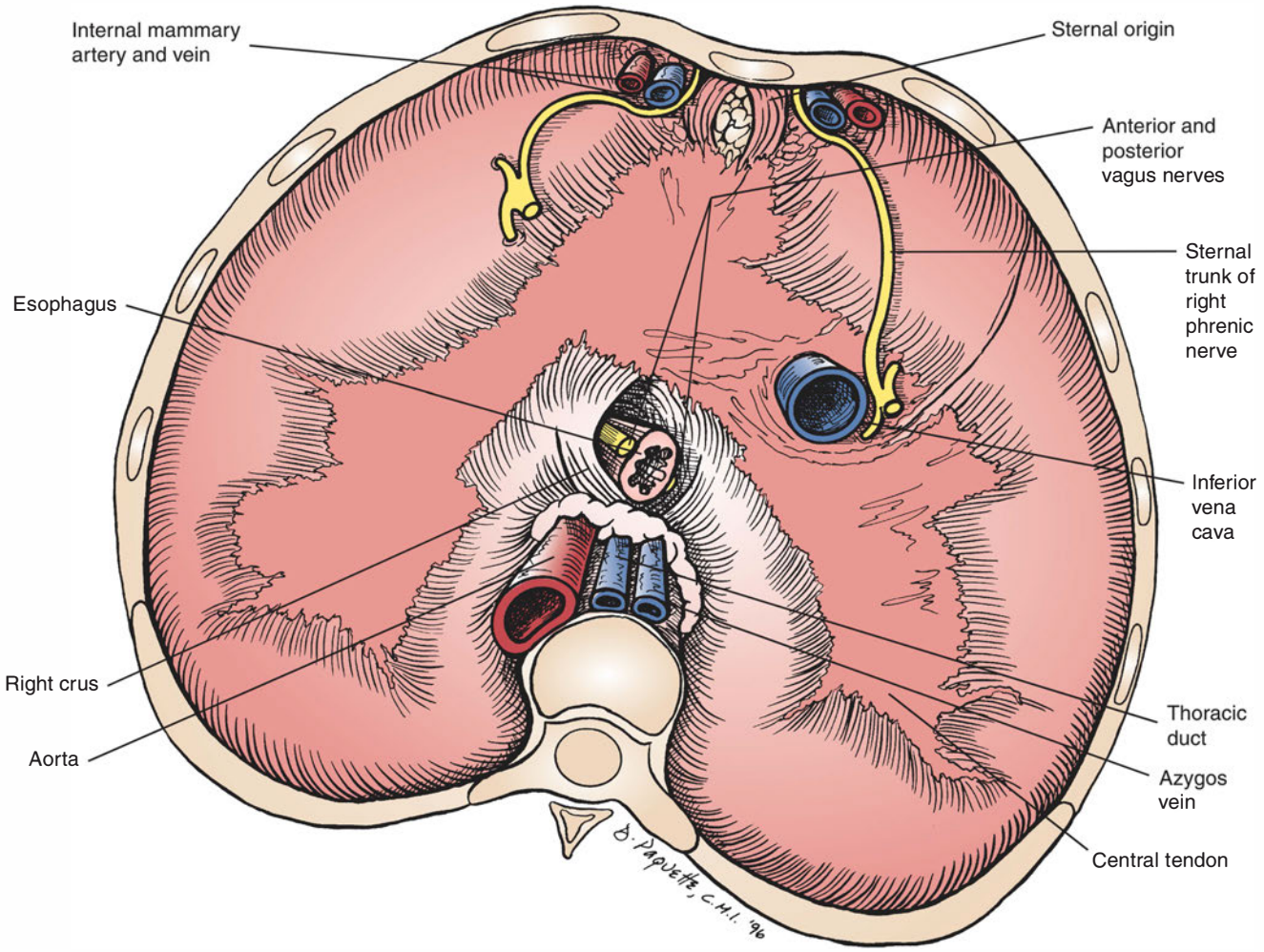


Fig. 26.1. Anatomy of the diaphragm as viewed from the thorax.

inferior phrenic veins, which drain into the IVC on the right, and both the IVC and the adrenal vein on the left.

Motor innervation of the diaphragm comes exclusively from the phrenic nerves (Fig. 26.2). These nerves arise from C3–C5, coming down through the thorax posteriorly, and moving anterolaterally over the pericardium. On the left, the phrenic nerve enters the diaphragm just lateral to the cardiac border and, on the right, the nerve enters the diaphragm just lateral to the caval hiatus. The phrenic nerves, supplying all motor and some sensory innervation of the diaphragm, are made up of four trunks: sternal, crural, anterolateral, and posterolateral. The nerves first give off the sternal branch, then penetrate the diaphragm and course along the inferior surface of the diaphragm. The peripheral portions of the diaphragm receive some sensory fibers from the intercostal and subcostal nerves, as well.

NORMAL FUNCTION

As the chief muscle of inspiration, the diaphragm descends during inspiration and rises during expiration. The right dome may reach as high as the level of the 5th rib and the left dome may ascend to the 5th intercostal space. The exact level of the diaphragm will vary with the phase of respiration, the patient's position (supine vs. upright), and the volume occupied by the abdominal viscera. Contraction of the diaphragm results in flattening of the domes, increasing the intrathoracic volume, and permitting negative pressure and entry of air into the chest.

For ideal respiratory mechanics, both hemidiaphragms ought to be functionally intact. Unilateral phrenic nerve dysfunction manifests via hemidiaphragm elevation with suboptimal ventilatory movements. Injury or paralysis of bilateral phrenic nerves can result in significant respiratory compromise, with limited capacity for compensation by the accessory respiratory muscles.

In addition to its important role in ventilation, contraction of the diaphragm also has a significant impact on circulatory function, with changes in intraabdominal and intrathoracic pressures aiding in appropriately timed increases in venous return. Further, contraction of the diaphragm may alter intraabdominal pressures such as to facilitate emesis, defecation, and bladder emptying.

SURGICAL CONSIDERATIONS

When operating on or near the diaphragm, familiarity with the path and anatomic location of the phrenic nerves is of great importance. Preservation of diaphragmatic function is best achieved through intraoperative awareness of phrenic innervation and careful placement of incisions when opening the diaphragm for access or excision. Depending on the goals and details of the specific operative procedure, one of several common incisions may prove useful

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Fig. 26.2. Anatomy of the diaphragm with the branches of the phrenic nerve as viewed from the abdomen. The sternal branch of the phrenic nerve is seen in Figure 26.1.

(Fig. 26.4). Circumferential incisions are placed parallel to the muscle edge, approximately 2 cm from the peripheral margin. Radial incisions may be used, coming from the central tendon outward, but the exact location must be selected such that only the most distal branches of nerve are at risk of transection. A midline incision may be made, either anteriorly or posteriorly, but care must be taken to ligate the left inferior phrenic vein to avoid bleeding from this vessel.

IMAGING AND FUNCTIONAL EVALUATION

Prior to operating on the diaphragm, appropriate diagnostic studies should be obtained to confirm presumed diagnoses and to characterize relevant anatomic and physiologic features. Chest radiography serves as a useful initial study for a variety of diaphragmatic abnormalities, and may be followed by additional tests for diagnostic confirmation and further elucidation of specific pathologic features and relevant anatomy. Cross-sectional imaging, via computed tomography (CT) and magnetic resonance imaging (MRI), may provide needed details regarding diaphragmatic defects and neoplastic processes and their relationships with surrounding structures.

When concern has been raised for functional deficits of the diaphragm, the test of choice is termed a "sniff test." This imaging study, performed under fluoroscopy, involves dynamic imaging of the diaphragm with observed sniffing. In this test, a dysfunctional hemidiaphragm should demonstrate paradoxical movement with the degree of paradoxical movement having clinical relevance. Dynamic breathing MRI can also be used to assess diaphragm function in the setting of concern for diaphragmatic paralysis. Phrenic nerve stimulation may be additionally helpful in evaluating diaphragm function in the setting of neuromuscular disorders.

SPECIFIC DISORDERS AND MANAGEMENT

Congenital Defects

Morgagni Hernia

The hernia of Morgagni is an anterior, retrosternal diaphragmatic defect, occurring between the xiphoid and the costochondral attachments of the diaphragm (Fig. 26.5). Relatively rare, this embryologic failure of myoblast migration accounts for less than 1 in 50 cases of diaphragmatic defects. With this defect, abdominal contents tend to herniate on the patient's right, with the left-most aspect of the defect occupied by pericardium. While this congenital defect is present at birth, symptoms are relatively minor, and, oftentimes,

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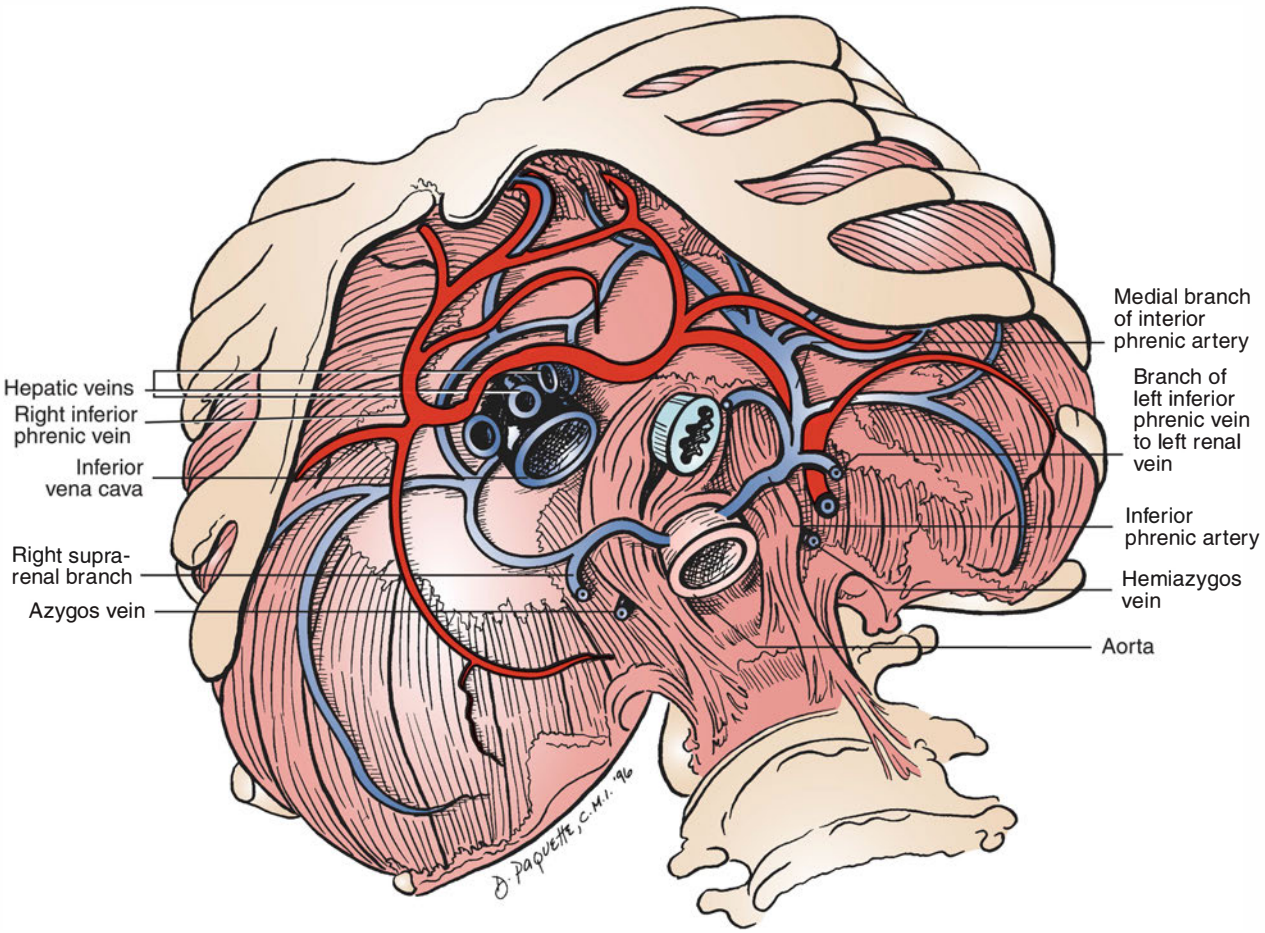


Fig. 26.3. Abdominal view of the diaphragm showing the arterial supply and venous drainage.

patients do not present until adulthood. Increased abdominal pressure, such as with obesity or pregnancy, tends to precipitate increased symptoms, frequently leading to the diagnosis. These symptoms may range from vague fullness to bowel obstruction, and in some patients the defect may be identified incidentally upon radiographic evaluation performed for unrelated indications. Regardless of the extent of symptoms, operative repair is performed for all appropriate surgical candidates in order to prevent complications of obstruction and ischemia.

Repair of Morgagni hernias is typically performed via an abdominal approach. Either a midline or subcostal incision will facilitate appropriate visualization and exposure. Laparoscopic repair is also well described. For those circumstances in which herniated structures appear to be fixed high in the chest, a transthoracic approach may be required for adhesiolysis and sac reduction. After gaining access, attention should be directed toward reducing contents of the sac using gentle traction, taking care to avoid injury to involved organs. The sac is then resected. Following reduction and resection of the hernia sac, repair of the diaphragm is performed. The defect may, potentially, be closed primarily if there remains a reasonable margin of muscular tissue around the defect. Closure is performed with heavy nonabsorbable suture placed in an interrupted fashion (Fig. 26.6). In the absence of a complete muscular rim, the defect will require attachment of the free muscle edge to the costal margin. One should also note that if a primary repair results in excessive tension, nonabsorbable mesh or a polytetrafluoroethylene (PTFE) patch should be placed to close the defect between the diaphragmatic rim and the chest wall. If the pleural space is entered, a chest tube should be placed at the end of the procedure.

Bochdalek Hernia

Congenital failure of the closure of the pleuroperitoneal canal results in a posterolateral diaphragmatic defect, permitting herniation of the foregut structures as they return into the abdominal cavity (Fig. 26.5). The majority of these defects tend to be left sided (>85%), and they are often associated with significant cardiac anomalies. Major morbidity results from the space occupation in the chest by the abdominal organs, ultimately hindering normal lung development. The subsequent pulmonary hypoplasia renders many of these patients critically ill, during the delivery and neonatal periods, ultimately causing substantial morbidity and mortality.

Diagnosis is typically made by prenatal ultrasound, which facilitates advanced planning for delivery, perinatal care, and operative intervention. Postnatal diagnosis is most often made after chest radiography demonstrates intestinal contents in the thoracic cavity. As mentioned, this structural defect is accompanied by physiologic abnormalities, often including some degree of pulmonary hypoplasia as well as pulmonary hypertension. A number of important recent advances have been made in improving the care for patients with this

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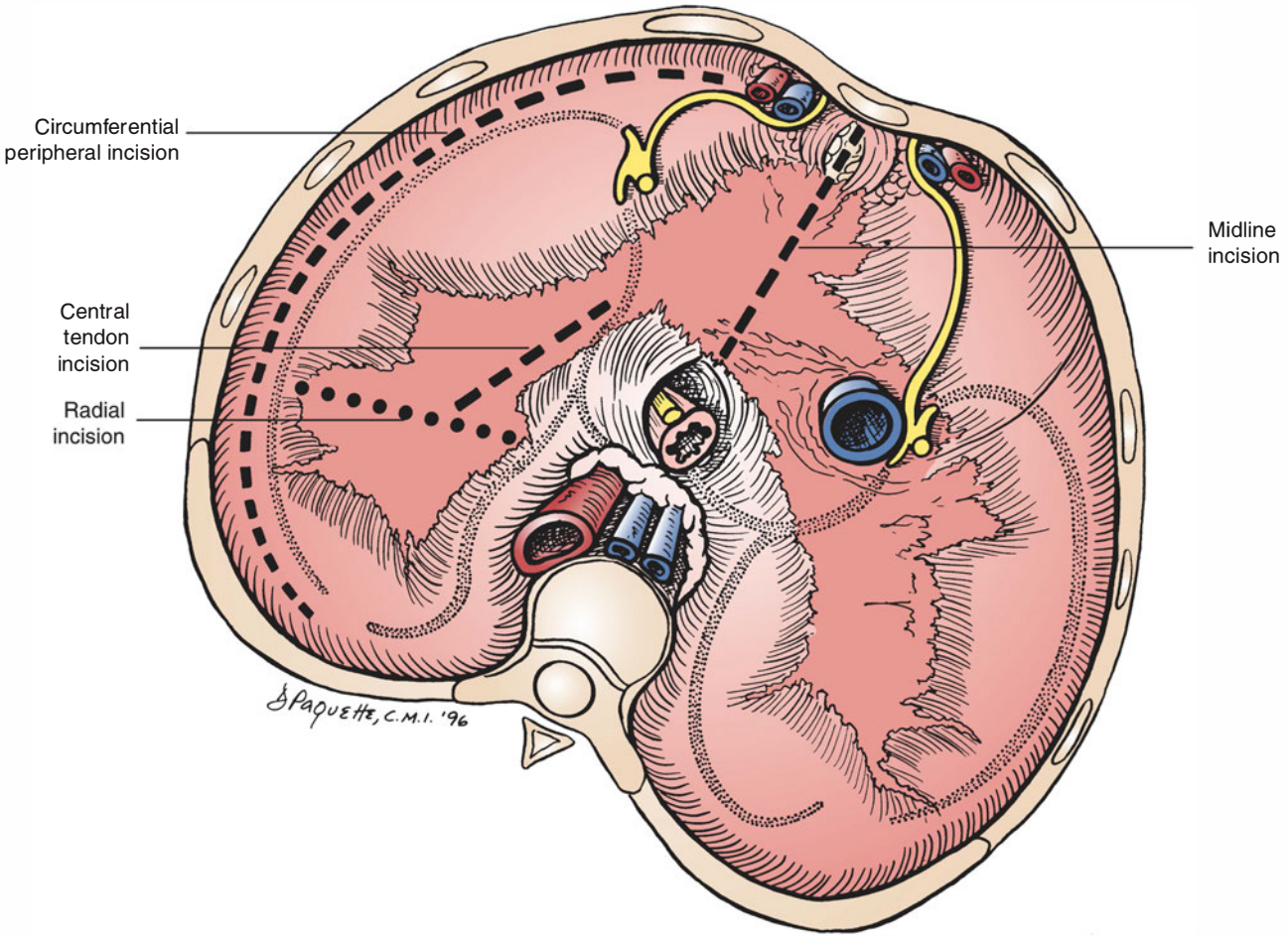


Fig. 26.4. Thoracic view of the diaphragm showing the common diaphragmatic incisions and their relationship to the branches of the phrenic nerve.

condition, including improved prenatal detection, extracorporeal membrane oxygenation (ECMO), and recognition and treatment of severe pulmonary hypertension. Lung protection strategies, such as the use of high-frequency oscillatory ventilation (HFOV) and adherence to airway pressure limitations, have been employed to limit barotrauma. However, despite these treatment advances, overall survival remains in the range of 60% to 70% dependent to a significant extent on the severity of the abnormality. Due to the high prevalence of associated anomalies, thorough evaluation should be undertaken to identify any other major congenital conditions prior to consideration of operative intervention. While plans for operative intervention should be initiated upon diagnosis, repair should not be performed emergently. Infants should be placed on mechanical ventilation, undergo nasogastric decompression, and have nutritional needs addressed, allowing stabilization in the neonatal intensive care unit preoperatively. Once the infant has been physiologically optimized and the presence of other major congenital anomalies has been investigated, operative repair may be considered.

A number of protocols have been described with regard to timing of operative repair. In general, preoperative goals include radiographic clearing of lung fields, optimization of tidal volume in the setting of minimal positive end expiratory pressure (PEEP), approximation of euvolemia, and demonstration of maximized right ventricular function. Repair may be performed on or off ECMO, and outcomes with each vary among institutions. The operative procedure is performed through a subcostal abdominal incision, and, after facilitating appropriate exposure, the hernia contents are gently reduced. Hernia sacs are present in a minority of cases, but should be uniformly sought and resected if present. Edges of the defect should be identified, and an assessment should be made regarding feasibility of primary repair, which should only occur in the setting of adequate tissue that can be brought together without tension. Prior to closing the defect, a chest tube should be placed under direct visualization. If primary repair is feasible, this can be performed with nonabsorbable interrupted sutures placed in a mattress fashion. Large defects may require prosthetic mesh that should be stitched in place with nonabsorbable, pledgeted sutures (Fig. 26.7). In the absence of a posterior muscular rim, it may be necessary to suture the prosthesis directly to the ribs and intercostal musculature. Once the diaphragm has been repaired, as the abdominal viscera are returned to their appropriate locations and attempts are directed toward closing the abdomen, inadequate abdominal domain may pose a dilemma. This can be dealt with by using mesh to bring the abdominal wall together without tension, by closing the skin only (resulting in a ventral hernia), or use of a silo. Even after a well-timed and carefully executed operative repair, patients may remain critically ill and in a tenuous cardiopulmonary state postoperatively. Ongoing efforts must continue to achieve the best possible outcomes for these infants.

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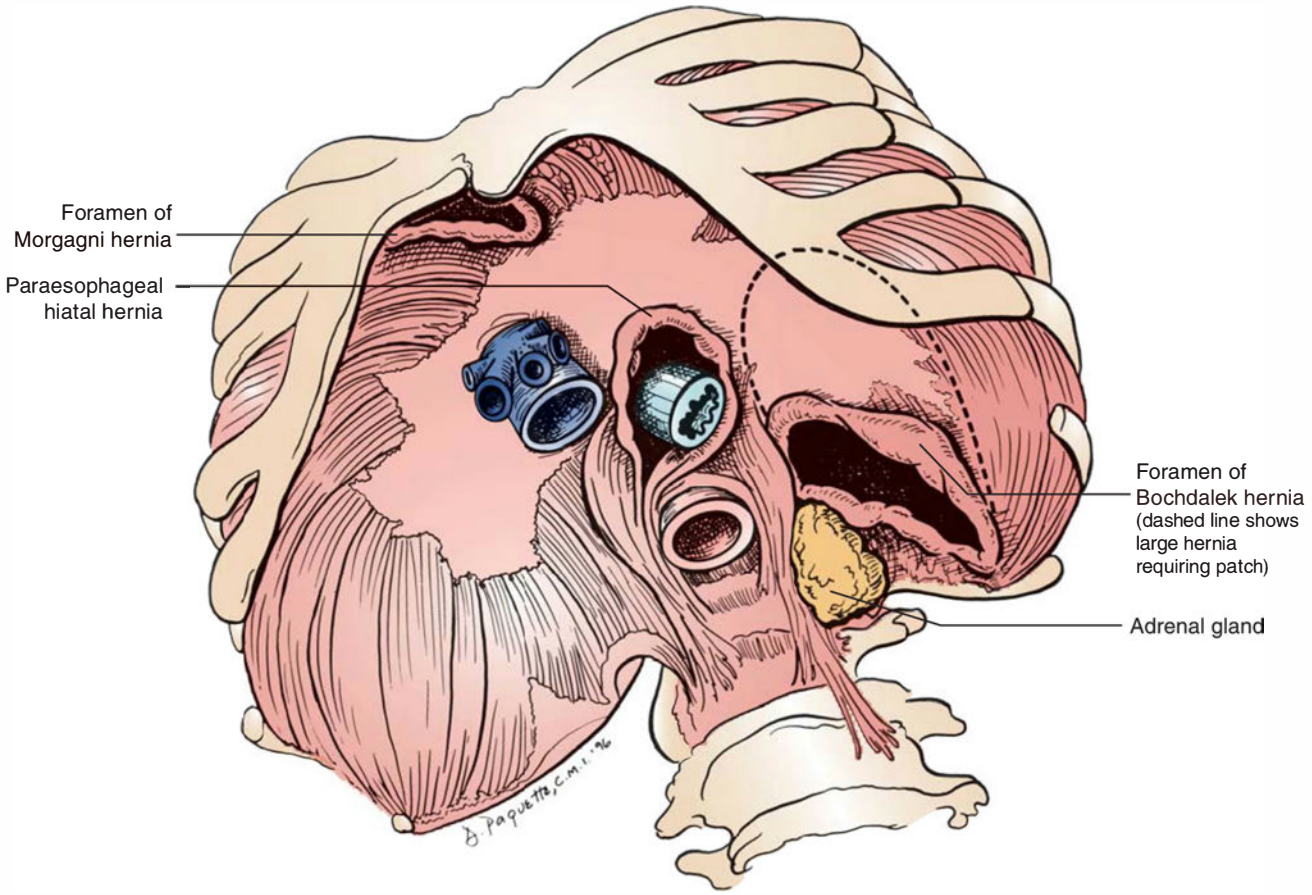


Fig. 26.5. Abdominal view of the diaphragm showing the location of the common hernias.

Acquired Defects

Hiatal Hernia

Hiatal hernias are categorized into four subtypes, with the most common being the type I sliding hernia. The type I hiatal hernia and its management are discussed elsewhere in this book, in terms of evaluation and treatment for gastroesophageal reflux disease. Type II, or paraesophageal, hernias involve a focal weakening of the phrenoesophageal membrane anterolateral to the esophagus, permitting herniation of the gastric fundus and antrum despite intraabdominal fixation of the distal esophagus and gastric cardia (Figs. 26.5A and 26.8B). Obstruction may then occur at the level of the diaphragm, with the stomach additionally at risk for volvulus and ischemia. Type III is comprised of both a type I and type II, while type IV involves generalized enlargement of the hiatus with multiple abdominal viscera in the chest.

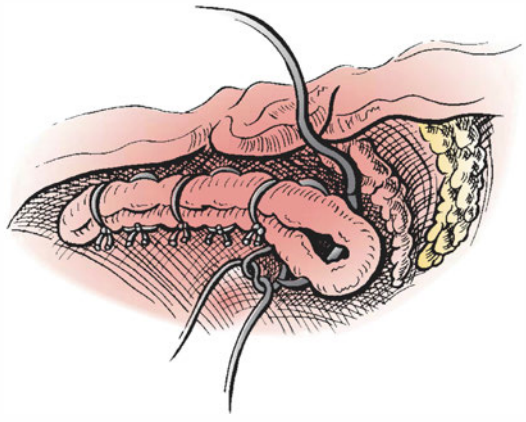


Fig. 26.6. Repair of hernia of Morgagni. Abdominal approach when an anterior rim of muscle is present.

Patients with paraesophageal hernias may report some substernal fullness, postprandial discomfort, or symptoms of reflux. Many of these patients may not be symptomatic at all, or may be diagnosed only when they present with complications of the hernia. Those patients presenting with volvulus or obstruction may be critically ill with severe pain and nausea and will require emergent operative intervention to prevent strangulation. Historically, it was widely recommended to repair all paraesophageal hernias, regardless of symptom profile. More recent attention has been given toward potentially observing asymptomatic patients, particularly those that are high risk for operative intervention.

In repairing paraesophageal hernias, several approaches have been described. Traditionally, this involved thoracotomy for hernia reduction, followed by laparotomy for Belsey or Nissen fundoplication, and, when appropriate, Collis gastroplasty. While this strategy is still employed, our preferred approach is via laparoscopy. Our technical approach to giant hiatal hernia repair requires consistent emphasis on several key operative points, tenets that hold the surgeon to the same principles of

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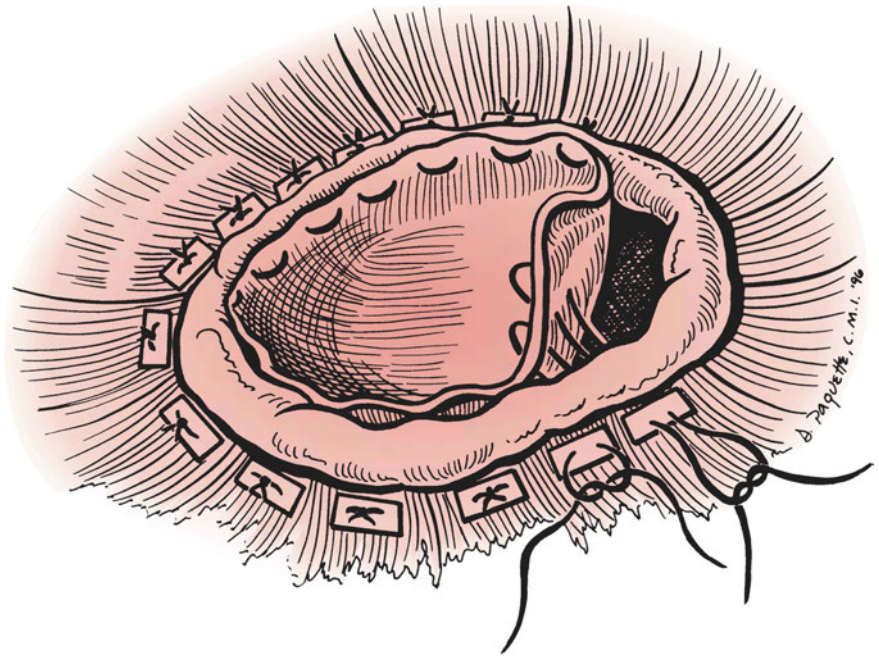


Fig. 26.7. Repair of a large hernia of Bochdalek with patch. The interrupted mattress sutures are buttressed with pledgets.

open surgery, and consequently result in comparable outcomes.

After gaining laparoscopic access, attention is directed toward reduction of the stomach. The stomach itself is not grasped directly; rather, the apex of the hernia sac is grasped and inverted by pulling caudally, and the sac is opened well away from the crural edge. Dissection is carried into the mediastinal areolar tissue plane, allowing the stomach to fall into the abdomen. Care is taken to maintain crural integrity without exposing the crural muscle fibers. Following division of the sac, the fat pad is dissected circumferentially. In this process, by staying right on the surface of the muscle fibers of the esophagus and stomach, the vagal nerves should be successfully preserved. The crura are then closed in an interrupted fashion with nonabsorbable #1 sutures and intracorporeally tied knots, reinforced with pledgets, for approximately two-thirds of the anteroposterior distance. Use of standard suture and needles allows for appropriately sized,

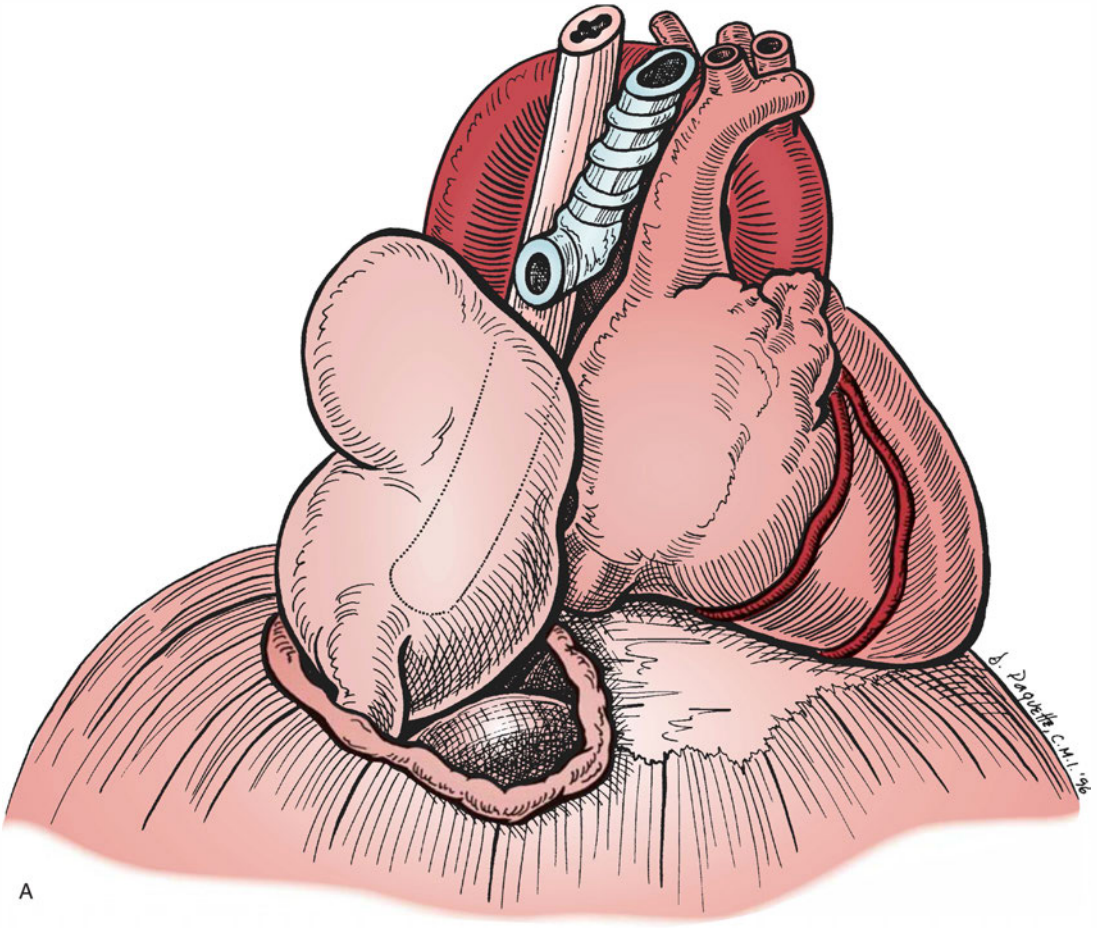


Fig. 26.8. Paraesophageal hernia. (**A**) Type II hernia with the duodenum and gastroesophageal junction anchored below the diaphragm. (**B**) Type III hernia with migration of the gastroesophageal junction and duodenum into the chest. (**C**) The completed Nissen fundoplication demonstrating the three sutures holding the wrap in addition to the placement of the sutures anchoring the fundus to the crura.

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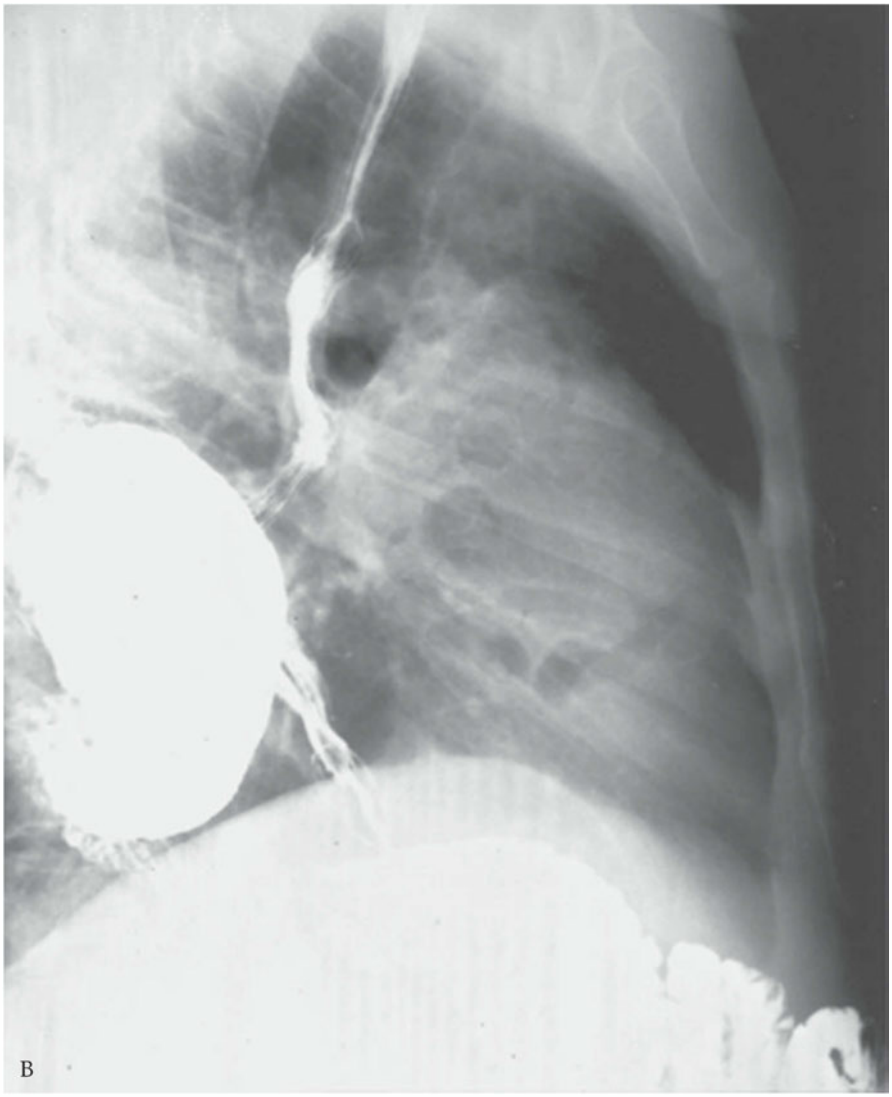


Fig. 26.8. (Continued)

full-thickness crural bites, replicating the crural closure achieved in open operations.

After closing the crura, the intraabdominal portion of esophagus is assessed for adequate length, and if found to be less than 2.5 cm, a wedge Collis gastroplasty is performed over a 46F bougie. The greater curve of the stomach is held anteriorly and inferiorly, and stapler brought through the left upper quadrant is positioned nearly perpendicular to the direction of the bougie. A second stapler load is typically necessary from this angle, followed by one or two stapler loads from the right paramedian location.

In our practice, a Nissen fundoplication is next performed over a bougie, constructed with three stitches of 2-0 silk, incorporating a moderate bite of esophagus into each stitch. Ensuring adequate (but not too much) tension is of key importance. After constructing the wrap, three stitches of 2-0 silk are used to anchor the top of the fundus to the crura (Fig. 26.8C). In some circumstances, we complete the repair with a biologic mesh buttressing of the closed hiatal defect, based on multiple studies suggesting a decrease in recurrence when mesh is employed. While open procedures remain efficacious for paraesophageal hiatal hernia repair, reports from centers of excellence have demonstrated that a minimally invasive approach can be performed safely and with comparable results, thus sparing patients the morbidity of a more invasive operation.

Trauma

The management of traumatic injuries to the diaphragm depends upon the timing of presentation; injuries presenting in the acute setting may be approached differently than those diagnosed after a period of chronicity. Such injuries may be secondary to blunt or penetrating trauma, with motor vehicle accidents accounting for the majority of blunt mechanisms and knife and gunshot wounds comprising most of the penetrating injuries.

Diaphragmatic injuries in the setting of blunt trauma require mechanisms associated with a tremendous amount of force, and are often accompanied by a number of additional and severe injuries. Acute rupture of the diaphragm from blunt injury occurs much more frequently on the left than on the right, upon transmission of force from the abdominal viscera upward. On the right, the liver serves to absorb some of the transmitted energy, thus resulting in fewer right-sided diaphragmatic injuries.

Penetrating trauma tends to be a more frequent cause of diaphragmatic injury than blunt trauma. As opposed to blunt mechanisms of injury, these cases typically occur with more focal (rather than diffuse) concomitant injuries. In patients with penetrating injuries to the chest, a plain radiograph obtained in the trauma bay may reveal abdominal viscera in the thoracic cavity, indicating a diaphragmatic rupture. However, defects may not always result in immediate herniation. For patients with penetrating injuries to the chest that do not meet criteria for operative exploration, close follow-up is useful in identifying any diaphragmatic injuries missed at initial evaluation. For patients with penetrating wounds to the abdomen, operative exploration is often mandated, and the diaphragmatic defect can be found intraoperatively. Careful evaluation of the diaphragm is essential to avoid late herniation through an unnoticed defect. Laparoscopy has been used both for evaluation and repair of diaphragmatic defects in this setting. The authors caution the use of laparoscopy for this application as a thorough and complete evaluation is often tremendously challenging, especially as it relates to full evaluation of the posterior portion of the diaphragm.

Patients undergoing exploration for acute traumatic injury are typically approached via midline laparotomy. Diaphragmatic repair, while important, should be performed following management of any life-threatening hemorrhage or other significant visceral injury. Areas of nonviable tissue should be excised. Traumatic diaphragmatic defects can usually be repaired primarily. This should be done with nonabsorbable interrupted sutures (often pledgeted) placed in a mattress fashion. In rare circumstances mesh may be necessary but this is typically not the case for acute diaphragmatic lacerations. If mesh is used and a phrenic nerve injury appears to have

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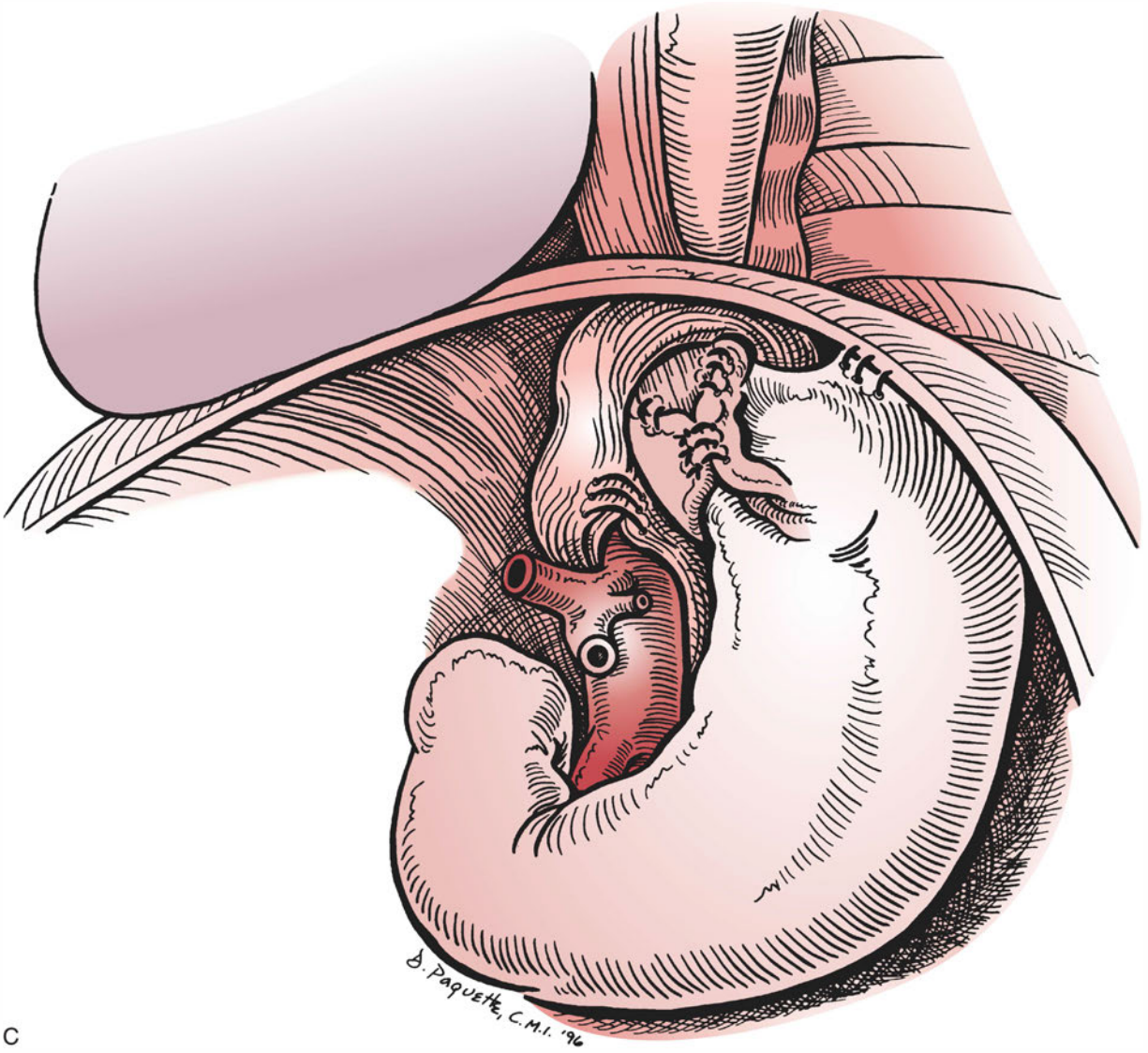


Fig. 26.8. (Continued)

occurred, care should be taken to ensure that the mesh is taut so as not to allow paradoxical diaphragmatic motion in the future.

The late or delayed identification of diaphragmatic injuries requires a different approach, as abdominal viscera may be involved by extensive adhesions in the chest. Even if patients have been asymptomatic for a prolonged latent phase, operative repair is necessary to prevent obstructive complications. The traditionally described approach for this operation was via thoracotomy in order to permit adhesiolysis and visceral reduction. However, in recent years, a number of techniques employing thoracoscopy and/or laparoscopy have been described. If the hernia contents appear above the level of the inferior pulmonary vein, an intrathoracic approach is typically safest. For patients with larger defects, thoracoscopy may prove challenging due to visual obstruction by the herniated viscera.

Regardless of the type of access used, the operative principles are the same. Abdominal contents are gently reduced, the hernia sac is excised, and the defect is repaired. If the defect comes together primarily without tension, this can be performed with interrupted nonabsorbable sutures; however, mesh patches may be necessary if a primary repair would result in undue tension thus increasing risk of disruption and recurrence.

Eventration and Paralysis

While diaphragmatic eventration and paralysis occur as two discrete pathologic entities, they share similar physiologic manifestations, and thus surgical treatment—when indicated—is similar in approach. Eventration is a congenital abnormality resulting from failed development of the muscular portion of the diaphragm, with maintenance of the normal attachments to the sternum, ribs, and spine. Grossly, the affected diaphragm appears thinned and attenuated, and is functionally ineffective during respiration. While more common on the left, this defect may be unilateral or bilateral, and may affect just a portion or an entire hemidiaphragm.

Clinically distinct from eventration, acquired paralysis or paresis of the diaphragm may occur in several clinical settings, with the most common etiologies being phrenic nerve trauma after cardiac surgery and encroachment of thoracic malignancies upon the phrenic nerve. In the postoperative setting following cardiac procedures, paralysis may be due to traction on the nerve, pressure from a retractor, thermal injury resulting from techniques used to cool the heart, or associated with the use of cautery in the vicinity of the nerve. This deficit, at times, may be

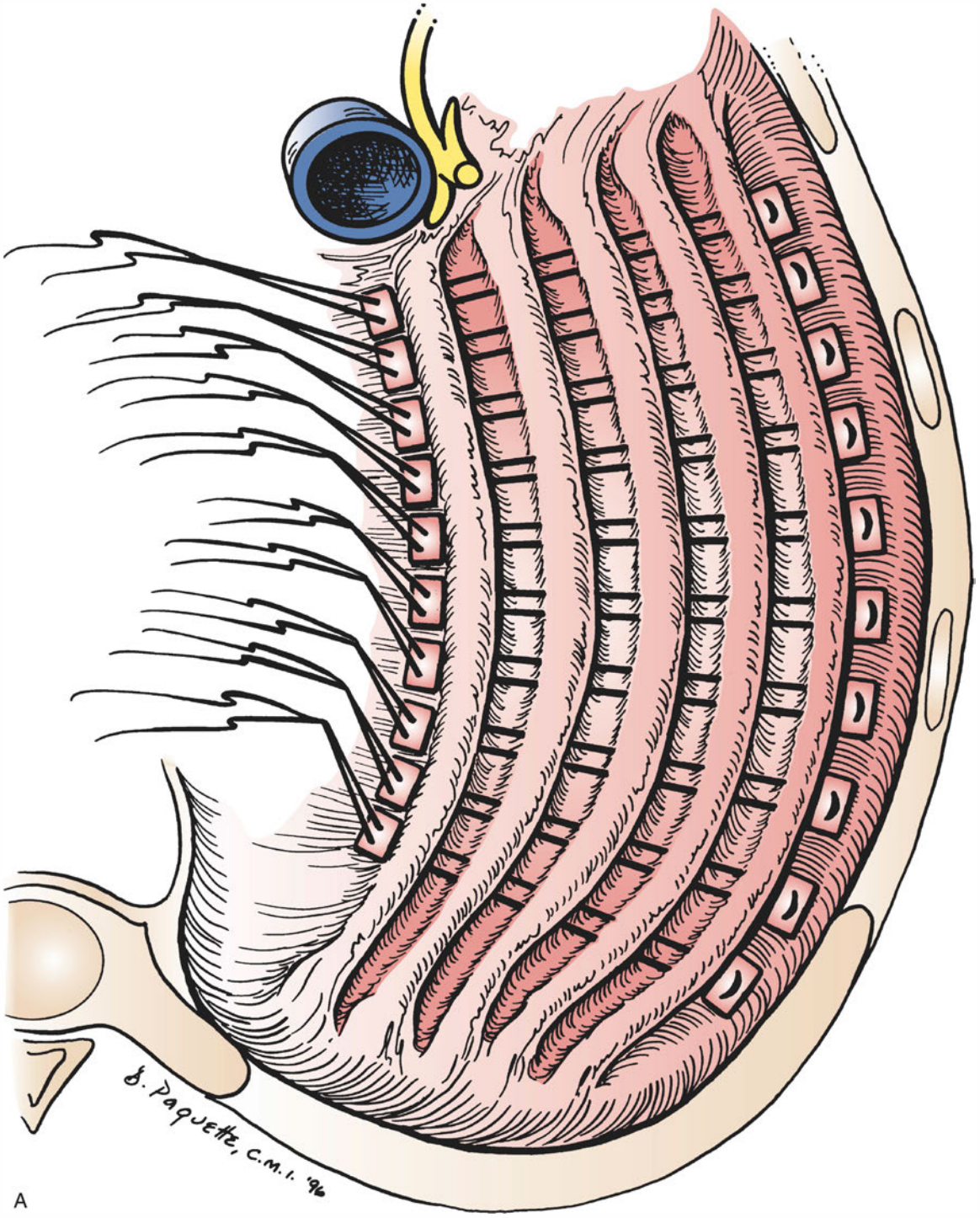


Fig. 26.9. Technique for plication of the *right* diaphragm in an adult via thoracotomy. (**A**) Sutures buttressed with Teflon pledgets extend anteriorly to the level of the vena cava. (**B**) Plicated diaphragm. (**C**) Repair of diaphragmatic eventration using a video-assisted thoracoscopic technique to plicate the diaphragm. ICS, intercostal space.

temporary in nature. Less common causes of phrenic nerve dysfunction include trauma, neuromuscular disorders, or infectious processes. A number of cases are also idiopathic, with no clear inciting factor prior to development of diaphragmatic dysfunction.

As both of these processes result in ineffective caudal excursion of the diaphragm during inspiration, symptoms may be similar but especially with eventration may not be present to any significant extent. Impaired ventilation leads to the dominant complaint of dyspnea. As portions of the lung adjacent to the dysfunctional diaphragm display alveolar hypoventilation, regional vasoconstriction occurs, with relative ventilation/perfusion mismatch. Mild hypoxemia may be present, inciting compensatory hyperventilation and patient perception of shortness of breath. Orthopnea may also be present as the individual's abdominal organs displace lung volume in a supine position. For patients with both of these diagnoses, diaphragmatic plication may be an effective therapy in achieving symptom relief.

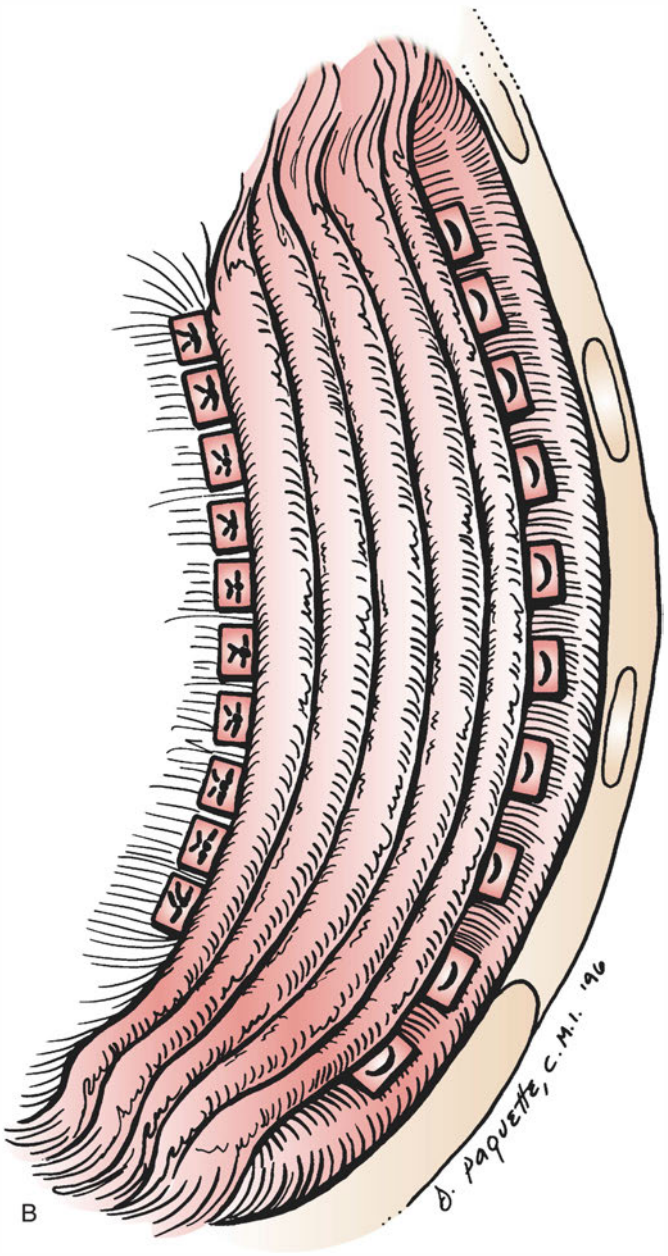


Fig. 26.9. (Continued)

Diaphragmatic plication has been shown to have efficacy in symptomatic patients with eventration as well as diaphragmatic paralysis. A number of operative techniques have been described, including those that are transabdominal as well as transthoracic. While early reports detailed open procedures, minimally invasive operations have been highly favored in recent literature. Regardless of the surgical approach, there are several generally accepted operative indications for plication. Patients should be symptomatic; no intervention is necessary for those individuals who experience no respiratory compromise as a result of their condition. There should be significant paradoxical motion of the affected hemidiaphragm seen on the sniff test. For patients who have phrenic nerve paralysis after cardiac surgery, operative intervention should follow a period of observation (typically 1 year) if it is reasonably tolerated, as many of these paralyses are transient and improve with time. Finally, patients ought to be appropriate operative candidates. Benefits of plication are modest, and, as with all interventions, the risk–benefit ratio should be carefully weighed for each patient.

Traditionally, diaphragmatic plication has been described through a thoracotomy via the 8th intercostal space. The lateral and posterior portions of the diaphragm are gathered in a pleated approach with interrupted 0 sutures that are pledgeted (Fig. 26.9A). Sutures are placed at 1 cm intervals and brought from the central portion (medially) of the diaphragm out to the peripheral margin of the diaphragm laterally (Fig. 26.9B). Completion of the plication affords the patient a taut diaphragm that will overcome the phrenic nerve dysfunction on that side by reducing the position during peak inspiration.

The thoracoscopic approach has also been described recently. For this approach a double-lumen tube is placed and the technical points are demonstrated in Figure 26.9C. The principles of the operation follow those of the open approach. This is certainly more challenging because of the steric issues encountered.

Our preferred approach to diaphragmatic plication is via laparoscopy. This approach works well for both left- and right-sided paralysis. We perform this procedure under general anesthesia with a single-lumen endotracheal tube. (Selective ventilation is not necessary.) Patients are placed in a supine position and secured to the operating table to allow for steep reverse Trendelenburg positioning during the procedure. Four 12-mm trocars are placed, with two in the midline and an additional two in the ipsilateral upper quadrant of the abdomen. Consequent to the pneumoperitoneum, the abnormal hemidiaphragm is displaced cranially. Electrocautery is used to make a small perforation in the dome, and the chest is entered. The resultant pneumothorax allows the hemidiaphragm to be pulled downward for plication.

For the plication, we use pledgeted nonabsorbable suture to place U-stitches into the diaphragm. The posterior portion is plicated first in an anteroposterior direction followed by a second plication line directed lateromedially, resulting in a final T-shaped plication (Fig. 26.10). In the process of performing the plication, the initial perforation at the dome is closed. We then place an 18F to 20F chest tube in the ipsilateral chest at the end of the procedure. Patients receive intense postoperative pulmonary toilet, and chest tubes are removed when drainage diminishes to less than 200 ml/day. The initial postoperative radiograph should demonstrate the plicated diaphragm at a lower position than the unaffected contralateral side. This will normalize with both diaphragms at the same level by 1-month follow-up. At 1 month postoperatively, both diaphragms

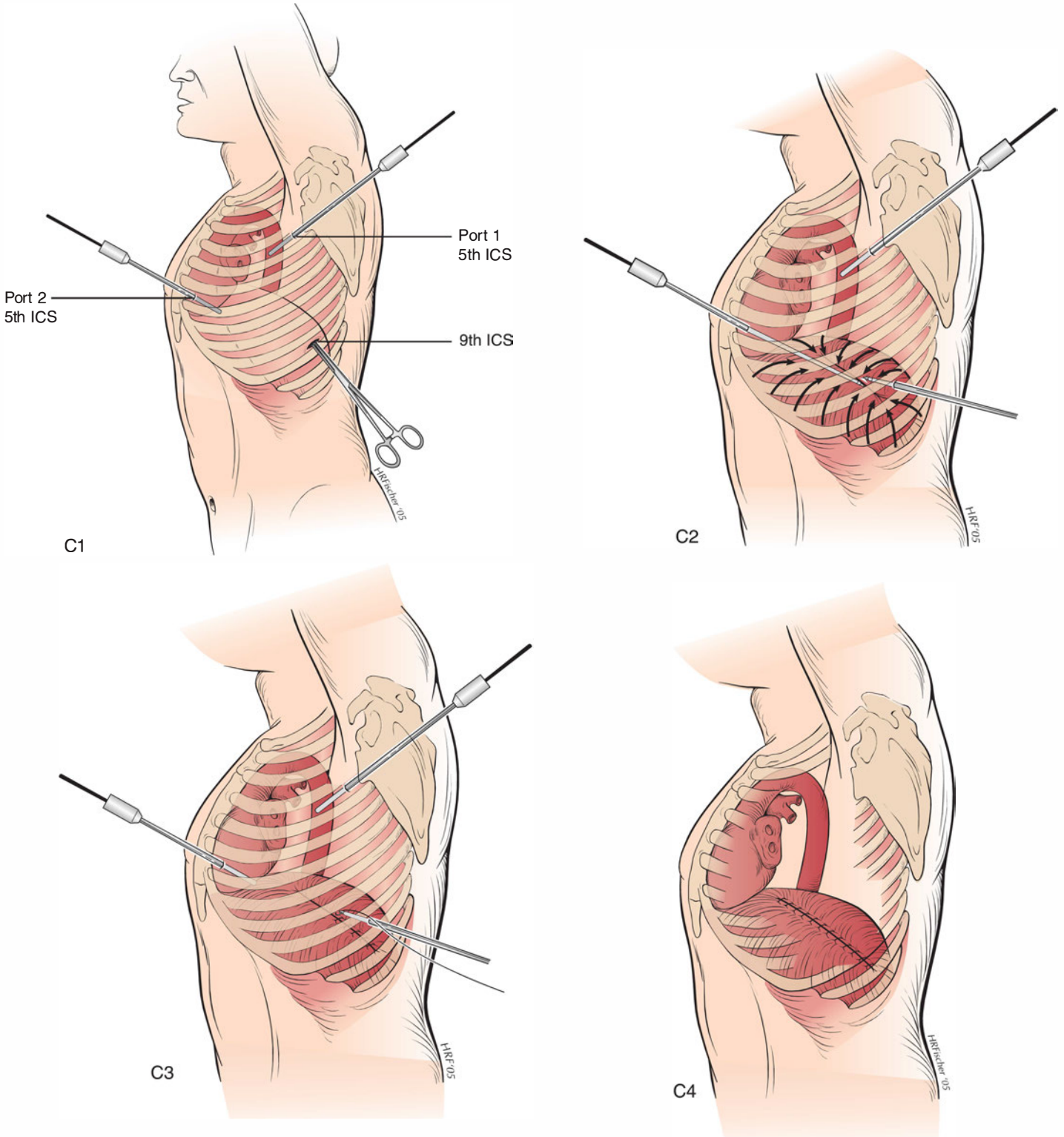


Fig. 26.9. (Continued)

should be at the same level. While there are a number of additional methods described for diaphragmatic plication, our outcomes—with regard to symptom relief, hospital course, and postoperative complication—have been excellent with this strategy.

Central Nervous Diaphragmatic Failure

Though less common than phrenic nerve paralysis, diaphragmatic dysfunction may be secondary to central neurologic disorders, including high cervical spinal cord injuries and primary alveolar hypoventilation. While spinal cord injuries are due almost exclusively to trauma, or rarely occur after an ischemic event, primary alveolar hypoventilation is a congenital disorder. Patients with this diagnosis fail to manifest an appropriate autonomic response to hypercarbia and

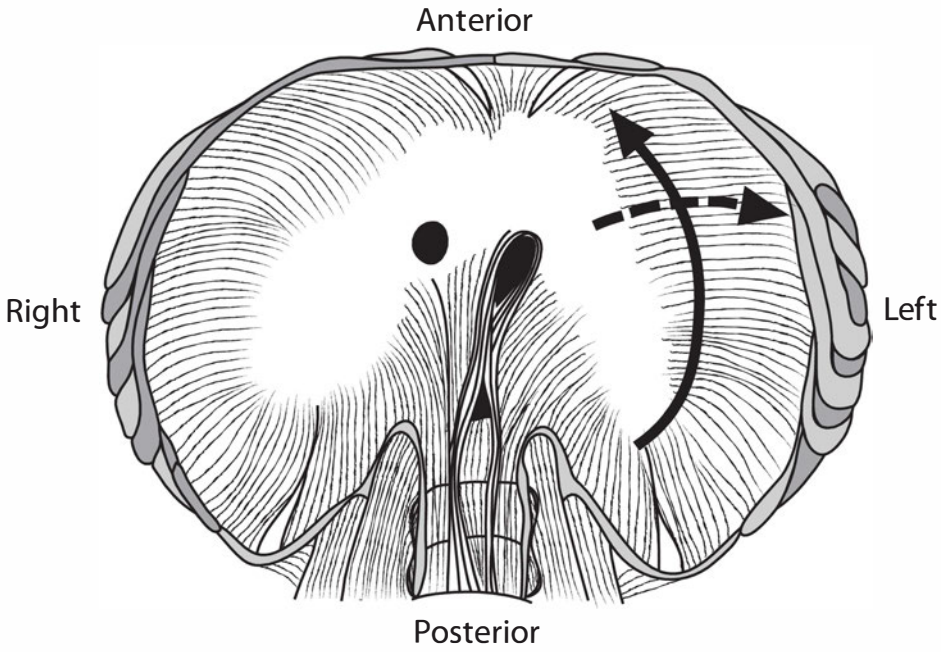


Fig. 26.10. Laparoscopic plication of the diaphragm. Schematic demonstrating the course of plication for the laparoscopic approach. Sutures are placed from a posterior to anterior approach and then a medial to lateral approach. (Reprinted with permission from Groth SS, Rueth NM, Kast T, et al. Laparoscopic diaphragmatic plication for diaphragmatic paralysis and eventration: an objective evaluation of short-term and midterm results. J Thorac Cardiovasc Surg 2010;139:1452, with permission.)

hypoxia, secondary to abnormal receptors in the medulla oblongata. These individuals are at great risk of apneic events during sleep; during wakeful periods, they continue to breathe via conscious efforts. Diaphragmatic pacing may serve as a potentially useful therapy for these patients. Pacing may also be beneficial for individuals with cervical spine injuries, and is most useful for those above the C3 level, as the phrenic nerve remains intact. If C3–C5 nerve roots are involved, less benefit is derived from pacing, and if the injury is below C5, there is no benefit to phrenic nerve pacing.

In order to pace the diaphragm, an electrode must be placed in contact with the phrenic nerve. This may be accomplished via open cervical or thoracotomy approach, or, more recently, thoracoscopic approaches have been described, as well. Patients should not be paralyzed intraoperatively, so that stimulation of the nerve can be recognized. The electrode should ultimately be placed against the phrenic nerve adjacent to the mediastinum. This is connected to a subcutaneously placed receiving unit, which in turn communicates with an external pacemaker.

Porous Diaphragm Syndromes

Porous diaphragm syndromes are an uncommon and diverse group of diseases that result from a common anatomic etiology. That etiology is a diaphragmatic defect that affords the translocation of substances from the peritoneal cavity into the pleural space. The most common site of the defect is in the tendinous portion of the diaphragm. These defects may be congenital, but more often are acquired. Thoracic involvement tends to be more common on the right side.

Two of the most illustrative examples of these syndromes include Meigs syndrome and catamenial pneumothorax. In Meigs syndrome (fibroma or fibroma-like benign ovarian tumor, ascites, and pleural effusion) there is a clear relationship between the tumor, ascites, and pleural effusion. Surgical removal of the tumor treats the syndrome, and the thoracic surgeon is involved in management of the pleural effusion until resolution of the hydrothorax. In catamenial pneumothorax, patients present with a spontaneous pneumothorax that occurs at the start of a menstrual cycle. Prevailing evidence suggests that ectopic endometrial implants along the diaphragm cause perforations in the diaphragm leading to retrograde filling of the thoracic cavity with air from the fallopian tubes. Treatment involves thoracoscopy (or thoracotomy) with closure of the diaphragmatic defect, if it can be identified, and pleurodesis. Menstrual periods must be chemically ablated for several months before and after the procedure to avoid a high incidence of recurrence.

Diaphragmatic Malignancies

Infrequent in occurrence, the majority of primary diaphragmatic tumors are benign, with a number of described histologies. These include fibromas, lipomas, neurofibromas, schwannomas, and leiomyomas, as well as bronchial, echinococcal, mesothelial, and teratogenous cysts. Among malignant tumors of the diaphragm, fibrosarcomas and solitary fibrous tumors are the most common. Such primary lesions of the diaphragm may be discovered incidentally on chest radiography, and can be further characterized with MRI and/or CT imaging.

More common than primary diaphragmatic tumors, malignancies originating elsewhere may involve the diaphragm by direct extension. This may be seen with thoracic malignancies, such as bronchogenic carcinoma, mesothelioma, and tumors of the chest wall or mediastinum, as well as in association with tumors originating from abdominal sites, such as the esophagus, stomach, or liver.

Masses of the diaphragm—both benign and malignant—should be resected. Those which are malignant should be removed with a wide margin of normal tissue, regardless of the size of the resultant defect. Use of prostheses or muscle flaps may be necessary to deal with large tissue defects. However, if a significant defect is repaired and the resultant diaphragm seems likely to have impaired phrenic nerve function, then the repair should be taut enough to avoid paradoxical motion upon inspiration.

Diaphragmatic Flap

While uncommonly employed as such, the diaphragm may be used as a pedicled flap for tissue coverage when needed in the chest. Specific applications may include reinforcement of bronchial stumps, as well as coverage of bronchopleural fistulas, esophageal leaks, and esophageal perforations. Although there are other options available for vascularized pedicled flaps, the diaphragmatic flap is a useful technique to have in one's surgical armamentarium.

In order to harvest a diaphragmatic flap, one should approach the diaphragm from the thoracic operative field. After initially incising the diaphragm, a light source may be introduced to the abdomen. Transillumination is particularly useful in identifying the phrenic artery in order to maintain the vascular supply of the pedicled flap. The diaphragm is next divided, beginning posteriorly and continuing anteriorly. In order to optimize vascularity, the width of the flap should be about

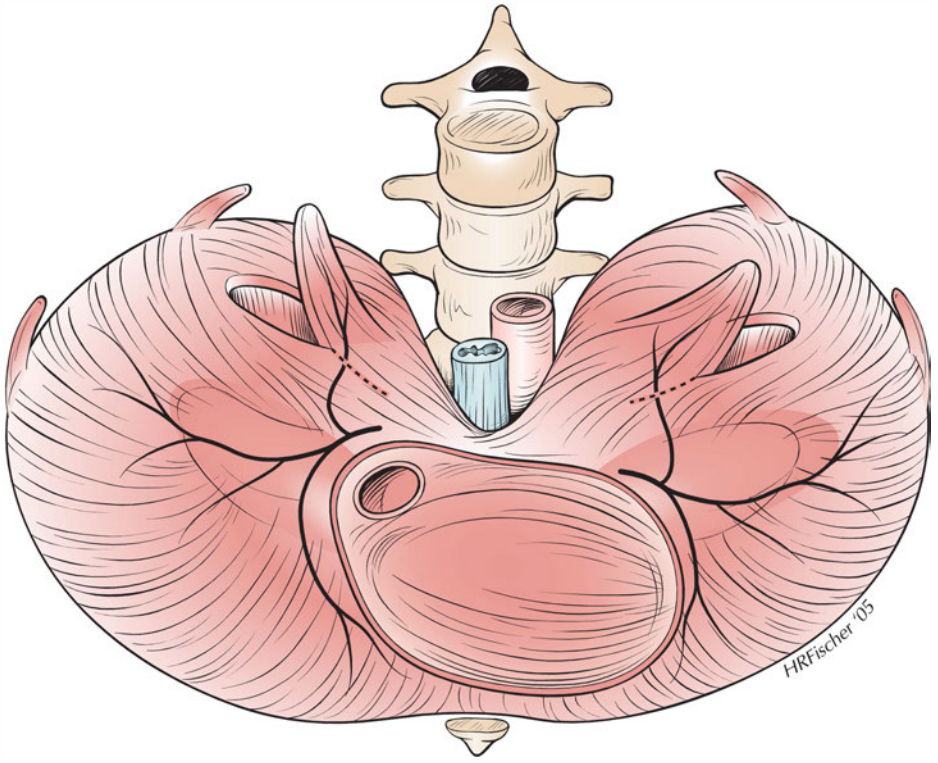


Fig. 26.11. Technique for harvesting a diaphragmatic flap.

one-fourth the length (Fig. 26.11). After bringing the flap to the desired location, and ensuring that adequate tissue has been harvested to complete the desired coverage without tension or torque on the flap, the defect in the diaphragm may be closed. This is typically performed with interrupted nonabsorbable sutures, bringing the newly created defect together primarily.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Thoracic surgeons do not spend a lot of time thinking about the diaphragm unless problems occur. Neoplasms are exceedingly rare yet the diaphragm certainly can be involved by malignancies from adjacent organs and partial resection may be required to completely resect a tumor from, for instance, the lower lobe of lung. In addition, for those patients with malignant pleural mesothelioma who undergo extrapleural pneumonectomy, resection of the diaphragm with patch reconstruction is a key component of the procedure.

Diaphragmatic paralysis from whatever cause usually results in significant dyspnea on exertion initially; yet, with time, the symptoms often improve to the extent that surgical correction is rarely required. Likely this is not due to restoration of function of the paralyzed hemidiaphragm but simply the result of a physiologic accommodation. In our experience eventration of the diaphragm rarely results in symptoms to such an extent that plication is necessary. Differentiating eventration from paralysis sometimes may be difficult but the distinguishing feature is significant paradoxical motion of the paralyzed diaphragm seen during the sniff test. In the patient with a paralyzed diaphragm plication will likely contribute little to symptom relief unless significant paradox is demonstrated.

Diaphragmatic pacing has been performed in a few specialized centers and the indications for the procedure are few, the main one being central hypoventilation syndrome. For diaphragm pacing to be useful the phrenic nerve must be intact and thus the procedure is not indicated for those with phrenic nerve injury. There has been some work done in reinnervating the diaphragm using intercostal nerve anastomosed to the phrenic nerve.

The porous diaphragm syndromes present unique challenges, each of which must be addressed on an individual basis. The exact cause of catamenial pneumothorax remains elusive since rarely is a specific lesion seen at the time of operation. Unless the monthly menstrual cycle is chemically interrupted, recurrence is highly likely with the onset of the first period following surgical repair and pleurodesis. Patients who present with ascites with persistent leakage into the chest also present difficult management problems as rarely is a specific communication found. Pleurodesis usually fails to resolve the problem and resolution usually only is achieved when the specific cause of the ascites is addressed.

The authors have done a superb job in summarizing all of the major issues related to the diaphragm that are of importance to the surgeon.

LRK

27 The Thoracic Duct and the Management of Chylothorax

Bradley M. Rodgers, Maxim Itkin, and John C. Kucharczuk

Chylothorax is the abnormal accumulation of fluid from the lymphatic system within the pleural space. Chyle is not only composed mainly of the lymphatic drainage from the intestine but also includes lymph from the lungs, liver, abdominal wall, and the extremities. The amount of lymph originating from the extremities as a component of chyle is negligible under normal circumstances. Chylothorax may be either congenital or acquired (Table 27.1). Congenital chylothorax is thought to occur either secondary to disruption of the thoracic duct during delivery or to congenital anatomic abnormalities of the duct, such as atresias. Acquired chylothorax can arise from multiple causes. Disruption of the thoracic duct may occur due to blunt thoracic or cervical trauma. Occasionally, the duct is injured in penetrating thoracic trauma, although the posterior location of the duct makes this form of injury quite uncommon. The thoracic duct may be lacerated during placement of left subclavian venous catheter, however, far more commonly is injury to the duct or its tributaries, occurring during thoracic operations. Chylothorax has been estimated to occur after 0.25% to 0.50% of all intrathoracic operations and has been described after nearly every type of thoracic surgical procedure but appears to be more common following cardiac procedures requiring considerable mediastinal dissection at the base of the heart or after esophagectomy.

Mediastinal neoplasms are responsible for the majority of chylothoraces that develop spontaneously in adult patients. Most of these are secondary to obstruction of lymphatic pathways from mediastinal lymphomas. Infections are an uncommon cause of chylothorax in the United States, but tuberculous lymphadenitis is a more common cause in many other countries. Miscellaneous causes for chylothorax include thrombosis of the subclavian vein or superior vena cava (SVC), usually secondary to long-dwelling intravenous catheters, child abuse, and pulmonary lymphangiomatosis (Table 27.1).

ANATOMY

The thoracic duct is the largest lymphatic channel in the body. It conveys the majority of the lymph within the body into the circulatory system. The duct arises embryologically as paired channels with numerous crossing anastomoses. In most instances, the paired structures fuse to form a singular vascular channel with the right duct persisting in the lower thorax and the left persisting in the upper chest (Fig. 27.1). Although up to 50% of individuals may have anomalous patterns of thoracic duct anatomy, in its most classic anatomic position the duct arises from the cisterna chyli anterior to the first or second lumbar vertebral body. The duct runs cephalad from the cisterna along the right side of the aorta and enters the chest through the aortic hiatus. The duct ascends in the right thorax, medial to the azygos vein and posterior to the esophagus. At the level of the fourth or fifth thoracic vertebra, the duct crosses anterior to the vertebral body and behind the esophagus to become a left-sided structure, dorsal to the aortic arch. The duct passes through the thoracic inlet posterior and to the left of the esophagus and forms an arch that rises 3 to 4 cm above the clavicle to the level of the sixth or seventh cervical vertebrae. It crosses anterior to the subclavian artery and the thyrocervical trunk as it extends laterally and terminates by opening into the angle of the junction of the left subclavian and the internal jugular veins (Fig. 27.2). A bicuspid valve at the lymphaticovenous junction prevents the reflux of blood into the duct at this junction. The duct is 3 to 5 mm in diameter at its origin in an adult, but its caliber diminishes in the mid-thorax to dilate again just proximal to its venous termination. The duct receives numerous lymphatic tributaries from the thoracic wall as it ascends through the chest, and there are multiple lymphaticovenous anastomoses between the duct and the azygos and intercostal veins.

Riquet described two major thoracic duct tributaries from the heart. The right efferent trunk primarily drains lymph from the right ventricle and ascends between the aorta and pulmonary artery, connecting to the thoracic duct high in the left chest. The left efferent trunk drains lymph primarily from the left ventricle and ascends behind the pulmonary artery, usually connecting with the azygos vein in the right chest. Injury to the right efferent trunk may account for many cases of chylothorax or chylopericardium after cardiac surgery.

Although this represents the most common anatomic configuration of the duct, there are numerous variations that may be of considerable surgical significance. The most common variation in anatomy is a duplicate duct, occasionally at the lower thoracic, but more commonly at the cervical level. The level at which the duct crosses the vertebral column may also be variable.

PHYSIOLOGY

There are multiple valves throughout the length of the thoracic duct, particularly in the cephalic end, that ensure unidirectional flow. The wall of the thoracic duct contains smooth muscle cells with an intrinsic contraction interval of 10 to 15 seconds. Movement of chyle through the thoracic duct is modulated primarily by the intrinsic contraction of the duct wall and the pressure gradient between the abdomen and thorax. The rate of lymph formation from the intestine and liver also affects the rate of flow through the duct. Flow through the thoracic duct varies between 0.38 and 3.9 ml/min.

The function of the thoracic duct is to transport ingested fat and lymphatic fluid from the abdominal viscera and lower body into the venous circulation. Approximately 60% to 70% of all ingested fat is absorbed by the intestinal lymphatics and transported by the thoracic duct. Fatty acids containing <10 carbon atoms are absorbed directly into the portal venous system, whereas

Table 27.1 Etiology of Chylothorax

Congenital
Traumatic
Ductal anomalies
Acquired
Traumatic
Surgical
Blunt trauma
Penetrating trauma
Child abuse
Nontraumatic
Malignant
Inflammatory
Idiopathic

larger fats are formed into chylomicrons and transported into the lymphatics. The thoracic duct is also the main pathway for return of extravascular plasma proteins and lymphocytes to the vascular space. Prolonged loss of thoracic duct lymph can lead to fat and protein malnutrition as well as to immunocompromise secondary to loss of T lymphocytes.

DIAGNOSIS

The diagnosis of chylothorax is suspected with the development of a pleural effusion in certain high-risk clinical settings, such as after esophagectomy. The diagnosis is confirmed by examination of the pleural fluid obtained by thoracentesis. In an individual consuming a normal diet, the diagnosis is usually quite evident, with a milky appearance to the fluid. Often, however, the patient has not received normal fats enterally before the development of the chylothorax, and the fluid in this clinical setting has the appearance of serum. Chemical analysis of the fluid reveals elevated triglyceride and total protein levels (Table 27.2). Cell counts reveal a marked predominance of lymphocytes, with numbers ranging from 400 to 7000/ml. Chronic pleural effusions secondary to tumors or infections may occasionally appear milky because of the accumulation of cholesterol in the fluid. This so-called pseudochylothorax can be differentiated from true chylothorax by the determination of the triglyceride level in the effusion. Most chylous effusions have a cholesterol/triglyceride ratio of <1, whereas nonchylous effusions have a ratio of >1. Pleural fluid with a triglyceride level of >110 mg/dl has a 99% chance of being chyle. If the triglyceride level is <50 mg/dl, the probability of a chylous effusion is only 5%.

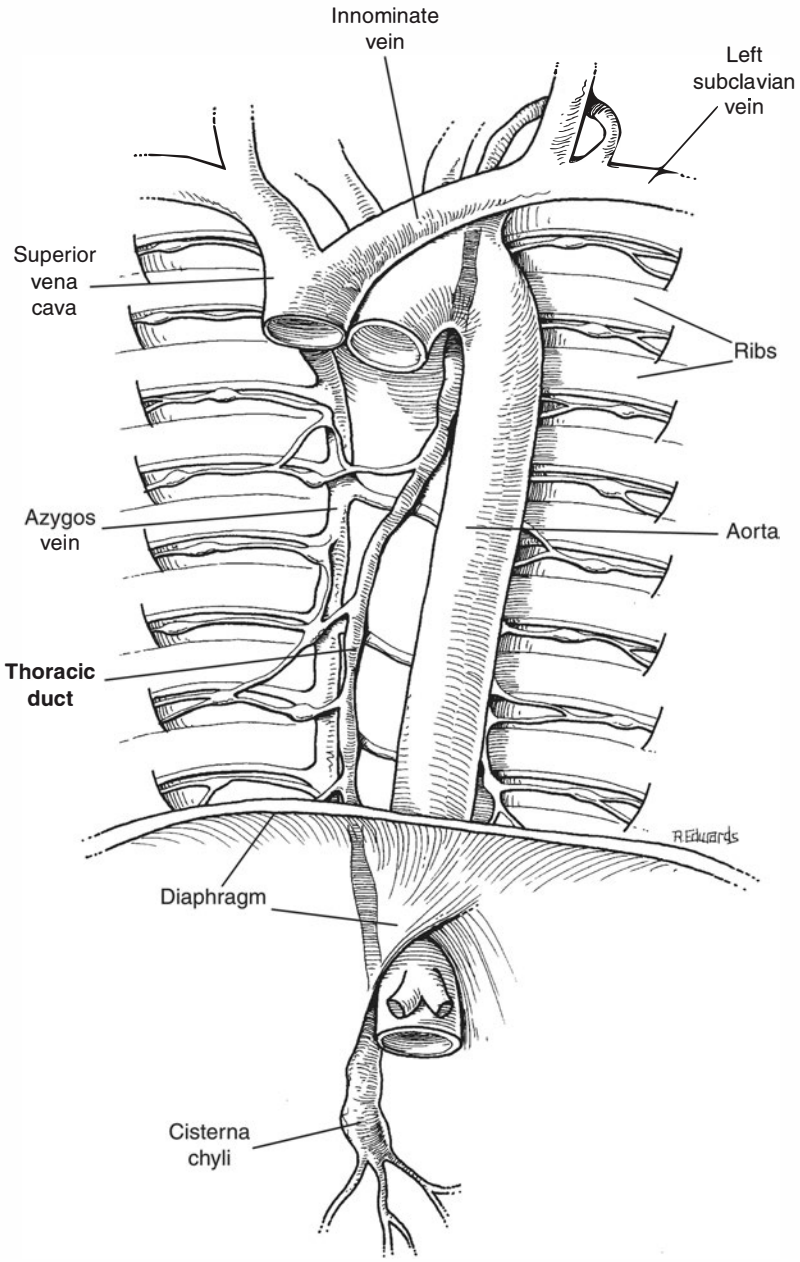


Fig. 27.1. The most common anatomic pattern of the thoracic duct. The duct enters the chest as a right-sided structure and crosses to the left chest at the fourth thoracic vertebra.

The thoracic duct may be visualized by standard lymphangiograms or by nuclear scintigraphy. Often these studies will demonstrate the anatomy of the duct and the level of the lymphatic leakage. They are rarely helpful, however, in the management of these patients and should not be considered a routine part of their evaluation.

CLINICAL PRESENTATION

Chylothorax primarily causes symptoms of respiratory insufficiency as the volume of the effusion gradually increases. Most cases of chylothorax are slow in their progression, and symptoms do not occur for several days to several weeks after the initial injury to the duct. Rapid accumulation of chyle with the production of severe respiratory symptoms is uncommon but is occasionally encountered after complete transection of the duct by traumatic or surgical injury. Chyle itself is bacteriostatic, probably because of its high fatty acid content, and symptoms of pleural infection are uncommon. Patients with longstanding chylothorax with loss of significant volumes of chyle often become hypoproteinemic. These patients also often develop lymphopenia from the loss of T lymphocytes in the chyle and may

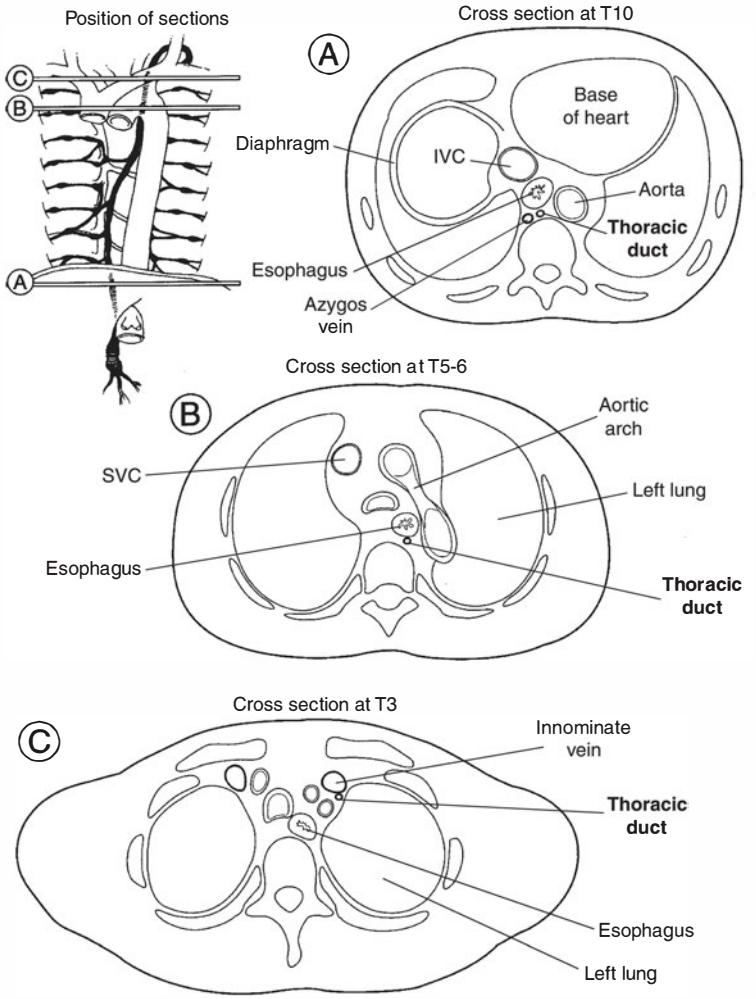


Fig. 27.2. The anatomic relationships between the thoracic duct and the mediastinal structures at various levels within the chest. (**A**) shows the most common location of the thoracic duct as it enters the chest at the T10 level, (**B**) shows the most common location of the thoracic duct in the mid chest at the T5-6 level, and (**C**) shows the most common location of the thoracic duct at the T3 level as it exist the chest and enters the neck. IVC, inferior vena cava; SVC, superior vena cava.

Table 27.2 Composition of Chyle

pH	7.4–7.8
Specific gravity	1.012–1.025
Lymphocytes	400–7000/dl
Culture	Sterile
Fat globules	Staining with Sudan red
Total protein	2.2–5.9 g/dl
Albumin	1.2–4.2 g/dl
Globulin	1.1–3.6 g/dl
Fibrinogen	16–24 g/dl
Total fat	0.4–6.0 g/dl
Triglycerides	>plasma
Cholesterol	65–220 mg/dl
Electrolytes	=plasma
Glucose	48–200 mg/dl
Cholesterol/triglyceride ratio	<1

become relatively immunocompromised. In fact, malnutrition and infection account for the majority of deaths after the development of a chylothorax.

MANAGEMENT

The control of chylothorax is thought to occur with the formation of pleural adhesions in the region of the chylous leak, thus preventing the flow of lymph from the thoracic duct or its branches. The management of patients with confirmed chylothorax, therefore, begins with attempts to completely drain the lymphatic fluid from the chest. In some cases, this may be accomplished by thoracentesis, whereas in most patients, with more rapid fluid accumulation, a tube thoracostomy is required. Measures are then instituted to reduce total thoracic duct lymphatic flow, initially using an oral diet containing fats primarily in the form of medium-chain triglycerides, which are absorbed directly into the portal venous system. Patients in whom significant lymphatic flow persists may require total elimination of enteral nutrition, with the institution of intravenous alimentation.

Several clinical reports have suggested that the use of somatostatin or its longer acting synthetic analog octreotide may stop persistent chylous accumulation. The effect of somatostatin on thoracic duct flow is probably secondary to its reduction of splanchnic blood flow and its reduction of intestinal fat absorption, reducing chylomicron synthesis. Somatostatin has generally been administered as an intravenous infusion, using 250 μg/h in adults and 3.5 to 10 μg/kg/h in children. The dose may be increased in stepwise manner to maximal response. Octreotide has generally been administered subcutaneously at doses of 100 μg twice or thrice daily for adults and 10 to 40 μg/kg per day in children (Table 27.3). Children and diabetic adults should be monitored for hyperglycemia or hypoglycemia, and adults are occasionally noted to have cardiac arrhythmias. The safety profile of these drugs, however, appears excellent, and they should be used early in patients with persistent chylous effusions to attempt to avoid protein and fat loss.

The length of time that it is reasonable to persist with conservative therapy is somewhat controversial and depends on the cause of the chylothorax and the volume of lymphatic loss. Some authors suggest intervention in cases of traumatic chylothorax when the daily loss of chyle exceeds 1,500 ml in adults or 100 ml per year of age

Table 27.3 Somatostatin/Octreotide Dose

Somatostatin	
Adult	250 μg/h IV
Child	3.5–7.0 μg/kg/h IV
Octreotide	
Adult	100 μg twice to thrice daily SC
Child	10–40 μg/kg/day SC

IV, intravenous; SC, subcutaneous.

in children for a 5-day period, when the output of chyle has not diminished over a 14-day period, or when nutritional complications appear imminent.

The mortality for patients with chylothorax was approximately 50% until Lampson described mediastinal ligation of the thoracic duct in 1948. This procedure remains the most commonly used operation for persistent chylothorax, although several alternative procedures have been developed in recent years. In patients with unilateral chylous effusions, the chest should be opened on the ipsilateral side, whereas in cases of bilateral effusion, the right chest should be chosen initially. Many authors recommend instillation of 100 to 200 ml of olive oil or cream into the stomach several hours before the operation to increase the fat content of the thoracic lymph and make the area of leakage from the duct itself more easily identifiable. After the induction of general anesthesia with orotracheal intubation, the patient is placed in a full lateral decubitus position. A posterolateral thoracotomy is performed, and the chest is entered through the seventh or eighth intercostal space. If the chylothorax has developed after a previous thoracotomy, the original incision is opened. The mediastinal tissues are examined carefully for the evidence of chylous leak. If such an area is identified, this region should be obliterated with nonabsorbable sutures, occasionally using polytetrafluoroethylene (Teflon) pledgets to compress larger areas of tissue. Whether or not a specific area of leakage is identified and controlled, the main thoracic duct should be ligated as it enters the chest through the aortic hiatus. To accomplish this, the esophagus is encircled and retracted anteriorly, and the tissues between the azygos vein and the descending aorta just cephalad to the aortic hiatus are ligated with nonabsorbable sutures (Fig. 27.3). In most cases, the actual thoracic duct may be identified in this area and ligation performed, whereas in other cases mass ligature of the tissues in this area is accomplished, without direct identification of the duct itself. The chest is closed in layers with a single large chest tube connected to a water seal. A retropleural approach to the thoracic duct has been described. The patient is placed under general anesthesia and positioned prone, and a segment of the right posterior eighth rib is resected after the periosteum is stripped. The posterior mediastinal pleura is dissected bluntly from the chest wall in this region, and the thoracic duct is identified medial to the azygos vein. The duct is obliterated with nonabsorbable suture material, and the incision is closed in layers without drainage.

Patients in whom the source of the chylothorax is suspected to be relatively localized, such as patients developing a chylous effusion after blunt or penetrating thoracic trauma, may benefit by thoracoscopic control of the lymphatic leak. Although the procedure may be performed under local anesthesia in these patients, the use of general anesthesia with unilateral ventilation greatly enhances the exposure of the mediastinum. The patient is placed in a full lateral decubitus position or rolled

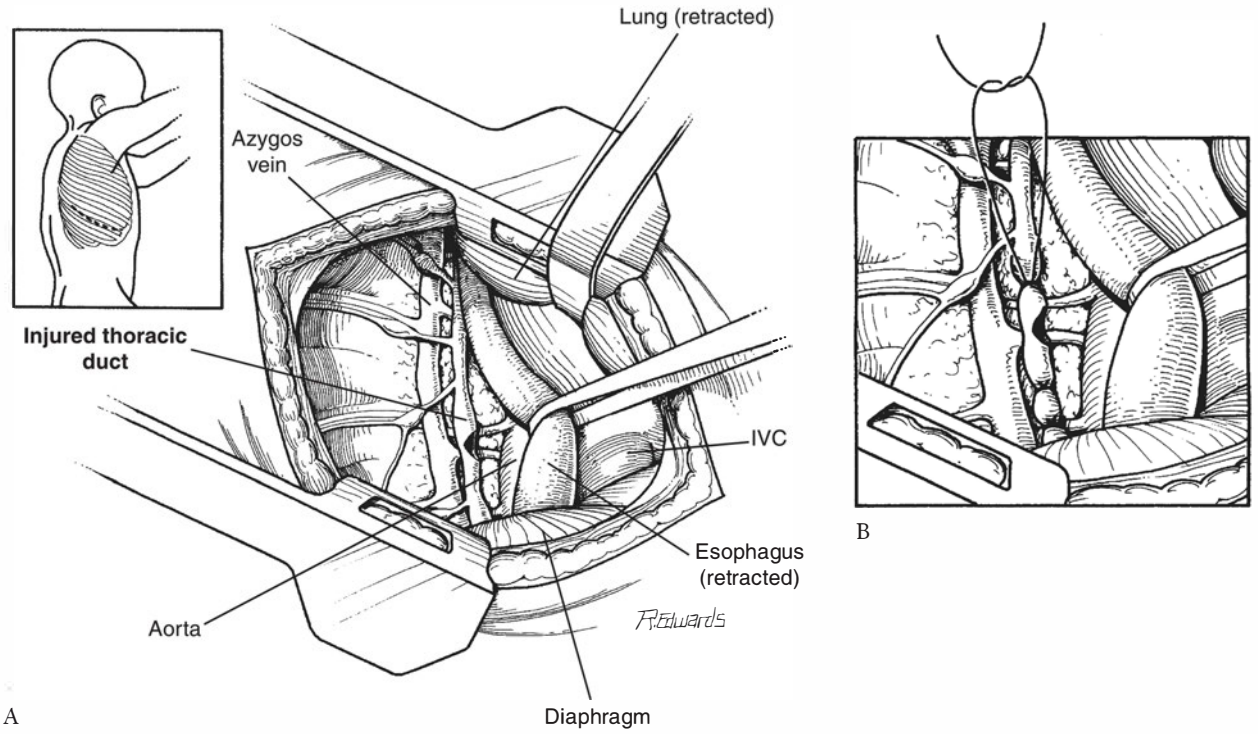


Fig. 27.3. Standard open management of chylothorax via thoracotomy. (**A**) The approach to the thoracic duct through a right posterolateral thoracotomy incision. The duct is exposed by anterior retraction of the esophagus. (**B**) The duct is ligated with nonabsorbable suture material as it enters the chest. (**Inset**) Thoracotomy incision (over the seventh or eighth intercostal space). IVC, inferior vena cava.

slightly forward to facilitate exposure of the posterior mediastinum. The initial trocar for the thoracoscope is placed in the sixth intercostal space in the mid-axillary line. A second trocar is placed posteriorly in the same or adjacent interspace (Fig. 27.4). The residual pleural lymph is aspirated, and the pleura overlying the posterior mediastinum is carefully examined from the level of the inferior pulmonary ligament to the innominate vein. In some cases, a defect in the parietal pleura will be discovered with lymphatic leakage through this region. We have preferred to use metallic clips on this tissue to obliterate the leak, although others have used direct suture ligation (Fig. 27.5). After the pleural leak is controlled in this region, the entire area is flooded with fibrin glue to form a seal over the parietal pleura. In most cases, the inferior pulmonary ligament is then divided with the electrocautery, and the thoracic duct is clipped or ligated at the level of the aortic hiatus. A single chest tube is placed through one of the trocar tracks while the other trocar sites are closed with subcuticular sutures.

If the origin of the chylous effusion is suspected to be more diffuse, such as in those patients who have undergone thoracic surgery or in patients with lymphangiomatosis or SVC thrombosis, the placement of a pleuroperitoneal shunt may be the best treatment strategy. In choosing to use a pleuroperitoneal shunt, the surgeon must decide between an internalized system, which is completely buried under the skin, and an exteriorized system, which leaves the pumping chamber exposed. The internalized system has the advantage of not requiring any maintenance of the catheter entrance or exit sites, but it does require a responsible patient or parent who is willing to compress the chamber as frequently as necessary in spite of some potential discomfort. The exteriorized system has proven quite helpful in small infants who may not tolerate chest wall compression well and has advantages in obese patients in whom the internalized chamber may be difficult to localize. This system facilitates the control of high-volume chylous leaks that require frequent pump compression. The disadvantage of the exteriorized system is the need for cleaning and dressing the catheter exit sites and the potential for infection through these sites.

Both the internalized and externalized systems may be placed under local anesthesia, although the use of general anesthesia facilitates the procedure. It is helpful in placing either of these systems

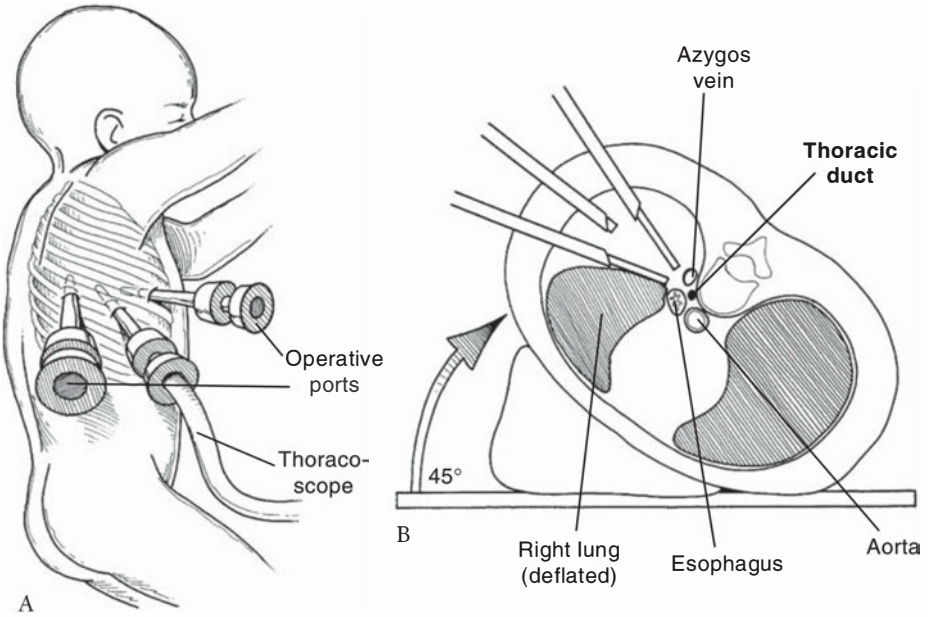


Fig. 27.4. The thoracoscopic approach to the thoracic duct and placement of thoracoscopic instruments. (**A**) The patient is placed in an exaggerated lateral decubitus position to expose the posterior mediastinal structures. (**B**) The thoracic duct is identified behind the esophagus between the aorta and the azygos vein.

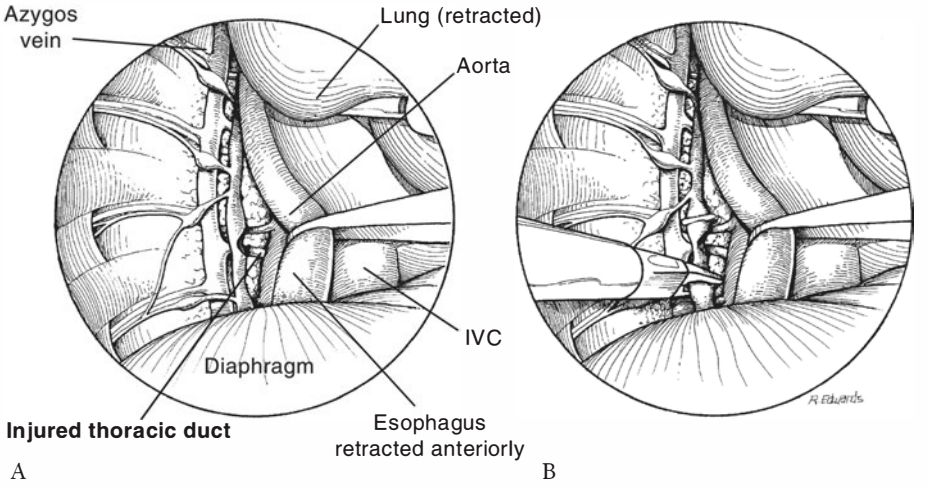


Fig. 27.5. Thoracoscopic management of chylothorax. The thoracic duct is exposed behind the esophagus (**A**) and occluded with metallic clips just above the level of the diaphragm (**B**). IVC, inferior vena cava.

to have some residual chyle within the chest at the time of the operation. This allows the surgeon to test the patency of the shunt system. If a chest tube is already in place, it may be clamped the night before the procedure to allow accumulation. The patient is placed in the supine position with the affected chest elevated 30 to 45 degrees with a wedge. The entire chest and abdomen of the affected side are prepared and draped. The pleuroperitoneal shunt system is placed on the body surface to determine the optimal position of the shunt and the incisions. It is helpful to mark the incisions with a skin pencil at this time. The pumping chamber for the internalized system should be placed in a subcutaneous pocket overlying the lower ribs of the anterolateral chest wall. The pleural catheter should be directed as far posteriorly as possible to allow optimal-dependent drainage. A 2-cm skin incision is made just caudad to the proposed intercostal entrance of the pleural catheter. A generous subcutaneous pocket is dissected caudad from

this area over the lower ribs. A 2-cm transverse incision is made overlying the anterior rectus sheath midway between the umbilicus and the xiphoid process (Fig. 27.6). The anterior rectus sheath is divided transversely, and the fibers of the rectus muscle are separated to expose the posterior rectus sheath and peritoneum. Two concentric purse-string sutures of 4-0 polypropylene (Prolene) are placed in the posterior rectus sheath. The pleuroperitoneal shunt system is filled with sterile saline by compressing the pumping chamber while the entire system is under fluid. Careful attention must be paid to orienting the pumping chamber in the proper direction because there are two unidirectional valves within the chamber. The pleural catheter is trimmed to appropriate length and placed across the intercostal space in tangential manner. This is the most critical portion of the operation, and care must be taken not to kink the catheter as it crosses the chest wall. A long clamp is then passed from the lower incision to the upper incision, through the subcutaneous pocket, and the distal catheter and pumping chamber are drawn into the pocket by traction (Fig. 27.7). The pumping chamber must be seated well within the pocket to ensure that neither of the incisions is directly over the compression chamber. Patency of the system is confirmed by repeatedly compressing the pumping chamber and observing for free flow of chyle through the distal tubing (Fig. 27.8). The peritoneal catheter is trimmed to appropriate length, and the posterior rectus fascia and peritoneum are opened within the purse-string sutures and the catheter passed into the abdominal cavity. Both purse strings are tied securely around the catheter. The patency of the system is checked once again by repeatedly compressing the pumping chamber to ensure free flow of fluid. The two incisions are closed with subcutaneous and subcuticular absorbable sutures.

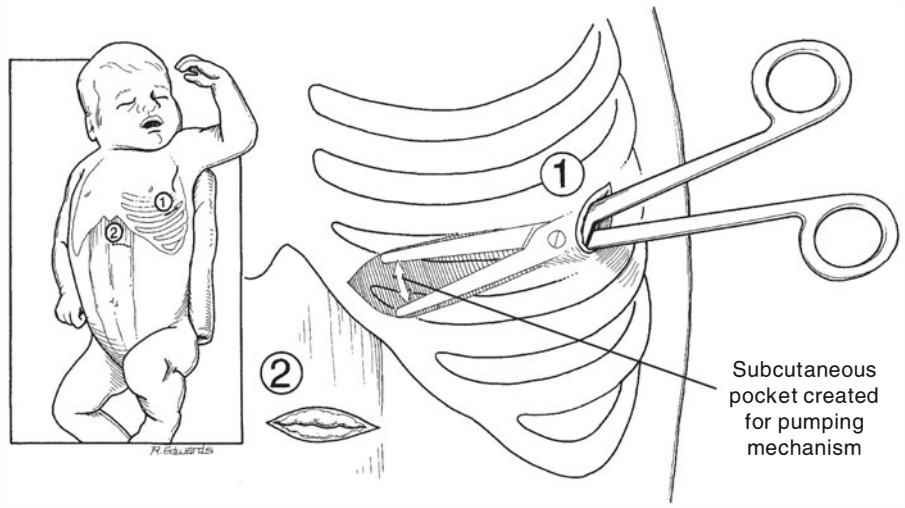


Fig. 27.6. Placement of an internalized pleuroperitoneal shunt. The patient's hemithorax is elevated slightly, and short incisions are placed over the eighth intercostal space in the posterior axillary line and the anterior rectus sheath. A generous subcutaneous pocket for the pumping chamber is developed from the thoracic incision. 1, incision for pleural end of shunt; 2, incision for peritoneal end of shunt.

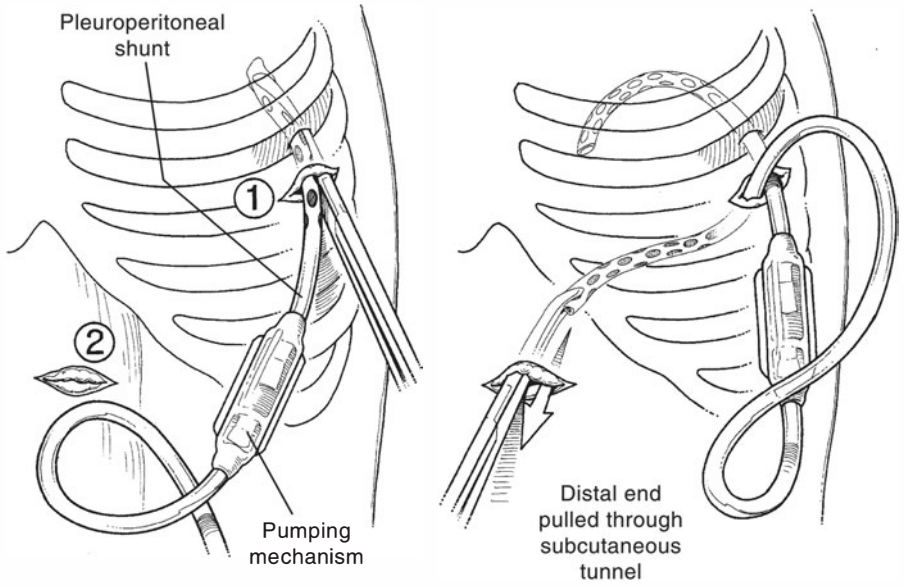


Fig. 27.7. Internal shunt system. The pleural catheter is passed tangentially across the intercostal muscle into the pleural space (1). The distal catheter and pumping chamber are advanced by a clamp inserted through the abdominal incision (2).

Planning for an exteriorized system starts as for the internalized system. The shunt is placed on the body surface to determine the best position for the incisions. Subcutaneous tunnels of approximately 6 to 10 cm in length are made for proximal and distal tubing. A 1-cm incision is made overlying the interspace through which the thoracic catheter will be placed, and a 2-cm incision is made overlying the anterior rectus sheath just above the level of the umbilicus. The thoracic catheter is drawn through the subcutaneous tunnel by passing a clamp in a caudad direction from the interspace incision. The Teflon cuff surrounding the catheter is placed at the level of the thoracic insertion (Fig. 27.9). The abdominal catheter is drawn through the subcutaneous tunnel by passing a clamp cranially from the rectus incision, and the cuff is positioned to lie at the level of the peritoneum. Two concentric purse-string sutures are placed through the posterior rectus fascia and peritoneum, as for the insertion of the internalized system, and the peritoneal catheter is inserted (Fig. 27.10). The two purse-string sutures are tied, and each is sutured to the Teflon cuff of the abdominal catheter. The thoracic cuff is not sutured to the thoracic fascia. The incisions are closed in layers after the system is tested by compressing the exteriorized pumping chamber (Fig. 27.11). The catheters are sutured to the skin at the exit sites, and sterile dressings are applied.

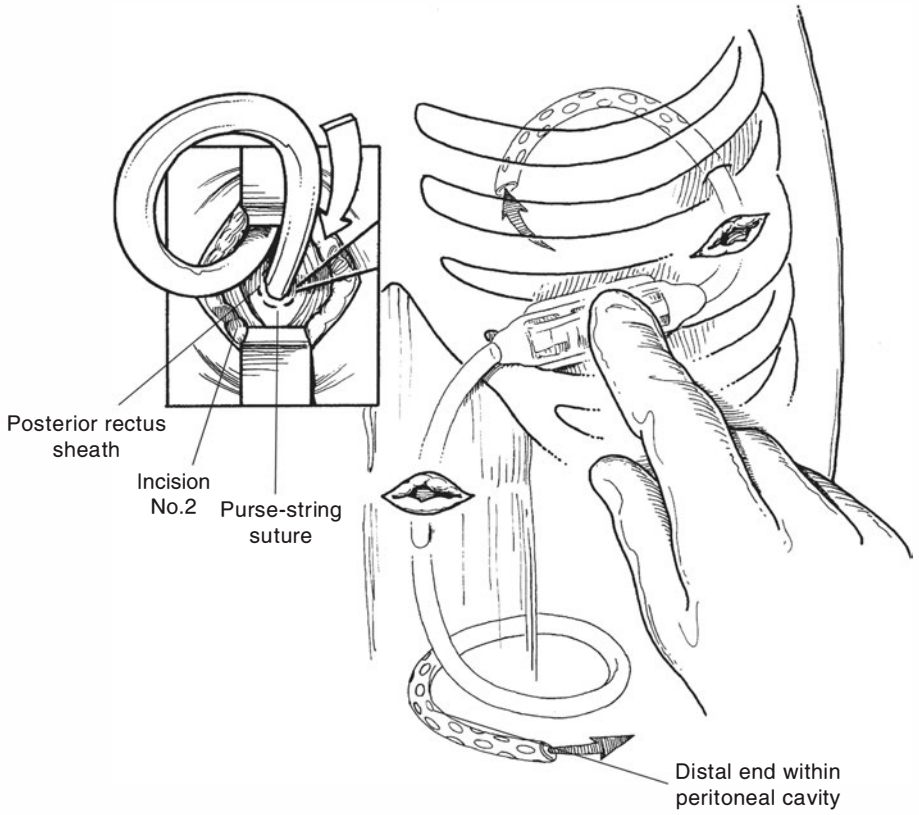


Fig. 27.8. Internal shunt system in position. The peritoneal catheter is placed through two concentric purse-string sutures in the posterior rectus sheath and peritoneum. Patency of the system is confirmed by repeated compression of the pumping chamber. (*Dark arrows* indicate direction of flow.)

Postoperatively, a schedule of pump compression is established that will allow complete evacuation of chyle from the thorax. An estimate of the frequency of compression necessary can be made by identifying the preoperative volume of lymphatic leakage and estimating that each compression of the chamber pumps approximately 2 ml of chyle from the chest to the abdomen. Usually, the patients are asked to compress the chamber for a certain number of times, 4 times a day. With the exteriorized chamber, the patient can easily tell when the chest is empty because the chamber will not fill with chyle after compression.

When the flow of lymph through the pleuroperitoneal shunt has diminished to a minimum, the patient is instructed not to pump the chamber for an interval of 2 weeks, after which a chest radiograph is obtained to be certain that there is no reaccumulation of fluid. If the chest is dry, the shunt is removed by simply withdrawing the pleural catheter. The abdominal catheter is dissected out, and the entrance through the posterior rectus fascia is closed with interrupted absorbable sutures to prevent visceral herniation.

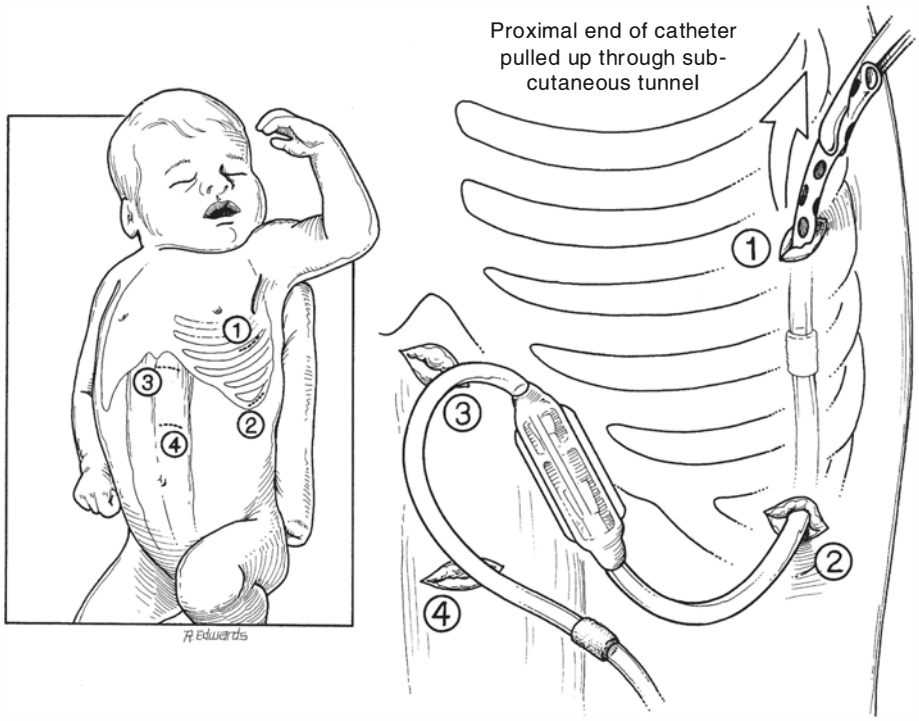


Fig. 27.9. Placement of an externalized pleuroperitoneal shunt. Long subcutaneous tunnels are developed for the pleural and peritoneal catheters. (Numbers indicate incision sites.)

THORACIC DUCT EMBOLIZATION

The rationale of thoracic duct embolization (TDE) was initially conceptualized and tested by Dr. Constantine Cope from the University of Pennsylvania based on the underlying assumption that the ability to visualize the lymphatic system and TD abnormality using lymphangiogram could increase the treatment success and a minimally invasive percutaneous approach could decrease morbidity and mortality.

Over the years, pedal lymphangiogram has proved to be the major obstacle for wider acceptance of TDE. Traditional pedal lymphangiography is both time-consuming and technically challenging, and requires specialized equipment that is not readily available. The recent development of the intranodal lymphangiogram opened the door to more widespread use of TDE from specialized academic centers to the community hospitals. Based on a simple ultrasound-guided puncture of the groin lymph nodes, this procedure is within basic skills of any interventionalist practicing ultrasound-guided procedures. Intranodal lymphangiography has been described as a technically less challenging alternative to conventional pedal lymphangiography.

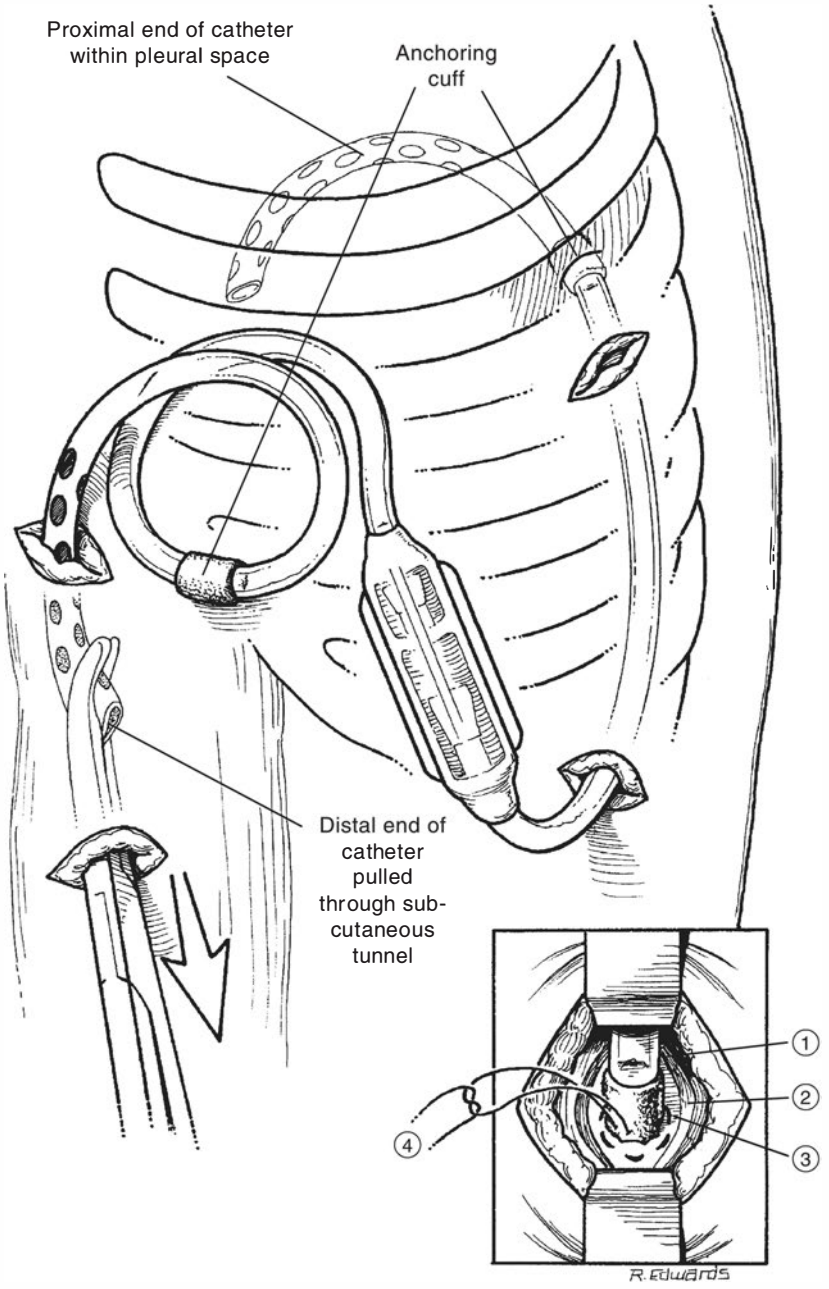


Fig. 27.10. The catheters are placed with the polyethylene terephthalate (**Dacron**) cuffs at the level of the intercostal muscles and the posterior rectus sheath (**inset**). The anterior rectus sheath is opened (1). The rectus muscle is split in the direction of its fibers (2). The posterior rectus sheath (3) and periotoenum are opened. The catheter is inserted and secured with two purse string sutures (4) one through the periotoneal and Dacron cuff, the other through the posterior rectus sheatlh and the Dacron cuff with care to avoid damage to the shunt.

INTRANODAL LYMPHANGIOGRAPHY

In this technique, an inguinal lymph node is directly accessed with a 25-gauge spinal needle (BD Medical, Franklin Lakes, NJ) under ultrasound guidance with the needle tip positioned in the hilum of the node (Fig. 27.12). Subsequently, an oil-based contrast agent (Ethiodol; Savage Laboratories, Melville, NY) is injected by hand at a rate of about 1 to 2 ml per 5 minutes. If successful, immediate opacification of the lymphatic vessels is observed under fluoroscopy (Fig. 27.13). A total volume of approximately 6 ml of contrast is injected into a lymph node in each groin to opacify the abdominal and pelvic lymphatics in an adult. To further propel the advance of the contrast in the lymphatic system, the injection of lipiodol is followed by injection of saline ("saline flash"). The injection of saline into lymph nodes is painful for the patient and for that reason intravenous sedation is recommended.

The feasibility of using intranodal lymphangiography for TDE has been well demonstrated. Using intranodal lymphangiogram, the abdominopelvic lymphatics, cisterna chyli, and thoracic duct may be visualized and subsequently the thoracic duct successfully accessed and embolized. This technique also significantly reduces the embolization time, mainly due to more rapid catheterization of the lymphatic system and faster transit time between the initiation of the lipiodol injection and opacification of the TD. Overall, intranodal lymphangiography for TDE appears to be easier, safer, and faster than the older method of pedal lymphangiography.

THORACIC DUCT EMBOLIZATION DESCRIPTION OF THE TECHNIQUE

After the cysterna chyli and/or its tributaries are visualized following a intranodal lymphangiogram, the lymphatic system is accessed transabdominally using a 21- to 22-G needle. To prevent the leakage of chyle from cisternal chyli, the access to the lymphatic system is performed through the lymphatic feeders below the cysterna chyli (Fig. 27.14). Using this access, a stiff 0.018 inch wire (V-18, Boston Scientific, Natick, MA) is advanced into the TD, followed by a 3-F microcatheter (Rapidtransit, Cordis Hialeah, FL).

Contrast is then injected through the catheter in an attempt to demonstrate the origin of the chyle leak. In traumatic chylothorax, the cause of the leak is most often a tear of the thoracic duct or leakage from a TD branch (Fig. 27.15A and 27.15B). In idiopathic chylothorax, the cause of the chylothorax frequently is occlusion of the upper part of the thoracic duct (Fig. 27.16).

If the TD appears to be normal and there is an unobstructed flow of the contrast into the left subclavian vein, the duct should not be embolized. In this situation, the diagnosis of chylothorax has to be questioned. To confirm the diagnosis, a food challenge test using a bolus of cream has to be performed and the level of the triglycerides in the effusion is measured. If the diagnosis is confirmed, other causes of the chylothorax have to be suspected, such as chylous ascites or a lymphatic malformation.

After the cause of the leak is identified, embolization of the thoracic duct is performed below the origin of the leak (Fig. 27.17). If the leak is identified as coming from a branch of the TD, selective catheterization of the branch can be attempted. The embolization is performed

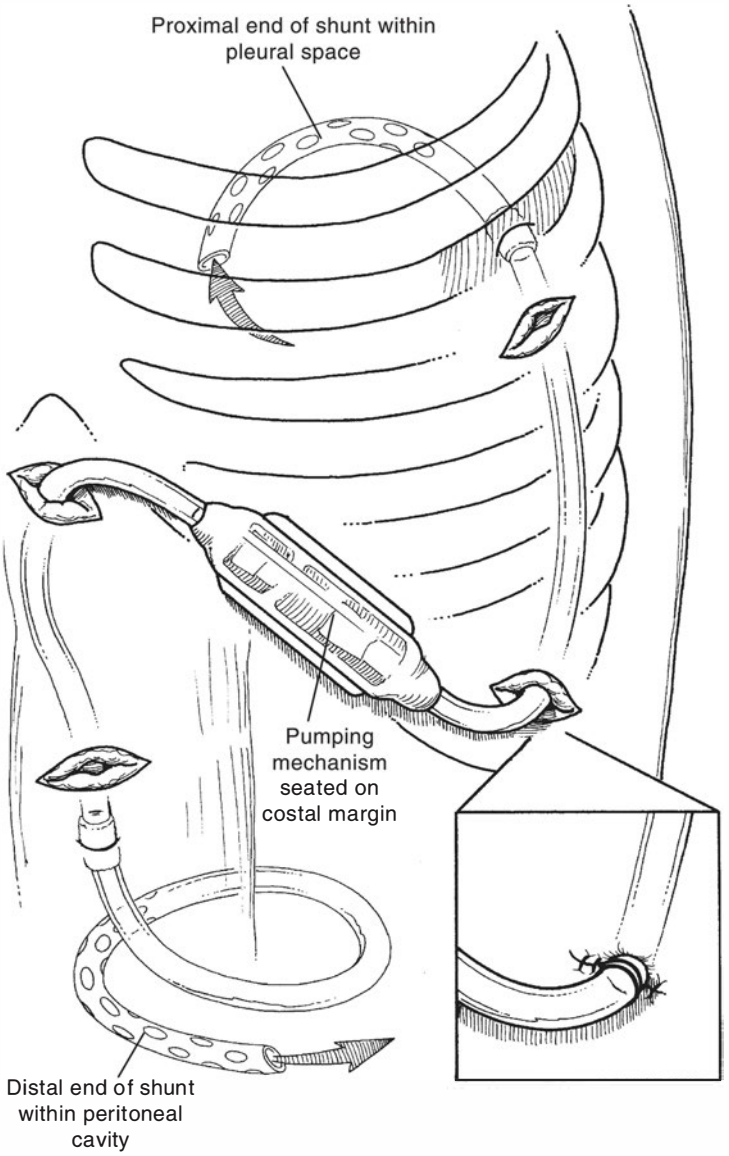


Fig. 27.11. Patency of the externalized system is confirmed by repeatedly compressing the pumping chamber. The catheters are secured at the skin exit site. (**Inset**) Securing suture wrapped around shunt tube.

using a combination of embolization coils and NBCA glue (Truefill, Cordis Hialeah, FL). If a leak is suspected from multiple small collaterals, then embolization with NBCA glue only can be performed.

In case TD catheterization is technically unsuccessful, TD needle disruption has been described. Using this technique, the retroperitoneal lymphatics are disrupted using a "twiddling motion with the needle." The idea is that traumatic disruption of the lymphatic vessels results in controlled bleeding into lymphatic vessels and the blood clots and/or local inflammation would close the leak.

Given the technical difficulty of the procedure, there remains interest in optimizing performance by improving guidance to aid in puncturing the thoracic duct. Recently, combining MRI with fluoroscopy to guide access to the thoracic duct has been described.

THORACIC DUCT EMBOLIZATION OF TRAUMATIC CHYLOTHORAX

Iatrogenic injury of the TD or its branches during thoracic, cardiac, or neck operation remains the most frequent cause of traumatic chylothorax. Traumatic chylous ascites also may present as chylothorax. The negative pressure developed in the chest during inspiration causes the chyle to migrate from the abdomen into the pleural space through pores in the diaphragm.

Chylothorax following a surgical procedure usually presents within the first 2 weeks following operation. The incidence of chylothorax complicating pulmonary resections can be as high as 4%, and recent report suggests that the incidence may be rising given the increased frequency of extensive resections and lymph node dissections performed by thoracic surgeons. Morbidity and mortality resulting from a chylothorax are significant. Chylothorax following esophagectomy is associated with a statistically significant increase in 30-day major complications (85% vs. 46%; $p < 0.001$) and death (17.7% vs. 3.9%; $p < 0.001$) when compared with patients with no chylothorax.

Overall, conservative therapy only resolves the chylothorax in <50% of cases with a reported mortality of 50% to 75%. Success of thoracic duct ligation in treating chylothorax following esophagectomy approaches 67%. Although these results are promising, the operative risk and morbidity, when compared with the safety and improved outcomes associated with TDE make it, at least in our hands, the preferred alternative to surgical ligation. The growing experience with TDE and the advent of intranodal lymphangiography supports its use as first-line treatment for traumatic chylothorax.

The largest series of TDE in treating traumatic chylous effusions detailed results in 109 patients. In 73 of 109 cases, the TD was catheterized, and TDE with endovascular coils and/or NBCA glue was performed in 71 of 73 patients. The leak resolved in 64 of 71 (90%) of these patients. In 18 of the 33 cases in which TD catheterization was unsuccessful, needle interruption of the thoracic duct below the diaphragm was attempted and resulted in resolution of the chylothorax in 72% of patients. Overall success based on intent to treat for the entire series was 71%. In 20 patients who had failed previous surgical ligation, embolization or interruption was attempted in 17 and was successful in 15 (88%) demonstrating the utility of TDE even after failed surgical intervention. The most common causes of operative TD ligation failure were "missed TD" (Fig. 27.18), incomplete ligation of the TD (Fig. 27.19), or leak from the "stump" of the ligated duct (Fig. 27.20). In some cases, persistent chylothorax was present in spite of lymphangiographic demonstration of complete ligation (Fig. 27.21). In these cases, injection of the cisterna chyle with NBCA glue completly alleviated the chylothorax. There were 3 (3%) minor complications observed in this study.

In the pediatric population, chylothorax is most commonly observed after cardiac surgery and has a reported incidence of 2% to 5%. TDE in the pediatric population is more challenging due to the small size of the lymphatic vessels. Generally, the technique of TDE in children is similar to adult with

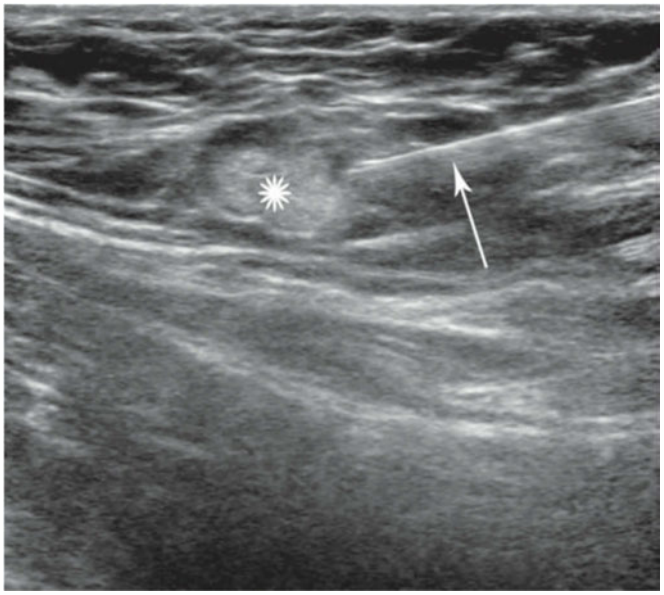


Fig. 27.12. Ultrasound image of the lymph node demonstrating the tip of the needle (*white arrow*) positioned in the lymph node hilum (*white star*).

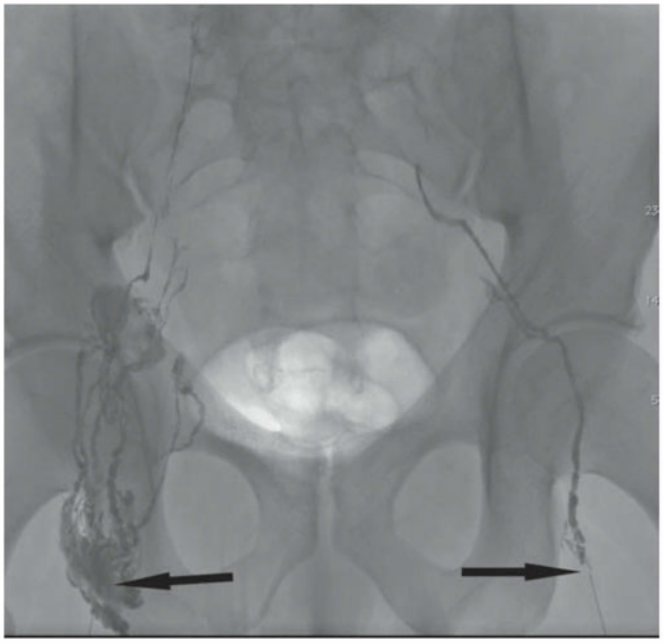


Fig. 27.13. Fluoroscopic image demonstrating the intranodal lymphangiogram. Tip of the needle is located in the lymph nodes (*black arrows*). On the right side, the lymphatic ducts of the right groin lymph nodes communicate directly with the mid-abdomen lymph nodes, bypassing the pelvic lymph nodes. In this quite common situation, the opacification of the cisterna chyli and TD happens within minutes.

one exception: in children with right-to-left shunt use of an oil-based contrast agent (lipiodol) has the potential to result in systemic fat embolism. Fortunately, it is possible to use water-based contrast in these patients. These initial results show promise for TDE to manage chylous effusions in the pediatric population. Developing significantly less technically challenging intranodal lymphangiogram should further facilitate the adoption of this technique.

THORACIC DUCT EMBOLIZATION FOR NONTRAUMATIC CHYLOTHORAX

Nontraumatic chylothorax is a relatively rare condition in which the leak develops without any precipitating traumatic event. The causes of the nontraumatic chylothorax include malignancy, congenital abnormalities, systemic diseases (e.g., SLE and Behcet disease), infection (e.g., tuberculosis), and an idiopathic group where seemingly no cause can be determined.

These cases present considerable diagnostic and therapeutic challenges when compared with those with a traumatic chylothorax. Generally, there are three types of underlying anatomical abnormalities that may result in chylothorax: (1) chylous ascites that presents as chylothorax; (2) occlusion of the upper part of the TD with subsequent development of the pleural collateralization and chyle leak; and (3) chylothorax associated with lymphatic malformation.

Recently, experience in 34 patients undergoing TDE for nontraumatic chylous effusions was reported. Thoracic duct catheterization and embolization were technically successful in 24 of 34 patients (70.6%). Overall, the clinical success rate evaluated based on "intention to treat" criteria was 53% (N = 18 of 34). In the group in which the TDE was technically successful (N = 24), the clinical success rate was 67.7% (N = 16). Interestingly, the clinical success rate was varied based on the lymphangiographic pattern. The greatest clinical success (88%) occurred in patients who were found to have an occlusion of the thoracic duct with multiple mediastinal collaterals and complete obstruction of passage of contrast from the thoracic duct into the subclavian vein (Fig. 27.16). The lowest success rate (16%) was observed in patients with thoracic duct on lymphangiography. It is possible that in these patients the sources are intestinal lymphatics, which communicated directly with the pleural space. This group does indeed presents significant challenges.

Over the years, an algorithm has been developed for the evaluation and treatment of these patients. If free fluid in the abdomen is identified on cross-sectional imaging, chylous ascites and passage of the chyle from the abdomen into the chest are suspected. In this case, sampling of the abdominal fluid and testing it for chyle are recommended. As mentioned earlier, the diaphragmatic openings that result in free communication of the fluid between abdomen and chest have been described. In these cases, the placement of the peritoneal-venous shunt is indicated as primary treatment. If chylous ascites is not identified, we perform heavily weighted T2 MR imaging of the chest and abdomen to define the anatomy of the thoracic duct to exclude lymphatic malformation and attempt to identify lymphatic malformations which could be culprit for chyle leak (Fig. 27.22).

To examine the patency of the thoracic duct, we perform an intranodal

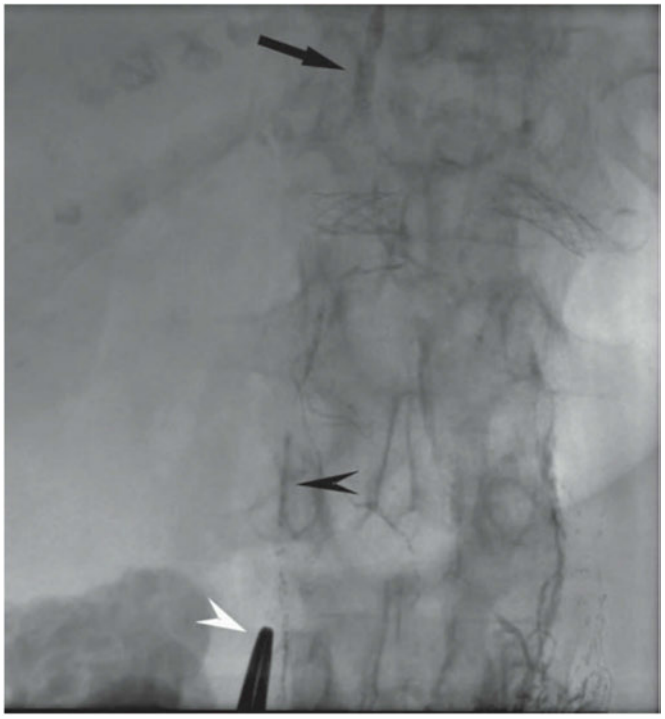


Fig. 27.14. Digital X-ray demonstrating the planning for the access of the lymphatic system. The tip of the hemostat (*white arrowhead*) marks the skin entry site through the lumbar lymphatic vessel (*black arrowhead*) that feed into cisterna chyli (*black arrow*).

lymphangiogram. In cases where the TD is occluded and multiple collaterals are identified (Fig. 27.16), TDE is performed. In those situations where the TD is patent and there is free flow of the contrast into subclavian vein, TDE should be avoided. If lymphatic malformation is identified on the MRI or lymphangiogram, sclerotherapy and/or direct injection of the lymphatic malformations with embolization material is performed.

LOW TERM OUTCOME OF THORACIC DUCT EMBOLIZATION

With the growing experience with TDE to treat traumatic and nontraumatic chylous effusions, late term complications of the procedure, although rare, are now being recognized. Occlusion of the TD and redistribution of the significant fluid flow (2 to 3 l/day) can potentially result in long-term sequelae such as lymphedema, chylous ascites, and protein-losing enteropathy. A recent case report described an incident of chylous ascites occurring in a patient with a chylothorax who had a failed attempt at

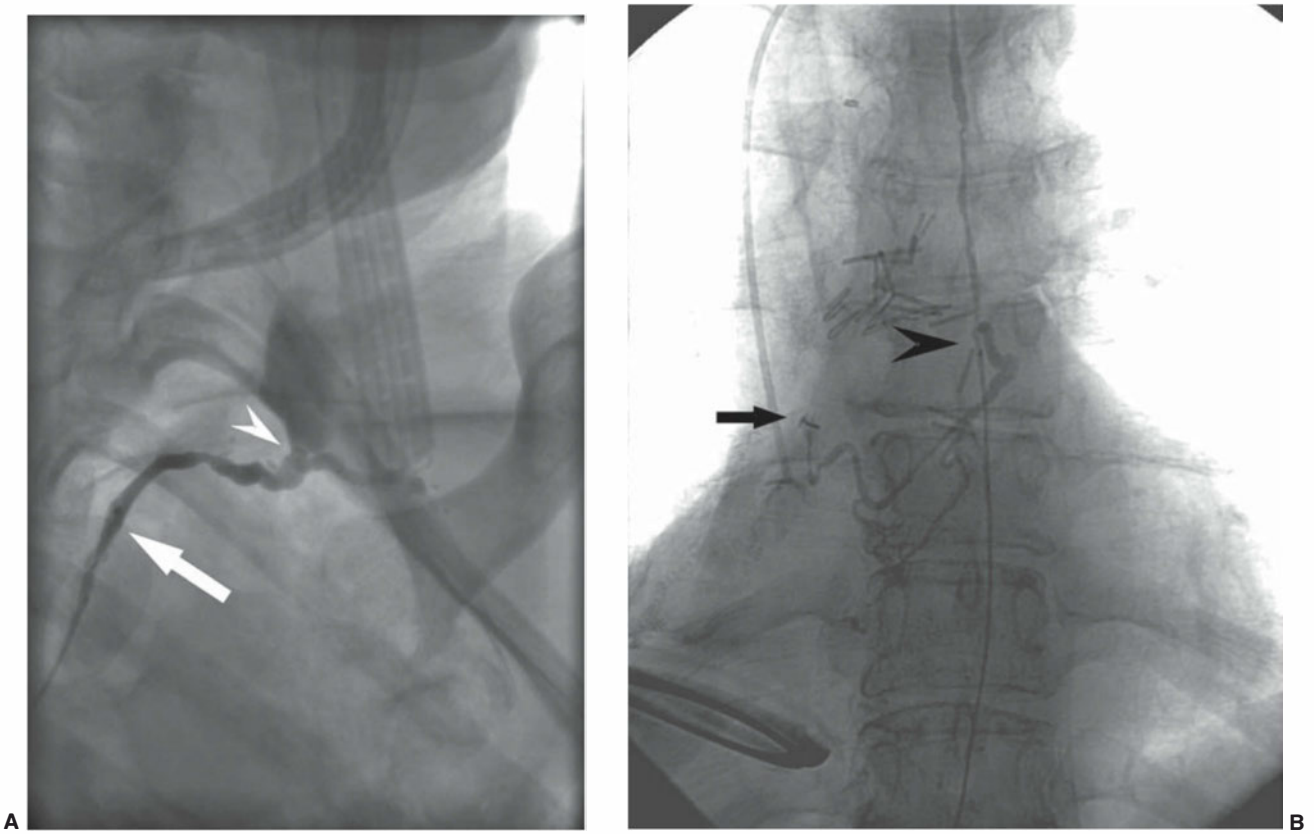


Fig. 27.15. (**A**) Injection with the contrast of the distal part of the TD through the microcatheter (*white arrow*) demonstrating the leak (*white arrowhead*) in patient with chylous leak after neck surgery. (**B**) Injection with contrast of the proximal part of TD through the catheter (*black arrowhead*) demonstrating the leak from the branch of the TD (*black arrow*). Surgical clip at the leakage point is noted. The surgeon saw the leak during the operation and unsuccessfully attempted to close it.

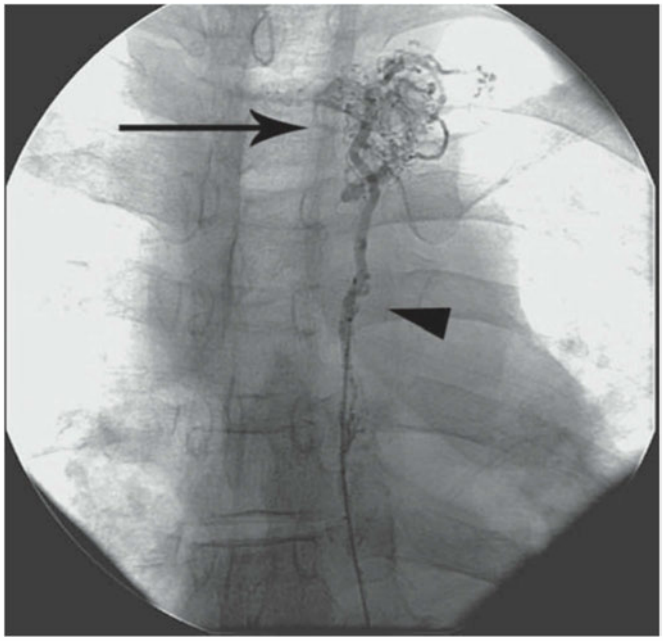


Fig. 27.16. Fluoroscopic image demonstrating complete occlusion of the distal part of the TD with the development of multiple collaterals (*black arrow*) through the catheter located in the middle part of the thoracic duct (*black arrowhead*).

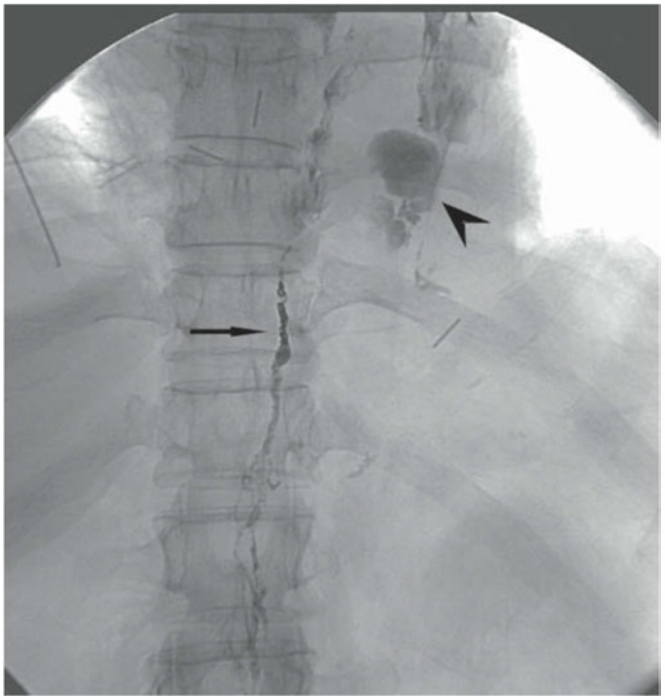


Fig. 27.17. Fluoroscopic image demonstrating embolization coils (*black arrow*) located at the distal part of the TD, below the chyle leak (*black arrowhead*).

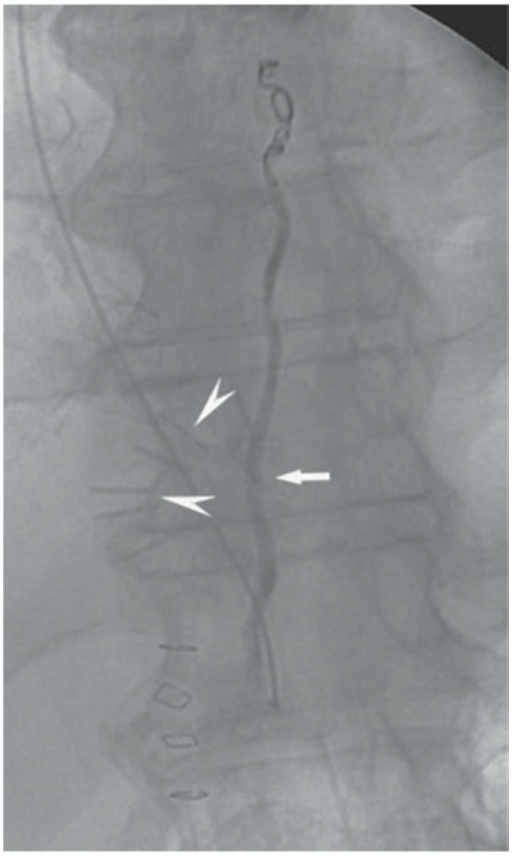


Fig. 27.18. Spot image of the glue cast in the TD (*white arrow*) post-embolization in patient with failed TD ligation. Surgical clips (*white arrowheads*) are located next to the TD.

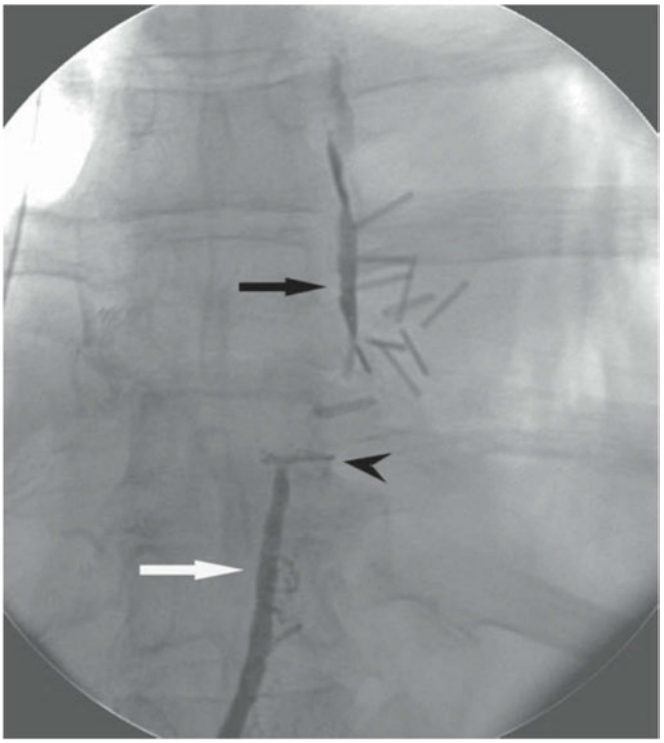


Fig. 27.19. Spot image of the distal part of the TD (*white arrow*) injection with contrast. Surgical clip (*black arrowhead*) partially occluded TD. The contrast passes through the occluded duct into distal part of the TD (*black arrow*).

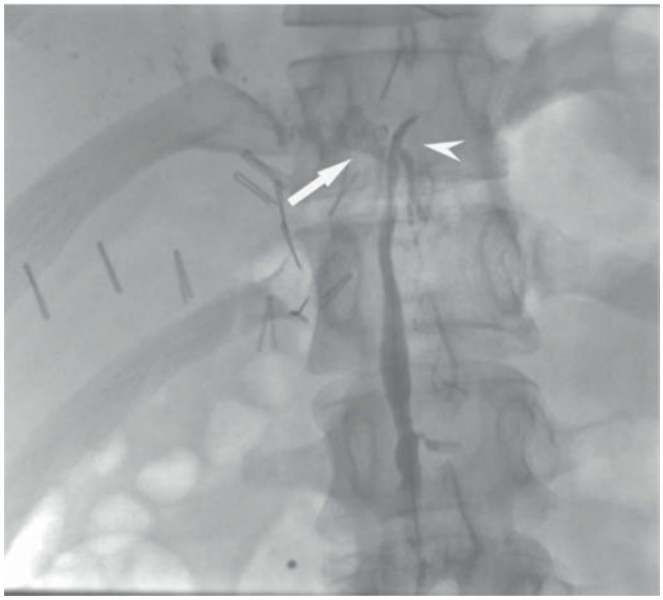


Fig. 27.20. Fluoroscopic image of the injection of the contrast into cysterna chyli through microcatheter with its tip located close to the "stump" of the TD (*white arrowhead*). There is an extravasation of the contrast out of the stump of the TD.



Fig. 27.22. Heavily weighted T2 MR image of the abdomen and chest. There is a high signal in the mid-abdomen corresponding to the multiple lymphatic malformations (*arrows*). Left pleural and pericardial effusions are noted.

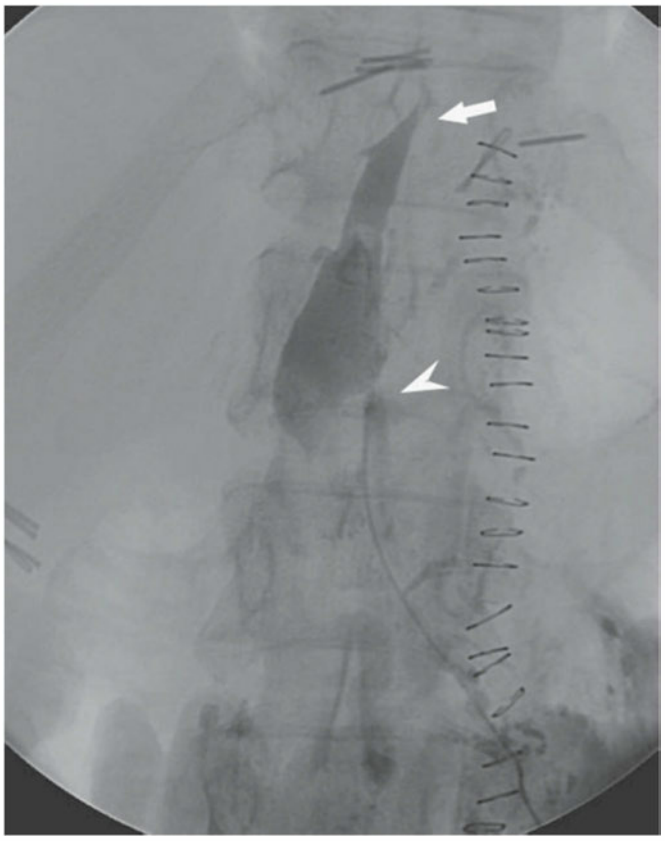


Fig. 27.21. Fluoroscopic image of the injection of the contrast into cysterna chyli through microcatheter (*white arrowhead*). There is a complete obstruction of the flow by successful TD ligation using surgical clips (*white arrow*). Injection of the NBCA glue into cisterna chyli stopped the leak.

cannulating the thoracic duct. This complication had not been observed in prior studies of TDE, which included cases of failed thoracic duct cannulation. In a long-term follow-up of patients who underwent TDE, chronic leg swelling and chronic diarrhea are complications that have been thought to be "probably related" to the procedure. These potential complications should be part of informed consent for the procedure.

The results of all of these invasive procedures for chylothorax are excellent. Sporadic case reports suggest that thoracic duct ligation is successful in controlling the chylous leak in approximately 85% of patients. Murphy indicated that pleuroperitoneal shunts are successful in 100% of patients with chylothorax after cardiac surgery and in 75% of patients with chylothorax secondary to caval obstruction. The success of thoracoscopic control of chylothorax appears comparable to that of these other procedures. Percutaneous techniques appear to be successful in approximately 70% of patients. Occasionally, a patient with diffuse lymphatic leak, such as a patient with caval obstruction or lymphangiomatosis, will fail to respond to these procedures. In these patients, a pleurodesis on the affected side will virtually always

control the chylous leak. This may be performed by thoracoscopy, using either pleural abrasion or talc pleurodesis, or by an open pleurectomy. TDE may prove to be the treatment of choice for chylothorax regardless of etiology due to its higher success rate and lesser morbidity. Over the past several years, the cumulative experience with TDE has increased significantly and the advent of intranodal lymphangiography offers the promise of increasing its use as the first-line therapy for chylothorax.

It is important to recognize that the development of chylothorax is not a benign condition. Treatment should be prompt and aggressive to minimize loss of protein, fats, and lymphocytes. The effusion must be drained, and the patient should be placed on a medium-chain triglyceride diet or total parenteral nutrition immediately. Somatostatin or octreotide should be instituted in those patients with drainage persisting for more than 3 days, and surgical therapy should be contemplated for effusions lasting 7 to 10 days. The choice of surgical procedure for the treatment of refractory chylothorax is determined by the experience of the surgeon and the cause of the chylothorax. Patients with direct injury to the duct from blunt or penetrating chest trauma may be the best candidates for posterolateral thoracotomy and direct control of the chylous leak as well as ligation of the main trunk of the thoracic duct. Patients with chylothorax arising secondary to thoracic surgical procedures in whom reexploration may be difficult or prohibitively stressful may be excellent candidates for the placement of a pleuroperitoneal shunt or percutaneous techniques, which may be performed under local anesthesia. Patients with long-standing and loculated chylothorax may benefit from thoracoscopic control, which enables the surgeon to disrupt the multiple loculations within the pleural space while controlling the leak of chyle.

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EDITOR'S COMMENTS

And I thought I knew a lot about chylothorax. The authors have written an absolutely superb chapter that should be mandatory reading for all thoracic surgical residents as well as practicing cardiothoracic surgeons. I am including even those surgeons who limit their practice to adult cardiac surgery because chylothorax rears its ugly head on occasion following cardiac surgical procedures not to mention how often it is seen following surgery for congenital heart disease. I would submit that it is far easier to avoid a problem with the thoracic duct than to fix a problem with the duct, not to mention the morbidity that results from a chyle leak. Should a chylothorax occur, however, the surgeon should be familiar with the problem and the potential solutions because time is of the essence.

I have not had any personal experience with somatostatin or octreotide in managing chylothorax, but other than the expense, either of these agents might be tried early in the course of a chylothorax especially if the patient is draining <500 ml/day. Having waited too long to ligate the thoracic duct on at least one patient, I now wait no longer than 7 days to intervene if the leak does not stop with conservative measures. The morbidity of hypoalbuminemia and immunosuppression in an early postoperative patient is simply too great to take any chances. We have been the major proponents of percutaneous techniques for sealing the duct, and the procedure is well described in this chapter. The advent of intranodal lymphangiography should make thoracic duct embolization much more accessible as no longer is pedal lymphangiography, a lost art, required. If the cisterna and its tributaries can be visualized, there is a high likelihood of successful embolization with the resolution of the chylothorax thus saving the patient a surgical procedure. One can make a rather convincing argument that an attempt at thoracic duct embolization should be the first intervention attempted in a patient with chylothorax.

The standard approach to ligating the duct remains a low right thoracotomy with ligation of the duct near the aortic hiatus, where it remains as a single trunk. Failure of this approach usually means that the duct was ligated too high in the chest, thus leaving a branch, or branches, patent. The thoracoscopic approach may be useful, although it is not easy, and the incidence of failure may be slightly higher than that achieved with an open procedure. For a left-sided effusion the duct may be approached via a left thoracotomy, as the authors point out. Ligation of the duct for a chylothorax caused by diffuse obstruction of mediastinal lymphatics by tumor, usually lymphoma, tends to have a significantly greater failure rate, and resolution of the effusion is usually accomplished by the treatment of the malignancy. It is in these situations that the pleuroperitoneal shunt may be of more help than simple ligation of the duct. Especially in those patients with higher output, rarely have we been successful simply by trying to effect pleurodesis.

The bottom line to all of this: Act quickly and decisively in managing an iatrogenic chylothorax. We recommend not waiting any longer than 7 days before proceeding with ligation or obliteration of the thoracic duct if the leak has persisted despite total parenteral nutrition.

LRK

28 Pericardial Procedures

Atasha Asmat and Nabil P. Rizk

ANATOMY AND PHYSIOLOGY OF THE PERICARDIUM

Anatomy

The pericardium consists of two layers. The inner layer, the visceral pericardium, is a single layer of mesothelial cells that is closely adherent to the myocardium. The outer layer, the parietal pericardium, forms a tough fibrous structure and is composed of dense collagen bundles and a small number of elastic fibers. The two layers are separated by a small amount of pericardial fluid.

The parietal pericardium attaches superiorly to the ascending aorta and the superior vena cava. From there, it continues across the superior vena cava, over the right border of the heart and the pulmonary veins, and encircles the inferior vena cava. After the parietal pericardium crosses the inferior vena cava, the inferior pericardium is densely adherent and essentially one with the diaphragm. The parietal pericardium then turns superiorly, just beyond the apex of the heart, and continues over the pulmonary veins, back to the aorta.

There are two sinuses within the pericardial space. They are formed by the pattern of pericardial reflections around the pulmonary vein and the vena cavae posteriorly. The transverse sinus is posterior to the ascending aorta and the pulmonary artery and is anterior to the atria and the superior vena cava. The oblique sinus is located directly behind the left atrium, centered between the pulmonary veins.

Physiology

The normal pericardium contains 15 to 50 ml of serous fluid. The pericardial fluid acts as a lubricant that reduces friction between the pericardial membranes during each heartbeat. Owing to the tough fibrous structure of the parietal pericardium, the pericardial sac is relatively noncompliant. As a result, alterations in the pericardium can have an impact on cardiac hemodynamics.

Pathophysiology

The pericardial sac is noncompliant but can expand if an effusion develops slowly, and it can accommodate large amounts of fluid before hemodynamic consequences occur. If a pericardial effusion develops acutely, however, rapid fluid accumulation overwhelms the ability of the pericardium to distend resulting in increased intrapericardial pressures. This can lead to impaired diastolic filling of the heart, caused by collapse of the right atrium and the right ventricle, and thus limit the stroke volume. As such, patients who develop pericardial effusions with hemodynamic compromise usually develop tachycardia, as it is the sole mechanism through which cardiac output can be increased.

The pericardium can also develop scarring and, consequently, become inelastic, resulting in major hemodynamic effects. A thick, rigid pericardium can prevent the respiratory variation in intrathoracic pressure from being transmitted to the cardiac chambers. Normally, with inspiration, negative intrathoracic pressure is generated; when pericardial constriction is present, this negative pressure is not transmitted to the heart. Consequently, pulmonary venous pressure and left-sided filling are reduced during inspiration. The thicker pericardium also results in the pericardial volume becoming static. As such, the reduction in left-ventricular filling during inspiration needs to be compensated for by increased right-ventricular filling, with septal shift toward the left ventricle. The opposite occurs during expiration. This interaction is referred to as ventricular interdependence. Finally, elevated atrial pressures result in rapid diastolic filling, which, because of the inelastic pericardium, abruptly decreases mid-diastole.

PERICARDIAL EFFUSIONS

Etiology

There are several disease processes that can cause pericardial effusions (Table 28.1). Secondary malignancies (usually from the lung or the breast) are the most common causes of pericardial effusions.

PERICARDIAL (CARDIAC) TAMPONADE

Pericardial (cardiac) tamponade is a medical emergency that occurs when the accumulation of fluid within the pericardial sac results in reduced diastolic filling of the heart and subsequent hemodynamic collapse. The development of tamponade is not related to the amount of fluid that accumulates, but rather to the rate at which the fluid accumulates and the capacity of the pericardium to distend. Therefore, even acute accumulation of small amounts of fluid may be sufficient to cause tamponade.

The first step in managing cardiac tamponade is to make the diagnosis. Patients may present with symptoms of worsening dyspnea, fatigue, or chest pain. Findings on examination can include sinus tachycardia, distended neck veins with elevated jugular venous pressure, and pulsus paradoxus.

When tamponade is suspected on the basis of medical history and physical examination, further evaluation including an electrocardiogram, chest X-ray, and echocardiogram is necessary. The electrocardiogram will usually demonstrate sinus tachycardia and low voltage. An enlarged cardiac silhouette may be evident on chest films. Echocardiography continues to play a major role in the diagnosis of cardiac tamponade. The features to note on echocardiography include the following:

- diastolic collapse of the right heart chambers
- respiratory variation in volumes and flows
- inferior vena cava dilatation

Management of Pericardial Effusions

A wide variety of therapeutic options are available to manage pericardial effusions. These options include simple observation,

Table 28.1 Major Causes of Pericardial Effusions

Malignancy
Metastatic
Primary
Trauma
Blunt
Penetrating
Iatrogenic
Metabolic
Uremia
Infection
Viral (coxsackie virus A9, mumps, Epstein–Barr virus, cytomegalovirus, varicella, rubella, HIV)
Bacterial
Fungal
Tubercular
Cardiac
Pericarditis
Post cardiac surgery
Aortic dissection
Radiation
Idiopathic

use of anti-inflammatory or antineoplastic chemotherapy, pericardiocentesis, percutaneous balloon pericardiotomy, and surgery. Because of the lack of prospective studies evaluating the efficacy of the different treatments, the optimal management of this condition remains a matter of controversy.

When determining the choice of treatment for pericardial effusions, a number of factors need to be considered. These factors include the presence or absence of tamponade, the underlying diagnosis or etiology of the effusion, the potential for recurrence, and the resources available at one's institution.

Pericardiocentesis

Pericardiocentesis can be performed at the bedside or under echocardiographic or fluoroscopic guidance. Local anesthesia is usually adequate. To begin, the area between the xiphoid process and the costal margin is infiltrated. An 18-gauge spinal needle attached to a three-way stopcock and syringe is inserted between the two structures and advanced toward the left shoulder at a 45-degree angle. The needle should be gently aspirated as it is being advanced. If the procedure is being performed without imaging, attaching the needle to a precordial electrocardiographic lead can help to identify when the myocardium is breached, as ST elevation, which resolves when the needle is withdrawn, will be noted. Once the pericardial space is entered, a guide wire is advanced, and a pigtail catheter is introduced by use of a modified Seldinger technique. Fluid that is removed can be sent off for further evaluation (biochemical, microbiologic, and cytologic examination).

Major complications of pericardiocentesis include laceration and perforation of the myocardium and the coronary vessels. Other complications that have been reported include air embolism, pneumothorax, arrhythmias, and puncture of the peritoneal cavity or abdominal viscera. The incidence of major complications ranges from 1.3% to 1.6%, and safety has improved with the use of echocardiographic or fluoroscopic guidance.

One advantage of pericardiocentesis is the avoidance of general anesthesia. It can also provide effective immediate improvement in unstable patients with cardiac tamponade. However, not all pericardial effusions are appropriate for pericardiocentesis—namely, small, posterior, and loculated effusions. Other disadvantages include high recurrence rates and reduced diagnostic yield. A retrospective study comparing outcomes after pericardiocentesis and surgery found a recurrence rate of 16.5% in patients who underwent pericardiocentesis, compared with 4.6% in patients who underwent surgery. The diagnosis of malignancy was confirmed in 59% of patients who underwent percutaneous procedures, compared with 62% of patients who underwent open drainage.

Percutaneous Balloon Pericardiotomy

Percutaneous balloon pericardiotomy is performed in the cardiac catheterization laboratory using local anesthesia and intravenous sedation, with fluoroscopic and echocardiographic guidance. The pericardium is entered with an 18-gauge pericardial needle by use of a standard subxiphoid approach. A guide wire is then advanced into the pericardial space, a catheter is introduced, and pericardial fluid is drained for laboratory studies. Radiographic contrast medium is then injected into the pericardial space to aid visualization. The catheter is removed, and the tract is dilated using a 10F–14F dilator. A low-profile balloon-dilating catheter containing some radiographic contrast medium is inserted over the guide wire and positioned across the parietal pericardium. To ensure adequate opening of the parietal pericardium, a series of balloon inflations are performed. A pigtail catheter is often left in the pericardial space for 24 hours and removed once complete resolution of the effusion has been confirmed by echocardiography. Chest X-rays are also performed after the procedure to exclude the possibility of a pneumothorax.

There have been several small studies describing the use of this modality as an alternative to a surgically created pericardial window. The largest trial, which was part of a multicenter registry, involved 50 patients. In that trial, the modality was considered successful for 46 patients, after a mean follow-up of 3.6 months. The procedure was deemed unsuccessful for four patients, either because of recurrence of effusion or tamponade, bleeding requiring surgical intervention, or persistent catheter drainage requiring surgery.

Subxiphoid Pericardial Window

Subxiphoid pericardial window is a surgical procedure that can be performed with the patient under either local or general anesthesia. Because of the lack of evidence in the literature, the optimal anesthetic management for patients with pericardial tamponade who require a subxiphoid pericardial window remains open to debate. The advantages of using general anesthesia include improved surgical evacuation of pericardial contents and improved patient comfort. In cases where general anesthesia is administered to patients with significant hemodynamic compromise, the patient should be prepped and draped, with the surgeon ready to begin, before induction of general anesthesia. Unpublished data have also suggested that there is no difference in outcomes between patients managed with local anesthesia and sedation and those managed with general anesthesia (Fig. 28.1).

For the subxiphoid approach, a vertical incision is made that begins at the xiphoid–sternal junction and extends inferiorly by 4 to 5 cm. The linea alba is identified and divided with care so that the peritoneum is not breached. To provide better exposure the xiphoid process is resected. The costal margin is elevated, allowing the diaphragm and peritoneal fat to be separated by use of a sponge stick, until the pericardium is identified. The pericardium is opened using shallow strokes of a blade to minimize risk of injury to the underlying myocardium. Entry into the pericardial space is confirmed when an initial gush of fluid occurs. When the

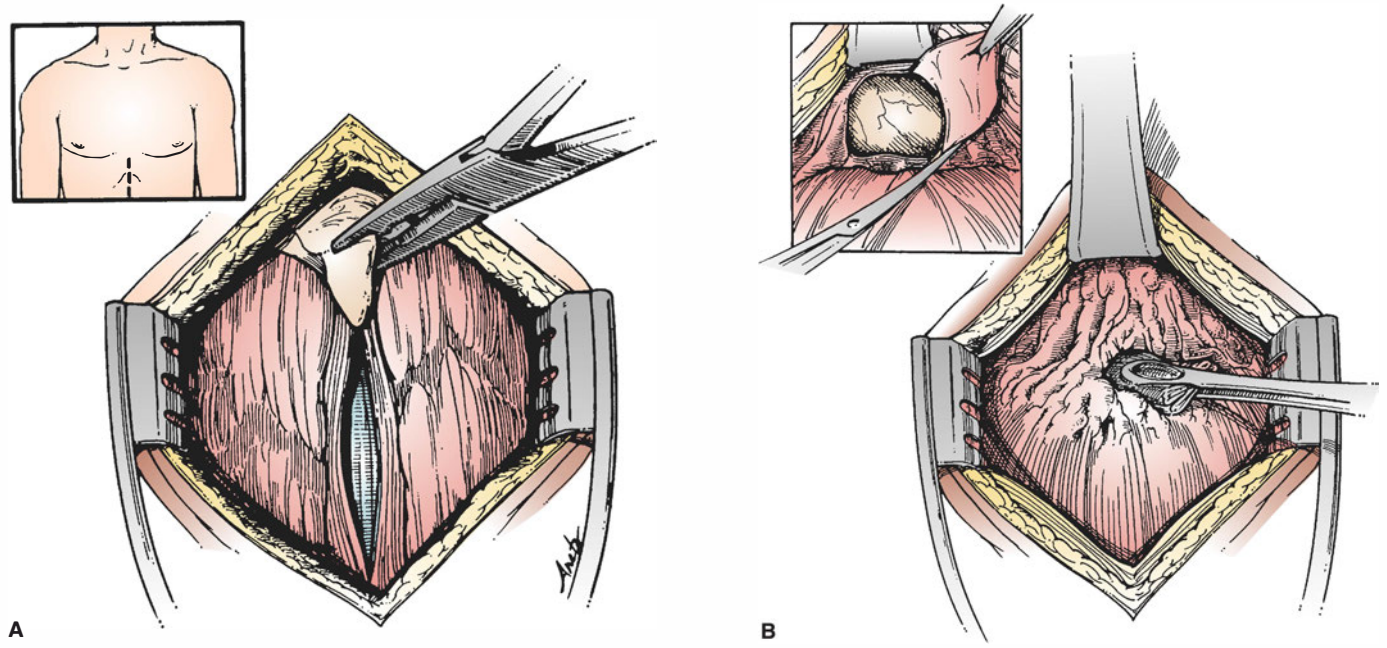


Fig. 28.1. Subxiphoid pericardial window. (**A**) An *upper midline* incision centered over the xiphoid process is performed, with sharp dissection down through the linea alba. The xiphoid can be resected for better exposure. (**B**) The sternum and costal margin are elevated with a retractor. A combination of blunt dissection with a sponge stick and sharp dissection with electrocautery is then used to dissect the soft tissue superiorly, ultimately exposing the inferior pericardium. Once exposed, the pericardium is incised with scissors or a knife, the pericardial sac is digitally explored, and a large portion of the inferior pericardial sac is excised. A right-angle chest tube is then placed into the sac, from a separate incision.

effusion is hemorrhagic, it can be difficult to determine whether cardiac injury has occurred. The surgeon should therefore be vigilant to monitor the patient's hemodynamic status at this time.

Once the effusion is drained and samples are sent for culture and cytologic examination, a finger is used to explore the pericardial space and to break up any loculations. A piece of pericardium is then resected (as large as is possible to safely remove) and sent for microbiologic assessment and histologic examination. One or two 28F right-angle chest tubes are placed through stab incisions on either side and inferior to the incision and positioned in the inferior aspect of the pericardial space. The linea alba is closed using interrupted nonabsorbable sutures, and the skin and the subcutaneous tissue are closed using absorbable sutures, in the standard fashion. The chest tubes are left in place until the drainage is less than 100 ml/day and when complete drainage of the pericardium has been confirmed by echocardiography.

In published case studies, the recurrence rates for patients who underwent open subxiphoid drainage range from 0% to 9.1%. The majority of these studies reported only a single method of drainage. One retrospective study found that the recurrence rate for patients who underwent open subxiphoid window was 1.1%, compared with 30% for patients who underwent pericardiocentesis. Similarly, another retrospective study found that the recurrence rate after open subxiphoid drainage was lower (4.6%) than that after percutaneous catheter drainage (16.5%).

Transthoracic Pericardial Window

Thoracotomy Approach

Open thoracotomy can be a useful approach for hemodynamically stable patients who present with a pericardial effusion. It can also be useful for those patients who require concurrent performance of additional procedures, such as lung biopsy. A left-sided approach is generally preferred, as a larger amount of pericardium is accessible on the left side. Patients are placed supine, with the left side elevated by approximately 30 degrees. Single-lung isolation is preferred to allow adequate visualization.

The incision is made in the inframammary crease and the fourth or fifth intercostal space is entered. The pericardium can be visualized once the lung is retracted laterally. The pericardium is opened with care taken not to injure the phrenic nerve or the underlying myocardium. When this approach is used a large piece of pericardium can be resected, to minimize the incidence of recurrence. A large, 28F chest drain is usually placed in the left pleural cavity and the thoracotomy wound is closed in the standard fashion.

Video-Assisted Thoracoscopic Approach

The video-assisted thoracoscopic surgery (VATS) approach is a safe and effective approach that is minimally invasive and allows concomitant exploration of the pleural cavity. Effective single-lung isolation is mandatory, and, therefore, this approach is not suitable for patients who are hemodynamically unstable. Although the approach can be performed on either side because of the proximity of the pericardium to the chest wall, a left-sided VATS approach is technically more challenging (Fig. 28.2).

After induction of general anesthesia the patient is placed in the lateral decubitus position. The incision for the camera port is usually made in the seventh or eighth intercostal space (depending on the patient's body habitus), in the posterior axillary line. A utility incision is then made anteriorly, under direct vision, in the fifth anterior intercostal space. To allow for the use of several different instruments through the same incision, the incision is made approximately 4 to 5 cm in length.

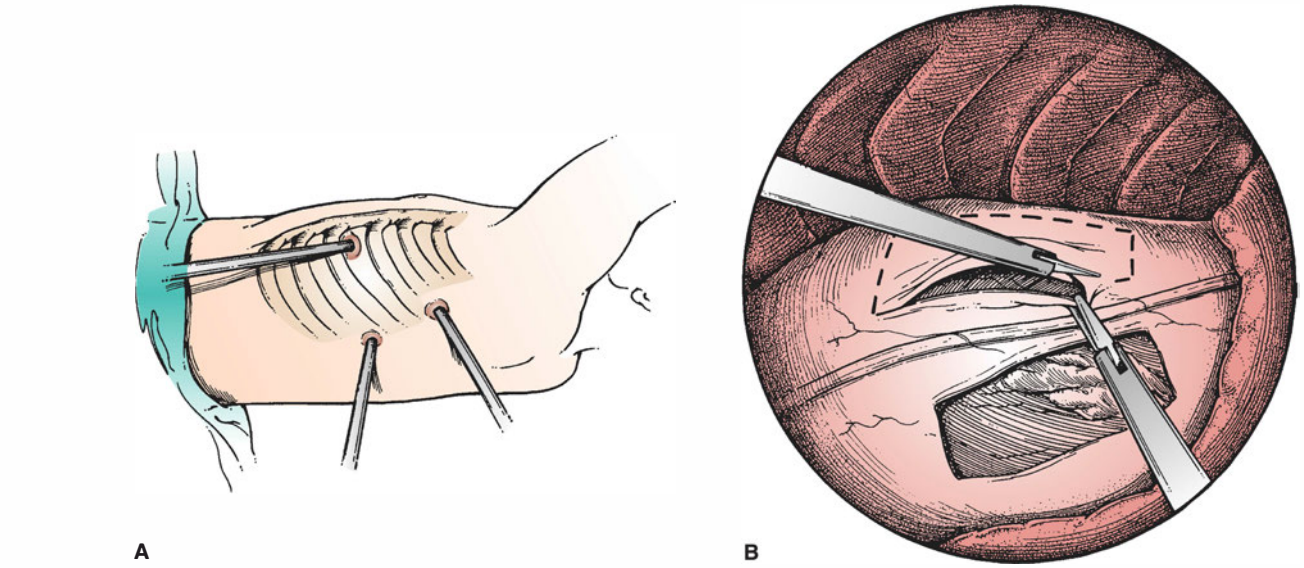


Fig. 28.2. Transthoracic pericardial window. (**A**) The patient is placed in a full right-lateral decubitus position, and three ports are placed into the left hemithorax (one in the eighth intercostal space posterior axillary line and two at approximately the midscapular line, in the sixth and ninth intercostal spaces). (**B**) Portions of the pericardium are resected, ensuring that the phrenic nerve is visualized and spared. At the end of the procedure, a chest tube is placed into the hemithorax via the posterior axillary incision.

Next, the phrenic nerve is identified, and an incision is made anterior to it. A generous pericardial window is created by resecting a piece of anterior pericardium. This is repeated posterior to the phrenic nerve as well. If the procedure is being performed on the right side, a single large pericardial window is created, since the phrenic nerve runs closer to the hilum.

Several studies have reported recurrence rates ranging from 0% to 8% after creation of a thoracoscopic pericardial window. A retrospective study that investigated outcomes after subxiphoid window and VATS pericardial window noted recurrence rates of 8% and 10%, respectively. That study also noted that time of anesthesia for thoracoscopy was significantly longer (117.1 vs. 81.1 minutes).

Pericardial Sclerosis

Intrapericardial sclerosis induces an inflammatory response that will obliterate the pericardial space. The pericardial space is accessed via percutaneous catheter drainage or after surgical drainage (subxiphoid window, thoracotomy, or VATS). The procedure is performed once drainage through the catheter has been less than 100 ml for more than 24 hours. Local anesthetic, such as lidocaine, is instilled through the catheter, followed by instillation of the sclerosing agent. The drain is kept clamped for 3 hours and then removed.

Sclerosing agents that have been used include tetracycline, doxycycline, bleomycin, and thiotepa. These various sclerosing agents appear to have equal effectiveness in minimizing recurrence rates after drainage of pericardial effusions. However, it appears that some agents may be associated with more complications. Doxycycline can result in intense pain with most patients who experience pain requiring intravenous analgesia. More than half of the patients who receive doxycycline also develop fevers higher than 38.5°C. Alternatively, thiotepa does not result in pain and very rarely generates a pronounced febrile response. In addition, it has been shown to be more cost-effective.

CONSTRICTIVE PERICARDITIS

Constrictive pericarditis can occur after any disease process that affects the pericardium (Table 28.2).

Signs and symptoms of constrictive pericarditis are analogous to those seen with fluid overload (elevated jugular venous pressures, ascites, and peripheral edema) and/or to diminished cardiac output in response to exertion (fatigue and dyspnea on exertion). Echocardiographic examination is essential for patients being evaluated for constrictive pericarditis. Findings include a septal bounce that is related to respiratory variations in the motion of the ventricular septum, and decreased transmitral flow velocity.

Table 28.2 Causes of Constrictive Pericarditis

Idiopathic
Cardiac surgery
Mediastinal radiotherapy
Connective tissue disorders
Infectious

Computed tomography may demonstrate increased pericardial thickness and calcification, but it is important to note that a normal appearance on computed tomography does not exclude the diagnosis of constrictive pericarditis. Gated cardiac magnetic resonance imaging allows direct visualization of the pericardium that normally has a low magnetic resonance imaging signal intensity. Features that are characteristic of constrictive pericarditis on cardiac magnetic resonance imaging include increased pericardial thickening and dilatation of the inferior vena cava.

Cardiac catheterization occasionally may be required to confirm the diagnosis of constrictive pericarditis for patients with nondiagnostic echocardiographic findings. The major hemodynamic findings include elevated right-atrial pressure, rapid x and y descents in the atrial pressure curves, and "square-root" signs in the right-ventricle and left-ventricle pressure tracings

(a dip-and-plateau pattern in the ventricular pressure curves caused by rapid early filling and limited late filling).

The most effective approach to manage constrictive pericarditis remains unclear as no prospective studies have compared outcomes in patients undergoing medical therapy with those undergoing operation. While the natural history of constrictive pericarditis remains unknown observational studies and case reports suggest that patients who do not undergo surgical intervention experience progression of symptoms and early death. Conversely, it has been reported that a large number of patients who underwent pericardiectomy experienced improvement in their symptoms, as well as improvement in their quality of life. Therefore, pericardiectomy remains the treatment of choice for patients with symptomatic constrictive pericarditis.

Pericardiectomy

Pericardiectomy is the accepted approach to manage patients with symptomatic constrictive pericarditis. However, opinions differ with regard to the ideal surgical approach, the extent of the pericardiectomy, and the need for cardiopulmonary bypass.

The two standard surgical approaches that have been used are (1) left anterolateral thoracotomy and (2) median sternotomy. There are few reports that address the issue of surgical approach, and the choice of approach appears to be left to the surgeon's personal preference. Left anterolateral thoracotomy provides excellent exposure of the left ventricle and, consequently, permits a more complete release of the left ventricle. Median sternotomy allows for a more extensive pericardiectomy to be performed with better exposure of the right ventricle and great veins. A retrospective study that compared the two surgical approaches in a cohort of 36 patients demonstrated that outcomes and mortality rates were similar for both approaches. Regardless of the approach used, all patients require invasive hemodynamic monitoring with an arterial and central venous catheter. In patients with significant hemodynamic compromise, a pulmonary artery catheter may also be required.

The extent of pericardiectomy to be performed is also controversial. A total pericardiectomy is defined as a wide excision of the pericardium, with the phrenic nerves defining the posterior extent, the great vessels (including the intrapericardial portion and the superior vena cava–right atrium junction) defining the superior extent, and the diaphragmatic surface (including the inferior vena cava–right atrium junction) defining the inferior extent of the pericardial resection. A partial pericardiectomy is defined as any excision that is less than a total pericardiectomy. A retrospective study that compared 338 patients who underwent total pericardiectomy with 57 patients who underwent partial pericardiectomy demonstrated that the risk of death was 4.5 times higher for patients who underwent partial pericardiectomy. This study also demonstrated better long-term survival and improved functional status (New York Heart Association functional classification) for patients who underwent total pericardiectomy.

The use of cardiopulmonary bypass as an adjunct to pericardiectomy remains a matter of debate. Some believe that the use of cardiopulmonary bypass facilitates surgical dissection by emptying the ventricular cavities (to define the appropriate plane of dissection), as well as by providing better access to the diaphragmatic surface of the left ventricle (thus allowing for a more extensive pericardiectomy). Cardiopulmonary bypass is also helpful in situations where inadvertent myocardial injury has occurred during dissection. The major disadvantage of cardiopulmonary bypass is the potential for increased bleeding with the systemic heparinization.

Median Sternotomy

The patient is placed in the supine position, with the arms tucked to the side, and a median sternotomy is performed in the usual manner. Once the sternum is divided, adhesions, if any, are dissected away, both pleural spaces are opened, and the right and left phrenic nerves are identified. The aim of this procedure is to entirely resect the pericardium between the phrenic nerves and to mobilize the remainder of the cardiac surfaces within the pericardium (Fig. 28.3).

There are often limited loculated pericardial spaces, particularly around the great vessels and the diaphragm. To attempt to identify the pericardial space, transverse incisions are created in these areas. While creating the initial flap, the surgeon must exercise caution to avoid causing injury to the coronary artery. The pericardium can be densely adherent in the region of the coronary arteries, and in this instance it may be safer to leave small islands of pericardium on the heart. Bone-cutting instruments may be necessary when areas of calcification are encountered. Calcified deposits that extend into the myocardium should be left in place and not removed.

The pericardium over the left ventricle should be freed first. If the pericardium over the right ventricle is freed first, increased preload (improved diastolic filling of the heart) and afterload (left ventricle still restricted by thick pericardium) can result. This can cause acute right-ventricular dilatation and failure, with devastating consequences. The pericardium is resected between the phrenic nerves, and the heart is dissected free from the thick, constrictive pericardium, all the way down to both atria. Once the pericardiectomy is complete, mediastinal and pleural drains are placed, and the sternum is closed in the usual manner.

Left Anterolateral Thoracotomy

The patient is positioned supine, with the left side elevated by 45 degrees and the left hand placed behind the buttocks. An inframammary incision that curves posteriorly to overlie the fifth rib is made. The chest is entered through the fifth interspace. Removing the fifth costal cartilage may provide better exposure. The left internal mammary vessels must often be divided, and the left phrenic nerve is carefully identified (Fig. 28.4).

Once again, the loculated spaces that are present near the diaphragm and great vessels are a convenient starting point for the pericardiectomy. The pericardium is dissected free, and a tongue of pericardium is left attached to the phrenic nerve along its length. The pericardial resection is then extended posterior to the phrenic nerve, onto the left atrium, and then is resumed anterior to the phrenic nerve, ideally continuing as far as the right atrioventricular groove. The necessary precautions to avoid causing injury to the coronary vessels and myocardium should be taken.

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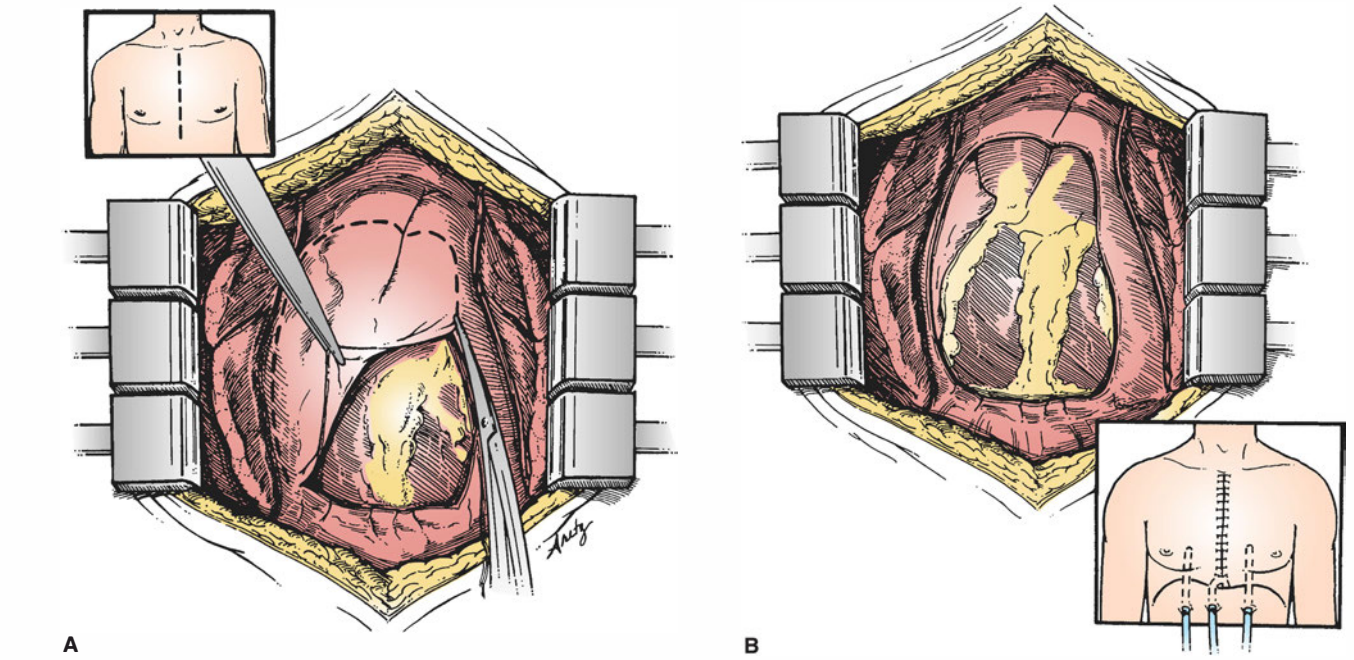


Fig. 28.3. Sternotomy and pericardiectomy. (**A**) A full sternotomy incision is performed, both pleural spaces are entered, and both phrenic nerves are identified. The pericardium is incised and freed first over the left ventricle. (**B**) A complete pericardiectomy includes removal of the pericardium extending laterally to both phrenic nerves and inferiorly from the diaphragm, extending superiorly to the aorta, pulmonary artery, and right atrium. Three chest tubes are placed before closure of the sternum (two pleural and one beneath the sternum).

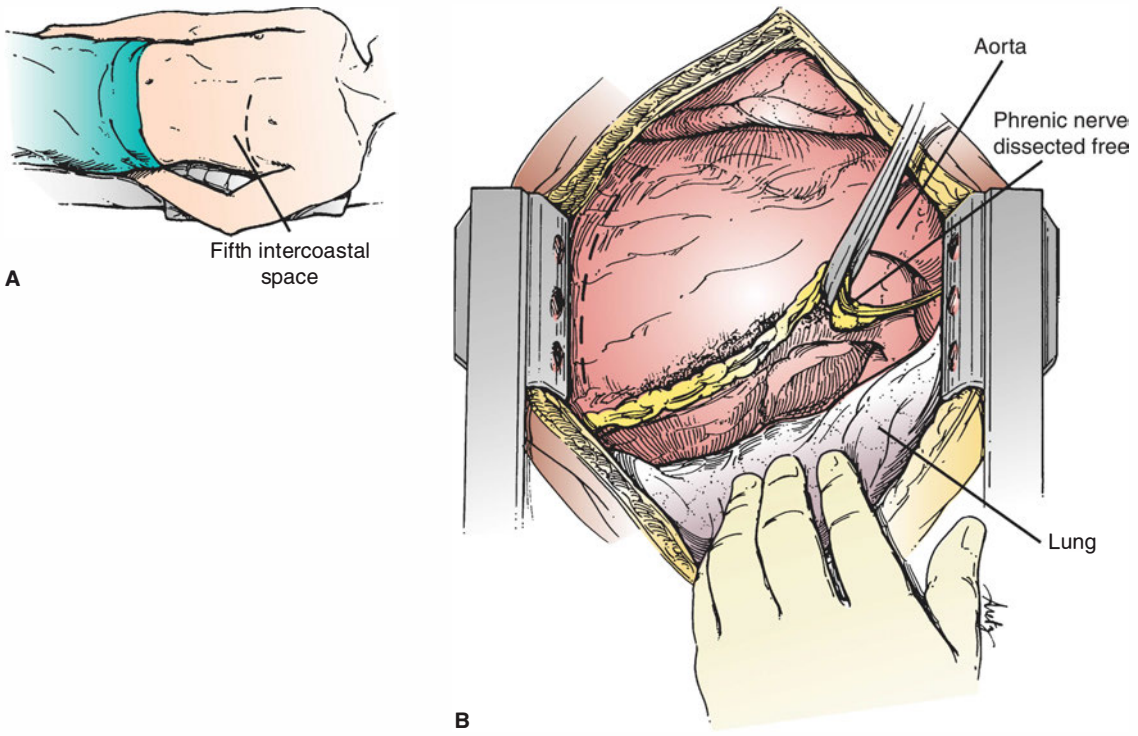


Fig. 28.4. Left anterolateral thoracotomy and pericardiectomy. (**A**) The patient is positioned supine, with the left side elevated by 45 degrees and the left hand placed behind the buttocks. An inframammary incision that curves posteriorly to overlie the fifth rib is made. The chest is entered through the fifth intercostal space. (**B**) The pericardium is dissected free, and a tongue of pericardium is *left* attached to the phrenic nerve along its length. The pericardial resection is then extended posterior to the phrenic nerve, onto the left atrium, and then is resumed anterior to the phrenic nerve, ideally continuing as far as the right atrioventricular groove.

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EDITOR'S COMMENTS

The management of a pericardial effusion depends to a great extent on the suspected etiology of the effusion and whether or not tamponade physiology is present. Simple pericardiocentesis may be the only procedure necessary if there is no evidence of tamponade and fluid for diagnosis is required. Previously, pericardiocentesis was carried out with the needle attached to an ECG to determine whether the needle was within the myocardium. The use of echocardiography to visually determine needle placement clearly is the more definitive procedure.

Despite the feasibility of catheter drainage of the pericardium as well as the development of balloon pericardiostomy, the definitive procedure for the treatment of a symptomatic malignant pericardial effusion remains the subxiphoid window. As the authors note, the procedure may be carried out with either local or general anesthesia but our preference has been to use general anesthesia. The surgeon must be present at the time of induction of anesthesia and the patient should be prepped and draped so that rapid access to the pericardium may be gained should there be a significant drop in blood pressure. Exposure to the pericardium is significantly enhanced by resecting the xiphoid process and applying downward pressure on the diaphragm. It is well known that the window itself is likely obliterated fairly rapidly and the therapeutic value of the pericardial window is the facilitation of visceral to parietal pericardial symphysis. It is this obliteration of the pericardial space that is likely the cause of the low recurrence rate following the performance of a subxiphoid pericardial window. The subxiphoid approach is far preferable to the left anterior thoracotomy approach, in my opinion. A right-angled chest tube should always be left in the pericardial sac following the subxiphoid window procedure with the tube placed to suction drainage. The operative procedure is further aided by the use of transesophageal echocardiography to define the location of the effusion and specifically whether there are loculations that need to be broken up and whether there is significant fluid in the posterior pericardium. A generous swath of anterior pericardium should be taken and submitted for pathologic examination.

The treatment of constrictive pericarditis is made all the more difficult because the distinction between a restrictive cardiomyopathy, which would not benefit from pericardiectomy, and constriction must be made. This distinction may rely on the imaging demonstration of thickened pericardium in addition to the echocardiographic or cardiac catheterization data. Depending on the extent of the constriction the procedure may be more safely carried out with the use of cardiopulmonary bypass, but this decision must be made on a case-by-case basis. It is also critically important, as the authors point out, to relieve the constriction of the left ventricle prior to releasing the right ventricle to avoid right ventricular dilatation. A total pericardiectomy is the optimal procedure.

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29 Surgical Management of Malignant Mesothelioma

Joseph S. Friedberg

INTRODUCTION

Albeit the most common primary tumor of the pleura, malignant pleural mesothelioma is a very rare cancer, only several percent as common as nonsmall cell lung cancer in the United States. The tumor is most commonly linked to asbestos exposure and typically occurs several decades after exposure. Despite the relatively recent mandates to protect the public from asbestos, it is expected that the incidence of mesothelioma will continue to increase for the foreseeable future.

The natural history of mesothelioma, which is almost always unilateral, is inexorable local progression with encasement of the lung and invasion of the chest wall, diaphragm, and/or mediastinum. The majority of patients will succumb to the disease less than a year from the time of diagnosis. Although the disease has a reputation for being a purely localized tumor, at least half the patients will have occult metastases at the time of their demise. The clinical manifestations of metastases have become more commonly noted in some patients who have undergone aggressive multimodal approaches and are fortunate enough to have an extended survival.

Patients will commonly present with dyspnea secondary to a pleural effusion. Other presenting symptoms, such as weight loss or pain from chest wall invasion, are even more ominous and tend to reflect more advanced or aggressive form of the cancer. Radiographic studies may reveal only the pleural effusion and no other detectable disease. Patients with a pleural rind will commonly demonstrate a contracture of the hemithorax as a result of decrease in the size of the rib interspaces, which can be easily seen on a plain chest film.

Unless a history of asbestos exposure is offered, mesothelioma may not be immediately considered and the diagnosis will be delayed. Further delaying the diagnosis is the common event of a negative cytology on fluid withdrawn by thoracentesis. Ultimately the recurrence of the effusion or the recalcitrance of the clinical complaints to attempted interventions will, hopefully, lead the patient to a physician experienced with mesothelioma. Ideally this will be a thoracic surgeon facile in dealing with pleural diseases.

Establishing a diagnosis and simultaneously palliating the symptoms caused by the effusion can be accomplished thoracoscopically through a single 1-cm incision. If the patient is potentially a candidate for an aggressive treatment protocol, then the ideal situation is for the surgeon who would be performing the therapeutic operation to perform the diagnostic/palliative procedure. The reasons for this are twofold. The first is that mesothelioma has a propensity to seed incisions. Hence, it is often desirable to excise the biopsy incisions at the time of the thoracotomy. Consequently, if the same surgeon performs both procedures, the incisions used for the biopsy procedure can be placed in line with a potential thoracotomy, such that they could be excised as part of the thoracotomy without compromise of chest wall integrity or the need for an additional incision. In addition, the surgeon who performs both procedures can decide between pleurodesis or placement of a long-term indwelling catheter as the best route of palliation that will not interfere with future operations or therapeutic interventions.

Pemetrexed-based chemotherapy is, ostensibly, the standard of care treatment for pleural mesothelioma. Beyond this chemotherapy, essentially all treatments—including surgery—remain investigational. Nonsurgical modalities that have been used to treat this malignancy include many combinations of systemic therapy, radiation therapy, and many types of immunotherapies.

To date, however, the treatments that have met with the greatest measure of success in extending life are surgery-based multimodal treatments. Again, hobbled by the scarcity of this cancer and the small number of patients undergoing operation for this cancer, large randomized prospective trials do not exist to validate the role of surgery in treating this cancer. It is, therefore, appropriate that any patient being offered surgery for this cancer should be the focus of a multidisciplinary panel discussion to assess the options. Patients undergoing operation for mesothelioma should, in addition to standard disclosures for informed consent, be aware that surgery is not the standard of care. Furthermore, as mesothelioma remains an incurable cancer, patients should understand that although the procedure is done with "curative intent," the realistic goal is extension of life beyond what would be anticipated with less aggressive approaches. Ideally, patients undergoing surgery for this cancer should be part of a clinical trial that will accrue data to further the field and help establish optimal treatments for future patients.

Patient selection for surgery-based treatments can vary from institution to institution, but the general principles observed by all groups are that the cancer must appear confined to one hemithorax and the patient must be at reasonable risk for the proposed procedure. Areas of controversy are typically stage-related inclusion/exclusion criteria, particularly N2 lymph nodes, and mesothelioma subtype. As a general rule, patients with nonepithelial histologies are much less likely to enjoy any significant period of remission compared to patients with epithelial subtypes. Some surgeons will have an age limit, whereas others do not. As pemetrexed-based chemotherapy is standard, essentially all surgery-based treatments will incorporate this, but whether it is given as a postoperative adjuvant, preoperative neoadjuvant, or both is also currently institution dependent. Finally use of other modalities, if any, combined with surgery, remains institution dependent.

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Because microscopic disease remains after any operation for a pleural malignancy, the goal of surgery in these multimodal protocols is to achieve a macroscopic complete resection. There are two approaches toward achieving this goal, operations that take the lung or operations that spare the lung.

The lung-sacrificing approach, extrapleural pneumonectomy, currently is the most common approach. This operation involves resection of the parietal pleural envelope en bloc with the lung, diaphragm, and pericardium. The pericardium and diaphragm are then reconstructed with prosthetic patches. This procedure has the advantages of standardized techniques/nomenclature, leaving the least amount of residual microscopic disease and, because the lung is absent, the ability to deliver full-dose adjuvant hemithoracic radiation. The obvious disadvantages are the risk and lifestyle consequences of pneumonectomy and the smaller pool of patients, many of whom are elderly, who are appropriate candidates for pneumonectomy.

Whereas extrapleural pneumonectomy enjoys uniformity in technique and nomenclature, lung-sparing surgery enjoys neither. Some surgeons intentionally leave behind gross disease, while others use it to achieve a macroscopic complete resection. Terms to describe this procedure include palliative pleurectomy, palliative debulking, pleurectomy, decortication, pleurectomy–decortication, and radical pleurectomy. Radical pleurectomy is the term used by the author and is defined as a procedure used to achieve a macroscopic complete resection that spares the lung and, whenever possible, the phrenic nerve, and as much of the diaphragm and pericardium as possible. The advantages of radical pleurectomy are the obvious benefits of lung preservation which, compared to pneumonectomy, has the potential to translate into less operative risk resulting in more patients being eligible for surgery-based treatment, better postoperative quality of life with greater reserve, and, consequently, more treatment options when the inevitable recurrence occurs. The disadvantages include lack of standardization, unique technical challenges, the need for judgment as to when the resection is complete, inability to incorporate standard adjuvant hemithoracic radiation, and, almost certainly, more residual microscopic disease than after an extrapleural pneumonectomy.

The modalities that have been used in an adjuvant and/or neoadjuvant capacity with operation include radiation, chemotherapy, and immunotherapy. The two principle modalities that have been used as intraoperative adjuvant therapies are hyperthermic chemotherapy lavage and photodynamic therapy. Again, with the exception of pemetrexed-based chemotherapy none of these, in any combination, is considered the standard of care.

Once a patient has been reviewed in a multidisciplinary conference and has elected to pursue a surgery-based treatment while understanding and recognizing other options, we initiate an evaluation to determine if the patient is a safe and appropriate candidate from an oncologic perspective. Our preoperative safety workup includes all of the usual studies and evaluations for a major pulmonary procedure, but with special emphasis on pulmonary function, cardiac function, and nutritional status. With respect to staging, our standard noninvasive preoperative workup includes a chest computed tomographic (CT) scan, brain imaging, and a positron emission tomography (PET) scan. MRI is utilized selectively when there is a question as to the extent of diaphragmatic or mediastinal invasion. We routinely perform a bronchoscopy and outpatient laparoscopy with peritoneal lavage to rule out radiographically occult disease. If the contralateral thorax has any questionable findings, we incorporate a VATS inspection of that hemithorax at the same time. Mediastinoscopy or endobronchial ultrasound (EBUS)-guided biopsies of the mediastinal lymph nodes also is commonly performed. Until our most recent study the status of the mediastinal (N2) lymph nodes had not demonstrated significant correlation with outcome. Now that we have established that mediastinal nodal involvement is correlated with outcome, although not an exclusion criterion like contralateral chest or abdominal disease, we also incorporate mediastinal staging with EBUS as part of our invasive staging workup.

OPERATIVE TECHNIQUES

Diagnosis, Palliation of Pleural Effusion, and Invasive Staging

Occasionally, a diagnosis can be established on the basis of fluid cytology from a thoracentesis or a closed pleural biopsy. More commonly, however, a surgical biopsy is required. This is best accomplished using a thoracoscopic technique and can be performed through a single 1-cm incision. After medical clearance the patient is brought to the operating room and once general anesthesia is induced, a bronchoscopy is performed. We have, on one occasion, discovered contralateral endobronchial metastases that served as a contraindication for any aggressive treatment options. The expected appearance of the airway, in the setting of a large effusion or bulky pleural disease, is extrinsic compression. If there is a significant amount of secretions in the airway, the surgeon should plan on performing a completion toilet bronchoscopy at the conclusion of the operation in order to maximize postoperative lung expansion.

Diagnosis and Palliation for Patients Presenting with Effusion

Once the patient is turned to the lateral decubitus position, the position of the double-lumen tube or bronchial blocker is confirmed and the lung on the operative side is isolated. After preparing the skin and positioning the patient, a potential thoracotomy incision is drawn on the chest wall. It is our practice to enter the chest through a serratus-sparing thoracotomy through the sixth interspace or the bed of the resected seventh rib. Starting in the anterior axillary line and working forward as necessary, the chest is sounded with a 22-gauge needle, along the potential future incision line, until fluid is encountered. This sample is saved for microbiology stains and cultures if there is any chance that the effusion is secondary to an infectious process. Having identified a safe site for entry, a local cutaneous and intercostal block is performed with a long-acting local anesthetic and a 10-mm incision is then created. The chest is then entered through this incision and fluid is aspirated to allow visualization. The fluid is collected for cytologic evaluation in the event that lesional tissue is not confirmed on the pleural biopsies. It is our practice to send an entire suction canister of pleural effusion for this purpose.

A 5 mm 30 degree thoracoscope is then introduced and the hemithorax is inspected. A small suction or blunt laparoscopic instrument can then be placed in through the same incision, alongside the thoracoscope, and loculations are disrupted and any remaining fluid is aspirated. Mediastinoscopy, or any other narrow, biopsy forceps are then introduced. The 30 degree angle allows the surgeon to maneuver the scope and the biopsy forceps without interference. In early-stage mesothelioma visual inspection within the chest may reveal only an injected-appearing pleura, in which case extensive parietal pleural biopsies are performed. More advanced stages will reveal popular or nodular lesions that will eventually coalesce into plaques coating the chest wall, lung, diaphragm, and mediastinum. It is critically important to never violate the visceral pleura or a persistent

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air leak can result. All biopsies should be taken from the parietal pleura. Biopsy specimens need to be sent to the pathologist for frozen section analysis and "lesional tissue" needs to be confirmed along with a sufficient quantity of specimen for the pathologist to run all additional tests required to render a final diagnosis. Although most diagnoses are currently made on the basis of morphology and the immunohistochemical profile, some institutions may still elect to perform electron microscopy that may require special handling of the specimen and this should be discussed with the pathologist. This is occasionally required when patients are having repeat biopsies for previous nondiagnostic surgical biopsies. Molecular or genetic analysis of biopsy specimens may also be undertaken in order to definitively establish a diagnosis.

At the conclusion of the specimen sampling, palliation of the effusion should be considered. If, based on the proposed treatment plan, the patient is going to have another procedure in the near future, or if the lung fails to fully expand, then a cuffed, silicone pleural catheter with a valve should be placed with the intent for this to remain in place on a chronic basis to deal with recurrent effusion. If the treatment plan does not anticipate further surgery, or the operative surgeon desires obliteration of the pleural space, then a chemical pleurodesis should be performed. One critical error is to instill talc into a chest cavity where visceral pleural–parietal–pleural apposition does not occur. In this setting, with entrapped lung, a residual pleural space will remain. If it gets infected, it can be very difficult to treat and if talc, a permanent foreign body, is in the chest cavity, this can spiral into an incurable empyema that will be the direct cause of the patient's death. If the patient will be a candidate for a surgery-based treatment, then the tube should be brought out in the same incision line, close enough to the VATS port that it can be incorporated in an excision at the time of thoracotomy. If the patient is not going to be a candidate or wishes not to have further surgery, then the tube should be brought out along the anterior axillary line at the level of the costophrenic recess. This will allow for better drainage. In either case, the tube should be directed posteriorly, to drain fluid at the diaphragm, and then extend to the end of this length paraspinally in a cephalad direction. The single VATS incision is then closed with absorbable sutures in a watertight manner. Depending upon the patient, this procedure may be performed on an outpatient basis. Commonly, however, these patients are best served by staying in the hospital overnight such that they can receive training on how to drain their tube in addition to determining an effective pain-control regimen. Finally, it is often valuable to obtain a CT scan following drainage of the fluid with the tube on suction. This will give the best indication of whether or not the disease appears to be a true pleural cancer or if there are pulmonary nodules more suggestive of metastatic disease. Often these nodules will not be visible on the preoperative CT scan because of the presence of fluid and lung compression.

Diagnosis for Patients Presenting with a Pleural Rind

If a patient has a thick pleural rind, then the diagnosis can be obtained without entering the pleural space. Based on the CT scan, a spot along a potential future thoracotomy incision is selected where there appears to be significant pleural tumor thickness. Under general anesthesia, the area is infiltrated with local anesthetic. Depending upon the thickness of the soft tissue between the skin and the target, the incision can be as small as 1 cm or may need to extend to 3 to 4 cm. The soft tissue is incised and the dissection is carried into the underlying interspace where a hard, white mass will be encountered. Using a scalpel, or a biopsy forceps, specimens are harvested. Great care needs to be taken to avoid full-thickness penetration of the tumor and violation of the underlying visceral pleura. Again, it is imperative to confirm the presence of lesional tissue. It would be the exception to have a free pleural space, in the presence of bulky pleural disease and the absence of an effusion, so there is unlikely to be a role for video thoracoscopy in these cases. If additional tissue is required, it is best to harvest tumor along the interspace rather than risk deeper biopsies that may enter the lung. Once the pathologist has confirmed the presence of lesional tissue and a sufficient quantity of specimen to establish a diagnosis has been obtained, the incision is closed after irrigating with sterile water or saline. With the incision filled with liquid, the area is observed during several breaths to assure that the fluid does not drain into the chest. This would indicate the presence of pleural patency and may require placement of a chest tube. The anesthesia team is then requested to deliver a prolonged Valsalva maneuver and the site is inspected for bubbles. If it appears that the lung has been entered, a soft drainage tube should be placed in the bed of the incision and treated like a chest tube. If there are no bubbles and the fluid level is static, then the incision can be closed in layers without any drainage.

INVASIVE STAGING STUDIES

Given the fact that surgery for mesothelioma is investigational and, in reality, palliative, we feel strongly that every effort should be made to rule out any extrathoracic disease. Beyond radiographic studies, we routinely perform invasive staging that includes bronchoscopy, EBUS, and laparoscopy. As indicated by the preoperative imaging, we selectively perform contralateral VATS and, on occasion, upper endoscopy. In our protocols mediastinal nodal involvement is not an exclusion criterion, but any extrathoracic disease does exclude the patient from a surgical approach. Laparoscopy is used to rule out diaphragmatic transgression and/or peritoneal metastases. We have, on several occasions, uncovered occult peritoneal metastases that were documented with cytologic analysis of a saline peritoneal lavage. In addition, we have discovered both false positive and false negative readings on the radiographic interpretation of diaphragmatic invasion. If a patient is suspected of having a fused abdomen, then we will forego the laparoscopy instead obtaining a dedicated abdominal/pelvic CT to complement the PET scan. If these reveal any concerning findings and the patient is committed to pursuing surgery, then we will go as far as performing a laparotomy to rule out abdominal disease prior to proceeding with a radical pleurectomy.

Assuming nothing is discovered on bronchoscopy (or contralateral VATS) the patient is placed in the supine position for the laparoscopy. The abdomen is entered through a 5-mm infraumbilical port. A 30-degree laparoscope is used. After confirming that there was no injury upon entry, a second 5 mm subcostal port is placed on the side of the tumor. At this point the entire peritoneal cavity is explored. The subcostal port is used to provide sufficient visualization of the undersurface of the diaphragm. Any suspicious areas are sampled for histologic analysis. Otherwise, random peritoneal biopsies are then performed. Following this 1 L of sterile saline is instilled, the abdomen is agitated, and the fluid is collected for cytologic analysis.

RADICAL PLEURECTOMY AND EXTRAPLEURAL PNEUMONECTOMY TECHNIQUES—AS PART OF A MULTIMODAL APPROACH

Introduction

There is currently no incontrovertible data to support surgery for mesothelioma, let alone a specific approach. What does

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appear clear is that a macroscopic complete resection is the goal, with residual gross disease after resection almost always translating into a poor oncologic outcome. As a result, if the surgeon elects to proceed with a lung-sparing procedure, it should be a radical pleurectomy with the goal of a macroscopic complete resection. It is commonly held that sparing the lung is an intraoperative decision, based principally upon the extent of disease within the fissure. The author has published evidence dispelling this notion and demonstrating that it is possible to achieve a macroscopic complete resection with lung-sparing surgery in essentially any patient, even those with very bulky disease, extension into the chest wall, and/or extensive involvement of the fissure. The exception is the patient with invasion of the major bronchovascular structures but, in over 100 consecutive cases, with many patients having tumor volumes in excess of 1,000 mL, this has not been encountered. Such a patient, assuming the cancer had selectively invaded the lung but not transgressed the diaphragm and/or mediastinum, would require an extrapleural pneumonectomy.

Whether or not lung-sparing surgery proves equal, inferior, or superior to extrapleural pneumonectomy remains to be established. What is clear, however, is that sparing the lung results in a slight broadening of the candidate pool for surgery-based therapies and, almost certainly, results in a superior quality of life to what would be expected with an extrapleural pneumonectomy.

Positioning and Incision

In addition to the standard preparation for a procedure of this magnitude, it is critical to confirm that any special equipment and/or personnel are available. Examples of such items would be an argon beam coagulator for chest wall cautery and prosthetic patches for pericardial and/or diaphragm reconstruction. Employment of any intraoperative adjuvant therapies, such as photodynamic therapy or heated chemotherapy, requires the requisite additional layers of preparation.

Prior to positioning the patient, a nasogastric tube should be placed as this will facilitate in identification of the esophagus and can also be left in place if it is elected to leave the endotracheal tube in place at the conclusion of the operation. Once it is confirmed that the NG tube is in the stomach, either by aspiration of gastric contents or by intraoperative palpation, it is our practice to instill 100cc of cream spiked with an ampule of methylene blue. This is to allow for the detection of any chyle leak during the operation, and allow for it to be repaired. The patient is then positioned in the appropriate lateral decubitus position and draped in a manner that will allow extension of the incision to the costal margin if necessary. All previous incisions should be marked and, if at all possible, incorporated into, and excised with, the thoracotomy incision.

The previous incisions are incorporated as ellipses. The cutaneous ellipse is grasped in one hand and the deeper portion is palpated. If the biopsy track is palpable, then it is traced—with a margin—into the interspace and can be amputated at the pleural level or left en bloc to be removed with the main specimen. If there is no palpable abnormality, then the ellipse is taken as a full-thickness skin and subcutaneous fat specimen down to muscle.

There is a distinct advantage to minimizing trauma to the chest wall musculature with multiple excisions, particularly if the patient is going to have a pneumonectomy and require watertight closure of the chest wall. Occasionally patients may have had biopsies through one or several incisions that cannot be incorporated into any reasonable incision or which would require a degree of reexcisions that may compromise chest closure. If there is high suspicion of tumor growth in the biopsy sites, then they need to be excised. If there is a question, or if it appears that resecting multiple incision sites could compromise chest closure, then specimens can be sent for frozen section analysis. In these situations we send the skin and subcutaneous tissue from the ellipse, with the question to be answered as to whether what is palpable is scar or tumor. Only if cancer is identified on the frozen section will we then follow the entire region of firmness through the chest wall musculature and remove the intercostal muscle in the region, leaving the skeletonized ribs. These soft tissue resections can, in fact, be quite sizable in order to resect back to normal-appearing tissue. Regardless, all excised tract sites should be marked with surgical clips for radiographic identification, should the final pathology reveal tumor and adjuvant radiation be indicated. Some groups preemptively radiate all incisions after surgery, but this is of unproven benefit at this time.

Our typical approach is to enter the chest through the sixth interspace, sparing and retracting the serratus anterior muscle, but dividing the latissimus dorsi. If there is enough room in the interspace to gain entry to the extrapleural plane then we will leave the seventh rib, typically shingling it posteriorly to provide sufficient exposure and ideally avoid rib fracture. If the hemithorax is contracted, with no appreciable interspaces, then we routinely resect the seventh rib and gain access to the extrapleural plane through the bed of the resected rib. Rarely, the sulcus will be so deep in a large patient that additional room is required. In those situations the seventh rib can be excised, even if it was possible to enter the extrapleural plane without rib resection. Some surgeons advocate elevating the chest wall and creating a flap large enough to make a counterincision in the ninth or tenth interspace. It is likely to result in more postoperative pain, but it is an option to keep in mind for a particularly difficult diaphragm dissection.

Radical Pleurectomy

General Approach and Strategy

Over the years the author has utilized multiple techniques in an attempt to develop a standardized approach to radical pleurectomy. What follows is a description of this procedure in its current iteration. The general strategy that has resulted in the most reproducible results involves mobilizing the entire cancer from the hemithorax, such that it is tethered solely to the lung, and then resecting the entire visceral pleura en bloc with the mobilized cancer. Understanding that every one of these cancers is different and the surgeon must, therefore, be flexible in the approach, the typical order of dissection is bony hemithorax, posterior mediastinum, superior mediastinum, anterior mediastinum, diaphragm, and lung.

Chest wall/posterior-superior mediastinal mobilization: The first step of the operation is to mobilize the tumor off the bony hemithorax, followed by the posterior and superior mediastinum. This initial portion of the operation is the same whether the surgeon is planning to perform a radical pleurectomy or extrapleural pneumonectomy. The extrapleural plane is identified and entered adjacent to the incision. It is developed bluntly as much as possible. Blunt finger dissection, working a broad front, causes cleavage in the correct plane. Sharp dissection is more likely to leave behind gross tumor. The argon beam coagulator is a good device for cauterizing the chest wall, from which capillary oozing can lead to significant blood loss. The chest wall is the safest portion of the operation and is a good chance for the surgeon to get a sense as to how the tumor interacts with the surrounding tissues. As the dissection is carried to the posterior reflection onto

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the posterior mediastinum, the surgeon can follow the intercostal veins from the azygos or hemiazygos veins as they traverse the mediastinum to safely transition from the chest wall to the posterior mediastinum. On the right side, the surgeon must take care not to get behind the esophagus and on the left it is the aorta that must be left in place as the pleura is separated from its medial surface. In the superior mediastinum, it is the subclavian artery on the left and the vena cava on the right of which the surgeon must be mindful. It is often helpful to employ a 10 mm 30 degree video thoracoscope for supplemental vision during the dissection to assure that the correct plane is identified and maintained, especially in the apex of the chest as the thoracic inlet and most superior mediastinum are dissected. On the right side, the azygocaval junction is typically approached both superiorly and posteriorly, having followed the azygos vein. This is an area where a venous injury can occur if the wrong plane is entered.

Anterior mediastinum: The anterior mediastinum is approached by sweeping off all pericardial fat in an anteroposterior direction, starting in the pericardiosternal recess. The surgeon must take care not to breach the anterior pleural reflection and enter the opposite hemithorax. This portion of the operation is highly variable. Occasionally, nearly the entire pericardium is covered with pericardial fat and removing this fat leaves the pericardium with a macroscopic complete resection. More commonly, the pericardial fat dissection gets the surgeon out of the anterior recess, but confronted with most of the pericardium directly involved with the cancer. Rarely, the tumor will separate, leaving normal-appearing pericardium. If that is not the case, then an attempt can be made to separate the layers of the pericardium, leaving the serous pericardium intact and resecting the fibrous pericardium en bloc with the cancer and mediastinal pleura. This is technically challenging, but often possible. If the pericardium is too extensively involved to achieve a macroscopic complete resection, then the surgeon has two options. If it is only a small area, then it can be resected and the surgeon may wish to sew in a prosthetic pericardial patch if there is a concern about potential cardiac herniation. If it is an extensive area, then the entire pericardium can be resected. As the lung is being left in place, the pericardium does not have to be reconstructed unless the surgeon is concerned the patient will be at risk for cardiac torsion. Regardless of approach, the goal remains achieving a macroscopic complete resection. As the dissection is extended posteriorly, ultimately culminating at the anterior hilum of the lung, the phrenic nerve can be identified at the level of the superior mediastinal dissection and skeletonized as it traverses the anterior mediastinum. Whether preserving the nerve and the diaphragm, without a pleura, helps with respiratory function postoperatively is a current area of investigation, but it is intuitively attractive and remains our current approach. It is unusual, even when encased in bulk tumor, to not be able to preserve the nerve.

Diaphragm: The diaphragm dissection is started in the costophrenic recess, attempting to bluntly separate the pleura from the underlying muscle. Rarely the pleura will separate from the underlying bare musculature. Often, however, this requires sharp dissection and is best accomplished with broad-tipped scissors, allowing the scissors to "find" the plane between the hard tumor and the soft underlying normal tissue. Limited areas of full-thickness invasion are resected, leaving only the peritoneum, and the diaphragm can be reconstructed primarily with heavy absorbable sutures. Sometimes, especially if the inseparable tumor is a central island and there is sufficient laxity in the remaining debrided diaphragm, the area can be lifted away from the abdomen and undercut with a thick tissue stapler. Care, obviously, must be taken to assure there are no viscera caught in the stapler and this is readily done by palpation. The staple line must then be oversewn with heavy absorbable sutures as the diaphragmatic muscle, comprising the staple line, can tear and result in a hernia. If there is extensive involvement of the diaphragm then it is resected, as it is during an extrapleural pneumonectomy, and reconstructed with a 2 mm Gore-Tex patch.

Lung: At this point in the dissection the entire tumor is tethered solely to the lung. The anesthesiologist is asked to connect the operative lung to an alternative oxygen supply with an in-line stack of PEEP valves that will allow it to be held under positive pressure ranging from 10 to 30 cm water. The tumor is then sharply incised, extending through the visceral pleura. The plane between the undersurface of the visceral pleura and bare lung parenchyma is then developed. Initially this is best accomplished with a forceps and fine scissors, until at least several millimeters beyond the incision have been liberated. At this point the edge can be better grasped and the bare lung parenchyma can be very gently retracted. Over the years, the instrument that has proved best suited for further developing the plane is a broad Cobb dissector. Initially there is often a torrential air leak when the visceral pleura is removed from the parenchyma but, literally, within minutes the leaks nearly abate if the plane was maintained at the interface of visceral pleura and parenchyma. It is also worth noting that the assistant should be cautioned about gentle suctioning at this point in the operation, as even the most ginger advance with a suction instrument can penetrate into the lung parenchyma when it is devoid of pleura. This will result in air leaks that do not seal very readily. Sometimes a cancer will be encountered where portions or even, rarely, the entire lung will not yield the subpleural plane. In these cases, the electrocautery can be used to open the plane that is visibly evident, but does not yield to simple sharp or blunt dissection.

The critical element in the lung dissection, and what is often considered a contraindication to lung-sparing surgery, is to remove the tumor from the fissures. This is safely accomplished by saving this part of the lung dissection for last. The cancer is tracked down into the fissure, following from both sides. An even level is maintained as the base of the fissure is approached. At this point the lung is deflated and the surgeon will be able to palpate the deepest portion of the fissure of which the cancer has formed a cast. In any patient with complete fissures, this will typically terminate on the surface of the interlobar pulmonary artery. Under direct vision, the veil-like investing tissue over the artery is sharply divided, thereby releasing the cancer from the fissure. This will often result in skeletonization of the pulmonary artery within the fissure (Fig. 29.1). Once the tumor is released in the fissure, the similar investing band of extrapleural tissue can be identified and divided around the hilum, thereby dividing the remaining

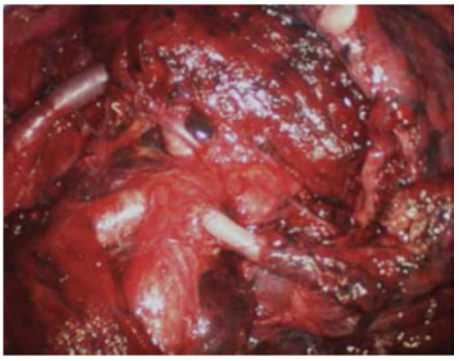


Fig. 29.1. Skeletonized pulmonary artery within the fissure.

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Fig. 29.2. Unified pleural specimen.



Fig. 29.3. Piecemeal pleural specimen.

attachments of the tumor to the patient. In cases where the cancer is pliable, a single large specimen may result (Fig. 29.2). Sometimes, however, the cancer may be so firm that in order to have enough room to work and still preserve the lung, the tumor is better removed piecemeal (Fig. 29.3).

Once the specimen is removed, the chest cavity should be inspected to assure that a macroscopic complete resection has been achieved. The video thoracoscope by providing light and magnification is very useful in helping to inspect all surfaces.

Lymphadenectomy: A thoracic lymphadenectomy is then performed dissecting all standard nodal stations. In addition, the author has been harvesting the posterior intercostal lymph nodes. The significance of these lymph nodes has not been definitively established and is an area of active investigation. For the purpose of our publications and analyses, given the lymph nodes are not described in any current staging schema, we have considered them as N1 lymph nodes as we have had multiple instances where these were the only positive lymph nodes. These nodes are accessed using the electrocautery to incise the posterior interspaces at the level of the rib heads. Often the nodes can then be bluntly delivered with a fingertip, like ejecting a pea from a pod, but sometimes it is necessary to reach into the interspace with a broad-tipped forceps. Care must be taken during this maneuver to avoid avulsing the intercostal vessels as the nodes are closely associated.

Postoperative care: Straight chest tubes are placed anteriorly and posteriorly and directed to the apex. If a fluted tube is not used, then additional holes should be cut to access air and fluid throughout the entire intrathoracic traverse of the tubes. A rongeur is helpful for making extra holes in the chest tubes, taking care to not cut more than a quarter the diameter of the tube to avoid kinking, and making sure the most proximal hole is cut through the radiopaque marker to assure all holes are intrathoracic. A right-angle tube or flexible fluted tube is also placed along the diaphragm, terminating in the posterior costophrenic recess. Depending upon standard criteria, as well as the surgeon's sense of the need for positive pressure to maintain full lung expansion, the patient can either be extubated or remain intubated and on pressure mode ventilation at the end of the operation. There are usually significant endobronchial bloody mucoid secretions present at the conclusion of these procedures; so the patient should undergo a completion toilet bronchoscopy at the conclusion of the operation, and again prior to extubation if the patient remains intubated. If the patient is extubated, then each chest tube should be placed on 20 cm of suction or higher levels if the lung is not fully inflated on the postoperative chest X-ray. There is a premium on achieving full lung expansion using the higher suction since this will, counterintuitively, help the air leaks seal more quickly if visceral to parietal pleural apposition can be achieved. If the patient is left intubated, then usually −10 suction applied to the chest tubes is adequate and can be increased when the patient is extubated. Because of the air leaks, volume calculations on the ventilator are unreliable and the ventilator needs to be adjusted empirically, based upon blood gasses. Typically, patients are hypocarbic, presumably from air being sucked through the lungs like gills. Having all three chest tubes connected to separate collection devices will allow the surgeon to assess leaks and drainage in a more useful manner. If the tubes are still leaking after 2 days, then the suction can be decreased as long as the lung remains fully inflated. Once on water seal, the leaks tend to stop quickly. Tubes can then be removed per routine criteria. Despite the enormity of the initial leaks, persistent air leaks are rare.

Attention to postoperative nutrition and pulmonary toilet are the critical issues postoperatively. Because the patients will have a compromised diaphragm, even the most motivated patients may have trouble clearing their secretions and may require one or several bronchoscopies in the postoperative period. There should be a very low threshold to performing bronchoscopy for secretion management.

Extrapleural Pneumonectomy

Extrapleural pneumonectomy involves the en bloc removal of the parietal pleura, lung, diaphragm, and pericardium, with prosthetic reconstruction of the diaphragm and pericardium. This procedure almost certainly remains the most commonly performed operation for mesothelioma and enjoys both the greatest standardization and largest reported experience. Beyond these advantages over lung-sparing operations, it undoubtedly leaves behind less microscopic disease. It does, however, engender the additional morbidity and quality of life changes that result from pneumonectomy. The author does not currently perform this operation for mesothelioma and would reserve it

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for a patient with epithelial mesothelioma who is a surgical candidate, but has a tumor demonstrating invasion of major bronchial and/or pulmonary vascular structures. The author's preference and confidence in lung-sparing surgery arises from the excellent clinical results that have been obtained using this technique in combination with intraoperative photodynamic therapy, and abandoning hemithoracic radiation as an adjuvant treatment. This intraoperative adjuvant treatment remains the subject of ongoing investigation and despite superior clinical results remains an investigational treatment, just like any other surgery-based approach to treating mesothelioma. Any surgery-based multimodal treatment that involves hemithoracic radiation, or an intraoperative adjuvant that might be injurious to the lung, will require an extrapleural pneumonectomy. Extrapleural pneumonectomy, like radical pleurectomy, should be performed only by surgeons who are comfortable with the operation and postoperative management of these patients and in centers with the appropriate resources and personnel.

The technique for extrapleural pneumonectomy is identical to that for radical pleurectomy for the bony hemithorax and superior and posterior mediastinum. Anteriorly, however, the pericardium is entered, allowing for exploration of the pericardial space and resection of the pericardium, which will ultimately be removed en bloc with the specimen. Once the pericardium is dissected to the level of the diaphragm, the diaphragm resection is commenced.

The diaphragm is then dissected by dividing the costal insertions and entering the plane between the diaphragmatic musculature and the peritoneum. This plane is then developed with blunt dissection, often with sponge sticks, separating the two structures. On the right side, great care should be taken to identify and divide the inferior phrenic veins to avoid avulsing them from the vena cava. On the left side care should be taken to preserve the rim of the diaphragmatic crus that forms the esophageal hiatus so as to avoid abdominal visceral herniation.

Following completion of these maneuvers the specimen is tethered only by the pulmonary hilar structures. On the right side, the pulmonary artery and the superior and inferior pulmonary veins are divided intrapericardially. On the left side, due to its relatively shorter length, the pulmonary artery is divided at its emergence from the pericardium. The veins are divided intrapericardially, same as on the right side. The bronchus, as with any pneumonectomy, should be taken as close to the carina as possible.

The Brigham group, to which much credit must be given for reducing this operation to a science, has a technique for patch reconstruction of the diaphragm that allows for visceral swelling after intraoperative bicavitary heated chemotherapy. Another technique is to fashion and sew in a 2 mm polytetrafluoroethylene (PTFE) patch with heavy nonabsorbable sutures, sewing to the residual rim of diaphragm and pericardium, occasionally encircling a rib if possible. Every effort should be made to make the patch as taut as possible to avoid paradoxical motion during respiration and also to facilitate radiation if it is planned as an adjuvant therapy. If radiation is planned, this should be discussed with the radiation therapist preoperatively as marking the perimeter of the diaphragm, and even the patch itself, with surgical clips may be useful for radiation planning.

The pericardium is reconstructed with a patch, commonly PTFE, and should be fenestrated to avoid tamponade, though these holes seem to seal off and patches are commonly found adherent to the heart, even in the setting of tamponade when the recurrence presents as a malignant pericardial effusion. On the right side it is critical to leave the caval openings generous, bearing in mind that the mediastinum will shift medially when the patient is taken out of lateral decubitus position and caval compression could result. It is the author's practice to be able to introduce one to two fingers through each caval opening.

It is prudent to buttress the bronchial stump after this operation. It is the author's preference to harvest the remaining pericardial fat that typically remains in the pericardiosternal recess. This can be taken as a superiorly based graft and sutured over the stump. Care must be taken not to breach the anterior pleural reflection into the opposite chest. Often these grafts are meager, compared to a standard pericardial fat pad, and must be crafted to achieve sufficient length. If this is not possible, then an intercostal muscle can be harvested, but this further compromises the integrity of the chest wall closure and with these rapidly filling chest cavities the surgeon must be vigilant for a large incisional seroma formation, which can be difficult to manage and disastrous if there is a wound infection. The Brigham group enters the abdomen as part of their intraoperative adjuvant therapy. In this setting omentum, arguably the best buttress, should be harvested to cover the stump.

Depending on the surgeon's routine and the available postoperative support, it is usually best, if extubation criteria are fulfilled, to remove the endotracheal tube in the operating room to avoid positive pressure on the bronchial stump. Another option is to change the double-lumen endotracheal tube to a single-lumen tube and leave the patient intubated overnight. When this option is selected, the patient is maintained on a short-acting sedative, like Propofol, and ventilated on a pressure-limited ventilation mode using the minimal pressures necessary to maintain adequate gas exchange. Unless oxygenation is an issue, PEEP does not need to be used. The patient can then be bronchoscoped the following morning, secretions cleared, and then extubated after sedation is held and pain control is assured. If the patient had a pericardial reconstruction, it is prudent to maintain a supine position and allow for only minimal rolling for the first day. Otherwise, having the patient up and ambulating immediately is critical for pulmonary toilet.

Postoperatively it is the author's practice to leave a single tunneled chest tube in place and connected to a balanced drainage collection system to monitor for bleeding, chyle leak, and to position the mediastinum. A single simple 3-0 monofilament stitch is placed through the center of the chest tube incision and left untied, to be tied when the tube is removed. The Brigham group has a routine for aspirating the pleural space through a red rubber catheter and then removing the tube. In either case, the surgeon must be cognizant and vigilant that both bleeding complications and chyle leak are more common after extrapleural pneumonectomy than a standard pneumonectomy. Even in the absence of a complication the hemithorax often fills much more rapidly after an extrapleural pneumonectomy than a standard pneumonectomy. For all of these reasons, the surgeon must be extra vigilant in following the postoperative chest radiographs and tension hydrothorax should be considered in the differential for a patient with rapid or insidious decompensation. The author tends to leave the tube in place until the drainage is clearly trending downward and, depending upon the size of the patient, is less than several 100 ml/day. This may take several days. Although controversial, it is our practice to maintain patients on antibiotics until the chest tube is removed and to instill a small volume of broad-spectrum antibiotics (such as several drops of Betadine) into the chest cavity just prior to tube removal. Otherwise, the postoperative care of the extrapleural pneumonectomy patient is similar to that of other pneumonectomy patients: adequate pain control, early and frequent ambulation, pulmonary toilet, and maintenance of a

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relative state of dehydration until the threat of postpneumonectomy pulmonary edema has passed.

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EDITOR'S COMMENTS

There are only a little over 3,000 cases of malignant pleural mesothelioma seen in the United States in any given year; thus the average surgeon likely will not see a case. As Friedberg points out this malignancy is best treated in centers that have an interest in the disease and are able to offer the full array of potential treatment options. The surgical procedures that he describes, with the exception of thoracoscopic biopsy, are perhaps best left for those surgeons who have a special interest and expertise in mesothelioma as he does. Prior to embarking on a treatment regimen the diagnosis of mesothelioma must be established with confidence and this usually means that tissue must be obtained as pleural fluid cytology only infrequently is able to yield an answer. Even with a tissue specimen at times differentiating mesothelioma from adenocarcinoma can be challenging. The use of tissue biomarkers may aid in confirming the diagnosis but no one marker has been shown to be the definitive one. An elevated level of serum fibulin-3 has been shown to be highly accurate in distinguishing those patients with malignant mesothelioma from those exposed to asbestos but without mesothelioma and from those with other malignancies or benign causes of pleural effusion. Currently, this serum biomarker has not been proven to be of value in the early diagnosis of malignant mesothelioma or for monitoring patients who have undergone therapy. Other potential biomarker candidates include osteopontin and mesothelin.

Patient selection appears to be the most significant determinant long-term outcome as those patients selected for a surgical-based multimodality approach tend to be those that have the highest likelihood of prolonged survival. These clearly are the "best" patients, that is those who are young enough to withstand a multimodality approach that involves a major operation, those with disease limited to a hemithorax, and those with favorable histology. Those patients who present with mesothelioma with pure epithelial elements are those who have shown long-term survival with resection. The sarcomatoid variants do poorly with any treatment and should rarely be candidates for resection and certainly not outside the confines of a rigorously designed investigational protocol. Attainment of a complete resection with negative margins is perhaps the most important determinant of long-term survival. Friedberg refers to this as removal of all gross macroscopic disease. I am somewhat surprised that Friedberg still considers patients with proven mediastinal nodal involvement as candidates for surgical resection.

As to whether radical pleurectomy can achieve as complete a removal of all gross macroscopic disease as well as extrapleural pneumonectomy remains to be determined in my opinion. I congratulate Dr. Friedberg on his attempts to define the optimal procedure to attempt to remove all gross disease while still retaining the lung. Clearly, if the lung can be saved while still achieving removal of all macroscopic disease the patient benefits by experiencing less morbidity and clearly a lower chance of mortality as compared to extrapleural pneumonectomy. In the best and most experienced hands the occasional perioperative death occurs following extrapleural pneumonectomy though this has clearly been attenuated based on the work done by the Brigham group.

Malignant pleural mesothelioma continues to produce significant therapeutic challenges made all the more difficult by the relative paucity of cases seen in any given year. As mentioned, these patients are best treated in centers that have a particular expertise and interest in the disease.

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30 Mediastinal Lymphadenectomy

Boris Sepesi and David Rice

INTRODUCTION

An adequate pathologic assessment of lymph nodes draining the site of the primary tumor is an important oncologic principle in nearly all solid organ malignancies. Complete histologic evaluation of lymph nodes provides significant prognostic information for overall survival and disease-free survival. Additionally, the overall number of resected nodes has become a quality measure of the adequacy of surgical resection. Pathologic lymph node status (pN) is a crucial component of the tumor, node, and metastasis (TNM) staging system and influences clinical and therapeutic decisions in patients with nonsmall cell lung cancer (NSCLC). Although the survival benefit of lymphadenectomy has not been proven in randomized trials, it is possible that a select group of patients with truly loco regional disease might benefit from a meticulous lymph node dissection beyond the merits such a dissection would have for stage classification. A surgeon's role in this process is irreplaceable, as the performance of lymphadenectomy requires a thorough anatomic knowledge of lymph node basins draining the primary tumor site as well as a technical skill and judgment for safe accomplishment of this task.

ANATOMY AND CLASSIFICATION OF INTRATHORACIC LYMPH NODES

The prognostic importance of the metastases to regional lymph nodes in patients with lung cancer has been recognized for over 50 years. The first comprehensive classification of the thoracic lymph node stations has been developed by Naruke et al.; this Japanese classification was used worldwide for almost four decades. In the 1990s, the American Thoracic Society (ATS) attempted to refine the anatomical descriptions of thoracic lymph nodes and in 1997, Mountain and Dresler published a modification of the ATS lymph node map, which was later implemented into the American Joint Committee on Cancer (AJCC) and Union for International Cancer Control (UICC) Lung Cancer Staging System. While this revised form of lymph node stations has been widely adopted in the United States, it was only sporadically used by the European surgeons, and the Japanese continued to favor the original Naruke's classification. An effort to consolidate worldwide lung cancer staging data began in 1998 by the International Association for the Study of the Lung Cancer (IASLC) with the establishment of the Lung Cancer Staging Project. In this project, data from over 100,000 patients were collected. The discrepancies between Naruke and Mountain–Dresler thoracic lymph node maps became significant during data analysis as some patients with N2 (or stage IIIA) disease according to Mountain–Dresler classification were staged as N1 (or stage II) disease in Naruke classification. Consequently, IASLC published the third detailed anatomic classification of intrathoracic lymph node stations, reconciling the differences between former Japanese and American lymph node maps (Table 30.1). Fourteen discrete lymph node stations have been recognized, which are grouped into the following seven zones: supraclavicular (level 1 nodes), superior mediastinal upper zone (level 2, 3, and 4 nodes), aortic nodes/aortopulmonary zone (level 5 and 6 nodes), inferior mediastinal subcarinal zone (level 7 nodes) and inferior mediastinal lower zone (level 8 and 9 nodes), N1 nodes hilar/interlobar zone (level 10 and 11 nodes), and peripheral zone (level 12, 13, and 14 nodes; Fig. 30.1).

INTRATHORACIC LYMPHATIC FLOW AND METASTATIC SPREAD

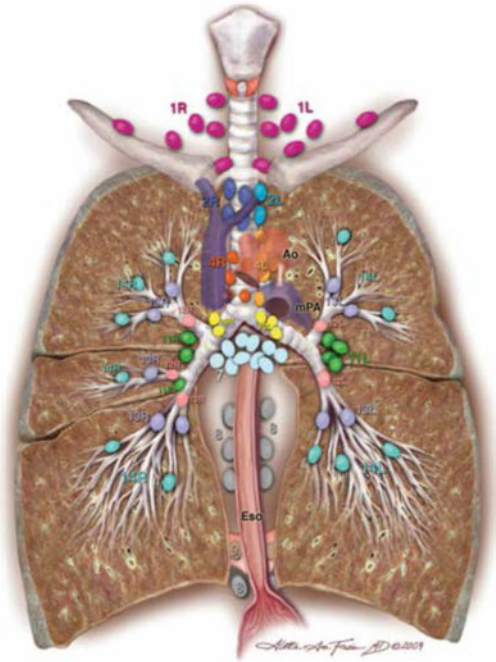
The anatomical description of the mediastinal lymphatic flow may be credited to Riquet et al. who, in a meticulous postmortem dissection utilizing dye injection of the lymphatics, identified intrathoracic lymphatic drainage pathways. The in vivo investigation of the thoracic lymphatic flow was later studied by Hata et al., who generated dynamic lymphoscintigrams by injecting ^{99m}Tc-labeled colloid into the segmental bronchial submucosa. He identified the following lobar and segmental lymphatic drainage patterns; *Right lung*: the apical and posterior segments of the right upper lobe drained via the hilar nodes (level 10), tracheobronchial nodes (level 4), and upper paratracheal lymph nodes (level 2) into the ipsilateral scalene nodes. Drainage from the anterior segment of the right upper lobe varied with approximately 50% of flow via subcarinal (level 7) lymph nodes into the right scalene nodes; occasionally, the lymph flow crossed the midline following the course of the innominate vein into the left scalene lymph nodes. The other half of time the flow from the anterior segment resembled the flow the other segments of the upper lobe. Lymphatic drainage from the middle lobe and the superior segment of the lower lobe resemble the upper lobe drainage ultimately ending in the ipsilateral scalene nodes either via lower paratracheal or subcarinal nodes. Again, in a minority of cases the flow from the middle and lower lobes was noted to cross to the left scalene nodes via subcarinal and left lower paratracheal nodes. *Left lung*: Lymphatic flow from the left lung was variable; however, certain patterns were identified. The apical posterior segment of the left upper lobe drained primarily via the subcarinal lymph nodes along the left vagus nerve to the left scalene nodes or along the recurrent laryngeal to the paratracheal lymph nodes. The anterior and lingular segments drained along the left nerve through the para-aortic nodes (level 5/6) to the ipsilateral scalene lymph nodes. Lymph from the basilar segments flowed via the subcarinal nodes to the pretracheal and contralateral paratracheal lymphatics to the right scalene nodes. Drainage from the superior segment was least constant and occurred via multiple pathways. The study demonstrated mostly consistent ipsilateral lymph drainage along mediastinal lymph node stations with an occasional contralateral drainage.

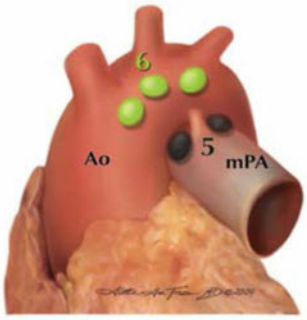
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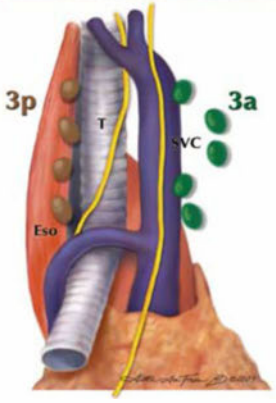
Table 30.1 IASLC Definition of Intrathoracic Nodal Stations

Nodal station	Description	Definition
Level 1 (right and left)	Low cervical, supraclavicular, sternal notch	*Upper border*: lower margin of cricoid cartilage *Lower border*: clavicles bilaterally, in the midline the upper border of the manubrium
Level 2 (left/right)	Upper paratracheal nodes	2R: *Upper border*: apex of the lung and pleural space, midline the upper border of the manubrium *Lower border*: intersection of the caudal margin of innominate vein with the trachea 2L: *Upper border*: apex of the lung and pleural space, midline the upper border of the manubrium *Lower border*: superior border of the aortic arch
Level 3	Prevascular and retrotracheal nodes	3a: Prevascular **On the right** *Upper border*: apex of chest *Lower border*: level of carina *Anterior border*: posterior sternum *Posterior border*: anterior to superior vena cava **On the left** *Upper border*: apex of chest *Lower border*: level of carina *Anterior border*: posterior sternum *Posterior border*: left carotid artery 3p: Retrotracheal *Upper border*: apex of chest *Lower border*: level of carina
Level 4	Lower paratracheal nodes	4R: paratracheal and pretracheal nodes extending, to the left lateral border of the trachea *Upper border*: intersection of caudal margin of innominate vein with the trachea *Lower border*: lower border of azygos vein 4L: left of the left lateral border of the trachea, medial to the ligamentum arteriosum *Upper border*: upper margin of the aortic arch *Lower border*: upper rim of the left main pulmonary artery
Level 5	Subaortic/aortopulmonary nodes	Subaortic nodes lateral to the ligamentum arteriosum *Upper border*: the lower border of the aortic arch *Lower border*: upper rim of the left main pulmonary artery
Level 6	Para-aortic nodes (ascending aorta or phrenic)	Nodes anterior and lateral the ascending aorta and aortic arch *Upper border*: tangential line to the upper aspect of the aortic arch *Lower border*: the lower border of the aortic arch
Level 7	Subcarinal	*Upper border*: the carina *Lower border*: Left: the upper aspect of the lower lobe bronchus on the left, Right: the lower border of the bronchus intermedius
Level 8 (left/right)	Paraesophageal nodes (below the carina)	Nodes adjacent to the wall of the esophagus and to the right or left of the midline excluding subcarinal nodes *Upper border*: Left: the upper border of the lower lobe bronchus Right: lower border of the bronchus intermedius on the right *Lower border*: the diaphragm
Level 9 (left/right)	Pulmonary ligament nodes	Nodes within the pulmonary ligament *Upper border*: the inferior pulmonary vein *Lower border*: the diaphragm
Level 10 (left/right)	Hilar nodes	Nodes immediately adjacent to the mainstem bronchus and hilar vessels including proximal portion of the pulmonary vein and the main pulmonary artery. *Upper border*: Right: the lower aspect of the azygos vein Left: the upper aspect of the pulmonary artery *Lower border*: Interlobar region bilaterally
Level 11	Interlobar nodes	Between the origin of the lobar bronchi
Level 12	Lobar nodes	Adjacent to the lobar bronchi
Level 13	Segmental nodes	Adjacent to the segmental bronchi
Level 14	Subsegmental nodes	Adjacent to the subsegmental bronchi

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Supraclavicular zone
- One Low cervical, supraclavicular, and sternal notch nodes

SUPERIOR MEDIASTINAL NODES

Upper zone
- 2R Upper Paratracheal (right)
- 2L Upper Paratracheal (left)
- 3a Prevascular
- 3p Retrotracheal
- 4R Lower Paratracheal (right)
- 4L Lower Paratracheal (left)

AORTIC NODES

AP zone
- 5 Subaortic
- 6 Para-aortic (ascending aorta or phrenic)

INFERIOR MEDIASTINAL NODES

Subcarinal zone
- 7 Subcarinal

Lower zone
- 8 Paraesophageal (below carina)
- 9 Pulmonary ligament

N1 NODES

Hilar/Interlobar zone
- 10 Hilar
- 11 Interlobar

Peripheral zone
- 12 Lobar
- 13 Segmental
- 14 Subsegmental

Fig. 30.1. IASLC nodal chart with stations and zones. (Reprinted courtesy of the International Association for the Study of Lung Cancer and with permission of Aletta Frazier, MD. Copyright © 2009, 2010 Aletta Ann Frazier, MD.)

A number of authors have studied the pattern of intrathoracic metastatic spread of lung cancer. Borrie initially demonstrated cancer occurrence along the bronchus intermedius on the right side and along the main fissure on the left; these lymphatic sumps are now referred to as the *Sumps of Borrie*. In a study of 359 patients who underwent mediastinoscopy, scalene node biopsy and thoracotomy, Nohl-Oser identified the pattern of regional metastatic spread. Right upper lobe tumors had a propensity to spread to ipsilateral mediastinal nodes (75%); contralateral mediastinal or scalene node involvement was rare in both right upper and lower lobe tumors (<5% to 7%). Left upper lobe tumors, however, had higher incidence of contralateral involvement in mediastinal and scalene nodes (10% to 13%) as did left lower lobe tumors (14% scalene and 25% mediastinal).

Intrathoracic lymphatic involvement of biopsy-proven N2 disease was characterized by Asamura et al. in a study of 166 patients. Right upper and lower lobe tumors had much higher propensity to spread into the pretracheal and ipsilateral paratracheal nodes, whereas right middle lobe tumors had the highest association with subcarinal lymphadenopathy (88%). On the left side, metastatic disease was most consistently found in the aortopulmonary window and para-aortic nodes (levels 5 and 6). Involvement of subcarinal (level 7) nodes occurred in the fifth of patients, most frequently when a tumor involved the lingular segment. Metastatic disease from lower lobe tumors was found with almost equal frequency in the subcarinal and aortopulmonary nodes. Although these studies demonstrate

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frequent orderly ipsilateral cancer spread from the intraparenchymal to hilar to paratracheal lymph node chains, skip metastases may occur in up to one-third of cases. The occasional drainage into the contralateral lymph nodes argues for a vigilant preoperative lymph node evaluation in patients with newly diagnosed lung cancer.

STUDIES CONCERNING LYMPH NODE DISSECTION

Sampling vs. Dissection

As mentioned previously, mediastinal lymphadenectomy is an important component of the surgical treatment of lung cancer. Importance of lymphadenectomy in accurate disease staging is self-evident; however, debate continues whether lymphadenectomy influences survival in lung cancer patients. Additional controversy in the resection and evaluation of mediastinal lymph nodes concerns sampling versus completely dissecting mediastinal lymph node stations. Previously, a randomized trial by Wu et al. demonstrated survival benefit for systematic nodal dissection in patients with resectable NSCLC; however, other randomized trials failed to show significant survival benefit. To further study the effect of mediastinal lymphadenectomy on survival in NSCLC, the American College of Surgery Oncology Group (ACOSOG) performed a multi-institutional prospective randomized trial (ACOSOG Z0030) to study the effect of mediastinal lymph node sampling (MLNS) versus complete lymph node dissection during pulmonary resection in patients with N0 or N1 (less than hilar disease) in NSCLC. The aim of this study was to compare the survival and recurrence pattern between the groups. Over a 5-year period (1999 to 2004), 1,111 patients were accrued into this randomized trial by 102 surgeons in 63 institutions. After excluding 88 patients, final intent-to-treat analysis was performed on 1,023 patients of whom 498 underwent MLNS and 525 who had mediastinal lymph node dissection (MLND). There were no significant demographic differences between the groups in tumor histology, Eastern Cooperative Oncology Group (ECOG) status, type, or completeness of resection. There was no difference in the median number of nodes removed by thoracoscopic technique or thoracotomy (15 vs. 19; $P = 0.17$). More nodes were removed during lobectomy compared with segmentectomy (18 vs. 14; $P = 0.006$). At a median follow-up of 6.5 years, there was no significant difference in the overall survival between the groups ($P = 0.25$). Similarly, the 5-year disease-free survival was no different (68% in MLNS group and 67% in MLND group; $P = 0.89$). Additionally, there were no differences found in local, regional, or distant recurrence between the two treatment arms. Authors concluded that lymph node dissection in and of itself does not improve long-term survival in patients with early stage T1 or T2 or nonhilar N0, N1 NSCLC who have pathologically negative mediastinal, or hilar lymph nodes after rigorous preresection lymph node sampling. It must be emphasized that these data cannot be extrapolated to T3 or T4 tumors, or to patients with N2 lymph node disease. Interestingly, the authors recommended that all patients with resectable NSCLC should undergo MLND, since complete nodal dissection does not increase morbidity or mortality but significantly aids in accurate pathologic staging.

Number of Lymph Nodes Harvested during Lymphadenectomy

To achieve maximum information about a given malignancy, histopathologic evaluation has to contain not only the description of the primary tumor but also the examination of resected lymph nodes. While complete lymphadenectomy is recommended for the treatment of lung cancer, what constitutes complete lymph node dissection is not well defined. The number of harvested lymph nodes has become a measure of the quality of surgery for certain tumors, such as colorectal cancer where a minimum of 12 lymph nodes have to be resected in order for the procedure to be considered adequate. In other malignancies (stomach, esophagus, lung), this number is not as clearly defined. Principally, the more lymph nodes examined the stronger the validity of tumor staging and less potential for stage migration effect. Data regarding the recommended number of harvested lymph nodes in NSCLC are derived either from retrospective studies or from consensus conferences. Current recommendation is for the removal of a minimum of six lymph nodes from mediastinal and hilar stations with at least one subcarinal lymph node. However, some authors recommend at least 10 lymph nodes from three nodal stations.

As part of the ACOSOG Z0030 trail, authors analyzed the yield of examined lymph nodes from each lymph node station, the frequency of obtaining at least one lymph node from a station, and the frequency of occult N2 disease by lymph node sampling and dissection. Lymph node sampling was performed in 555 patients, dissection in 556; final analysis was performed in 498 patients with sampled nodes and 524 with dissected nodes. Overall, a median of 18 lymph nodes were resected per patient for both left- and right-sided cancers. The median number of nodes from N1 stations was 5 for right-sided and 6 for left-sided tumors ($P = 0.134$). A median 3 nodes were harvested from subcarinal (level 7) station and 4 from 4R station. At least six lymph nodes were resected in 99% of patients with 90% of patients having more than 10 nodes resected from three lymph node stations. Four percent ($N = 21$) were found to have occult N2 disease, which was not identified by the rigorous pretreatment nodal sampling. While the majority of nodes were harvested at thoracotomy with only 29 patients having thoracoscopic lymphadenectomy, there was no difference in the number of harvested nodes. As expected, the number of harvested nodes was not associated with tumor type, age, or gender. Most importantly, an increase in N stage (upstaging) was noted with more thorough lymph node harvesting ($P = 0.043$), supporting the concept of systematic lymphadenectomy in accurate staging. The argument for comprehensive lymphadenectomy and thus accurate pathological staging is further underscored by the demonstrated survival benefit of adjuvant chemotherapy in stage I to III patients in the International Adjuvant Lung Cancer Collaborative Group Trial (IALT). Lung Adjuvant Cisplatin Evaluation (LACE), a meta-analysis of five largest randomized trials of adjuvant chemotherapy, also demonstrated improved survival in stage II and III patients.

Lymphadenectomy via Thoracoscopy vs. Thoracotomy

With the increasing use of video-assisted thoracic surgery (VATS), the question arises whether the efficacy of MLND is the same with VATS as with thoracotomy. D'Amico et al. utilized National Comprehensive Cancer Network's NSCLC Database to compare the adequacy of MLND during lobectomy by thoracotomy and VATS with respect to the number and location of all N1 and N2 lymph node stations resected or sampled; at least three lymph node stations had to be sampled or resected to be included in the analysis. The number of N2 lymph node stations and a total number of lymph nodes N1+N2 were also compared. Over a 3-year study period, they identified 4,215 patients with a new diagnosis of NSCLC of which 851 underwent lobectomy for stage I to III

disease. Final analysis was performed on 388 patients of whom 199 had VATS and 189 open lobectomy. There were no significant differences between groups in patients performance status, Charlson comorbidity score, tumor histology, utilization of positron emission tomography, or mediastinoscopy; however, VATS resection was more prevalent in patients with stage I disease ($P = 0.002$). The assessment of mediastinal lymph nodes was equivalent in both groups (median 3; $P = 0.12$). Similarly, the assessment of N2 lymph node stations was similar between thoracotomy (mean 2.91) and thoracoscopy (mean 3.15) groups. Moreover, the total number of N1 + N2 lymph node stations sampled was no different ($P = 0.06$). The authors concluded that mediastinal lymph node assessment of N2 lymph node stations and combined N1 + N2 stations in patients who underwent thoracoscopic lobectomy was as effective as in patients who underwent lobectomy via an open thoracotomy approach. The study did not evaluate other oncologic outcomes such as local, regional, or distal recurrence, or survival. Table 30.2 summarizes studies comparing the effectiveness of nodal harvest utilizing thoracotomy and thoracoscopy.

Survival and Local Recurrence

Whether mediastinal lymphadenectomy improves survival or local recurrence in patients with NSCLC remains an area of controversy. Wright et al. conducted a meta-analysis of randomized controlled trails to evaluate the influence of surgery in NSCLC. A pooled analysis of mortality over the first 4 years was made from three trails. While none of the individual trials found a significant difference in survival between the node dissection and sampling groups, the meta-analysis identified a significant reduction in the risk of death (HR 0.78; $P = 0.005$) in the group who had complete MLND. Ultimately, the definitive answer for this question may be difficult to achieve, since most studies are limited by small numbers of patients, differences in surgical technique, and adjuvant treatment algorithms. The Will Rogers phenomenon (the stage migration effect) is probably the most significant factor influencing patients' survival. The benefit of complete lymphadenectomy likely varies in patients with N0, N1, and N2 lymph node status. Hypothetically, patients with negative nodes (N0) should not derive any benefit from lymphadenectomy, whereas others (N2) may benefit from more accurate staging and recommendations for adjuvant therapy. In patients with N1 disease and limited N2 disease, there is a likelihood that lymphadenectomy not only improves staging accuracy but has also a therapeutic effect.

The theoretical argument that performing less than a complete mediastinal lymphadenectomy during lung cancer surgery might leave cancerous cells behind in nonresected lymph nodes has lead to concerns regarding whether lymph node sampling might lead to higher rates of local recurrence. Lardinois et al. studied the effect of lymph node dissection versus sampling on local recurrence in 100 patients with stages 1 to 3 who underwent R0 resection. Local recurrence was observed in 40% of patients who underwent nodal sampling versus 16% of patients who underwent dissection. Higher local recurrence was seen after sampling in both stage 1 (45% vs. 13%; $P = 0.02$) and stage 2 (55% vs. 17%; $P = 0.09$) patients; stage IIIA patients had nonsignificant difference in local recurrence between the groups. In patients with pathologically node-negative mediastinal nodes (N0 and N1 patients), observed local recurrence was 46% after sampling compared with 13% after dissection ($P = 0.004$). This study, however, was nonrandomized and nodal dissection or sampling was at the discretion of an individual surgeon. The ACOSOG Z0030 randomized trial did not demonstrate increased local recurrence in patients who underwent nodal sampling.

Morbidity and Mortality of Mediastinal Lymphadenectomy

One of the arguments regarding MLNS versus complete nodal dissection involves a potential increase in morbidity and/or mortality caused by MLND. A greater theoretical potential exists for injuring the mediastinal vascular structures, nerves, and tracheobronchial tissue during systematic nodal dissection; this may result in increased chest tube drainage or hospitalization.

The trial ACOSOG Z0030 evaluated morbidity and mortality associated with MLND compared with lymph node sampling of two comparable groups of total 1,100 patients with T1 and T2 disease. The median estimated blood loss was the same between the groups (200 ml); however, the median operative time was 15 minutes longer in the dissection group ($P < 0.0001$), as was the total chest tube drainage (1,459 ml vs. 1,338 ml; $P = 0.056$). These small differences are unlikely to be clinically significant,

Table 30.2 Summary of Trials Comparing Nodal Harvest via Thoracotomy and VATS

Author (year)		*n*	N1 mean #	N2 mean #	N1 # stations	N2 # stations
Sugi (2000)	VATS	48	8.2	13		
	Open	52	8.4	13.4		
Watanabe (2005)	VATS	191	15.2	19.3	–	–
	Open	151	9.8	19.6	–	–
Scott (2010)	VATS	66	15 (N1 and N2)	7 (N1 and N2)		
	Open	686	19 (N1 and N2)	7 (N1 and N2)		
Denlinger (2010)	VATS	79	4.8	2.4*		0.6 (STA 7)*
	Open	464	5.4	3.7		1.2 (STA 7)
D'Amico (2011)	VATS	199	–	4.8 (N1 and N2)	3.2	
	Open	189	–	4.4 (N1 and N2)	2.9	

*$P < 0.05$.

and indeed, there was no difference in either chest tube duration or median length of stay between the groups. Similarly, there was no difference in rates of postoperative atrial arrhythmias, pneumonia, chylothorax, myocardial infarction, recurrent nerve injury, or bronchopleural fistula. Overall, operative mortality was 1.37% and was similar for sampling (2%) compared with dissection (0.76%; $P = 0.157$). This large, prospective, multi-institutional randomized study, which established modern benchmarks for thoracic surgery, effectively disproved the notion that complete mediastinal node dissection was more morbid than nodal sampling.

MEDIASTINAL LYMPH NODE DISSECTION

Right-Sided Lymphadenectomy

The principles of mediastinal lymphadenectomy are the same for both open and thoracoscopic techniques. The goal of right-sided MLND is to remove nodes from stations 2R, 4R, 7R, 8R, and 9R—lymph nodes 10R to 14R are usually removed during lobar resection. Dissection of level 2R and 4R lymph nodes begins with opening of the mediastinal pleura midway between the right border of the superior vena cava (SVC) and the right vagus nerve, extending the incision from the superior aspect of the azygos vein, cephalad, to the level of the right subclavian artery. Lymphatic tissue is grasped and carefully dissected away from the right anterolateral aspect of the trachea taking care to avoid the right recurrent laryngeal nerve as it courses around the right subclavian artery (Fig. 30.2). The nodal packet is then retracted posteriorly and bluntly dissected away from the right side of the SVC. One should be aware of the existence of small venous branches from the posterior aspect of the SVC, which can easily be avulsed and should be divided between clips when present. Full mobilization of the azygos vein enhances the ability to dissect all nodal tissue medial to the azygos–SVC junction. A thorough dissection of the right paratracheal stations involves removal of nodal tissue to the left of the midline of the trachea. Care must be exercised in this regard as it is possible to injure the left recurrent laryngeal nerve if the dissection is overaggressive. After the removal of the level 2 and 4 nodes, dissection may be carried underneath the azygos vein anterior to the right main pulmonary artery and right mainstem bronchus; this allows dissection of the level 10R nodes. The exposure of the subcarinal (level 7) lymph node basin begins with opening the mediastinal pleura from the inferior border of the azygos vein, across the right mainstem bronchus, posterior to the hilum and inferiorly to the level of the inferior pulmonary ligament, which is fully mobilized; the lung is retracted anteriorly (Fig. 30.3). Level 7 nodes are in a close relationship with the inferomedial aspect of the right and left mainstem bronchi of the esophagus, and posterior aspect of the pericardium. The arterial supply to the level 7 nodes comes from the right (usually one) and left (usually 2) bronchial arteries, which usually traverse the anterior aspects of the mainstem bronchus. Dissection usually begins on the inferior border of the lymph nodes allowing the entire packet to be elevated off the pericardium. Posterior retraction of the esophagus allows further development of the plane between the nodal packet and the esophagus. Dissection proceeds along the inferior border of the right main bronchus

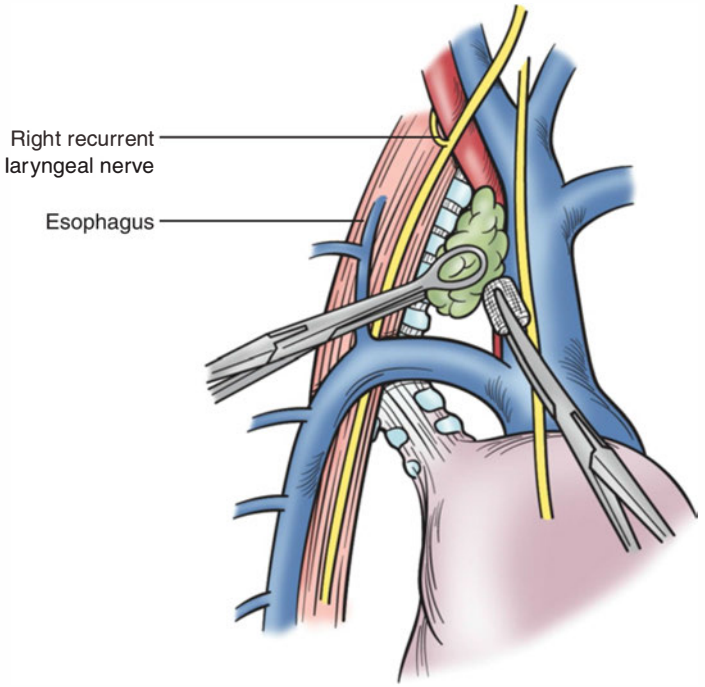


Fig. 30.2. Dissection of right paratracheal nodal packet (stations 2R and 4R).

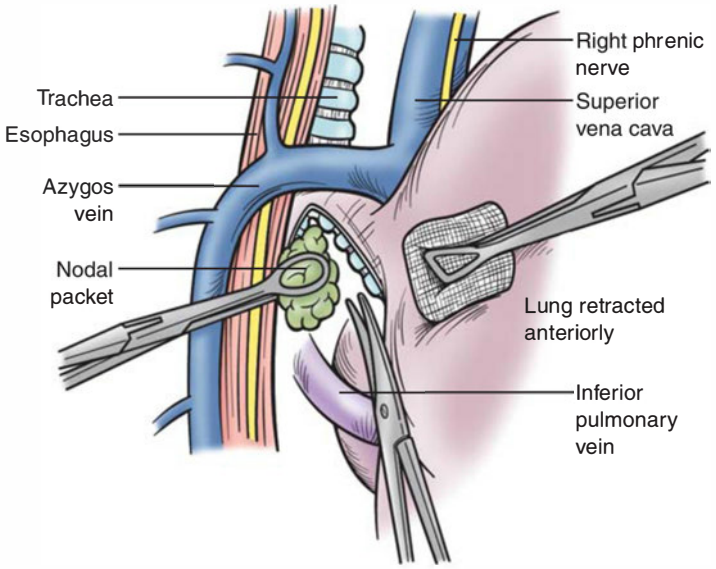


Fig. 30.3. Dissection of the subcarinal (station 7) nodal packet.

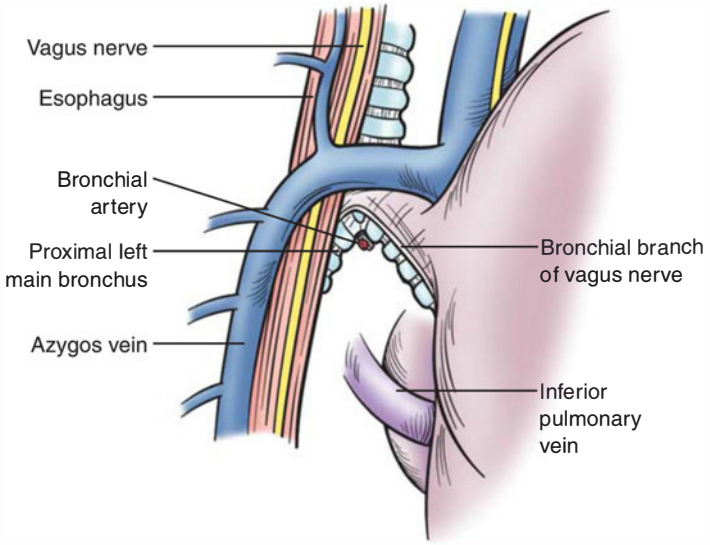


Fig. 30.4. The subcarinal nodes are supplied by branches from the bronchial arteries, which should be clipped during dissection.

to the level of the carina. At this point, the vascular supply to the nodal packet should be searched for and clipped or ligated before dissecting the nodes away from the carina (Fig. 30.4). Grasping lymph nodes with a ring forceps allows retraction of the entire lymphatic chain, which is commonly removed en bloc. Vagal branches to the lung may be divided and usually may transiently stimulate the cough reflex. Further hemostasis may be achieved with gentle packing with or without hemostatic agents such as Surgicel® (Ethicon, Inc., a Johnson & Johnson company, Somerville, NJ) or thrombin with gelfoam. Full mobilization of the inferior pulmonary ligament exposes nodal stations 8 and 9. Level 9 nodes lie within the ligament and are readily removed. Level 8 nodes found in the periesophageal space are likewise dissected free and removed avoiding the injury of the main trunk of the vagus nerve. Nodes anterior to the SVC (3a) and posterior to the trachea (3p) are not routinely removed as part of a right-sided MLND but should be excised if enlarged or suspicious on preoperative imaging studies.

Left-Sided Lymphadenectomy

Left-sided mediastinal lymphadenectomy removes nodal stations 4L, 5L, 6L, 7L, 8L, and 9L. Starting just below the thoracic inlet, the mediastinal pleura is incised posteriorly to the left subclavian artery. The subclavian artery is retracted anteriorly to expose the trachea. Left paratracheal lymph nodes (level 2) are grasped with ring forceps and dissected free. Care is taken to preserve the left vagus nerve running medial to the subclavian artery with the left common carotid artery and descending anteriorly onto the aortic arch below, which the recurrent laryngeal nerve branches and ascends in the tracheoesophageal groove superiorly (Fig. 30.5). Next, with the lung retracted inferiorly, the mediastinal pleura anterior to the aortic arch and parallel to the vagus nerve is incised superiorly exposing the level 5 nodal basin posterior to the ligamentum arteriosum between the aortic arch and left pulmonary artery. While keeping the left recurrent laryngeal nerve protected, these nodes are grasped and dissected free. Level 6 nodes, anterior to the ligamentum arteriosum and medial to the vagus and recurrent nerve, are then swept superiorly with the periaortic fibrolymphatic tissue. Removal of level 5 and 6 nodes allows the superior retraction of the aortic arch and inferior retraction of the left main pulmonary artery exposing the inferior aspect of the trachea; this facilitates dissection of the level 4 lower paratracheal lymph nodes. With esophagus retracted posteriorly and lung anteriorly, the subcarinal space is exposed by extending the pleural incision from the inferior border of the aortic arch to the inferior pulmonary vein, posterior to the hilum. Subcarinal level 7 nodes are removed beginning along the medial border of the left mainstem bronchus continuing inferiorly and crossing the medial aspect of the right mainstem bronchus. Lymphatic tissue is dissected free from the pericardium posteriorly. At the level of the carina, bronchial arteries are ligated and further hemostasis is obtained with judicious use of cautery or gentle temporary packing. Inferiorly, the left pulmonary ligament is incised and level 9 nodes are readily ligated together with level 8 paraesophageal lymph nodes.

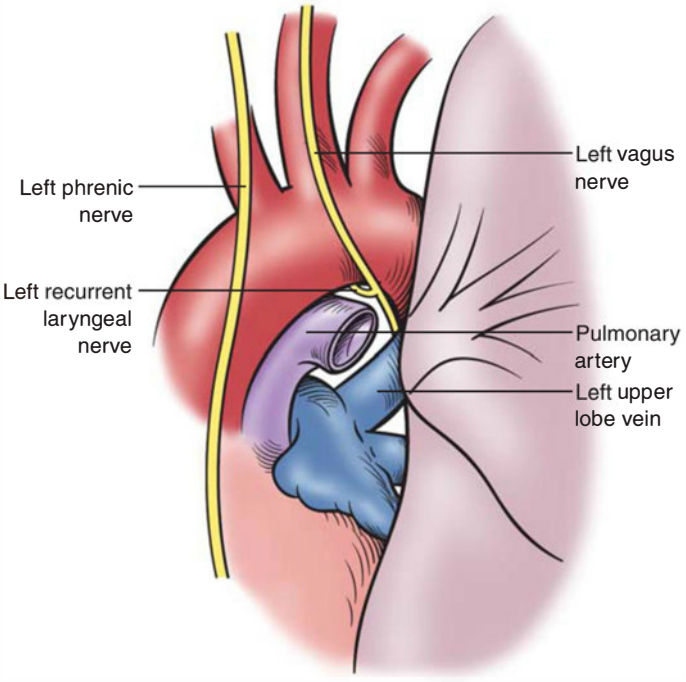


Fig. 30.5. Anatomic relationships of the left phrenic nerve, left vagus, and recurrent laryngeal nerves to the left main pulmonary artery and aortic arch.

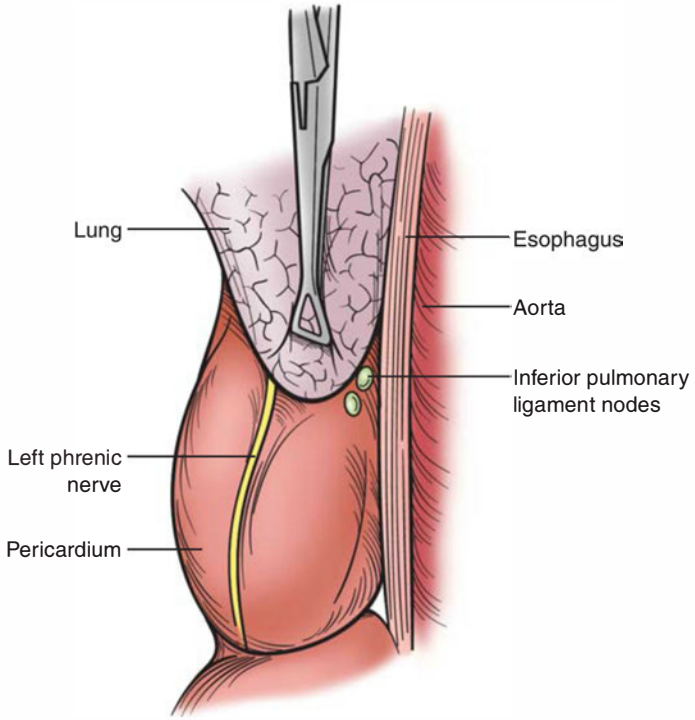


Fig. 30.6. Dissection of left inferior pulmonary ligament nodes (station 9L) and left paraesophageal nodes (station 8L).

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EDITOR'S COMMENTS

Though the procedure itself may not be therapeutic, there is no question that a systematic mediastinal lymph node dissection as part of any operation for non-small cell lung cancer provides the optimal staging information. With no additional morbidity resulting from mediastinal node dissection, there really is no reason simply to perform nodal sampling. The systematic dissection requires little additional effort and results only in a couple of minutes of additional operating room time. And for certain stage disease node dissection ultimately may prove to be of therapeutic value. The authors have done a beautiful job of laying out the data supporting mediastinal lymph node staging and describing how the procedure should be accomplished. A lobectomy without complete lymph node staging to include both N1 and N2 nodes is an incomplete operation and leaves many questions unanswered. Despite that knowledge, there are still many pulmonary resections performed today that do not include assessment of the mediastinal lymph nodes.

Mediastinal lymph node dissection on the right side results in the removal of a nicely contained nodal packet since the superior mediastinal compartment, bordered by the subclavian artery superiorly, the trachea posteriorly, the superior vena cava anteriorly, and the azygos vein inferiorly, is so nicely defined. The same cannot be said for the left side where the level 5 and 6 nodes are removed separately in addition to the level 2, 4, and 7 nodes. It is actually easier, and probably more effective, to assess the left paratracheal and tracheobronchial angle nodes with mediastinoscopy. Approaching the left paratracheal lymph nodes at thoracotomy requires dissection in the aortopulmonary window potentially putting the left recurrent nerve at risk of injury. It should also be noted that a mediastinoscopy does not obviate the need to remove other nodes from the left side or to complete a lymph node dissection on the right.

To achieve the most accurate staging information, it is critically important for the surgeon to precisely label lymph nodes so the pathology report may be correctly interpreted. I find it optimal to label nodes according to the level based on the IASLC map so there is no question as to the location or whether or not a given node is N1 or N2.

LRK

31 Surgical Management of Empyema

Naveed Z. Alam and Raja M. Flores

Empyema, or empyema thoracis, can be defined as a purulent fluid collection in the pleural space. The etiology is varied and includes contamination from adjacent organs, most commonly the lungs, or direct inoculation by trauma or iatrogenic interventions. It is an ancient disease that often requires therapy that harkens back to ancient times.

HISTORY

Empyema thoracis was first described in the 5th century BC by Hippocrates. He wrote that the most common cause was the entrance of foreign bodies into the lungs, either by inhalation or drinking. They may also be caused by parapneumonic effusions if the chest is not cleared by expectoration within 14 days. Symptoms were described as fever, rigor, and thoracic pain and signs included copious sweating, cough, red eyes, bent nails of the hands, and anorexia. On physical examination, Hippocratic physicians asked patients to sit on a chair and then shook them. By placing their ears to the chest wall (immediate auscultation), they could hear a typical rippling sound, similar to that produced by a bottle half-filled with fluid. If no sound was heard, the empyema was noted on the side of the chest that was more swollen and painful.

Management was conservative initially, with the application of plant roots and honey as well as physiotherapy in the form of warm baths and inhalation of vapors with subsequent shaking of the patient to achieve eruption and expectoration of the pus. If this failed, then surgery was pursued. "First, cut the skin between the ribs with a bellied scalpel; then wrap a lancet with a piece of cloth, leaving the point of the blade exposed a length equal to the nail of your thumb, and insert it. When you have removed as much pus as you think appropriate, plug the wound with a tent of raw linen, and tie it with a cord; draw off the pus once a day; on the tenth day, draw all the pus, and plug the wound with linen. Then make an infusion of warm wine and oil with a tube, in order that the lung, accustomed to being soaked in pus, will not be suddenly dried out. When the pus is thin like water, sticky when touched with a finger, and small in amount, insert a hollow tin drainage tube. When the cavity is completely dried out, cut off the tube little by little, and let the ulcer unite before you remove the tube (Papavramidou)."

This remarkable treatise on the management of empyema with open tube drainage as its hallmark defined the standard of care for the disease for more than two millennia. During the world influenza pandemic of 1918 to 1919, there was a crisis. Draftees in crowded military camps were particularly susceptible to the streptococcal pneumonia and empyema that often accompanied influenza. A survey by the Surgeon General of the Army found a mortality rate of 30% for empyema drainage at base hospitals with deaths frequently occurring within 30 minutes of the procedure. This prompted the appointment of Major Evarts A. Graham to head a commission to study the problem. Dr. Graham cautioned against early drainage due to the risk of open pneumothorax and advocated operation only after the development of frank pus as this is usually accompanied by an abscess isolated from the remaining pleural cavity. He also recommended closed drainage rather than open drainage. Adoption of these changes to practice decreased the mortality rate to close to 5%.

CLASSIFICATION

Empyema follows a natural progression that has been classified by the American Thoracic Society into three distinct stages. In the exudative stage, or acute phase, the inflamed pleural membranes swell and discharge a thin effusion with associated bacterial contamination. The fluid is clear and has a low cell count. As the disease progresses to the fibrinopurulent stage, or transitional phase, deposition of fibrin occurs resulting in loculation formation and turbid, or frankly purulent, fluid. In the final stage, the organizing or chronic phase, in-growth of fibroblasts, and associated collagen fibers render the lung trapped and relatively functionless.

ETIOLOGY

As mentioned above, empyemas generally result from either contamination from a contiguous septic organ or direct inoculation during trauma or iatrogenic maneuvers. The most common cause is parapneumonic. Other contiguous sources include esophageal ruptures, deep cervical abscesses, paraspinal infections, and subphrenic collections. Rarely, hematogenous spread from distant sources can result in empyema, particularly in the immunocompromised population.

Post traumatic empyemas are associated with either penetrating injuries or the presence of a hemothorax. As the hemothorax represents an ideal growth medium for bacteria, any instrumentation of the chest cavity, particularly in less than ideal conditions, increases the risk of development of an empyema. Chest tubes inserted in nonsterile conditions, multiple reinsertions of chest tubes, excessive manipulations of chest tubes, residual blood from incompletely evacuated hemothoraces, and prolonged drainage are factors all associated with the development of empyema. Other factors in blunt trauma associated with empyema include the presence of a hemopneumothorax, suggestive of parenchymal injury and ongoing bacterial contamination of the pleural space.

Iatrogenic causes of empyema range from minor interventions, such as needle biopsies and thoracentesis, to postoperative occurrence following lung or esophageal surgery. The management of a postpneumonectomy empyema with or without associated bronchopleural fistula is particularly challenging.

BACTERIOLOGY

The spectrum of causative organisms of empyema varies somewhat with geography; hence, local data are often required to guide

therapy. In developed countries, the most common organisms isolated in adult community acquired empyema are *Streptococcus milleri*, *Streptococcus pneumoniae*, and anaerobes. In hospital-acquired empyema, *Streptococcus aureus*, including methicillin-resistant *S. aureus* (MRSA), is the most commonly isolated organism. In pediatric empyema, the most common organism is *S. pneumoniae*.

COMPLICATIONS

The complications of empyema occur with increasing likelihood as the disease progresses, with the majority developing in the chronic or organizing stage. Pulmonary fibrosis due to increased scar tissue within the lung and pleura can develop. Similarly, scarring of the chest wall and intercostal muscles can lead to contraction. Complications associated with local drainage may occur as well as the infection follows a path of least resistance to suppuration. Contiguous contamination can result in a myriad of complications. In the pursuit of the path of least resistance to suppuration, patients can develop empyema necessitatis with drainage through the skin. Alternatively, the infection can travel to the lung parenchyma, resulting in bronchopleural fistulae. Similarly, mediastinitis, pericarditis, subphrenic abscesses, and osteomyelitis of the ribs or vertebrae can develop.

RADIOLOGY

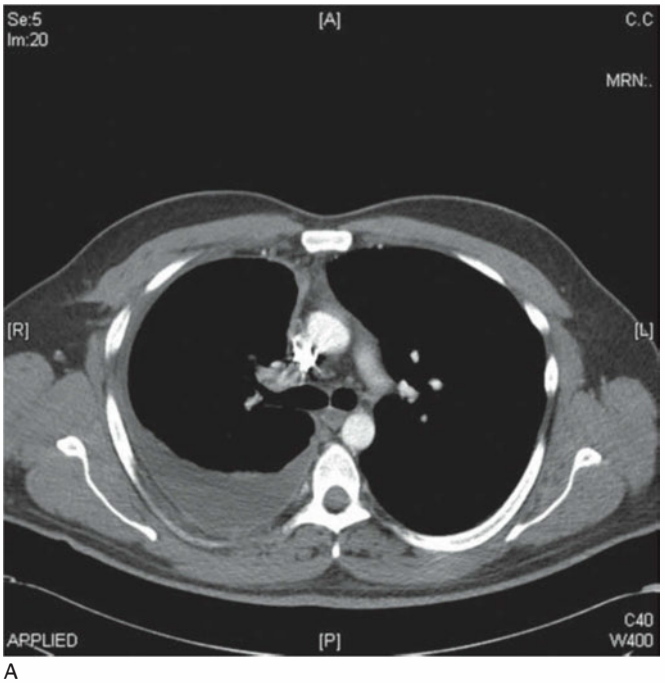
Radiological investigations facilitate diagnosis, enable planning of therapeutic approaches, and allow monitoring of progress. Ultrasound of the thoracic cavity can demonstrate septations and loculations and assist in making thoracentesis safer. Chest radiography is the first step in the evaluation of any suspected pleural pathology. Computed tomography (CT) is also very valuable in patients who do not have simple effusions. It permits visualization of the precise location of loculations, which can be helpful at the time of surgery, particularly with minimally invasive approaches.

MANAGEMENT

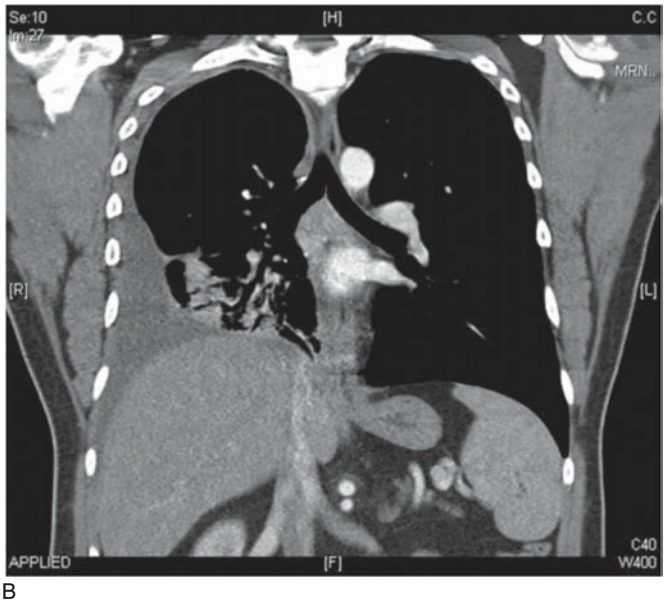
The goals in the management of empyema are to evacuate the infected material and insure re-expansion of the lung parenchyma with no residual space. The thoracic surgical adage, "No space, no problem" is particularly helpful to keep in mind in managing this disease process. A residual space will serve as a nidus of infection; hence, it needs to be obliterated. The earlier the intervention in the course of the disease process, the less invasive it needs to be. In the acute or exudative phase, simple drainage with even small-bore catheters along with appropriate antibiotic therapy will often be adequate. As the disease progresses to the fibrinopurulent or transitional phase, the development of loculations and adhesions makes complete drainage with simple small-bore catheters less likely to be successful. Figure 31.1A and 31.1B demonstrates a transitional phase empyema. There are no apparent loculations, but the CT images suggest that there is some sequestration of fluid and early fibrin deposition. In this setting, larger tubes or concurrent administration of fibrinolytics may be helpful, particularly in patients who are too unwell to tolerate general anesthesia.

The location of the empyema also plays a role. Collections that are posterior or paravertebral are particularly difficult to drain adequately via the percutaneous route. The narrow size of the intercostal spaces in the paravertebral location makes the placement of large-bore thoracostomy tubes difficult and uncomfortable. Similarly, collections adjacent to the mediastinum are also difficult to address percutaneously, as seen in Figure 31.2. Often these patients are better served by early video-assisted thoracoscopic surgery (VATS) drainage as are those whose loculations are so complex that adequate drainage is not able to be performed percutaneously (Fig. 31.3A and 31.3B).

The chain of intervention from least to most invasive then proceeds from small-bore catheters to chest tubes to VATS evacuation and decortication to thoracotomy for decortication to open window thoracostomy.



A



B

Fig. 31.1. (**A**) Axial computed tomographic mediastinal windows. Heterogenous density of fluid, suggestion of fibrin deposition. (**B**) Coronal computed tomographic mediastinal windows. No rind visible on visceral pleura.

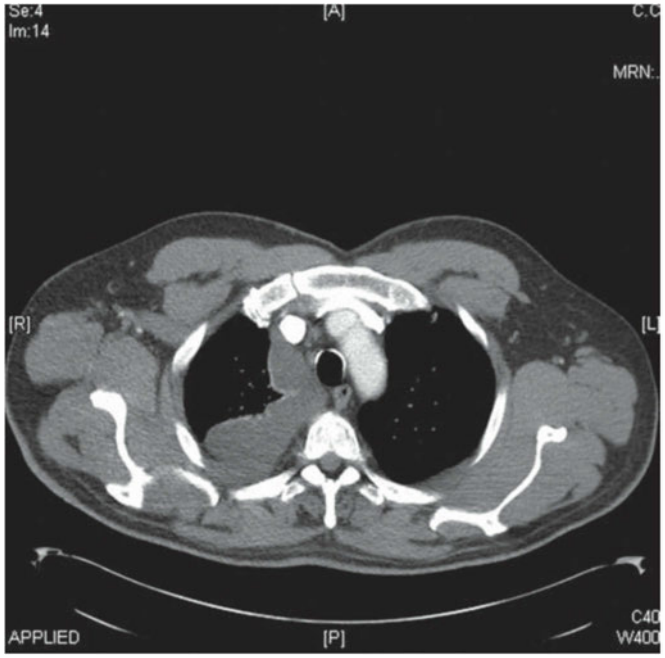


Fig. 31.2. Axial computed tomographic mediastinal windows. Medial collection with an impression of rind on the visceral pleura.

TUBE THORACOSTOMY

When considering chest tube or intercostal catheter placement, the first principles of drainage should be adhered to, that is, that the drainage be prolonged, adequate, and dependent. There is some controversy in terms of the size of catheter that should be used for initial management of empyema. Frankly, this discussion generally takes place among respiratory physicians or pulmonologists, but it behooves the surgeon to be aware of the research that is taking place in this forum. Although there is no randomized data comparing small-bore to large-bore catheters in the management of loculated empyema, results from the Multicenter Intrapleural Streptokinase Trials (MIST) did collect data on the size of the catheters placed. They reported no significant difference in the frequency of surgery or the mortality based on the size of the catheter placed (from <10F to >20F). It should be noted that the choice of size of the catheter placed was at the discretion of the treating physician and that those that had the largest tubes placed had more frank pus and higher lactate dehydrogenase levels.

With increasing numbers of small-bore catheters being placed by interventional radiologists, and often with multiple catheters being used to evacuate separate loculations, it may be that improvements in the placement of the small-bore catheters are rendering them as good as large-bore catheters and the trend is for many physicians to advocate their usage. It makes intuitive sense that multiple well-placed small catheters can manage the empyemas that would have been managed with a large-bore catheter. And as demonstrated in the MIST study, there is no change in the number of patients that still require surgical intervention.

FIBRINOLYTICS

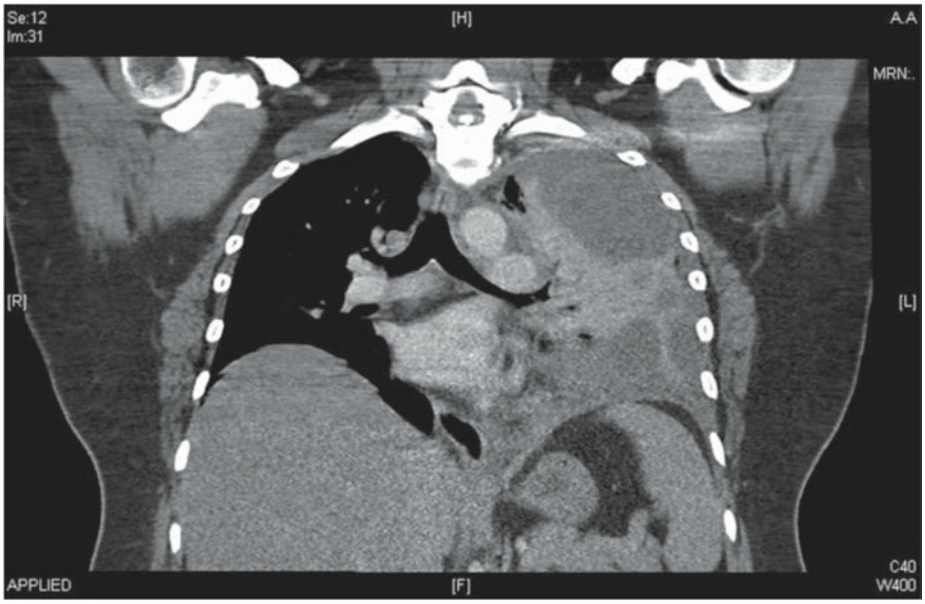
As noted above, the MIST trial was a double-blind randomized study of 430 patients. It showed that the administration of intrapleural streptokinase did not reduce mortality or the need for surgical salvage. This evidence for lack of benefit has led to other studies using other agents. The most recent reports focus on the concurrent administration of tissue plasminogen activator (t-PA) and DNase used together. A randomized trial reported that concurrent use of these two agents decreased the surgical referrals and the hospital stay as compared with placebo. This may influence the future management of early empyema as this combination of agents becomes more readily available.

DECORTICATION—OPEN VS VIDEO-ASSISTED THORACOSCOPIC SURGERY

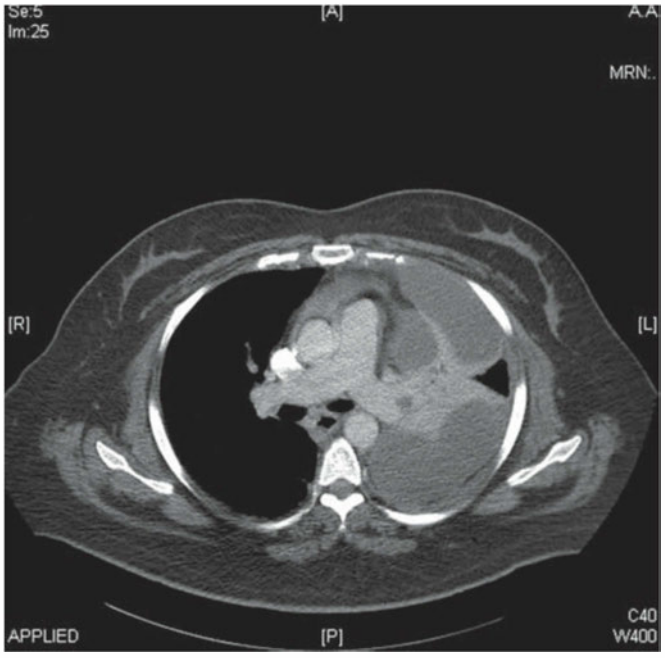
If simple drainage and antibiotic therapy fail to control sepsis, then surgical intervention is warranted. The choice of operation depends on the stage of the empyema, the underlying condition of the patient, and the competencies of the surgeon. The goals of the operation remain evacuation of infected material and obliteration of the pleural space by rendering the visceral and parietal pleural layers into apposition. A decortication consists of removing the infected material and freeing the lung parenchyma from the thick rind that often develops on the visceral pleural surface. This latter aspect of the operation is critical. Failure to re-expand the lung to fill the thoracic cavity will almost certainly result in persistent or recurrent sepsis. In addition, decortication always results in air leak from damage to the lung parenchyma when establishing and remaining in the correct plane to separate the rind from the visceral pleura. With complete lung expansion and chest tube drainage, the air leak will resolve in short order with minimal cause for concern. However, air leak in the setting of a persistent space will perpetuate the air leak and further contributes to ongoing sepsis and likely will mandate re-operation.

The surgical principles of decortication remain the same, regardless of approach (open or VATS). The infected material needs to be evacuated, the adhesions of the lung to the chest wall, mediastinum, and diaphragm need to be taken down, and the underlying lung parenchyma needs to have the thickened pleural rind removed enabling expansion of the lung, allowing it to fill the hemithorax and obliterate any residual space. The last steps should be copious irrigation before careful placement of drainage tubes. These steps were traditionally performed through an open procedure. At thoracotomy, intermittent ventilation of the operative side facilitates decortication as the inflated lung parenchyma provides gentle countertraction to the surgeon's instruments. A combination of blunt and sharp dissection is required. We have found the use of "peanuts" and Cobb elevators to be very useful in establishing the appropriate plane of dissection. As a last resort, criss-crossed incisions, as seen on a well-cured and smoked ham, can be made into the thickened visceral pleura to enable lung expansion.

In the early days of VATS, many thoracic surgeons felt that a true decortication could not adequately be performed by this method if there was a thick rind trapping the lung. With increasing experience in VATS techniques, 30-degree cameras, and cooperation with our anesthesiology colleagues, many surgeons now perform the vast majority of their decortications via VATS technique. The advantages are clear.



A



B

Fig. 31.3. (A) Coronal computed tomographic mediastinal windows. Multiple complex loculations on all surfaces. **(B)** Axial computed tomographic mediastinal windows. Heterogenous fluid density suggesting fibrin deposition.

These patients are often compromised and the recovery from VATS is clearly more rapid than with a thoracotomy. The main disadvantage is that the operation is often tedious and time-consuming. While generally performing VATS without lung isolation is impossible, it is the very rind that traps the lung that enables VATS decortications to be performed with the aid of intermittent ventilation of the operated side. While the lung is being ventilated, gentle blunt dissection with endoscopic elevators and "peanuts" or sharp dissection with scissors enables the removal of the peel. As in open surgery, key maneuvers include dissecting the lower lobe completely free of the diaphragm and dividing all adhesions circumferentially. Irrigation can often be performed early in the operation to facilitate exposure. Mechanical pulse lavage systems can be helpful in this regard.

In a comprehensive review comparing VATS to open decortication and to chest tube drainage alone for empyema, the authors evaluated 68 articles and chose 14 as providing best evidence for practice. They found that VATS offers superior outcomes to chest tube drainage alone and equivalent outcomes to open surgery. Moreover, they found that VATS decreases hospital stay, postoperative complications, and patient morbidity as compared with thoracotomy. The suggestion was that VATS should be considered before thoracotomy in patients requiring decortication.

SPECIAL CIRCUMSTANCES

Patients who have failed conservative management but are too unwell to tolerate general anesthesia present unique challenges. In these patients, long-term drainage needs to be established. One option is a well-placed large-bore chest tube. If the tube has been in place for 1 to 2 weeks, the lung should be fixed in position to the chest wall and that enables conversion of the closed drainage system to open drainage. Gradual removal of the chest tube, 1 to 2 cm a week, can be performed with the resulting cavity in the chest closing around the tube. In certain settings, a rib resection can be performed to enhance placement and long-term comfort of these empyema tubes. Another option is to perform an open-window thoracostomy by removing multiple ribs in the posterior and dependent portion of the hemithorax. Again, this can only be performed when the surrounding lung has adhered to the chest wall and is not in danger of collapse but this adherence is facilitated by the inflammatory process that results from the intrapleural infection.

In the post lung resection setting or in any situation where a residual space in the hemithorax exists due to underlying structural lung disease, it is important to achieve the goal of space obliteration. This can be accomplished by transposition of vascularized tissue into the thoracic cavity. Various chest wall muscles have been used for this depending on the location of the space in the hemithorax and the prior surgery performed. Serratus anterior, latissimus dorsi, and pectoralis major are good candidate muscles as long as they can be mobilized on their pedicles to the appropriate area. Greater omentum has also been reported with success when transposed through the esophageal hiatus or an enlarged foramen of Morgagni.

SUMMARY

Empyema thoracis has been well described for millennia. In addition to antibiotic and supportive management, the treatment

principles are twofold—evacuation of infected material and re-expansion of the lung with complete obliteration of any residual space. In early stages, this can often be accomplished by tube thoracostomy alone. In later stages, this surgical intervention likely will be required.

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EDITOR'S COMMENTS

If there is one take-away message from this chapter it must be that in matters of the chest, space is the enemy. Especially in the situation of an empyema not only must the infected fluid or the pus be evacuated but any residual space must be obliterated or surely infection will recur. Draining an infected collection early potentially avoids the need for more invasive procedures as the authors point out. If recognized early, it is perfectly appropriate to place small pigtail catheters under radiographic guidance as long as they affect complete drainage. In these early infected collections ordinarily, there is no problem with complete lung re-expansion that adequately fills the pleural space. For the more organized collections where thick pus is present, a large bore thoracostomy tube clearly is the treatment of choice but the tube must be positioned so that complete drainage occurs. Depending on the appearance on the chest radiograph "blind" chest tube insertion may be less valuable than proceeding with a VATS drainage procedure with precise tube placement.

The more difficult problem is the management of a residual space with incomplete lung expansion and making the decision as to whether to proceed with conversion to open drainage or operation with decortication. This really depends on the radiographic appearance and the duration of the infection as well as the patient's overall condition. Once the lung is "stuck," which occurs fairly rapidly in the face of an intrapleural infection, the chest tube may be disconnected from water seal drainage, a gauze pad applied to the end of the now-open tube, and a radiograph obtained to assure that the lung has not "dropped." If indeed the lung stays up, then the tube may be shortened and, as the authors point out, advanced over a period of time allowing the space to close over the tube. This can be a prolonged, uncomfortable process with the patient having to return for a number of outpatient visits. The open drainage tube needs to be in a dependent position. Open drainage may also be facilitated by an open window thoracostomy as long as the "window" is placed in a dependent location. This obviates the need for an uncomfortable tube sitting between ribs. If the open window is obstructed by the scapula, there is a tendency for it to close prematurely. If the optimal location for the window is near the scapula, we have resected a portion of the scapula to prevent it from interfering with the window. We also have not hesitated to go back and revise the window if it appears to be closing prematurely.

If the lung appears entrapped by a visceral pleural peel, then a decortication to allow for complete lung re-expansion is required. Whether one does this with a VATS approach or an open thoracotomy, the key is to completely remove the peel and to do this requires entering the appropriate plane between the peel and the visceral pleura. To do this most effectively, the peel has to be "mature," that is, fibrous and not simply fibrinopurulent. Entering the decortication plane is not easy; I use a 15-scalpel blade with the lung expanded to provide counter traction. Care must be taken to go deep enough, but not too deep as to enter lung parenchyma. Using a combination of sharp and blunt dissection and maintaining a broad sweep, the peel is gradually removed. We trim off the peel as it is freed up so that we are not working in a deep hole under a "hood." The areas of difficulty tend to be within the fissures. The lower lobe must be freed off the diaphragm as the authors point out. At times, the peel is so thin and so adherent that the only option is the "ham" the lung, which is creating cross hatching along the entire lung surface with cuts deep enough but not so deep that they enter the lung parenchyma. As in most things in life, in pleural infection timing is critical. The timing of initial drainage, the timing as to the duration of drainage, and the timing of further interventions, if needed.

LRK

32 Resection of Superior Sulcus Tumors

Christine Lau and G. Alexander Patterson

The term "superior sulcus" is really a misnomer in that there is no specific anatomic structure that corresponds but the term has come to refer to those primary lung tumors that occur in the apex of the lung that involve the chest wall and often other structures that reside in the thoracic outlet. These lesions can present with pain in the nerve distribution of the eighth cervical and first and second thoracic nerve roots. In addition, Horner syndrome may be present as a result of involvement of the stellate ganglion of the sympathetic chain. Such patients have the classic presentation described by Pancoast–Tobias syndrome. Patients with C8–T1 nerve root involvement may also present with typical neurologic findings of the "ulnar hand." However, the majority of patients with apical lung tumors present with nondescript but persistent shoulder or upper chest pain. Many of these patients are seen by orthopedic surgeons or chiropractors and are treated as if the problem is strictly a tendonitis or other orthopedic problem, and the only imaging studies obtained are shoulder films that when viewed in retrospect clearly show the lesion. It is not uncommon for patients to be treated for many months prior to a correct diagnosis being made. Posteroanterior and lateral chest radiograph may demonstrate nothing more than apical pleural thickening. Computed tomography (CT) and magnetic resonance imaging (MRI) usually depict the lesion clearly.

ANATOMIC CONSIDERATIONS

The insertion of the anterior, middle, and posterior scalenus muscles on the first and second ribs, respectively, divides the thoracic inlet into three compartments (Fig. 32.1). The anterior compartment contains the platysma and sternocleidomastoid muscles, the external and anterior jugular veins, the inferior belly of the omohyoid muscle, the subclavian and internal jugular veins and their major branches, and the scalene fat pad. The middle compartment includes the anterior scalene muscle with the phrenic nerve lying on its anterior aspect, the subclavian artery with its primary branches except the posterior scapular artery, the trunks of the brachial plexus, and the middle scalene muscle. Finally, the posterior compartment, which lies posterior to the middle scalene muscle, includes the long thoracic and external branch of the spinal accessory nerves, the posterior scapular artery, the sympathetic chain and stellate ganglion, vertebral bodies, intervertebral foramina, and intercostal nerves.

CLINICAL CONSIDERATIONS

The anatomic location and the pattern of invasion of the apical lung lesion determine the presenting symptoms and signs. Anterior apical tumors generally present with chest wall or shoulder pain. Hand or arm swelling suggests subclavian vein invasion on the left or brachiocephalic invasion on the right. The phrenic nerve may be involved as it crosses the scalenus anticus muscle. These anterior lesions usually do not involve the brachial plexus.

Tumors invading the middle compartment of the thoracic inlet may present with signs and symptoms related to compression or infiltration of the middle and lower trunks of the brachial plexus, which manifest clinically as pain radiating to the shoulder and upper extremity. Often these tumors spread along the fibers of the middle scalene muscle.

Tumors located posteriorly are usually located in the costovertebral groove and commonly present with some or all the signs and symptoms of the Pancoast–Tobias syndrome, because of the involvement of the C8 and T1 nerve roots, the posterior aspect of the subclavian and vertebral arteries, the sympathetic chain, the stellate ganglion, and prevertebral muscles. These tumors have a propensity to spread along the nerve roots up to the spinal canal through the intervertebral foramina. In addition, vertebral bodies may be involved by direct extension.

DIAGNOSTIC CONSIDERATIONS

Unfortunately, the diagnosis of superior sulcus tumor is often made late because symptoms are not specific, neurologic findings are sometimes absent, and routine imaging of the chest and shoulder are often not revealing. Often symptoms are mistakenly attributed to arthritis or other inflammatory conditions of the shoulder or cervical spine. The radiologic findings can be subtle because these lesions are often hidden behind the first rib and clavicle. Posteroanterior and lateral chest X-ray have little role in evaluation of the superior sulcus tumor. Superb imaging of the lesion and its local extension can be obtained using modern computerized tomographic techniques. High-resolution images with three-dimensional volume averaging allow precise location of extent of the tumor, nerve involvement, and spinal invasion. Specific nerve root or vascular invasion is better assessed by MRI. The easiest way to obtain a tissue diagnosis is by fine-needle aspiration biopsy. Transbronchial biopsy may be considered, but these lesions are so peripheral that this option is rarely useful. If there is a suspicion of extensive pleural invasion or pleural metastases, video-assisted thoracic exploration may be utilized for diagnostic purposes. All patients should undergo a thorough search for hematogenous metastases. Positron emission tomography (PET) imaging should be a routine part of the evaluation to assess both regional lymph node involvement and distant metastatic disease. Because these are T3 lesions, we employ mediastinoscopy and supraclavicular lymph node biopsy in every patient. Patients with N2 or N3 nodal involvement are not candidates for resection. The initial investigation of the potentially operable patient also includes preoperative cardiopulmonary functional tests and other investigations required for any major pulmonary resection.

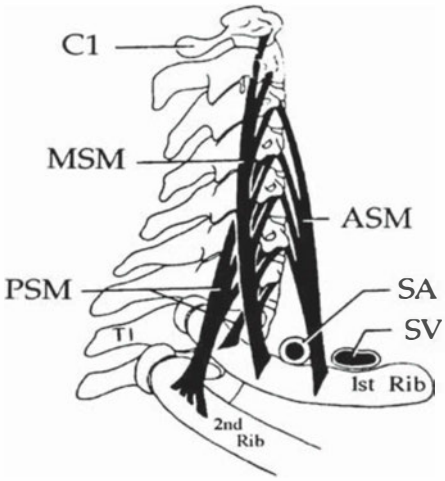


Fig. 32.1. The insertion of the anterior scalenus muscle (ASM), middle scalenus muscle (MSM), and posterior scalenus muscle (PSM) on the first two ribs divides the thoracic inlet into anterior, middle, and posterior compartments. C1 indicates the first cervical vertebral body. SA, subclavian artery; SV, subclavian vein.

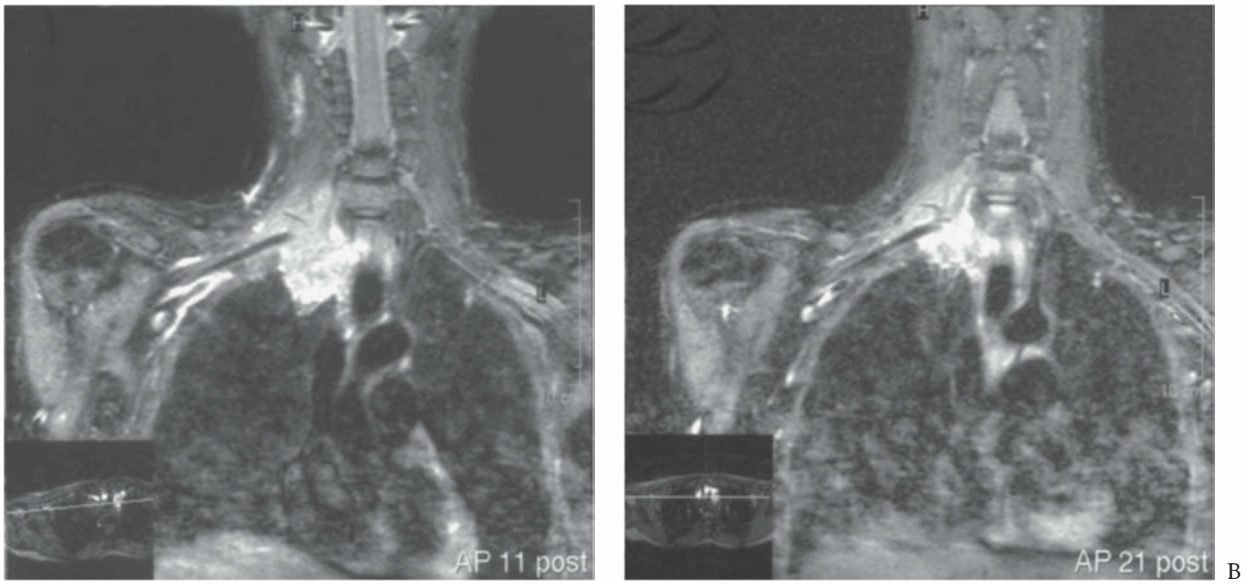
INDICATIONS

Determination of specific root involvement is important. T1 invasion may be evident only from pain and paresthesia in the medial aspect of the upper arm. C8 invasion will be associated with loss of strength in intrinsic muscles, lack of thumb opposition, and numbness in the fifth finger and medial half of the ring finger. These observations can be confirmed by electromyography, but clinical examination is usually sufficient. Phrenic nerve invasion is detected by elevation and immobility of the ipsilateral diaphragm. Vascular invasion is usually apparent by good-quality contrast CT scans, but additional information can be obtained by MRI. If vascular invasion is suspected, arteriography may be helpful, and Doppler ultrasound will demonstrate associated cerebrovascular arthrosclerotic changes, which might affect the decision regarding operability. Radiologic determination of vascular and nerve root invasion is sometimes equivocal. Apposition of the tumor to major structures does not confirm invasion or preclude surgical exploration to determine resectability (Fig. 32.2). Spinal invasion or extradural extension is often evident on CT scans, but occasionally MRI is necessary to rule out subtle spinal involvement.

These investigations are not merely academic. Extent of disease and, therefore, likelihood of complete resection must be determined preoperatively. Loss of the T1 nerve root is inconsequential, but resection of T1 and C8 will leave a severely impaired "ulnar hand," which the patient may not accept. Subclavian vein or arterial invasion is not a specific contraindication to resection. The vein can be resected without reconstruction. Arterial resection will require primary reconstruction or interposition grafting. Vascular invasion describes a T4 lesion and predicts a decreased long-term survival. Limited involvement of the vertebra such as a transverse process or partial vertebral body involvement is not a contraindication to resection. In fact, these resections can be conducted with local resection of the affected bone to obtain tumor-free margins. Patients with more extensive single or double vertebral involvement may be candidates for complete resection. Recent advances in techniques of vertebral resection and spinal stabilization make such resection feasible. Limited experience with such resections has been reported from a small number of experienced centers, and early results are encouraging.

Absolute contraindications to resection include N2 or N3 disease, extensive vascular invasion, brachial plexus involvement more extensive than C8 and T1, and multiple-level vertebral involvement with extension into the spinal canal.

After the 1924 and 1932 reports of Henry K. Pancoast describing the tumors of the thoracic apex, the entity was considered incurable until the report of Chardack and MacCallum in 1956 and the description of Shaw and Paulson in 1961. Radiation therapy given preoperatively over a 10- to 20-day period at a dose of 30 to 45 Gy had been the standard approach since the Shaw



A B

Fig. 32.2. (A) Preinduction magnetic resonance imaging (MRI) of a patient with a right superior sulcus tumor highly suspicious for the involvement of subclavian vessels. **(B)** Postinduction (chemoradiation) MRI of the same patient still suspicious for vascular invasion. The patient was explored initially through a supraclavicular approach. The vessels were found to be uninvolved, and a complete resection was performed using a standard Shaw–Paulson approach.

and Paulson report. In recent years, demonstration of tumor response following induction chemoradiation gave rise to an interest in its application in patients with superior sulcus tumors. A multicenter cooperative group trial demonstrated that cisplatin and etoposide and 45 Gy of radiation improved the rate of complete resection, pathologic complete response, local recurrence, and intermediate-term survival compared with historical controls treated only with induction radiotherapy. For this reason and because of the success of combined-modality therapy in stage IIIA lung cancer, induction chemoradiation has become the standard of care for superior sulcus tumors in most centers. It has also been pointed out by some authors that induction therapy should include higher doses of radiation as it has been shown that this does not increase perioperative morbidity but may result in a higher rate of complete response and ultimately long-term survival.

PERIOPERATIVE PATIENT MANAGEMENT

Preoperative preparation is as for any major pulmonary resection. A double-lumen endotracheal tube or bronchial blocking catheter for left-sided lesions is helpful. Standard monitoring lines include an arterial line in the contralateral radial artery. Two venous lines should be placed to allow for rapid volume replacement as needed.

SURGICAL TECHNIQUE

Different operative approaches are necessary depending upon the location of the primary tumor. The surgeon must be familiar with these various approaches. The goal of the operation is en bloc resection of the upper lobe along with involved ribs and other structures, including transverse processes, the lower roots of the brachial plexus, the stellate ganglion, and the upper dorsal sympathetic chain.

There are three approaches most commonly employed for these lesions. The posterior approach described by Shaw and Paulson is ideal for lesions situated posteriorly. The anterior cervicothoracic approach described by Dartevelle is ideal for the management of anterior lesions. The hemiclamshell approach is less commonly employed but is useful for anterior or posterior lesions.

We believe that whatever approach is selected, an initial cervical exploration is warranted. This is particularly true when a posterolateral Shaw–Paulson resection is anticipated. With the patient supine, shoulders elevated, neck extended, and head turned to the contralateral side, a transverse incision is made immediately above the clavicle. The platysma muscle is divided, as is the clavicular head of the sternomastoid muscle. The supraclavicular fat pad is excised and submitted for frozen-section examination to rule out the involvement of supraclavicular lymph nodes. The phrenic nerve is elevated away from the scalenus anticus muscle, so that the muscle may be divided. This exposes the subclavian artery and the lower roots of the brachial plexus. Anterior displacement of the brachial plexus exposes the scalenus medius muscle, which is then divided, taking care to preserve the long thoracic nerve. By mobilizing and inspecting these structures through a small cervical incision, tumor extent can be assessed and a judgment made regarding the possibility of subsequent complete resection before exposing the patient to the morbidity of a major posterior thoracotomy and rib resection. In addition, this anterior superior mobilization facilitates subsequent dissection and resection through the posterolateral thoracotomy.

Posterolateral Approach (Shaw–Paulson)

The patient is placed in the lateral decubitus position, leaning slightly forward. The upper arm is loosely supported by folded sheets and is free to move as the scapula is elevated. The skin preparation is carried out from the base of the skull (included are the spinal processes above C7) and down to the iliac crest and to the midline posteriorly and anteriorly.

Incision

A limited posterolateral incision is made, dividing the latissimus muscle, and entry is made into the chest through the fourth or fifth interspace. The pleural space and hilum are examined to exclude the presence of metastatic disease. This initial exploration also permits assessment regarding anterior (several centimeters away from the lesion) and inferior (one rib and one interspace) margins of resection.

Subsequently, the incision is extended superiorly between the spinous processes and the medial border of the scapula to the level of the seventh cervical vertebra (Fig. 32.3). The trapezius muscle is divided along the full length of the incision. The rhomboid muscles from superior to inferior are then divided in the line of the incision. The rhomboid muscles insert into the medial border of the scapula. Care should

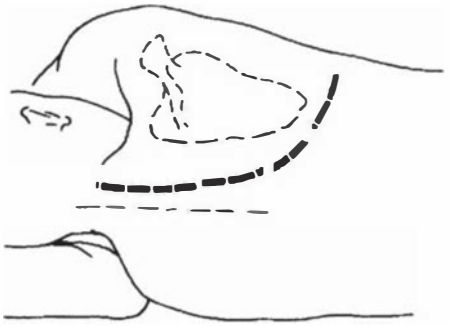


Fig. 32.3. The posterolateral thoracotomy incision. The pleural cavity is entered through a limited posterolateral thoracotomy, and resectability is determined. If the lesion is deemed resectable, the incision is extended posteriorly to the level of the C7 spine.

be taken to avoid injury to the dorsal scapular nerve and the satellite scapular artery, which run down the medial border of the scapula. The division of the rhomboid muscles elevates the medial border of the scapula from the chest wall.

A large Finochietto retractor is then placed with its lower blade in the interspace incision and the upper blade on the tip of the scapula. Opening the retractor elevates the scapula off the chest wall and exposes the subscapular musculature. These muscles are then divided with cautery up to the level of the first rib.

Chest Wall Resection

The chest wall resection is completed first, allowing the lung to be completely mobilized and permitting a safer subsequent pulmonary resection. All involved chest wall should be resected en bloc. Extrapleural dissection without rib excision, mentioned only to be condemned, results in incomplete resection and almost certain local recurrence. The lowermost rib to be preserved is identified, and an interspace incision is made along its superior border, extending from the anterior margin of resection to the transverse process posteriorly. The division of the ribs is started anteriorly. Intercostal muscles are divided with electrocautery.

Using rib shears, the ribs are divided anteriorly in succession from inferior to superior (Fig. 32.4). Traction on the previously divided anterior margins of the involved ribs exposes the anterior aspect of the first rib. For posterior tumors, the anterior aspect of the first rib can easily be encircled and divided with angled rib shears. The anterior and middle scalene muscles, previously divided from above, are exposed. The posterior scalene muscle is

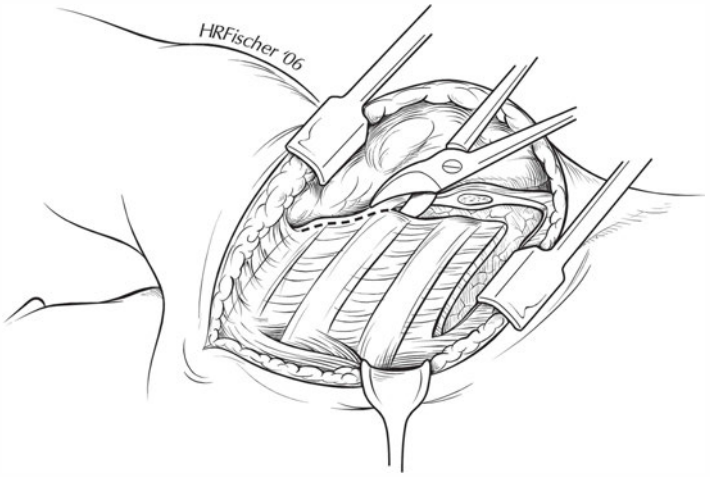


Fig. 32.4. Using rib shears, the ribs are divided anteriorly in succession from inferior to superior after the neurovascular bundle is ligated or clipped. (Adapted from Urschel HC, Cooper JD, eds. Atlas of Thoracic Surgery. New York: Churchill Livingstone; 1995:185, Fig. D.)



Fig. 32.5. The rib may be disarticulated from the transverse process in the costotransverse angle if the parietal pleura and not the ribs or vertebrae are invaded by the tumor.

divided where it crosses the lateral border of the first rib. The superior margin of the first rib is then exposed by careful superior mobilization of the subclavian vein, artery, and inferior aspect of the brachial plexus. At this point, the operation is continued posteriorly.

The erector spinae muscle is incised along its anterior border and retracted posteriorly from the first rib to the lowermost resected rib. This exposes the angle of the invaded ribs and transverse processes. Hemostasis can be obtained by packing the space between muscles and bony structures. If there is no radiologic evidence of rib or spinal invasion, the transverse processes can be left intact. The costotransverse process joint is opened, and the heads of the ribs are disarticulated by placing a periosteal elevator in the joint and levering forward (Fig. 32.5). This elevates the head of the rib from the spine. If rib invasion is present, leaving the transverse processes compromises the margin of resection. Therefore, the transverse processes (Fig. 32.6) and (if rib heads are involved) lateral cortex of the vertebra are amputated using an osteotome (Fig. 32.7). With completion of the bony resection at each level, the intercostal bundle is ligated and divided with the intercostal nerve ligated at the level of the neural foramen. Occasionally, significant venous hemorrhage is encountered from the orifice of the intervertebral foramen. Loose packing with hemostatic material can be used, taking care to avoid excessive pressure, which can result in migration of material into the spinal canal or occlusion of the anterior spinal artery. This posterior portion of the resection is continued superiorly until the angle of the first rib is reached.

Dissection of the Brachial Plexus

At this point, the first thoracic nerve (T1) below and the eighth cervical nerve (C8) above the neck of the first rib are visualized. The head of the first rib is then disarticulated from the spine. Usually, the T1 nerve is involved as it crosses the first rib. The nerve root is ligated or clipped and divided as it emerges from the intervertebral foramen. If the eighth cervical nerve is not involved, every effort should be made to protect it (Fig. 32.8). If the tumor also involves the eighth cervical nerve, the nerve

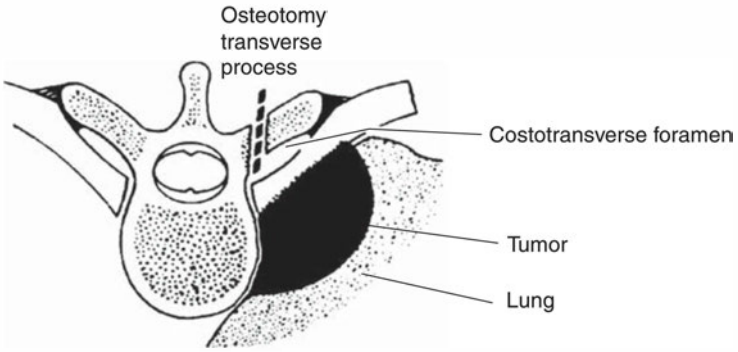


Fig. 32.6. The osteotomy is performed at the level of the transverse process if the tumor erodes the rib posteriorly.

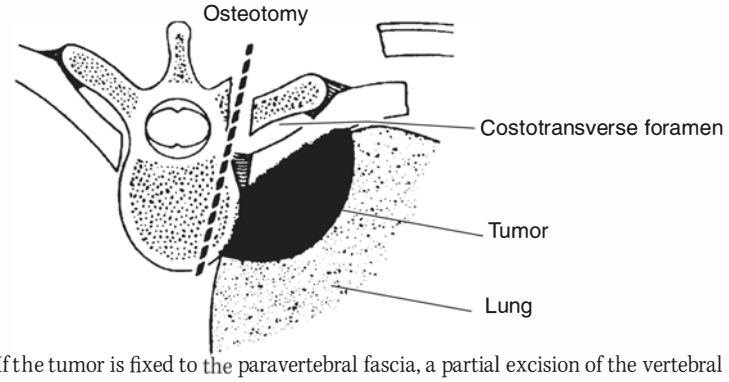


Fig. 32.7. If the tumor is fixed to the paravertebral fascia, a partial excision of the vertebral body can be done as illustrated here.

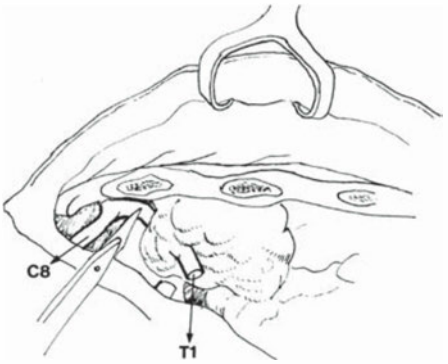


Fig. 32.8. Division of the first thoracic nerve root (T1) as it emerges from the intervertebral foramen and before its fusion with the eighth cervical root (C8).

root can be divided at the intervertebral foramen and the lower trunk of the brachial plexus divided lateral to the area of invasion. The nerve roots are ligated or clipped to prevent cerebrospinal fluid (CSF) leak. If a CSF leak is noted, the foramen should be covered with a pedicled flap of erector spinae muscle in an effort to help seal the leak.

Dissection of Subclavian Vessels

Dissection of the subclavian artery can usually be carried out in a subadventitial plane (Fig. 32.9). Branches such as the internal mammary artery and the thyrocervical trunk are ligated and transected as necessary. If the subclavian artery is invaded, it should be cross-clamped (after adequate systemic heparinization, e.g., 0.5 mg/kg) proximal and distal to the involved segment and reconstructed either by an end-to-end anastomosis or by interposing a polytetrafluoroethylene graft (6 or 8 mm). If the subclavian vein is involved or occluded by tumor, the segment of vein should be excised and the proximal and distal ends suture ligated. No attempt should be made to bypass the excised segment of vein. Management of the subclavian vessels can be difficult through the posterior approach, particularly if the tumor extends medially. In this circumstance, it is sometimes necessary to resect a segment of the anterior scalene muscle and phrenic nerve.

Vertebral Body Resection

Limited invasion of the body of the vertebra mandates vertebrectomy for complete resection. Recent advances in spinal instrumentation allow complete resection of tumors involving vertebral bodies or neural foramina (Fig. 32.10). This is a reasonable strategy for limited vertebral invasion (one or two vertebral bodies) without extension into the spinal canal. For more extensive spinal invasion, there is no justification for resection.

For en bloc resection of the involved vertebrae with the chest wall and lobe, a transcervical as well as a posterior midline approach is used, or alternatively a cervicothoracic transmanubrial approach plus a midline posterior incision can be used. The anterior incision is used to assess resectability. Tumor-bearing structures are removed with lateral division of the involved nerve roots, ribs, and partial midline division of the vertebrae. A standard lobectomy is performed leaving the lobe in place. After this, the posterior midline approach is used for unilateral laminectomies, nerve root division within the spinal canal, and midline vertebral body division. After the resection is complete, spinal stabilization is performed.

Pulmonary Resection

The chest wall specimen is still attached to the upper lobe. Anatomic upper lobectomy is the preferred parenchymal resection even if the lesion is small. The lobectomy is performed through the defect created by the chest wall resection or through the original exploratory intercostal incision (Fig. 32.11). Complete mediastinal node dissection or, at the minimum, nodal sampling is mandatory.

Postresection chest drainage is accomplished through two separate chest tubes, one of which must be placed at the apex of the pleural space. For chest wall resections down to the fourth rib, the scapula covers the chest wall defect, and therefore prosthetic reconstruction is not necessary. Muscle, subcutaneous tissue, and skin closures are accomplished in a standard manner.

Tatsamura proposed an approach for apical lung tumors that invade the thoracic inlet in which an incision is started at the level of the spinous process of the second or third thoracic vertebra,

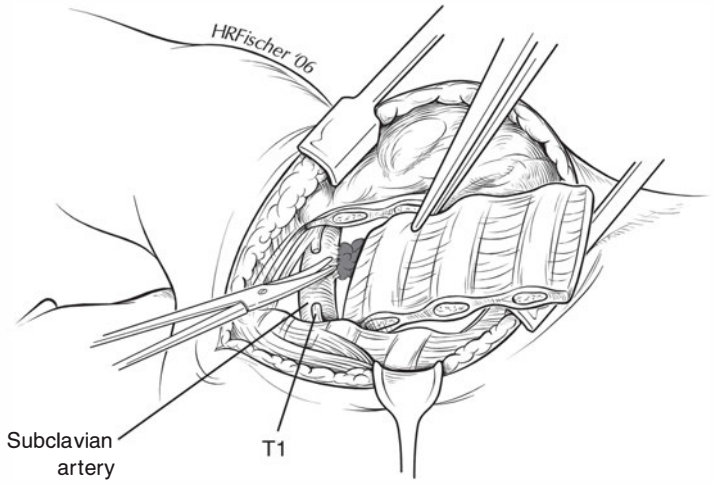


Fig. 32.9. Dissection of the subclavian artery from the tumor can usually be carried out in a subadventitial plane. (Adapted from Urschel HC, Cooper JD, eds. Atlas of Thoracic Surgery. New York: Churchill Livingstone; 1995:187, Fig. H.)

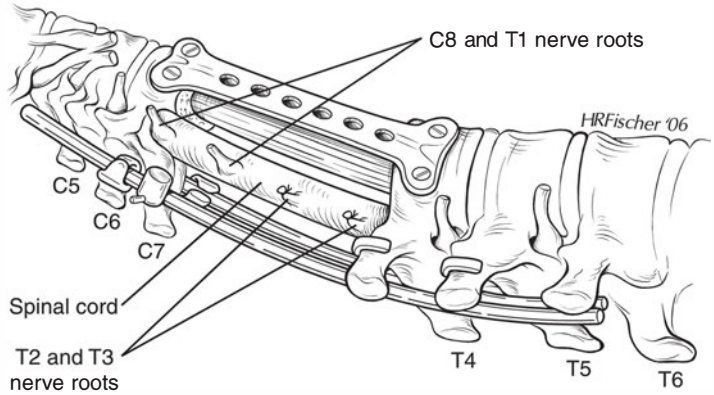


Fig. 32.10. Multilevel thoracic vertebrectomy and laminectomy, reconstruction with methylmethacrylate, placement of anterior locking plate and screw construct, and posterior fixation with hooks and rods. (Reprinted with permission from Gandhi S, Walsh GL, Komaki R, et al. A multidisciplinary surgical approach to superior sulcus tumors with vertebral invasion. Ann Thorac Surg 1999;68:1778.)

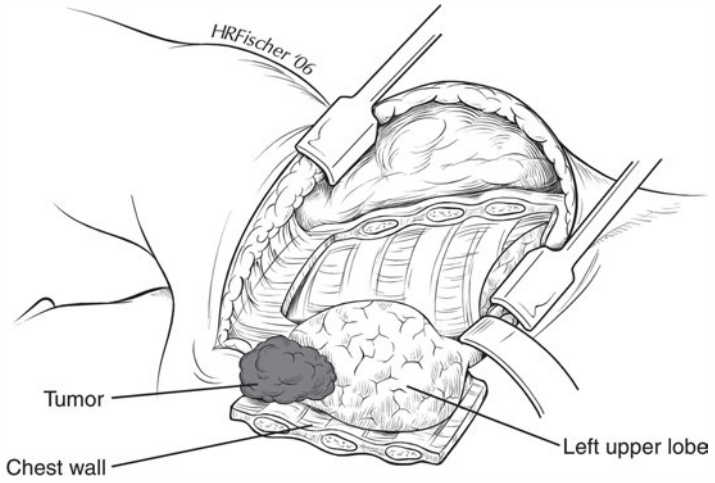


Fig. 32.11. En bloc removal of the chest wall and lobe. (Adapted from Urschel HC, Cooper JD, eds. Atlas of Thoracic Surgery. New York: Churchill Livingstone; 1995:188, Fig. I.)

continued downward along the paravertebral line around the tip of the scapula, and continued upward to above the nipple level, following the axillary line up to the level of the sternoclavicular joint (Fig. 32.12). The management of the thoracic inlet through this incision is the same as for the Shaw–Paulson and anterior approaches.

Anterior Approaches

Transclavicular Approach

This approach is ideally suited for anterior lesions. The patient is placed in the supine position with the shoulders elevated, the neck hyperextended, and the head turned away from the involved side. The operative field extends from the mastoid down to the xiphoid process and from the midaxillary line laterally to the hemiclavicular line.

An L-shaped incision is made that includes an oblique presternocleidomastoid incision extended horizontally below the clavicle and lateral to the deltopectoral groove (Fig. 32.13). By raising a subplatysmal flap laterally, one can resect the supraclavicular fat pad and rule out supraclavicular N3 disease. The sternocleidomastoid muscle is freed up inferiorly from the clavicle and manubrium to create a myocutaneous flap, which is then reflected back to yield a full exposure of the neck and the thoracic inlet. Superior mediastinal invasion is assessed by inserting a finger along the lateral aspect of the tracheoesophageal groove. Extension of the tumor into the thoracic inlet is carefully assessed. Resection through this approach requires visualization of the thoracic inlet immediately posterior to the medial clavicle. Dartevelle and colleagues described resection of the medial clavicle. However, medial clavicular resection can result in significant morbidity and limitation of shoulder motion postoperatively. A number of authors have described techniques that preserve the sternoclavicular joint and elevate the entire clavicle from medial to lateral. After resection is complete, the clavicle–sternal unit is rigidly fixed back to the sternum.

Dissection of the Subclavian Vein

Division of the internal, external, and anterior jugular veins makes visualization of the venous confluence at the origin of the innominate vein easier. On the left side, the thoracic duct should be identified, ligated, and divided. Division of the internal jugular vein improves exposure of the subclavian vein. If the subclavian vein is involved, it should be resected after proximal control and distal control have been achieved. The phrenic nerve is evaluated and preserved whenever possible. The anterior scalene

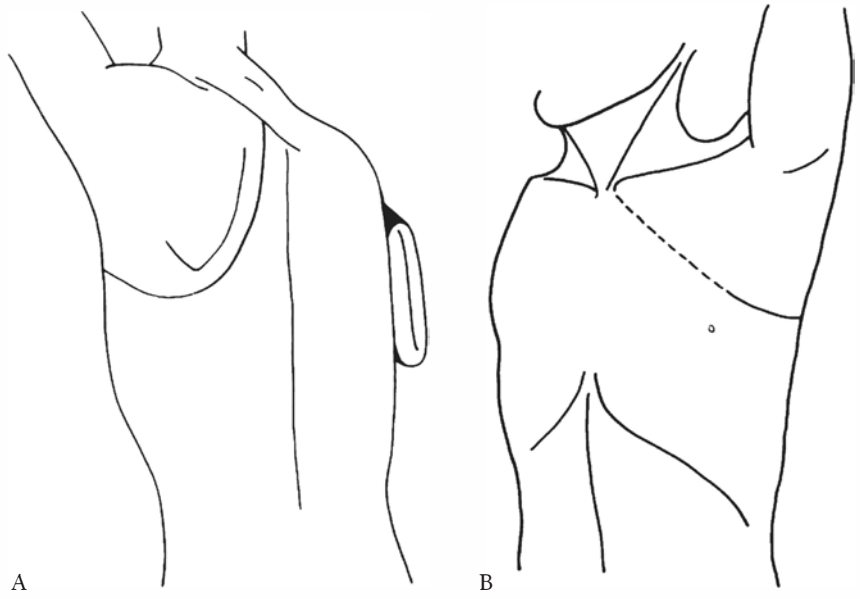


Fig. 32.12. Semidorsal (**A**) and semiventral view (**B**) of the incision. (Reprinted with permission from Tatsamura T, Sato H, Mori A, et al. A new surgical approach to apical segment lung diseases, including carcinomas and inflammatory diseases. J Thorac Cardiovasc Surg 1994;107:32.)

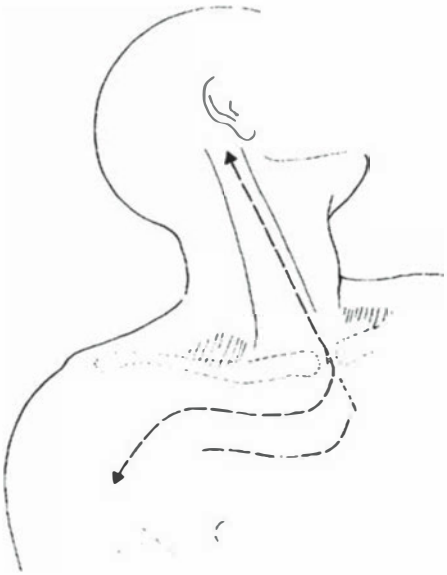


Fig. 32.13. Right transcervical incision. The patient is placed in the supine position with the neck hyperextended and the head turned away from the involved side. An L-shaped skin incision is made from the angle of the mandible down to the sternal notch. This is extended horizontally under the internal half of the clavicle and prolonged into the deltopectoral groove or into the bed of the second/third intercostal space, as indicated by the extension of the lesion.

muscle is then divided either at its insertion on the scalene tubercle of the first rib or well away from the tumor (Fig. 32.14). If the tumor has invaded the superior aspect of this muscle, the muscle should be divided at the insertion on the anterior tubercle of the transverse processes of C3 through C6.

Dissection of the Subclavian Artery

As for the posterior approach, the subclavian artery is mobilized by dividing branches as necessary. The vertebral artery is sacrificed only if involved and provided that no significant extracranial occlusive disease was detected on preoperative Doppler ultrasound examination. Usually, the artery can be dissected away from the tumor. If invaded, the artery should be resected and reconstructed as described for the posterior approach (Fig. 32.15).

Dissection of Brachial Plexus

The middle scalene muscle is divided above its insertion on the first rib or higher as indicated by the extension of the tumor. Depending on the extent of the tumor, especially if there is invasion of the middle compartment of the thoracic inlet, the

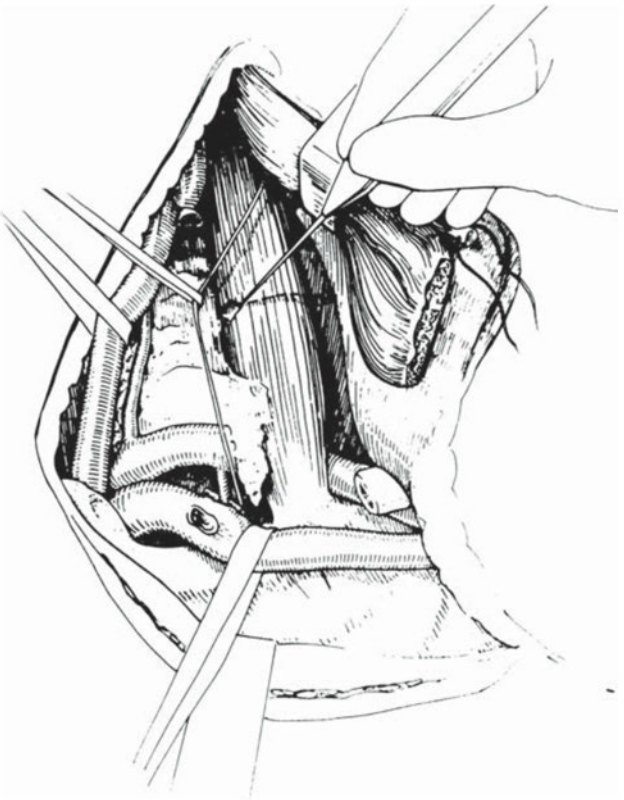


Fig. 32.14. After the division of the insertion of the anterior scalenus muscle on the scalene tubercle of the first rib, the subclavian artery is exposed. If the phrenic nerve is not invaded it must be preserved.



Fig. 32.15. The subclavian artery is divided after proximal and distal control is achieved.

muscle may have to be taken by dividing the attachments to the posterior tubercles of the transverse processes of the second through seventh cervical vertebrae. The nerve roots of C8 and T1 are then easily identified and dissected distally until they coalesce to form the lower trunk of the brachial plexus. Thereafter, the prevertebral muscles are detached along with the dorsal sympathetic chain and stellate ganglion from the anterior surface of the vertebral bodies of C7 and T1. This permits visualization of the intervertebral foramina. The T1 nerve root, if involved, is divided proximal to the tumor at the level of the intervertebral foramen. Although the tumor may extend well superior into the brachial plexus, neurolysis is usually achieved without division of any nerve roots above T1. Damage to the lateral and long thoracic nerves should be avoided to prevent a winged scapula.

Chest Wall Resection

The chest wall is divided anteriorly. For true anterior lesions, this may require resection of the lateral border of the sternum down to the lowest rib of resection. For lesions more lateral in location, the costal cartilage can be divided from the sternum. The intercostal space is divided laterally and posteriorly well beyond the margins of tumor involvement. The resected ribs

are then divided or disarticulated from the spine through tumor-free margins.

Pulmonary Resection

An anatomic upper lobe en bloc resection can be performed through the chest wall defect. The anterior superior exposure mandates resection from anterior to posterior, that is, on the right division of the superior vein, arterial branches, and bronchus; on the left, superior vein, upper lobe bronchus, and arterial branches. It is rarely necessary to reposition the patient after closure of the anterior wound and complete the lobectomy through a standard posterolateral thoracotomy. However, it is very difficult, if not impossible, to visualize the inferior pulmonary ligament and divide it safely through this anterior superior defect without videothoracoscopic equipment.

Chest Wall Closure

If the medial clavicle is resected, the sternomastoid muscle is fixed to the upper edge of the sternum. An anterior skeletal chest wall prosthesis is advised if the second rib and mandatory if the third rib is included in the resection. Marlex methylmethacrylate, Gore-Tex, and Prolene mesh are useful substitutes. Standard closure and chest drainage is employed.

Hemiclamshell or Trap-Door Incision

The hemiclamshell or trap-door incision combines a partial sternotomy and anterior thoracotomy (Fig. 32.16). The patient is positioned supine, usually with the ipsilateral operative side elevated.

Our preference is to begin the procedure with an incision in the inframammary crease and enter the chest through the third intercostal space. An assessment of operability and judgments regarding resectability are then performed. The incision is then extended superiorly through the upper sternum extending along the anterior border of the sternomastoid muscle. The internal mammary arteries are ligated and divided. A retractor or sternal hook is then placed, elevating the chest wall superolaterally. This allows exposure of the upper half of the superior mediastinum and the apex of the thoracic cavity. The superior vena cava and ipsilateral innominate vein are then dissected laterally until the subclavian vein is exposed. The medial clavicle is removed for better exposure of the subclavian vessels and brachial plexus. The involved ribs are divided at the costochondral or costosternal junctions, and the appropriate intercostal space is entered below visible tumor. The posterolateral aspects of the involved ribs are then divided, and the specimen is released within the chest cavity superiorly, remaining attached to the apical fascia. The dissection and management of the subclavian vein, artery, and brachial plexus are similar to those described for the transclavicular approach, and a lobectomy is completed.

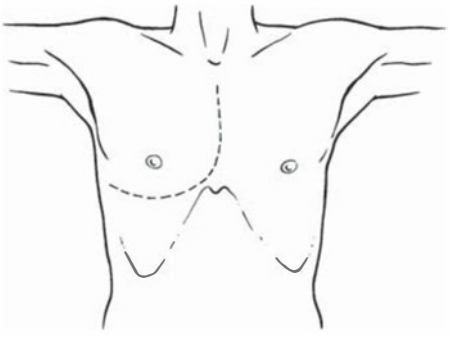


Fig. 32.16. The hemiclamshell incision, made with the patient lying supine. (Reprinted with permission from Bains MS, Ginsberg RJ, Jones WG, et al. The clamshell incision: an improved approach to bilateral pulmonary and mediastinal tumors. Ann Thorac Surg 1994;58:30.)

Masaoka Incision

The Masaoka incision involves an upper median sternotomy combined with an incision in the anterior fourth intercostal space below and a transverse cervical incision at the base of the neck superiorly (Fig. 32.17). Dissection then proceeds as described.

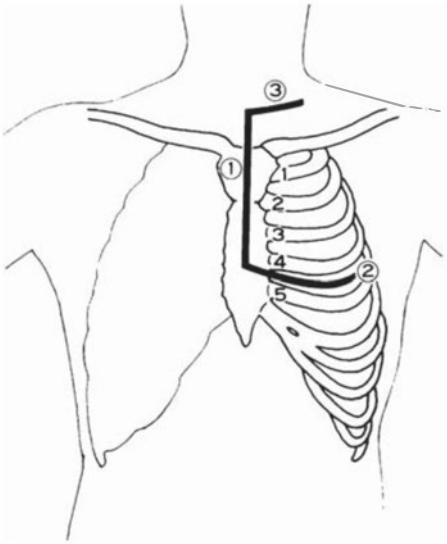


Fig. 32.17. The Masaoka approach includes a proximal median sternotomy (1) extending to the fourth anterior intercostal space below (2) and the base of the invaded neck above (3, transverse collar incision). (Reprinted with permission from Masaoka A, Ito Y, Yasumitsu T. Anterior approach for tumors of the superior sulcus. J Thorac Cardiovasc Surg 1979;78:413.)

SURGICAL COMPLICATIONS AND POSTOPERATIVE CARE

The potential complications that may occur after resection of apical lung tumors are similar to those for any major pulmonary resection. There are several complications unique to these resections. A CSF leak, if noted intraoperatively, must be sealed. This may require neurosurgical consultation, foraminotomy, and direct dural repair. If identified postoperatively by clear fluid drainage from chest tubes, aggressive management including re-exploration should be undertaken. Consequences of a persistent CSF leak such as subarachnoid or ventricular air embolism and meningitis warrant aggressive management.

The possibility of Horner syndrome and nerve deficits secondary to division of nerve roots should be discussed with the patient preoperatively. Resection of the lower trunk of the brachial plexus (C8 and T1) results in an atrophic paralysis of the forearm and intrinsic muscles of the hand (Klumpke–Déjérine syndrome). This can be a disabling situation for the patient. Hemothorax may occur as a result of chest wall resection along with the difficulty and risks of securing small veins at the level of the intervertebral foramina. Chylothorax is also a possible complication and can be avoided by individual ligation of the thoracic duct and its branches. If chylothorax is identified and persists, aggressive management including thoracic duct ligation is mandatory. If the subclavian vein has been resected, the ipsilateral forearm should be elevated to facilitate venous drainage and minimize edema. The radial pulse must be monitored to assess the patency of the revascularized subclavian artery.

CONCLUSIONS

Tumors of the pulmonary apex are challenging lesions to treat because of their close association and often involvement of surrounding structures within the thoracic inlet (brachial plexus, subclavian vessels, or spine). A multimodality approach consisting of induction chemoradiation followed by resection is the standard for patients deemed preoperatively to have resectable disease. As for any lung cancer resection, negative resection margins should always be the intent as this has been shown to be associated with significantly improved survival. The choice of incision is important. Anterior approaches have allowed complete resections of anterior apical lesions involving the subclavian vessels. Newer techniques of spine resection and stabilization enable

complete resection in patients with posterior tumors with vertebral involvement.

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EDITOR'S COMMENTS

Despite the title of the chapter there does not exist, in my opinion, a superior pulmonary sulcus or at least I have never been able to identify this as an anatomic structure. But the term has persisted. Perhaps it would be better to label these as "apical lung tumors" and recognize that not all apical lung tumors are so-called Pancoast tumors. The important distinction involves the recognition of those apical lung tumors that involve structures in the thoracic inlet so that preoperative chemoradiotherapy can be given, because this regimen has been shown in a large intergroup trial to be associated with improved survival when compared with historical controls. This preoperative regimen has now become the standard of care for these lesions, supplanting the time-honored, but poorly tested, radiation therapy alone given in 30-Gy fractions over a 10-day period. Preoperative radiation therapy had been given for these lesions since the initial description by Shaw and Paulson despite a lack of data demonstrating efficacy.

Establishing the diagnosis of an apical lung tumor can be difficult, and patients often have been followed for many months with shoulder pain attributed to musculoskeletal problems before a chest radiograph and chest CT scan are obtained. The apex of the lung is a difficult area to interpret on the chest radiograph, and this area accounts for a large percentage of lesions missed by radiologists. Persistent shoulder pain in a smoker without a specific inciting cause should be looked at with great suspicion and ideally a chest radiograph and CT scan should be obtained early following presentation of such a patient. Apical lung lesions are the one area where MRI scans, and especially the coronal reconstructions, have added to our diagnostic capabilities and enhanced our operative planning expertise. MRI provides a significant improvement in our ability to visualize the brachial plexus as well as the brachiocephalic vessels and allows for a determination as to the probability of invasion of these structures.

A needle biopsy, either an aspiration or a core biopsy, should be performed to establish a histologic diagnosis and I agree with the authors' recommendation of sampling the supraclavicular fat pad prior to proceeding with resection, even when this area is not clinically suspicious. PET scanning may point to this area, but because of the close proximity with the primary tumor, it may not be possible to distinguish nodal disease from the primary tumor. My preference is to perform a bronchoscopy and mediastinoscopy with supraclavicular fat pad biopsy as a separate procedure before initiating preoperative chemoradiotherapy, because if N2 or N3 disease is present the patient is not a candidate for resection and the "curative" radiation therapy course will differ from that given as a preoperative regimen.

As opposed to the authors, it is my feeling that the anterior cervicothoracic approach through an L-shaped incision along the anterior border of the sternocleidomastoid is the preferred approach for all apical lung tumors including posterior ones. One should keep in mind that the distance between anterior and posterior at the thoracic inlet is only a few centimeters. The anterior approach offers significantly improved access to the brachiocephalic vessels and the brachial plexus and still allows for complete chest wall excision and potentially vertebral body resection. Either the medial portion of the clavicle may be excised or, as we prefer, division of the manubrium and anterior portion of the first rib with elevation of the intact sternoclavicular joint and hemimanubrium. As opposed to the posterior approach, where the rib is disarticulated from the transverse process and then levered off the vertebral body, from the anterior approach the head of the rib is taken first and the neck subsequently separated away from the transverse process. We reserve the hemiclamshell incision for very large upper lobe lesions involving the thoracic inlet, feeling that the cervicothoracic approach works well in essentially all cases. The anatomic pulmonary resection can always be accomplished through the anterior incision, and we have never found it necessary to reposition the patient for a posterolateral thoracotomy in order to carry out the lobectomy. Again it should be emphasized that an anatomic lobectomy is the required parenchymal resection even though it might be tempting to simply do a wedge excision since only a very peripheral portion of lung parenchyma is usually involved. This temptation to try to get away with less than an anatomic lobectomy should be avoided.

(*continued*)

EDITOR'S COMMENTS (*continued*)

With respect to some of the smaller technical details, we use bipolar cautery if bleeding is encountered at the level of the neural foramen and avoid any packing with hemostatic material. CSF leaks should be assiduously avoided by careful ligation of the neurovascular bundle prior to division. If a leak is detected, it is insufficient to simply pack the area and a neurosurgery colleague should be consulted, to directly repair the dural rent by exposing the dural tear via a foraminotomy. It is optimal to recognize a CSF leak at the time of the initial operation but if a CSF leak is recognized postoperatively, a skull film should be obtained to see whether air is present in the ventricles. It may be necessary to send fluid for a glucose assay to verify that indeed the chest drainage is CSF. With a CSF leak, the patient will usually have a headache. Once a leak is demonstrated, a neurosurgeon should be involved. Conservative management involves placing a spinal drain, but as the authors point out, optimal treatment involves taking the patient back to the operating room for a direct dural repair. A CSF leak that is not dealt with aggressively exposes the patient to the risk of meningitis, something that should clearly be avoided.

A potential pitfall for the surgeon inexperienced in working around the spine and especially at the level of C8 and T1 is not carrying the dissection back to the level of the vertebral foramina where both nerve roots may be seen originating and subsequently forming the lower cord of the brachial plexus. If the dissection is not carried back to that level, it is possible to divide the lower cord of the brachial plexus mistaking it for the T1 nerve root. The head of the first rib sits between the C8 and T1 nerve root and is the key anatomic landmark for identifying these structures. Dividing the lower cord of the brachial plexus leaves the patient with a useless hand as the loss of the intrinsic muscle function results in a clawed hand.

LRK

33 Thoracic Sympathectomy (Sympathicotomy)

Seth D. Force

INTRODUCTION

Thoracic sympathectomy has undergone significant change since its initial description over 100 years ago. Originally described utilizing a posterior approach, the procedure has changed over the years from a supraclavicular approach and transaxillary thoracotomy to multiport thoracoscopy and, most recently, single-port thoracoscopy. Indications for a thoracic sympathectomy include the treatment of reflex sympathetic dystrophy (RSD), Raynaud's Disease, chronic pancreatic pain, and, most commonly, hyperhidrosis. Hyperhidrosis presents as severe sweating of the face, hands, axillary regions, feet, or any combination of these and is present in at approximately 2% of the population.



ANATOMY

The thoracic sympathetic chain exists as a paired group of, up to, 12 interconnected ganglia that correspond to each thoracic nerve. The chains lie in either pleural space on the ventral surface of the ribs just lateral to the costo transverse process articulation (Figs. 33.1 and 33.2). The sympathetic chains take a more lateral position as they travel more inferiorly in the chest. Each ganglion lies below the corresponding named rib thus the stellate, or T1 ganglion, lies just below the first rib and is frequently covered by cervicothoracic fat pad (Fig. 33.3). This anatomy, however, is not always constant and in a minority of patients ganglia may be fused or located at or slightly above their corresponding rib. Attention to anatomy is critical for defining the correct level of the desired sympathectomy. The first rib can always be identified by the subclavian artery that courses over it and by the fact that it lacks a corresponding intercostal muscle on its superior border.

Aside from the main sympathetic chain, smaller accessory nerves of Kuntz can be found in up to 60% of patients (Fig. 33.4). These nerves were originally described as only crossing the second rib but, more recently, they have been shown to crossover any of the first four thoracic ribs. Currently, there is some argument as to the role of the Kuntz nerves in pathologic processes such as hyperhidrosis; however, most thoracic surgeons make an attempt to find and transect these nerves.

Various approaches to division of the sympathetic chain have included resection of the individual ganglia as well as division of the interganglionic nerve trunk. This difference in the technique has led to confusion and misnomers when surgeons discuss the intended procedure in the literature and with patients. To clear this confusion, a consensus has been reached, which considers a *sympathectomy*, to be excision of a ganglion, or ganglia, over a specific rib or ribs (designated by the letter G/level of treatment). A *sympathicotomy*, on the other hand, consists of division of the sympathetic chain without removal of ganglia (designated by the letter R/level of treatment). This is the most common practice for conditions such as hyperhidrosis even though most studies improperly call this a sympathectomy. Current nomenclature defines the level of sympathicotomy at the corresponding rib where division of the nerve chain occurs. Thus, an R3 sympathicotomy would be defined as the division of the sympathetic chain over the third rib.

PREOPERATIVE EVALUATION/ NONSURGICAL MANAGEMENT OF THE HYPERHIDROSIS PATIENT

Preoperative Evaluation

Hyperhidrosis is defined as the excess production of sweat by the eccrine glands. Although specific volumes of sweat production have been suggested, to define hyperhidrosis, the typical patient has visibly moist hands or axillae and often has sweat dripping from these areas. A family history of hyperhidrosis can be found in up to 50% of patients and, although by definition patients must have symptoms for over 6 months, most patients have had significant symptoms for years.

Hyperhidrosis can be further separated in primary/idiopathic or secondary. Primary hyperhidrosis is usually limited to the face, hands, axillae, and feet with palmar–plantar sweating being the most common variant. Secondary hyperhidrosis can be localized or diffuse and is due to various conditions ranging from hyperthyroidism to neurologic injury and side effects from medications. A thorough history and physical examination is required for all patients being evaluated for increased sweating since surgical correction is considered only for patients with primary hyperhidrosis.

Nonsurgical Management

Nonsurgical management of primary hyperhidrosis includes the use of oral and topical agents, the application of botulinum toxin (Botox), and the use of electrical currents known as iontophoresis. Oral agents used are anticholinergic drugs and include glycopyrrolate, oxybutynin, and propantheline. These drugs work by muscarinic receptor blockade but have the disadvantage that their action is not specific to the areas of increased sweating. Common side effects include dry mouth and eyes, blurred vision, and urinary retention.

Iontophoresis involves the use of an electrical current that flows through a water bath where hands or feet are placed. Clinical success rates can be as high as 80%, and the treatment effects can last anywhere from 2 months to a year. Side effects of iontophoresis are mild and usually well tolerated. Topical agents usually involve an aluminum-based chemical ointment or cream, which can be applied to hands, feet, or axillary regions. Results of topical agents are variable and

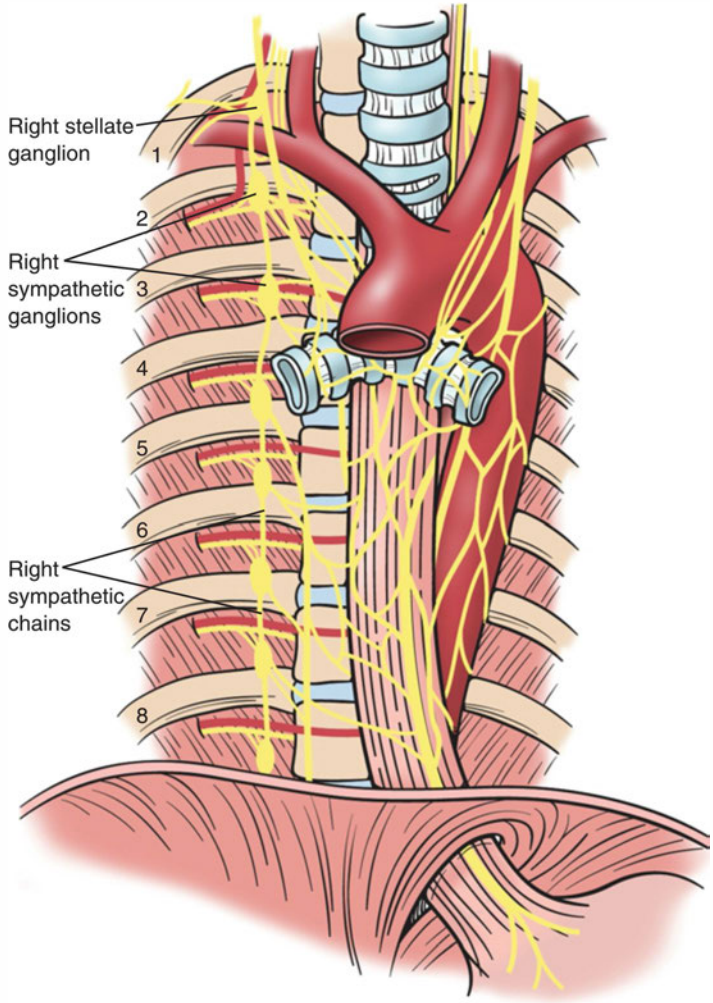


Fig. 33.1. Anatomic drawing of sympathetic chain.



Fig. 33.2. Intraoperative picture of right sympathetic chain.

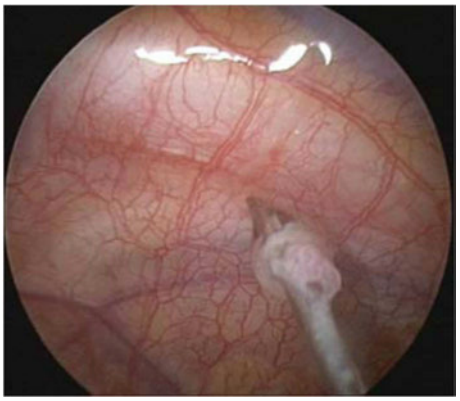


Fig. 33.4. Intraoperative picture of accessory nerve of Kuntz.

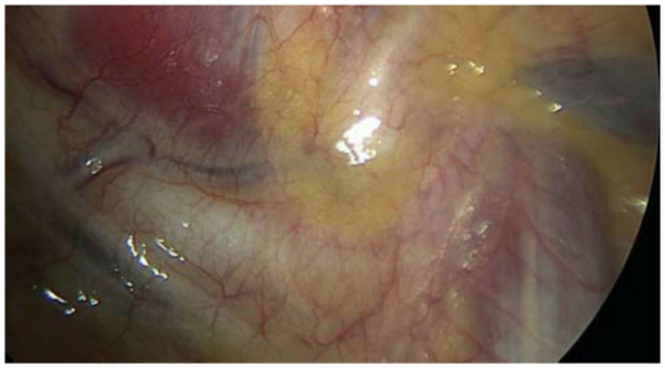


Fig. 33.3. Intraoperative picture of right stellate ganglion.

side effects are usually related to local skin reactions. Botulinum toxin injections for palmar, axillary, and pedal hyperhidrosis have also been used with good success rates but this treatment is not always well tolerated by patients and requires repeated injections over time.

SURGICAL MANAGEMENT

Surgical management has evolved from transaxillary and supraclavicular incisions to small incision single-port thoracoscopy. Recent controversy has focused on which levels to perform the sympathicotomy, how many levels to treat, and whether to divide or clip the sympathetic nerve. The ideal site of nerve division takes into consideration trying to achieve the best results with the lowest incidence of side effects, primarily compensatory sweating (CH). Although several different opinions exist, the recent consensus includes division of the sympathetic chain over the third rib (R3) for palmar hyperhidrosis and R3, R4 or R4, R5 for palmar–axillary or axillary hyperhidrosis. Facial blushing can be treated by an R2 division above the level of the second rib but care must be taken to avoid injury to the stellate ganglion.

Although most surgeons choose to divide the sympathetic nerve with electrocautery, clipping of the nerve has also been described. The primary reason given for sympathetic nerve clipping is to allow for clip removal and sympathicotomy reversal in the case of poor results or undesirable side effects. However, this

reversal has been found to be inconsistent and must take place within the first 2 weeks of the procedure to have a good chance at working. Other techniques such as thoracoscopic sympathetic block have been described. This technique utilizes injection of a local anesthetic into the sympathetic chain to create a temporary sympathetic block to replicate the effects of a sympathicotomy.

Surgical Technique

Thoracoscopic sympathicotomy is performed under general anesthesia utilizing a single-lumen tube. Patients are placed in a supine position with their arms abducted to 90 degrees and care is taken to avoid injury to the brachial plexus. The chest and both axillae are prepped into the surgical field but there is usually no need to remove any axillary hair for the procedure. An area at the edge of the axillary hairline is then injected with a local anesthetic and a 5-mm incision is then made in this area with a No. 15-blade scalpel. Next, ventilation is held and a mosquito clamp is used to dissect into the pleural space over the top of the fourth rib. A 30-degree 3-mm thoracoscope is then placed into the pleural space and ventilation is resumed. A Veress needle (Ethicon, Inc., Somerville, NJ) is then placed alongside the thoracoscope, and the pleural space is insufflated to 10 mmHg with CO_2. The sympathetic chain is then identified and usually the CO_2 can be stopped. Visualization at this point is also added by decreasing the tidal volume used to ventilate the patient. Once the desired level of sympathicotomy and all accessory nerves are identified, a 2-mm electrocautery is placed through the incision, next to the scope, and the sympathetic nerve is divided over the desired rib. The transected ends of the nerve are then separated by 1 to 1.5 cm to prevent regrowth (Fig. 33.5). All accessory nerves overlying the target ribs are then divided in the same manner. The electrocautery is then removed and replaced with a 2 or 3 mm suction, which is used to evacuate all air and CO_2 from the pleural space. The incision can be closed with a single absorbable stitch and the same procedure is performed on the contralateral side. An alternative approach is to use two thoracoscopy sites and a slightly larger scope (5 mm) until the surgeon is familiar with the anatomy and the technique. The patient is then extubated and taken to the recovery unit where a chest radiograph is performed. Results are usually realized immediately after surgery. Patients should be able to be discharged from the hospital the day of the procedure.



Fig. 33.5. Intraoperative picture of right side sympathicotomy.

The presence of an azygous lobe on the right can make the procedure a little more difficult. This situation can be dealt with by making one more 5-mm incision, on the right, and placing a 3-mm grasper through the incision to retract the azygous lobe away from the sympathetic nerve. Adhesions encountered primarily or secondary to previous surgery can be dealt with in a similar manner. The additional grasper can be used to retract the lung while the 2-mm cautery is used to divide the adhesions. Care should be taken to avoid injury to some of the larger intercostal veins, which can be difficult to control with the small cautery.

Results, Complications, and Side Effects

Results for palmar and axillary hyperhidrosis tend to be good with over 85% of patients expressing satisfaction with their outcomes. In the author's own series, sympathicotomy was successful in 99% of patients undergoing operation for palmar hyperhidrosis. Results tend to be slightly better for patients undergoing surgery for palmar or palmar–plantar hyperhidrosis than for axillary or palmar–axillary hyperhidrosis. Patients who undergo operation for facial blushing, on the other hand, only report excellent results in only 30% to 60% of cases.

The most common complication of thoracoscopic sympathicotomy is postoperative pain that can present as discrete pain at the thoracoscopy sites or as a band-like tightness, which can be troubling to patients if they are not forewarned. Pain is usually treated effectively with oral analgesics and anti-inflammatory drugs. Intravenous ketorolac can be used intraoperatively in patients who do not have contraindications to nonsteroidal anti-inflammatory drugs, to help minimize postoperative pain. Chronic postoperative pain syndromes are rare and occur in $<1\%$ of all patients. Nerve injury can occur and most commonly involves injury of the intercostal brachial nerve at the site of the thoracoscopy incision. This can lead to numbness on the lateral aspect of the breast and medial aspect of the biceps area and is usually self-limited. Injury to the brachial plexus has been reported and is usually due to excessive abduction of the arms. Care should be taken during position of the patient to only abduct the arms enough to provide adequate exposure to the axillae.

Pneumothorax has been reported in up to 5% of patients after thoracoscopic sympathicotomy. This can be due to failure to evacuate all CO_2 from the pleural space or from lung injury. Most patients can still be discharged on the day of the procedure as long as chest radiography demonstrates that the pneumothorax is not expanding and the patient is asymptomatic. Rarely, a persistent air leak is identified intraoperatively. This is more common in reoperations or in patients who require a pneumolysis for adhesions. A small pleural drain can be placed through the thoracoscopy incision and often removed several hours later, or the following day, once the air leak has resolved.

Bleeding complications occur in $<5\%$ of patients and can be due to injury to an intercostal vessel, superior vena cava, subclavian artery, or aorta. Care should be taken to ensure that the thoracoscopy incision is made at the third intercostal space and that dissection is performed over the fourth rib. Venous braches may be found coursing over the sympathetic chain. Small veins can be divided easily with electrocautery but larger veins should be avoided since injury to this structures can be difficult to control with cautery alone. If a larger vein is found to be obscuring the sympathetic nerve, the site of sympathicotomy can be moved slightly cephalad or caudad to avoid vein injury.

Horner's syndrome, which includes ptosis, miosis, and anhidrosis on the affected side, is due to injury of the stellate ganglion, which is located just below the first rib. Most occurrences of Horner's syndrome are transient and are likely due to edema caused by manipulation during the sympathicotomy. Permanent Horner's syndrome has been reported in as many as 5% of patients, but the acceptable incidence should be $<1\%$. Aberrant location of the stellate ganglion may, rarely, be a cause for injury but, more commonly, injury is

due to incorrect site of sympathicotomy due to an error in rib counting. Care should be taken to identify the proper anatomic landmarks and if uncertainty still exists then the sympathicotomy can be moved slightly caudad.

Cardiac arrhythmias can occur but are rare, occurring in fewer than 1% of patients undergoing surgery for hyperhidrosis. The presumed mechanism is stimulation or disruption of the cardiac pacemaker's fibers that are supplied by the lower cervical and upper thoracic sympathetic ganglia. Bradycardia, tachycardia, intraoperative cardiac arrest, and ST and QT segment changes have all been reported. Sympathicotomies limited to R3 or R3/R4 may only disrupt a small portion of the nerve fibers innervating the heart and therefore permanent dysrhythmias should be rare.

The most common side effect of thoracic sympathicotomy is compensatory hyperhidrosis. CH can occur in up to 30% of patients following operation for hyperhidrosis and is defined as an increase in sweating in the trunk, groin, and thigh areas. CH can be mild-to-severe and usually is the reason for lower satisfaction rates. The mechanism of CH is felt to be due to either compensatory changes created by disrupting sweat gland function in the target areas or changes with hypothalamic feedback; however, the exact cause remains unknown. Several risk factors have been associated with an increased risk for CH and include the level of sympathicotomy, number of levels divided, obesity, and the presence of significant preoperative sweating in the trunk, thigh, and groin areas. Currently, there is no optimal treatment for CH but options include the same modalities used for nonsurgical treatment of hyperhidrosis. Patients should be counseled extensively about the risk of CH and in patients at significant risk for CH, thoracoscopic sympathetic block, prior to sympathicotomy, may be a reasonable first step to assess the severity of compensatory sweating.

Gustatory sweating and phantom sweating can occur in up to 40% of patients. Gustatory sweating manifests as facial sweating that occurs while eating and its pathophysiology is poorly understood. Phantom sweating presents as a sensation that a patient is, or is about to start, sweating in the target areas. It is usually self-limited and is best treated by making patients aware of the phenomenon preoperatively.

Recurrent sweating can occur in up to 10% of patients, years after their operation, depending on specific target area. Plantar hyperhidrosis has the highest recurrence rates and can be seen within a year after surgery. Reoperation with division of accessory nerves or sympathicotomy one or two levels below the original sympathicotomy or sympathectomy has been reported with good results. The risk of recurrent sweating should be discussed with patients as part of their preoperative counseling.

OTHER INDICATIONS FOR SYMPATHICOTOMY

Division of the sympathetic chain can also be used to treat Raynaud's syndrome, RSD, chronic pancreatic pain, and cardiac abnormalities such as long QT syndrome or certain tachycardia syndromes. There is a paucity of literature reporting results for these diseases and results tend to be less successful than those reported for hyperhidrosis. That being said, early pain reduction in patients with severe pain due to chronic pancreatitis can be high as 80% with a bilateral sympathicotomy from R4 to R10. This procedure usually requires the use of a double-lumen endobronchial tube and is facilitated by placing the patient in the lateral decubitus position.

CONCLUSION

The popularity of thoracic sympathicotomy has grown in the last decade as advances in technology have allowed for smaller, single-port, thoracoscopic access and as our understanding of results, complications, and side-effects has increased. The majority of patients report excellent results with thoracoscopic sympathicotomy for palmar, palmar–plantar, and axillary hyperhidrosis. Nevertheless, patients still report problems with recurrences and severe compensatory hyperhidrosis. This later problem, in particular, can be worse to a patient than their original hyperhidrosis. Problems with severe CH are compounded by the fact that currently there is no optimal treatment for this phenomenon. Planning for thoracic sympathicotomy should involve a detailed discussion with the patient about the expected results, complications, and possible side effects.

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EDITOR'S COMMENTS

The advent of thoracoscopic sympathectomy, more properly known as sympathicotomy as Dr. Force points out, has focused attention on the previously unrecognized significant incidence of hyperhidrosis that exists within the population. The procedure has evolved from one where a double-lumen endobronchial tube was placed and three incisions were made to one that may be performed with a 2-mm thoracoscope and a 2-mm electrocautery. In addition, there have been significant modifications made in the number of levels where the sympathetic chain is divided as well as a recognition that a ganglion does not need to be resected. Some surgeons have further modified the procedure by choosing to simply place a clip on the sympathetic chain avoiding division of the chain. Measuring digital skin temperature during the sympathicotomy may be used to assess the efficacy of the procedure. A rise in digital temperature should occur soon after the sympathetic chain is either clipped or divided.

When we first began doing this procedure for palmar hyperhidrosis, we were impressed with immediate relief patients received and how incredibly pleased most of them were, having suffered for years with the embarrassment that came every time they would go to shake someone's hand. In addition, patients were even more pleased when their plantar sweating was significantly diminished as well. We recognized early on that for axillary hyperhidrosis and even more for facial sweating that overall results were not as favorable. Thus, patient selection is critical to obtain the best results. What we did not anticipate was the very high incidence of compensatory hyperhidrosis (CH) that occurs. In fact, I would go so far as to say that it is the rare patient who does not have CH. That is how commonly it is seen. Fortunately, most patients experience relatively minor CH and clearly express that it is far preferable to palmar or axillary sweating. But there is a significant and real incidence, albeit small, of patients who experience CH to such a degree that their lifestyle becomes significantly worse than what they experienced prior to operation and there is little that can be done for them. There have been attempts at reversal of the sympathicotomy with reanastomosis but this requires a technical tour de force and rarely is successful. Patients should be well informed prior to operation that it is highly likely they will experience some manifestation of CH and there is a small chance that it could be severe. Recognizing potential risk factors, as pointed out by Dr. Force, should ideally prompt a trial procedure such as the injection of local anesthesia to create a sympathetic block in an attempt to assess the likelihood of severe CH.

LRK

34 Operative Correction of Pectus Excavatum and Pectus Carinatum

Christopher S. Muratore, Thomas F. Tracy, and Larry R. Kaiser

INTRODUCTION

Pectus excavatum (funnel chest) and pectus carinatum (keel chest) are the most common chest wall deformities. Pectus excavatum is an anterior chest wall deformity that presents as a posterior depression and frequently associated with rotation of the sternum and the lower costal cartilages. It occurs in up to 1 in 300 to 1 in 1,000 births with a 4 to 1 male predominance. Protrusion of the body of the sternum, pectus carinatum, presents less frequently than excavatum and occurs more commonly in boys as well. The etiology of the pectus anomalies has not been established but the association between pectus excavatum and other musculoskeletal abnormalities, particularly scoliosis and Marfan's syndrome, suggests that abnormal connective tissue plays a role. A genetic predisposition is supported by a family history of chest wall deformities in approximately 40% of patients. The clinical presentation of pectus deformities varies from a noted depression or protrusion in the neonatal stage that persists throughout childhood and becomes progressive in adolescence. Most commonly, the pectus deformity becomes apparent and progressive with the onset of puberty and the axial growth spurt. Although there is limited physiological dysfunction noted during this initial timeframe, patients may become symptomatic from a cardiopulmonary perspective as they become more active. A universal concern of significance to all pectus patients is the cosmetic disfigurement, which is associated with a serious loss of self-esteem and can affect social behavior. The anterior chest disfigurement appearance becomes a major issue for adolescents at a time of great vulnerability during phases of puberty that is often characterized by excessive physical, social, and emotional changes. Patients with these deformities experience feelings of shame and try to hide their chests as reflected by their choice of clothing or body posture and avoidance of social activities and sports. This compounds the cardiopulmonary symptoms; patients become more withdrawn perhaps adding to their physical symptoms, which has been documented well beyond adolescence into adulthood. The purpose of this chapter is to review the indications for operative repair, to the preoperative evaluation, and to describe the common techniques for operative correction. The discussion focuses on the contemporary minimally invasive Nuss procedure, the classic open Ravitch procedure, and alternative procedures used for secondary operations and adult patients.



CLINICAL PRESENTATION

Children with pectus excavatum present with a wide spectrum of chest depression deformities from a mildly depressed sternum to a severe case in which the sternum almost abuts the vertebral bodies. The depression is created by two components. The first is posterior angulation of the body of the sternum beginning just below the insertion of the second costal cartilage. The second component is posterior angulation of the costal cartilages–sternal junction due to a seeming overgrowth and deformity of the costal cartilages. The sternum itself appears normal; the posterior angulation is due to the abnormal cartilage growth and position. The deformity maybe broad and shallow or narrow and deep, and asymmetry might exist as well. Pectus excavatum is generally well tolerated in infancy and childhood. Older children, however, may complain of pain in the area of the deformed cartilages or of precordial pain during or after exercise. Some patients might have palpitations, presumably transient atrial arrhythmias and might be associated with mitral valve prolapse. The physiologic impact of pectus excavatum has been the topic of much debate including the evolution of cardiopulmonary testing, which is well documented in the literature. Generally, this has included extensive pulmonary and cardiovascular function evaluation including echocardiographic studies. More recently, pulmonary function tests combined with exercise stress and metabolic testing have been employed, but not uncommonly no physiologic abnormality is detected. Provocative testing with exercise may be the most accurate way to determine the level, if any, of physiologic derangement. Pectus carinatum patients rarely are symptomatic, and no consistent abnormalities related either to cardiac or pulmonary function have been observed. Most commonly, carinatum deformities are less noticeable until adolescence and tend to progress during the pubertal growth spurt.

PREOPERATIVE EVALUATION

Parents bring their child for surgical evaluation at various times. Often, a young child will present with the congenital pectus variant without symptoms or cognitive psychosocial concerns. These patients and families need reassurance and careful follow-up on a yearly basis. Another time for surgical evaluation comes with the prepubertal and pubertal growth spurt. The axial growth during puberty is a common time for worsening of the pectus excavatum depression. The two most popular surgical corrections to date are the modified Ravitch procedure (open technique) and the minimally invasive repair made popular by Nuss (Nuss procedure). The popularity of the minimally invasive Nuss procedure and widespread usage of social media has brought many families and children in for early evaluation. The patient workup for each surgical approach can be assessed at the initial consultation and tailored based on the family's concerns and intentions as well as their desire for one procedure over another and modified by the surgeon's preference for surgical repair technique. In some centers,

the Ravitch procedure, or some modification, is preferentially performed after the axial growth spurt has concluded. This allows for surgical correction of the deformity with minimal concern for recurrence from continued axial growth. Most often this procedure is performed in the mid-to-late teens. The Nuss procedure can be performed earlier, although not recommended to be performed too early, as continued growth after bar removal will create a similar pectus recurrence scenario. It is our practice to counsel families and patients about the timing of operation in the early-to-mid teens during the axial growth spurt if they are interested in the Nuss procedure as the duration of the bar placement is 2 to 4 years (Fig. 34.1).

For those patients with a mild-to-moderate deformity who are sedentary, it is recommended that an exercise program, improved posture, and breathing exercises be prescribed and patients are reevaluated at 6 to 12 months follow-up intervals. Patients with clinically severe deformities at initial presentations can be carefully evaluated with a three-step process.

In addition to a thorough history, physical assessment of symptoms, the initial consultation permits education about the operative options if indicated and allows one to gauge the sincerity of a family's and child's wishes to proceed with surgical correction. The preoperative workup includes cross-sectional imaging with computed tomography (CT) scan or magnetic resonance imaging (MRI) of the chest to calculate the Haller index, a measurement of pectus depression severity calculated by dividing the transverse thoracic distance by the anterio posterior (AP) distance from posterior sternum to anterior vertebral body, pulmonary function studies including spirometry, and cardiology evaluation including echocardiogram and electrocardiogram. More recently, pulmonary function testing and stress treadmill cardiac evaluation can be combined as a measure of metabolic function providing an index of baseline cardiopulmonary performance. Surgical correction is generally indicated if the patient presents with symptoms, has a severe pectus excavatum based on the clinical evaluation including CT or MRI Haller index >3.2, evidence of cardiac or pulmonary compression on the CT, MRI, or echocardiogram, evidence of mitral valve prolapse, arrhythmia, or restrictive lung disease. An important additional criteria depending upon stage of pubertal development and social maturation is the degree of significant body image disturbance, which can influence both the child and family. A major consideration is the hesitation on the part of commercial payers to cover the cost of the pectus repair

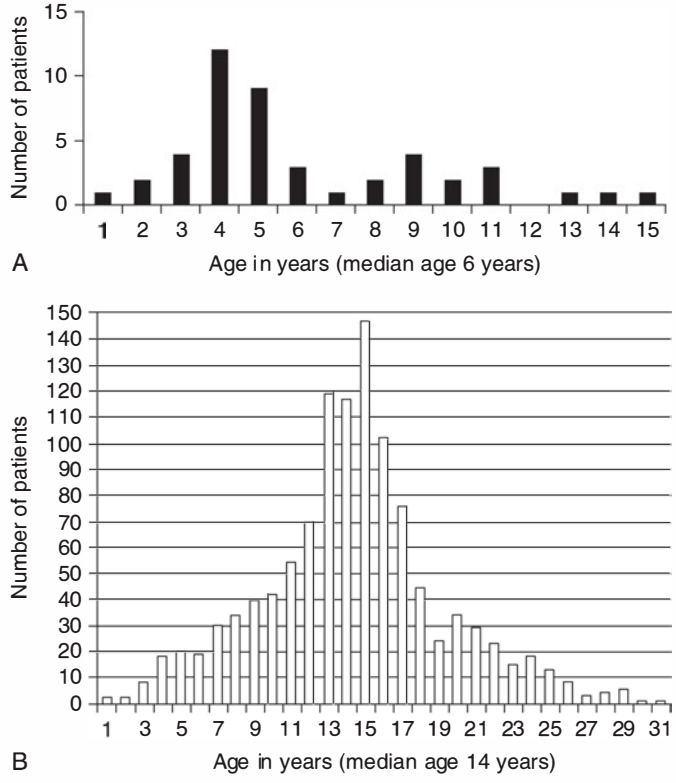


Fig. 34.1. (**A**) Age at primary surgery first decade: 1987–1996. (**B**) Age at primary surgery 1987–2008. With permission from Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252(6):1072-1081.

SURGICAL CONSIDERATIONS

In 1998, Nuss reported a "minimally invasive" correction of pectus excavatum by insertion of a convex steel bar under the sternum. The technique is possiblly based on the malleability and flexibility of the anterior chest wall. Although it requires no cartilage incision or resection and no sternal osteotomy, it is a misnomer to consider it minimally invasive. This technique requires advanced skill, training, and careful pre- and intraoperative planning for successful outcome. It is of utmost importance to take a careful history including any allergy history to the components used in the traditional stainless steel bar. Should there be a history or family history of allergy to metal or Nickel, patch testing should be done. If an allergy is identified and confirmed, a titanium bar can be used instead of a stainless steel bar. The titanium bar, however, is not as malleable as steel and consequently must be bent at the factory with computer-assisted design manufacturing technology, which is readily available from the manufacture. A copy of the patient's preoperative axial cross-sectional imaging scanning can be provided. Titanium is approximately four times as expensive as steel and therefore should not be used for routine cases where metal allergy is not an issue. The correct size bar needs to be determined in advance and this is accomplished by measuring the distance from the mid-axillary to mid-axillary line across the deepest point of the depression. The bar needs to be 2.5 cm or 1 inch shorter than the distance from the right-to-left mid-axillary line. The correct size and shape of the bar are critical to success of the operation and resulting elevation of the sternum. The stainless steel pectus support bar is bent in the operating room at the time of the operation to conform to the patient's chest configuration; conversely, the titanium bar needs to be prebent at the manufacturing facility based on the custom measurements.

A preoperative discussion with the patient and family regarding the postoperative pain management is crucial. This has been the topic of some debate as evidenced by numerous reports published in the literature, which favor epidural versus patient-controlled analgesia (PCA). The pain control strategy also needs to be

modified in consultation with the anesthesia and pain management specialists at each institution. It is our preference in consultation with the pediatric anesthesiologist ahead of time to consider a thoracic epidural; however, a one-time lumbar intrathecal injection of morphine has proven equally efficacious. We also favor PCA as this reinforces the motivated patient to be an active participant in their postoperative pain management and recovery.

Nuss Procedure

The operation is performed under general anesthesia (Fig. 34.2). Careful attention to pressure points and padding is essential and both arms are abducted comfortably so as not to cause injury. Some surgeons perform the procedure from the patients right to left, whereas others perform it from left to right. Moreover, other surgeons make a third incision in the subxiphoid region and apply a towel clip or a bone hook to help elevate the sternum for passage of finger or dissecting bar under the sternum. It is our practice once under anesthesia to measure the left-to-right mid-axillary distance again, to confirm the proper bar choice and then to bend the bar appropriately such that a slight overcorrection will be fashioned in the sternal region leaving a 2- to 4-cm flat section in the middle of the bar for sternal support. A 5.0-mm trocar is inserted either inferior to the site of bar placement or superior but in either position to allow for proper orientation and visualization of the mediastinum and passage of the bar across the mediastinum. Using CO_2 insufflation, a 5.0-mm, 30-degree scope will help accomplish this visualization and safety. Once the deepest portion of the pectus depression is appreciated, bilateral subcutaneous incisions can be created just off the mid-axillary lines and carried to the height of the pectoral ridges bilaterally. Employing unilateral thoracoscopy, the pectus introducer bar can be passed through the subcutaneous tunnel to the pectoral ridge and carefully introduced into the thoracic cavity under direct visualization (Fig. 34.3A–34.3F). At this point, the deepest portion of the sternum can be appreciated thoracoscopically and a plane of dissection can be created in that location. Often the deepest portion of the sternal location is immediately adjacent to the pericardium. As an added safety maneuver, first, a transmediastinal tunnel can be created just superior to that deepest part of the deformity to aid in lifting the sternum off the cardiac structures. To minimize the risk of catastrophic mediastinal injury, specifically cardiac injury, we recommend that the tip of the pectus introducer bar be maintained under strict visualization at all times. A gentle sweeping technique helps facilitate separation of the substernal areolar tissue and advancement of the pectus bar while the thoracoscope allows careful visualization posterior to the sternum to the opposite hemithorax. Once the pectoral ridge on the contralateral side is reached and confirmed by the assistant, the pectus bar is then pushed out of the contralateral hemithorax and into the contralateral subcutaneous tunnel. At this point, the bar can be pushed completely across the mediastinum and the assistant surgeon can help elevate the sternum and anterior chest by simultaneously lifting the bar. Modifications of the technique involve the use of bilateral thoracoscopy or using a subxiphoid window in certain situations to help elevate the sternum for the dissection or passage of the bar. Although one support bar is used in the majority of cases, it is at this point of the procedure where one might decide a second bar would be necessary to more effectively elevate the deepest part of the sternum if less than a desirable amount of anterior movement has been accomplished, keeping in mind the bar must reside under the sternum and not the xiphoid. The assistant then ties an umbilical tape to the pectus introducer bar and under thoracoscopic guidance, the umbilical tapes are pulled backward through the substernal tunnel to the original site of introduction. The previously prepared definitive pectus support bar is then tied to one of the umbilical tapes and again under strict thoracoscopic visualization and guidance is drawn through the substernal tunnel with the convexity facing posteriorly. Once the bar is in position, it is then rotated 180 degrees using the bar flippers simultaneously on each side. The end of the bar is then inserted into the stabilizer, which fits perpendicular to the bar and is stabilized to the chest wall with a series of 0 PDS sutures or surgical steel wire placed in figure-of-eight fashion around the bar and stabilizer to include rib and fascia. This minimizes the chance of postoperative bar displacement as was experienced in the experience with the Nuss procedure when the stabilizer was not routinely used. Attention is then turned toward wound closure with careful consideration for hemostasis and multilayer closure of the deeper tissues over the bar followed by subcutaneous closure and finally cutaneous closure.

The Nuss procedure also may be employed for correction of a pectus carinatum deformity. As for the repair of the excavatum deformity following evaluation of the degree of the deformity, a stainless steel bar is bent appropriately so that it will

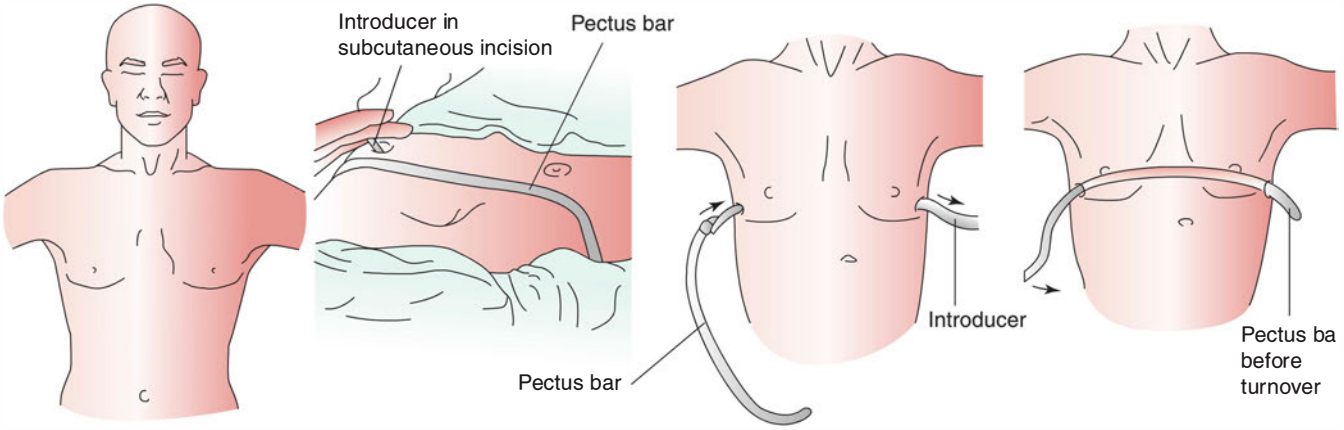


Fig. 34.2. Illustration of the Nuss operative procedure.

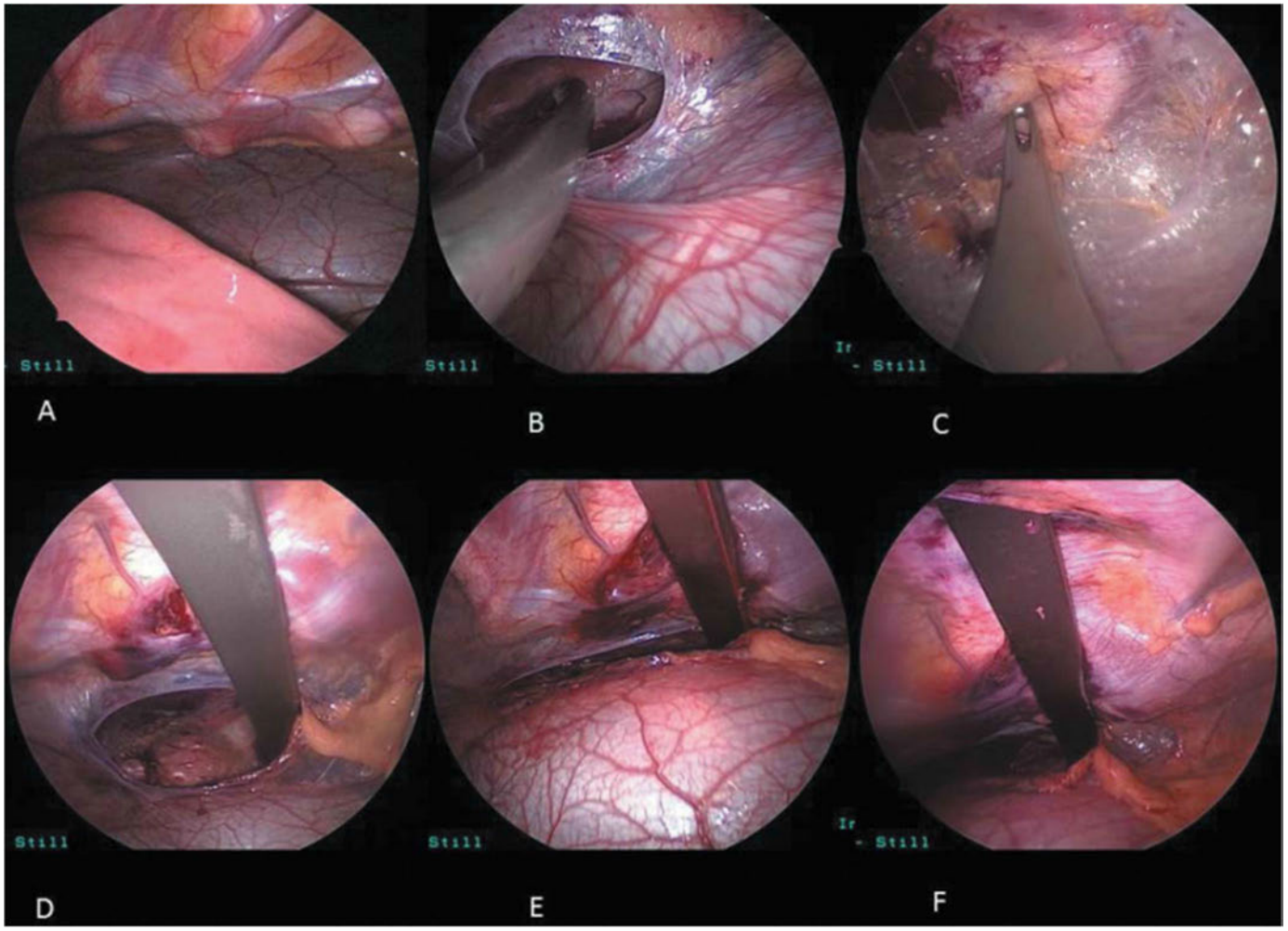


Fig. 34.3. Right-sided thoracoscopic view of (**A**) the site of lowest sternal depression, (**B**) introducer bar "tip up" establishing the areolar plane between sternum and pericardium, (**C**) complete visualization of the "tip up" concept across the substernal areolar plane at the contralateral pectoral ridge, (**D**) introducer bar passed through the contralateral pectoral ridge to the subcutaneous pocket and lifted with assistance to "mold" the anterior chest, (**E**) the pectus bar passed into position prior to "flipping," and (**F**) final position of the pectus bar lifting the lowest sternal depression at completion of bar fixation.

adequately depress the sternal deformity. The bar is placed through a subcutaneous tunnel and secured in place to the chest wall with stabilizers as described above. The bar should remain in place for at least 2 years according to those experienced with the Nuss procedure. The use of a dynamic compression system that uses a custom-made aluminum brace and avoids an operative procedure altogether also has been described for the treatment of the carinatum deformity. The brace simply pushes the deformity toward a more normal position over time, allowing bone reorganization to occur to allow the sternum to remain in the corrected position.

The Ravitch Procedure and Modifications

An alternative to the Nuss procedure and the procedure that has classically been described as the ideal repair for pectus excavatum deformity is the Ravitch procedure or some modification of this procedure that was originally described by Mark Ravitch. The classic Ravitch procedure involves bilateral subperichondrial resection of all deformed costal cartilages generally from costal cartilage 3 through 7 as well as an anterior wedge-shaped osteotomy to elevate the sternum. The sternal osteotomy is then supported with a retrosternal strut for 6 months to 1 year to secure it in a firm anterior position while the perichondrium heals and ossifies (Fig. 34.4).

Technique

A transverse incision is generally placed below and within the nipple lines at the site of the future inframammary crease. Skin flaps are then mobilized superiorly to just above the extent of the deformity using electrocautery to maintain meticulous hemostasis. The pectoralis major muscles are then reflected off the sternum and costal cartilages on both sides along with portions of the pectoralis minor. The correct plane can be challenging to identify; however, once the medial aspect of the muscle is separated using electrocautery from the sternal attachment, an empty knife handle can be used directly anterior to the costal cartilage to elevate the muscle. This is performed sequentially on all involved costal cartilages on both the right and left side, which then allows for placement of right angle retractors which are pulled anterior for additional exposure. Care must be taken to identify the proper avascular plane and avoid entry into the intercostal muscle bundles. This is not easy especially toward the upper costal cartilages, considering that one is working under a large skin flap that makes visualization and instrument placement

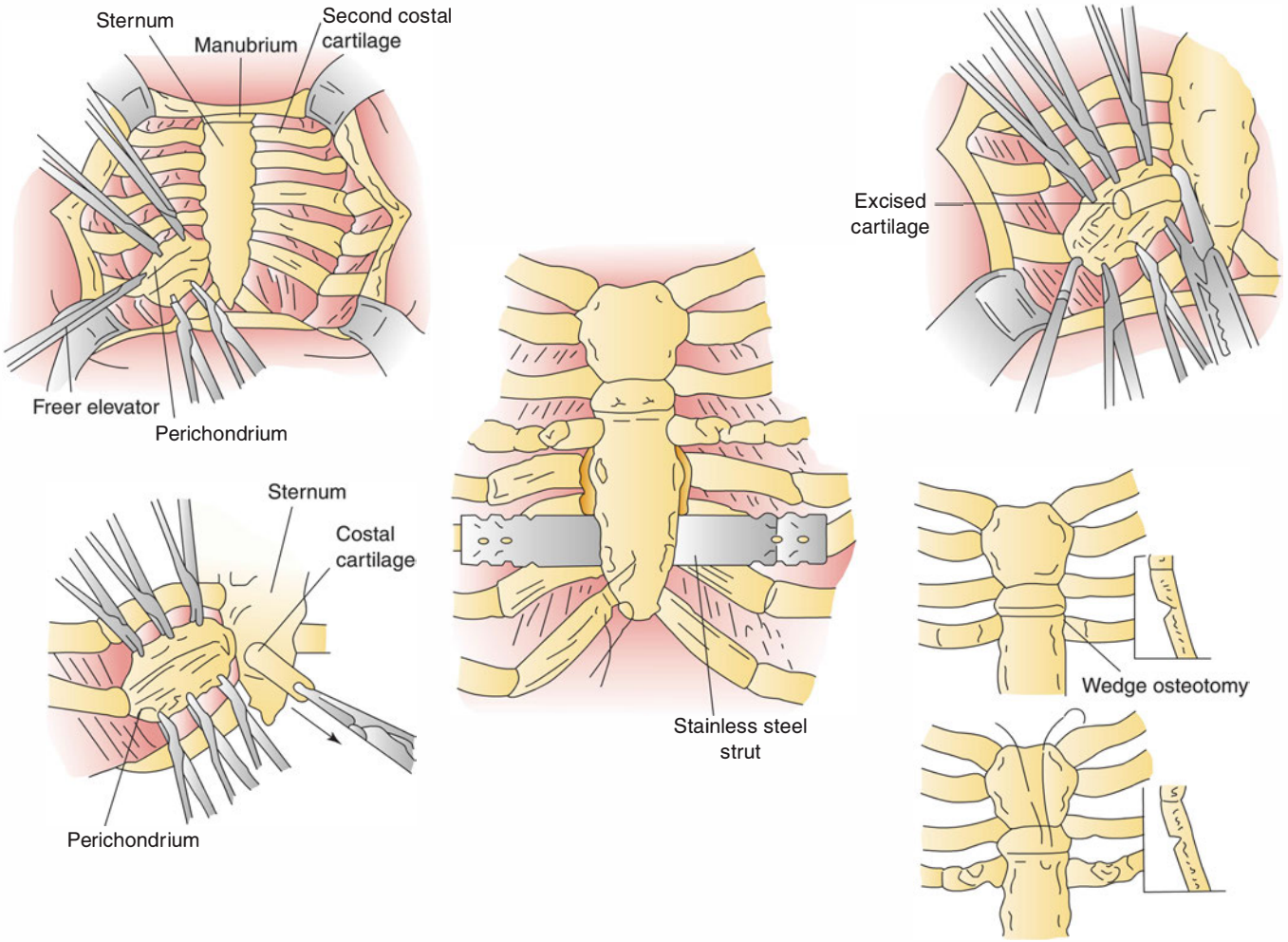


Fig. 34.4. Illustration of the modified Ravitch procedure.

difficult. Rectus abdominus muscle insertions are also divided as needed to expose the inferior cartilages.

Once fully exposed, the subperichondrial resection of the costal cartilages can be achieved by incising the perichondrium anteriorly. A plane can be created between the perichondrium and costal cartilage allowing for complete subperichondrial resection. The difficulty is in establishing the correct plane since it is very easy to incise through the perichondrium and into the cartilage. The subperichondrial dissection is most easily done with a freer-type elevator commonly found on neurosurgical instrument sets. The edges of the incised cartilage should be grasped with mosquito clamps to provide countertraction to facilitate the dissection. The most difficult part occurs in coming around the curvature of the cartilage to carry the dissection posteriorly. Care should be taken to avoid penetrating the posterior perichondrium as this layer should remain in place. The cartilage must be disconnected from the sternum carefully by placing a perichondrial elevator posterior to the costal cartilage as it is divided sharply or simply disarticulated. This prevents injury into the mediastinum. The divided cartilage can then be held with an Alice clamp or similar instrument, elevated out of the perichondrial bed and the costal cartilage can be completely removed from the perichondrium at the costochondral junction. This is executed bilaterally from costal cartilages 3 through 7 and facilitated with the understanding that costal cartilage anatomy for numbers 3 and 4 are flat and broad, circular for numbers 4 and 5, and the 6th and 7th are generally narrow and deep. Depending on the extent of the deformity, it may not be necessary to remove every cartilage between the third and the seventh. Once complete subperichondrial resection of all involved costal cartilage is achieved and hemostasis is assured, a wedge osteotomy can be created anteriorly generally just above the level of the third costal cartilage, but more precisely above the level of the deformity so that the anterior movement allowed by the osteotomy corrects, and potentially even slightly overcorrects, the excavatum deformity. Two transverse sternal osteotomies are created through the anterior cortex so as to fashion a wedge osteotomy with the intervening wedge segment and the anterior cortex removed. The base of the sternum and rectus muscle flaps are elevated with towel clips, and the posterior plate of the sternum is fractured behind the wedge osteotomy. The size and shape of the xiphoid process determines whether all or a portion of it needs to be excised, the sternum is elevated anteriorly almost to overcorrection, and a retrosternal metal strut is placed posterior to the sternum to support and fix the sternum in the corrected position. The retrosternal strut is secured with sutures to the ribs bilaterally to prevent strut migration, and the anterior incisions of

the perichondrium can be closed with absorbable sutures though realistically many of these do not allow for closure. A suction drain is brought through the inferior skin flap and placed in a parasternal position prior to the pectoral muscle flaps being secured back to the midline position on the sternum. Usually, a second suction drain is placed under the skin flap and the skin is then closed in layers with absorbable suture and a cosmetic subcuticular skin closure completes the procedure. The suction drain is removed once the drainage is <15 ml over an 8-hour shift. The retrosternal struts are generally removed 6 to 12 months after repair, which allows for enough time for solid fixation of the sternum in the correct position and for fibrosis of the chest wall where the subperichondrial resection of cartilage were carried out. The retrosternal strut or struts are removed generally as an outpatient procedure through small lateral incisions placed over each end of the strut.

In this period of time, the retrosternal metal strut that is in place differs significantly from that required with the Nuss procedure, where the retrosternal stainless steel bar is required to remain in place for 2 to 4 years to allow for costal cartilage and chest wall remodeling.

A modification of the Ravitch procedure initially described by Eric Fonkalsrud involves much more limited cartilage resection significantly reducing both the time and extent of the operative procedure. This modification requires only that small pieces of parasternal and costochondral cartilage be removed in a subperichondrial fashion as opposed to the entire costal cartilage at each level. This limited cartilage resection accomplishes the same purpose in that it allows for the sternum to be completely mobilized and elevated anteriorly achieving the desired correction once an anterior wedge osteotomy is made. In addition, the medial and lateral resection of pieces of cartilage at each level allows the cartilages to "drop back" to a normal anatomic location. The carinatum deformity may also be corrected using this technique. In these patients, once the limited cartilage resection is done at each level a simple transverse osteotomy is made across the anterior sternum at the desired level, and a wedge of autologous costal cartilage is placed within the osteotomy to depress the sternum to the desired location.

When correcting an excavatum deformity once the sternum is elevated anteriorly, a metal strut can be placed retrosternally to hold the corrected sternal position in place and subsequently removed in a minor procedure 1 year later as described above. Similarly, to stabilize the repair for a carinatum deformity a sternal bar is placed on the now posteriorly located sternum and a suture placed through the sternum and attached to the bar. Alternatively, modern orthopedic fixation using low profile plates and screws may be used for sternal fixation in either excavatum or carinatum deformities, thus avoiding the need for placement of a bar that needs to be removed. Several systems exist for this type of fixation specifically those designed for sternal closure following standard median sternotomy (SternaLockBlu system, Biomet, Inc. Warsaw, IN)

POSTOPERATIVE MANAGEMENT

In conjunction with our pediatric anesthesiologists, it is our preference to perform a one-time lumbar intrathecal injection of morphine at the time of operation. Patients are extubated in the operating room and recovered in the intensive care unit for the specific purpose of maintaining optimal pain control and relaxed breathing. Our general pain management strategy combination includes a narcotic, an anti-inflammatory, and a muscle relaxant. Initially, pain control is maintained with continuous and demand PCA narcotic infusion. Patients remain on bed rest immediately postoperatively with the head of the bed elevated. On postoperative day 1, the anti-inflammatory analgesic, Ketorolac (Toradol), and muscle relaxant cyclobenzaprine (Flexeril) are added to the pain control regimen. For patients not tolerating a clear liquid diet, IV diazepam may be substituted for the oral muscle relaxant, if necessary. Patients are given strict instructions regarding bending at the waist only and are out of bed to a chair and ambulate with assistance at least 3 times daily. Incentive spirometry use is encouraged every hour while awake and remains mandatory. The oral diet is advanced as tolerated. By postoperative day 2 to 3, as diet is advanced, the oral combination acetaminophen and hydrocodone (Vicodin) is instituted and the demand or continuous infusions of narcotic is weaned. Ibuprofen is substituted for the Ketorolac. Physical activity in the hospital, initially under the supervision of a physical therapist, is progressive but limited by bending at the waist and consists of regular ambulation including stair climbing and a range of motion exercises of the arms and shoulders. Movement strategies to promote comfort at home are taught. Patients are discharged home with the combination of an oral narcotic analgesic, a nonsteroidal anti-inflammatory agent, a muscle relaxant, and a stool softener to assure analgesia and comfort is adequately addressed. Deep breathing exercises and incentive spirometry continues at home as does regular ambulation and use of stairs. Patients and parents are instructed to limit all strenuous activity, including strenuous exercise, bending, and/or lifting object from the floor. The wearing of backpacks is initially prohibited. Our length of inpatient stay averages between 5 and 7 days and the first outpatient visit generally occurs around postoperative day 14. At that time, pain control and range of motion are assessed and adjusted. Oral narcotics, muscle relaxants, and anti-inflammatories are weaned as tolerated, and specific exercises designed to increase the range of motion are advanced.

COMPLICATIONS

Complications resulting from either surgical approach should be rare but include wound infection and pneumothorax. The most frustrating complication resulting from pectus repair is recurrence of the deformity. In large series with adequate follow-up, recurrence is reported to occur as frequently as 5% to 15%. Progressive deterioration of the repair overtime is described particularly during the interval of rapid growth at puberty, if the operation has been performed too early in younger patients. The use of strut fixation and bar stabilizers to avert migration and displacement of the bar with growth and delaying repair until the child is well into pubertal growth spurt might help delay or avoid recurrence (Figs. 34.5 and 34.6). Growth of the chest wall during pubertal and axial growth spurt while the bar is in place may also aid in remodeling the anterior chest wall and thoracic cage and prevent subsequent recurrence. Rigid fixation is uniformly applied in patients with Marfan's Syndrome because of their well-recognized high risk of recurrence.

A summary of complications resulting from the minimally invasive Nuss procedure is noted in Tables 34.1 and 34.2. Early complications include pneumothorax and hemothorax, well recognized in this group. Late complications are minimal but are generally associated with bar displacement most of which occurred prior to the use of bar stabilizers that fixate the bar to the chest wall. Several studies have been published that attempt to evaluate both the minimally invasive Nuss and open Ravitch procedures both in terms of outcomes and specifically looking at complications. A 2010 meta-analysis performed by Wales et al.

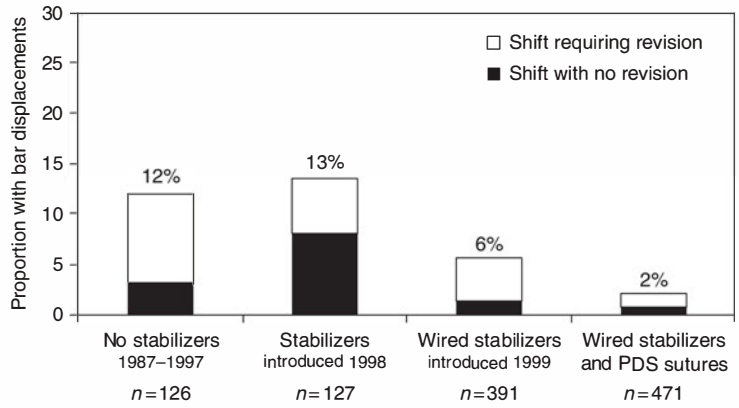


Fig. 34.5. Improvement in bar displacement rate following introduction of bar stabilizers and pericostal sutures. With permission from Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252(6):1072-1081.

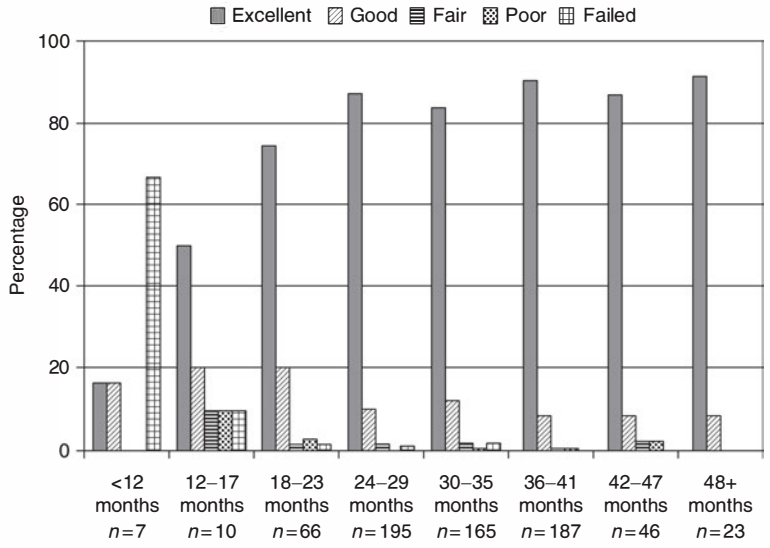


Fig. 34.6. Long-term results by length of time bar kept in situ showing that removal before 24 months has a higher recurrence rate. With permission from Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252(6):1072-1081.

Table 34.1 Early Postoperative Complications of Primary Repair

Pneumothorax with spontaneous resolution	64.7% (n = 727)
Pneumothorax with chest tube	4.0% (n = 45)
Horner's syndrome	15.5% (n = 174)
Drug reaction	3.2% (n = 36)
Suture site infection	1.0% (n = 11)
Pneumonia	0.5% (n = 6)
Hemothorax	0.5% (n = 6)
Pericarditis	0.5% (n = 5)
Pleural effusion (requiring drainage)	0.3% (n = 3)
Death	0%
Cardiac perforation	0%

With permission from Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252(6):1072-1081.

Table 34.2 Late Postoperative Complications

Bar displacements—total	64/1123 (5.7%)
Bar displacements requiring revision	45/1123 (4.0%)
Overcorrection (none required surgery)	41/1123 (3.7%)
Bar allergy (3 required bar removal)	35/1123 (3.1%)
Recurrence	11/1123 (1.0%)
Bar infection—total	6/1123 (0.5%)
Bar infection—required early removal	3/1123 (0.3%)
Hemothorax (post traumatic)	4/1123 (0.4%)
Lactosorb stabilizer inflammation	4/1123 (0.4%)

With permission from Kelly RE, Goretsky MJ, Obermeyer R, et al. Twenty-one years of experience with minimally invasive repair of pectus excavatum by the Nuss procedure in 1215 patients. Ann Surg 2010;252(6):1072-1081.

employed a systematic review to compare complication rates from both the Nuss and Ravitch procedures. This paper found nine noncontrolled studies (eight retrospective cohort series) and one prospective study published between 2001 and 2009 that directly compared the open Ravitch operation with the minimally invasive Nuss procedure. Overall, there was no difference noted between the techniques with regard to overall complication rates. Secondary outcomes such as duration of hospitalization, duration of surgery, time to ambulation, postoperative pain management, and patient's satisfaction were assessed and demonstrated a procedure-specific complication profile.

Specific complications, such as rate of reoperation was higher after the Nuss procedure compared to the Ravitch procedure and related mainly to bar migration particularly in the era prior to bar stabilizers being routinely used. Bar migration has been significantly diminished with the widespread usage of bar stabilizers. Postoperative pneumothorax and hemothorax occurred with slightly greater frequency in the Nuss group; however, there was no difference between the two groups in requirement for blood transfusion. The operative duration was nearly 70 minutes longer in the Ravitch group. There was no difference with regard to length of hospitalization or time to ambulation. Postoperative pain management strategies were

numerous and thus pain-related complications are very difficult to ascertain. Combinations of intravenous, oral, and epidural analgesia were used with similar results. Finally, only two studies used any sort of assessment to measure patient satisfaction. Unfortunately, they used different instruments to assess satisfaction and thus a definitive conclusion remains difficult. Recently, investigators associated with the Nuss group published their patient satisfaction surveys as part of their 21-year experience that includes nearly 800 patients who completed over 1,400 surveys. Over 90% of patients reported that they were either "very happy" or "happy" with their results. Only 1% of patients were "not at all happy" and over 90% of parents were "very happy" or "happy" with their child's outcome.

SUGGESTED READINGS

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EDITOR'S COMMENTS

The authors have described a learned approach to chest wall deformities. They have done an excellent job at discussing the two major procedures done. There certainly has been a lot of interest in the minimally invasive Nuss procedure. However, the recurrence rate is high for this procedure and the bands need to be placed for a significant period of time. The discussion about the specific technologic aspects of both repairs certainly gives the reader an opportunity to understand the best approach for an individual patient.

An area that was not discussed occurs fairly commonly in my practice. Patients with Marfan syndrome often have chest wall deformities and aortic dilation. This needs to be taken into account prior to the chest wall procedure. Our preference is to fix the aortic root if needed prior to any major sternal procedure. In addition, one has to be very careful in these patients in terms of preventing cardiac compression after closure of the deformed sternum.

ILK

35 Repair of Paraesophageal Hernia

Arjun Pennathur, Matthew J. Schuchert, and James D. Luketich

INTRODUCTION

Paraesophageal hernias represent subtypes of hiatal hernia. The most common form of hiatal hernia is the simple or sliding (type I) hiatal hernia (95%), in which the gastroesophageal (GE) junction migrates above the diaphragmatic hiatus, frequently associated with incompetence of the lower esophageal sphincter (LES). The remaining forms of hiatal hernia can be classified as paraesophageal (5%). Type II paraesophageal hernias are rare but are characterized by the position of the GE junction below the diaphragm, with a portion of the fundus and greater curvature of the stomach migrating through a hiatal defect alongside the esophagus. In type III paraesophageal hernias, the GE junction and fundus are displaced superiorly, with protusion through a hiatal defect (Fig. 35.1). Type IV hernias are defined as herniation of the entire stomach, omentum, and/or other intra-abdominal organs such as the transverse colon, and spleen into the mediastinum. The characteristic anatomic defects of paraesophageal hernias include enlargement of the diaphragmatic hiatus and abnormal laxity of the gastrosplenic and gastrocolic ligaments, allowing migration of the stomach (and other abdominal contents) into the chest. Giant paraesophageal hernias (GPEHs) are defined by the presence of greater than one-third of the stomach within the chest.

The actual incidence of paraesophageal hernia is unknown. It is estimated that paraesophageal hernias make up 3% to 15% of all hiatal hernias, producing an estimated incidence of 15 to 45 per 100,000 individuals within the general population. Symptoms of paraesophageal hernia frequently include obstructive symptoms, such as dysphagia, and reflux symptoms (heartburn, regurgitation, etc.). Chest pain (especially postprandial) is a common finding and is frequently mistaken for anginal symptoms. Postprandial distress, nausea, bloating, and anemia are also commonly encountered. A relatively asymptomatic but insidious symptom is occult gastrointestinal bleeding. Anemia is reported in 20% to 30% of patients with paraesophageal hernias in published series, but the rate of blood loss is slow and is rarely associated with hemodynamic compromise. A significant fraction of patients with paraesophageal hernia are asymptomatic or complain of only minor symptoms. The exact proportion of such patients is difficult to estimate for obvious reasons. Interestingly, it has been estimated that up to 89% of patients denying symptoms will actually describe some symptoms when questioned carefully.

Symptoms can be progressive, however, and can potentially result in catastrophic complications. Mortality from strangulation may be >50%, depending on the patient's age and the potential for delay in diagnosis. This has led many to argue that symptomatology is not a reliable predictor of who might progress to acute complications, and that elective surgery should be performed on most patients with a GPEH, in particular, those with organoaxial rotation as seen on a barium esophagram. Prompt elective repair after diagnosis has been recommended to avoid the development of such complications. When the repair is performed electively, excellent control of symptoms (>90%), with a low perioperative mortality of <1% to 2%, can be achieved.

Despite the foregoing observations, the beliefs held by surgeons are still largely based on small patient series and anecdotal case reports. There is little doubt that patients presenting with obstructive symptoms, bleeding, or both should undergo elective hernia repair. However, surgical correction of truly asymptomatic or minimally symptomatic paraesophageal hernias is controversial. Some studies suggest that asymptomatic patients have an 85% annual probability of remaining asymptomatic. This implies that about one in six patients will develop new symptoms, which can then be treated with elective repair. The probability that a patient's initial presentation would require an emergency operation has been estimated to be only 1% per year. Although mortality of an emergency operation has been reported as high as 50%, a pooled analysis of the largest series demonstrated an aggregate operative mortality rate of 17%. In a larger analysis of a nationwide database, the mortality rate for emergency operation for paraesophageal hernia (n = 1035) in 1997 was only 5.4%. Thus, for a 65-year-old asymptomatic patient, who has an 18% lifetime risk of developing life-threatening symptoms (1% per year) requiring emergency surgery (5.4% mortality), the overall lifetime risk of death with observation is approximately 1%, comparable to the expected 1% to 2% mortality of elective repair. Decision analysis models have failed to demonstrate a gain in quality-adjusted life expectancy with elective laparoscopic repair of asymptomatic paraesophageal hernias when compared with watchful waiting.

When symptoms arise, the only effective treatment of paraesophageal hernias is surgical repair. The surgical technique involves reduction of the herniated contents back into the abdomen, excision of the hernia sac, and closure of the hiatal defect. These procedures can be performed transthoracically or transabdominally with laparotomy or laparoscopically. In extremely high-risk patients with minimal symptoms, an anterior gastropexy can be performed to help prevent the development of incarceration or organoaxial volvulus. In the absence of reflux symptoms, the need to perform an antireflux procedure has been debated. However, up to 60% of patients with type III hernias have also been shown to have hypotensive LES pressures and abnormal 24-hour pH-monitoring studies. In addition, with the repair of large paraesophageal hernias, there is significant dissection performed around the hiatus, the phrenoesophageal ligament is divided, and the lower esophageal antireflux mechanism is affected; thus, many surgeons (including us) recommend routinely performing an antireflux procedure during elective repair. The use of partial (Belsey, Dor, and Toupet) or complete (Nissen) wraps has been reported, each with good results. The choice of wrap is determined by the individual patient's underlying anatomy,



Fig. 35.1. Barium esophagram demonstrating a type III paraesophageal hernia.



Fig. 35.2. Barium esophagram demonstrating acquired shortening of the esophagus.

symptom complex, and esophageal motility in addition to the surgeon's preferences.

PREOPERATIVE EVALUATION

Preoperative assessment includes a barium esophagram in order to definitively establish the diagnosis (Fig. 35.1). The barium esophagram is a critical component of evaluation and demonstrates the paraesophageal hernia; it can also give an estimation of esophageal length in addition to providing information on the presence of strictures or other sequelae of GE reflux. Upper endoscopy is also critical and allows assessment of the gastric anatomy, location of the GE junction, assessment for short esophagus (Fig. 35.2), esophagitis and other complications of reflux disease, and potentially excludes other abnormalities such as Barrett's esophagus, or neoplasia. It is also important to consider performing esophageal function studies. Esophageal manometry can be particularly useful for the evaluation of esophageal motor function, the presence of peristalsis, evaluation of the amplitude of contractions, and may influence the choice of fundoplication; in addition, it may allow the assessment of esophageal length (Maziak et al.,). However, it should be noted that manometry and 24-hour pH studies may not be feasible because the degree of anatomic distortion frequently interferes with the reliable performance of these studies. When performed, it should be done with caution in view of the anatomic distortion, and in a few centers the catheter is placed with endoscopic guidance. Patients also undergo a complete cardiopulmonary evaluation for clearance and risk assessment prior to repair of the paraesophageal hernia. Active pulmonary infection is treated prior to surgery, and patients are advised to stop smoking.

SURGICAL APPROACHES

Traditionally, the repair of a GPEH has been performed through an open laparotomy or thoracotomy. Increasingly, the repair of GPEH is being performed using minimally invasive techniques with a laparoscopic approach. In this chapter, we discuss the laparoscopic approach and the transthoracic approach for repair of GPEH.

Laparoscopic Repair of Giant Paraesophageal Hernia

In the operating room, after intubation, we start with the esophagogastroduodenoscopy (EGD) performed by the surgeon for confirmation of previous endoscopic findings. It is important not to insufflate too much air at the beginning of the procedure, since this will interfere with the laparoscopy. Standard laparoscopic port placement is used as shown in Figure 35.3. To facilitate the ease of mediastinal dissection, the abdominal port incisions can be shifted slightly cephalad in the case of a GPEH. The left lateral segment of the liver is retracted anteriorly with a 5 mm flexible retractor (Snowden Pencer, Genzyme, Tucker, GA) and secured to a stationary holding device (Mediflex, Islanda, NY). Dissection is begun by identifying the sac, dissecting the sac, and subsequently inverting the mediastinal hernia sac and pulling it back into the abdomen alongwith the herniated stomach (Fig. 35.4). Avoidance of direct traction on the stomach by using a "hand over hand" technique results in less trauma to the stomach

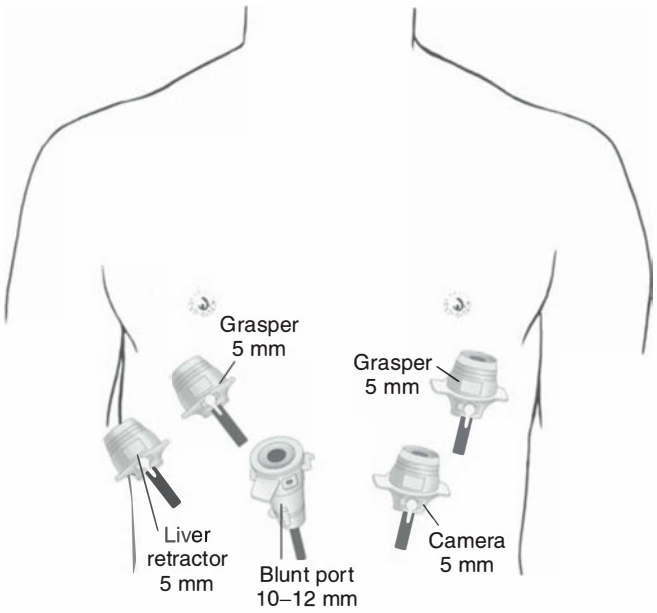


Fig. 35.3. Standard port placement for paraesophageal hernia repair and laparoscopic antireflux procedures.

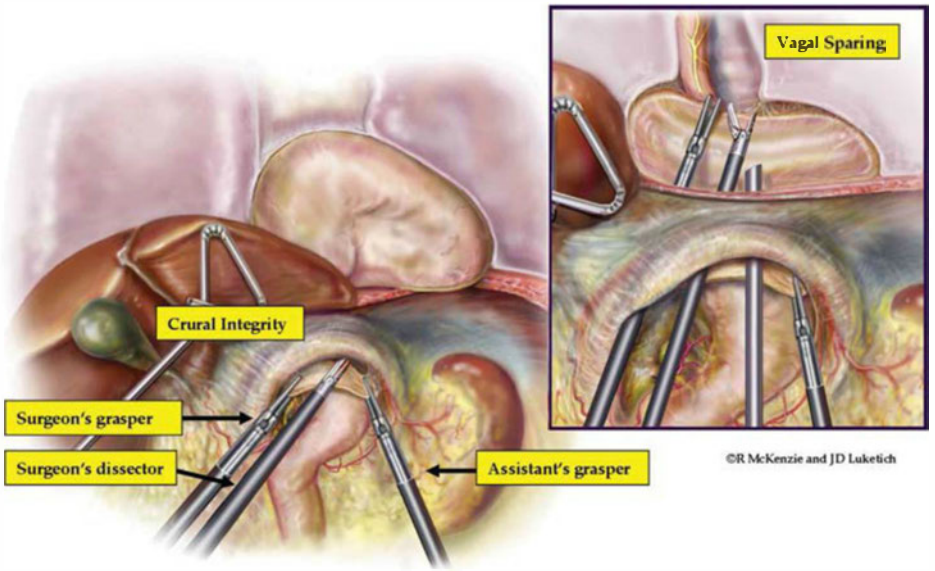


Fig. 35.4. Reduction of the hernia sac without direct grasping and traction on the stomach itself.

while reducing the herniated intrathoracic stomach. The crural reflection of the sac is incised, and the flimsy mediastinal attachments to the sac are carefully taken down by the harmonic scalpel (Ethicon, Cincinnati, OH) or the ultrasonic shears (U.S. Surgical Corp, Norwalk, CT). (Fig. 35.5). Care is taken to identify and preserve the proximal anterior and posterior vagus nerves during this portion of the dissection. Similarly, the adjacent pleura must be identified and swept laterally away from the plane of dissection. The surgeon and the anesthesiologist must communicate closely during this portion of the procedure because a sudden drop in blood pressure or significant increase in inspiratory pressures may indicate the development of a tension pneumothorax, which can be readily treated by placement of a pigtail catheter or a chest tube. Once the sac is completely freed up it, along with the contents, is reduced back into the abdomen and the redundant sac may be carefully excised avoiding injury to the vagi (Figs. 35.5 and 35.6). The complete dissection and reduction of the hernia sac with its contents from the chest to the abdomen is a critical component of successful surgery for GPEHs. With the delivery of the sac into the abdomen, the herniated stomach (and other herniated intra-abdominal structures) is reduced back into the peritoneal cavity. This portion of the case is also critical in achieving maximal esophageal mobilization and a tension-free repair. Inferior traction on the gastric fundus and epiphrenic fat pad is then performed to allow complete dissection of the right and left crus. A retrogastric/retroesophageal window is then created to expose the posterior portion of the left crus. At this point, the division of the short gastric vessels is performed in order to fully mobilize the fundus of the stomach.

Assessment of Short Esophagus

In the setting of a GPEH, careful identification of the GE junction after fat pad excision frequently reveals a shortened esophagus. The esophageal fat pad is carefully and completely mobilized medially, sweeping the anterior vagus to the right of the esophagus (Fig. 35.7). The distal esophagus is then mobilized at the level of the diaphragmatic hiatus circumferentially, and the surgeon then determines whether esophageal shortening is present. The proper assessment of esophageal length can be tricky because there is a tendency to overestimate the intra-abdominal length of the esophagus because of the diaphragmatic elevation caused by the pneumoperitoneum as well as the downward traction that is applied to the stomach. If the GE junction does not remain below the diaphragmatic hiatus with an adequate segment of tension-free intra-abdominal esophagus (ideally 2 to 3 cm), then further mediastinal mobilization of the esophagus should be performed, extending higher into the mediastinum to the inferior pulmonary vein, and in extreme cases, the dissection can be carried significantly higher if needed to gain additional esophageal length. The esophageal length should again be reassessed, with the goal of an adequate, tension-free intra-abdominal esophageal segment of 2 to 3 cm. If after extensive mediastinal mobilization, there is inadequate esophageal length, we then perform a Collis gastroplasty prior to fundoplication. This can be done with an end-to-end anastomosis (EEA) stapler (Figs. 35.8–35.10) or a GIA stapler. At present, we prefer to perform the Collis gastroplasty with a wedge gastroplasty technique, using a GIA stapler (Figs. 35.11 and 35.12).

As mentioned earlier, we routinely perform a fundoplication to prevent reflux since it is highly likely that the preceding dissection has disrupted the function of

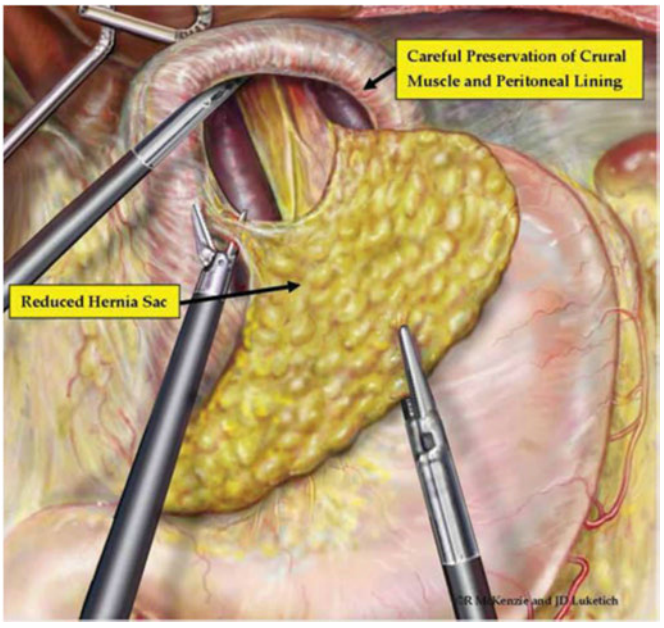


Fig. 35.5. Complete reduction of the herniated stomach back in the abdomen, with complete reduction of the hernia sac.

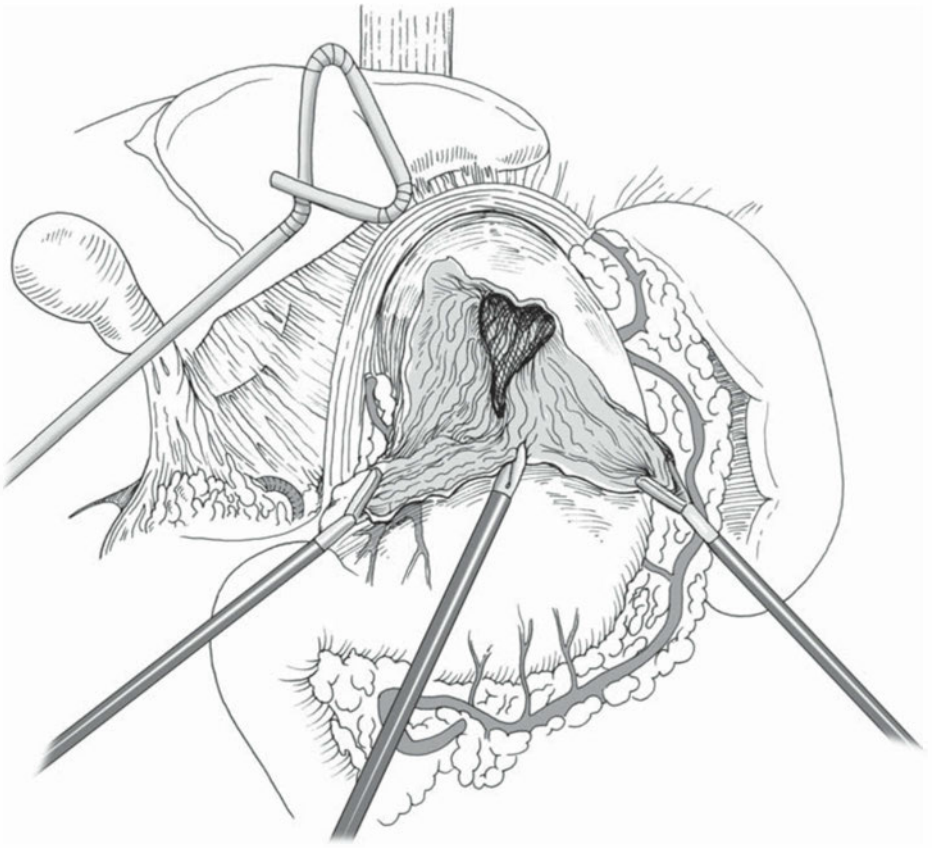


Fig. 35.6. Excision of paraesophageal hernia sac.

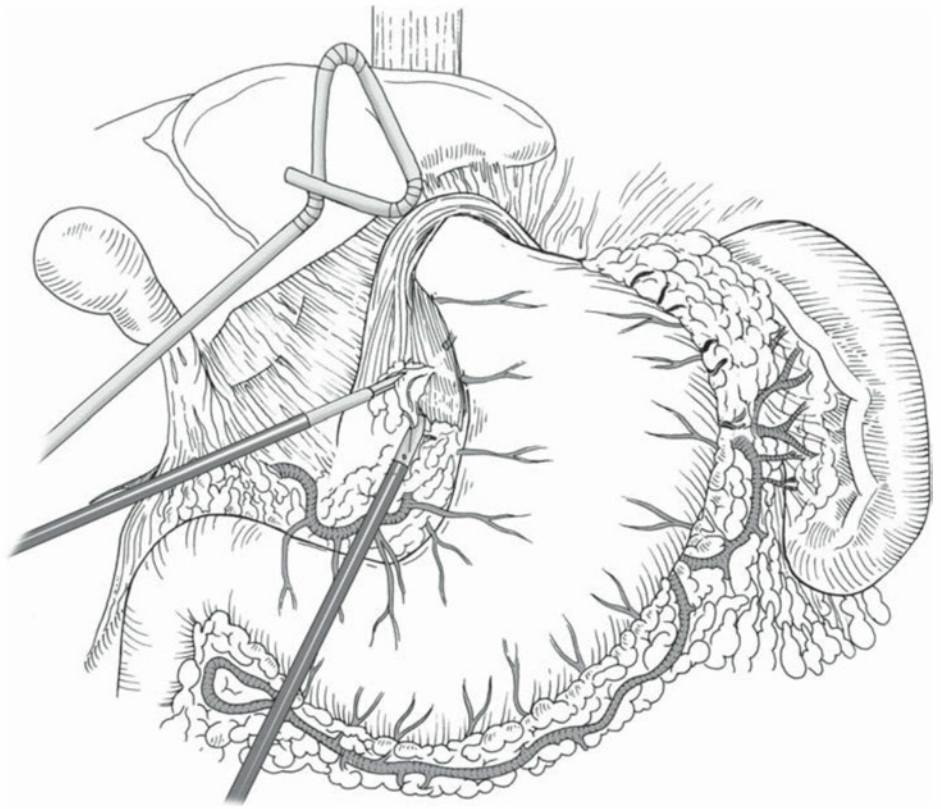


Fig. 35.7. Mobilization of the esophagogastric fat pad.

the LES. The choice of the type of fundoplication depends on the results of physiologic testing. In general, we prefer a floppy Nissen fundoplication (Fig. 35.13). This is performed after the surgeon places an esophageal bougie (54F), with direct laparoscopic vision of the distal esophagus, the GE junction, and the stomach. A floppy Nissen fundoplication is performed, and the bougie is removed. In brief, after complete mobilization of the distal esophagus and stomach, and determination of the location of the GE junction after dissection of the GE fat pad, an atraumatic instrument is passed in the retroesophageal window. The fully mobilized fundus is grasped and brought through the retroesophageal window with proper orientation. When the fundus is fully mobilized, there should be no or minimal tension and no tendency for the fundus to retract back through the retroesophageal window. A "shoe shine maneuver" is then performed to evaluate the tightness of the wrap, its mobility, and length. Subsequently, a floppy Nissen fundoplication is performed with space behind the wrap and the esophagus to allow the passage of an atraumatic instrument. Both the Collis gastroplasty and the fundoplication may be omitted in some high-risk patients, and instead a gastropexy is performed to anchor the stomach to the abdominal wall. While some have performed this as a single point or with the placement of a gastrostomy tube, we perform the gastropexy with several interrupted heavy mattress sutures anchoring the stomach to the diaphragm and the anterior abdominal wall. We start these sutures anchoring the stomach to the left crura and then to the anterior abdominal wall.

Approximation of the Crura

During the dissection, it is critical to maintain the peritoneal lining in an effort to preserve the integrity of the crura, and fully mobilize the crura. The crura are fully mobilized by dividing the phrenogastric, phrenosplenic, and retrogastric attachments. This mobilization, along with complete mobilization of the hernia and sac, allows the crura to be approximated primarily without excess tension in most cases. In some patients where crural tension is still present, inducing a small, left-sided pneumothorax may yield a "floppy diaphragm sign", which allows a much easier tension-free primary repair. This should be done in close coordination with anesthesia, and subsequently, the surgeon can place a small pigtail catheter and eliminate the pneumothorax. This catheter can then be removed

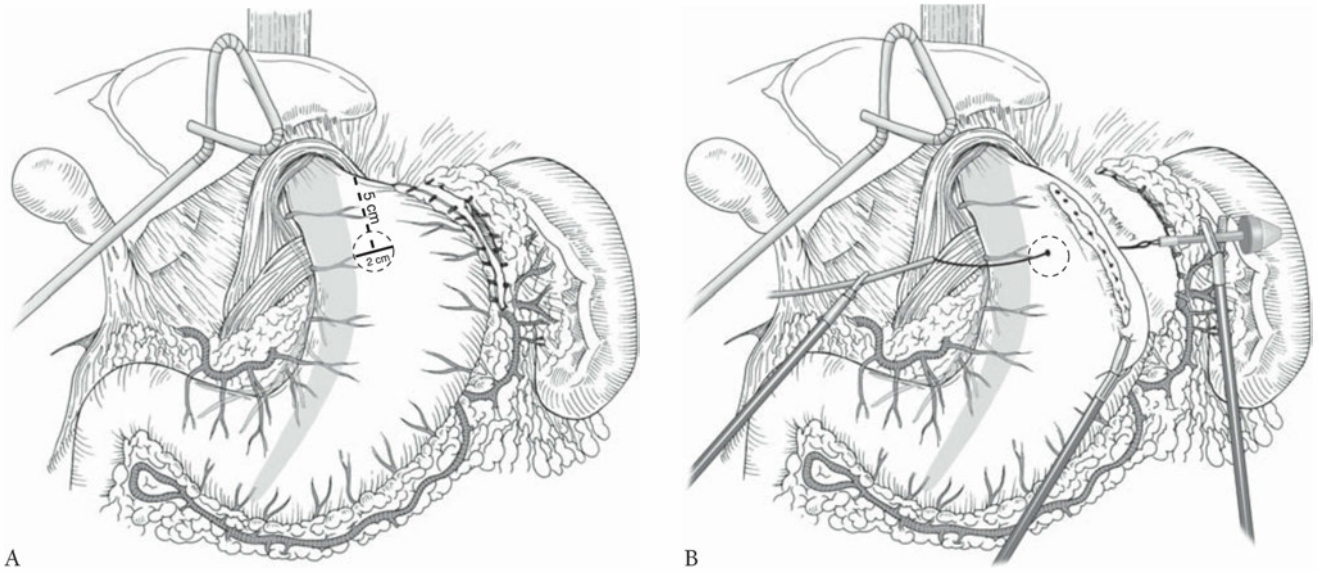


Fig. 35.8. Position of bougie (**A**) and delivery of EEA anvil (**B**) during Collis gastroplasty.

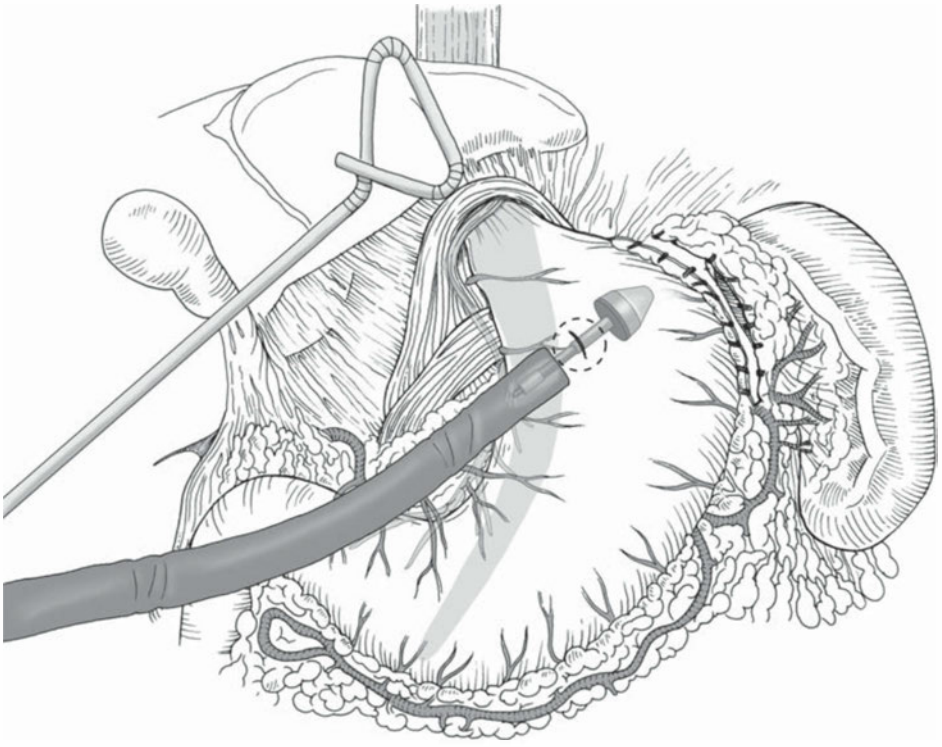


Fig. 35.9. Creation of circular gastric staple line with a 25 mm EEA stapler.

very early in the postoperative course. In unusual cases of an excessively large defect, a patch of Surgisis (Cook, West Lafayette, IN) is used to reinforce the closure. The crura are reapproximated posteriorly to complete the surgical procedure. The crural repair is performed with typically two or three heavy nonabsorbable braided sutures (0 Surgidac) placed posteriorly. We do not routinely use pledgets for the crural repair. We then reassess the hiatus, and if there is a significant anterior space and the hiatus appears wide, we place crural sutures anteriorly. The hiatus is reevaluated and the surgeon should be able to easily introduce the grasper with a 1 cm gap between the esophagus and the crura, with care being taken not to cause excessive narrowing of the hiatus, which may result in postoperative dysphagia.

Before closing, endoscopy is routinely performed with intraluminal insufflation to rule out esophageal or gastric leaks. A nasogastric tube is carefully placed under direct laparoscopic guidance. A barium swallow is obtained on postoperative day 1 to evaluate the repair and verify the absence of a leak; the nasogastric tube is typically removed on postoperative day 1. If no leak is present, clear liquids are started. If a clear liquid diet is tolerated, the patient is usually discharged to home on postoperative day 2. Advancement to a soft diet occurs over the subsequent 1 to 2 weeks.

Transthoracic Repair of Giant Paraesophageal Hernia

While our current primary approach for primary elective repair of paraesophageal hernia is a transabdominal laparoscopic approach, a transthoracic approach is favored by some surgeons and is a well-established technique (Maziak, Pearson; Patel, Orringer; Altorki, Skinner). The transthoracic approach is a particularly useful approach in patients with a long-standing GPEH, and those who may have a "hostile" abdomen due to multiple previous surgeries. It is useful in patients who have had multiple previous transabdominal antireflux procedures, or who have a recurrent paraesophageal hernia. The work-up for the patient who is being evaluated for a transthoracic approach is similar to that for the laparoscopic approach described earlier. A relative contraindication is previous left thoracotomy for inflammatory diseases such as empyema treated with decortication.

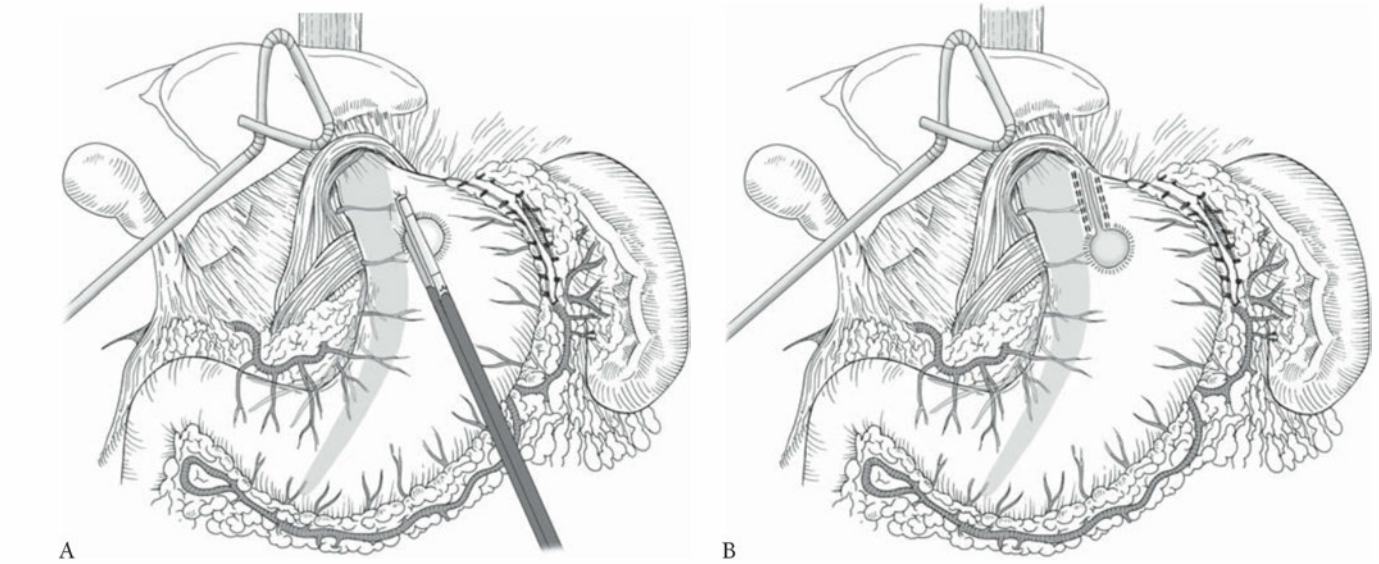


Fig. 35.10. (**A**) Completion of the neo-esophagus using a linear stapler. (**B**) Completed EEA gastroplasty.

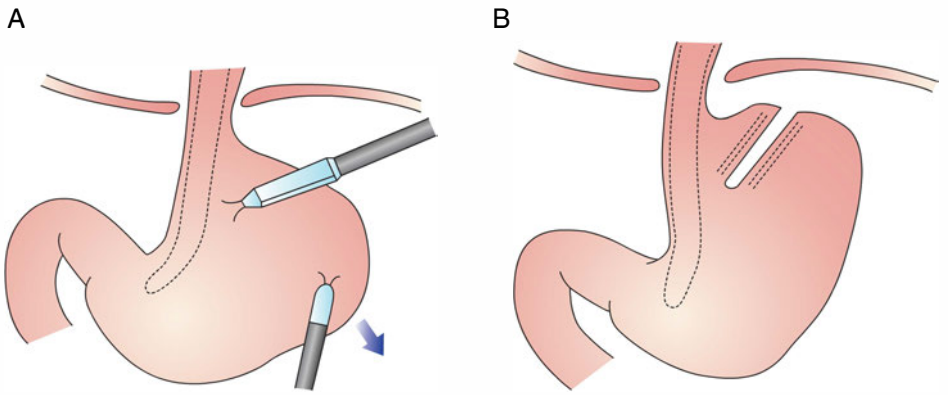


Fig. 35.11. Laparoscopic wedge gastroplasty. (**A**) The fundus is retracted inferiorly (in the direction of the *blue arrow*). The alignment of the bougie along the lesser curve is shown. The stapler is introduced through a left upper port. (**B**) The first staple line has been completed and brought snuggly to the edge of the bougie (Adapted from Luketich JD, Maddaus MA. Laparoscopic gastroplasty. In Pearson FG, Patterson GA, eds. Pearson's Thoracic and Esophageal Surgery. 3rd ed. Philadelphia, PA: Churchill Livingstone/Elsevier; 2008:326-336).

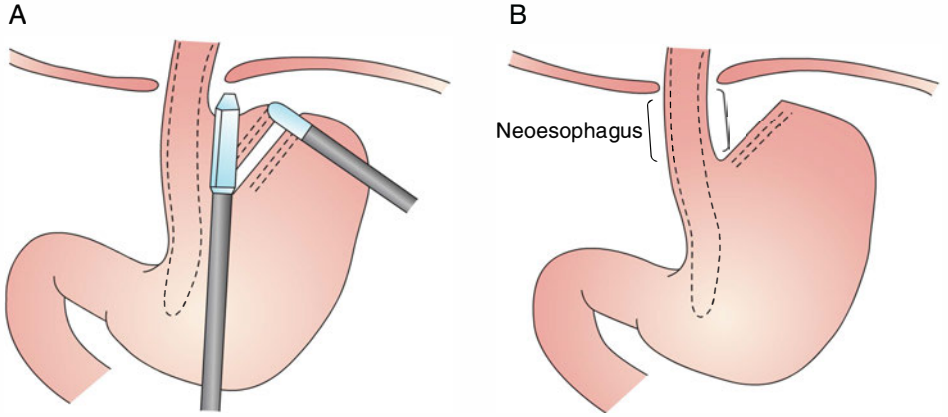


Fig. 35.12. Laparoscopic wedge gastroplasty. (**A**) The gastric wedge is transected with a stapler from the right upper quadrant. The staple line has to fit snuggly against the bougie. (**B**) Schematic of the completed gastroplasty (Adapted from Luketich JD, Maddaus MA. Laparoscopic gastroplasty. In Pearson FG, Patterson GA, eds. Pearson's Thoracic and Esophageal Surgery. 3rd ed. Philadelphia, PA: Churchill Livingstone/Elsevier; 2008:326-336).

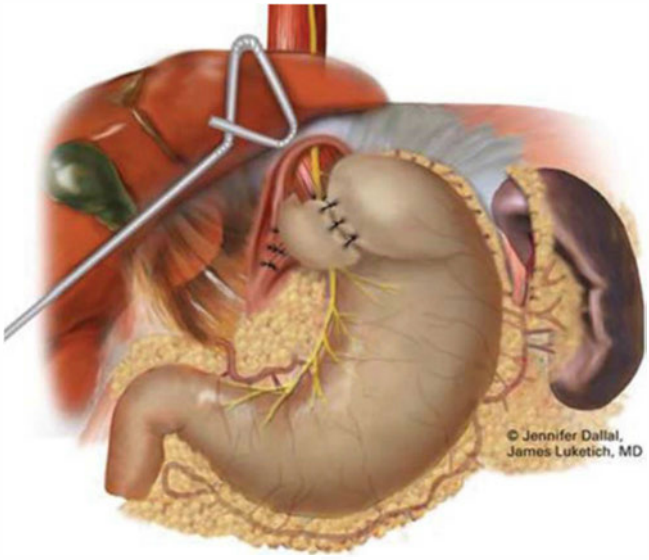


Fig. 35.13. Creation of floppy Collis–Nissen fundoplication.

In particular, for those patients who are being evaluated for a revision procedure due to recurrent paraesophageal hernia, a complete and comprehensive work-up should be undertaken. In these patients, the original indication and history along with the operative report should be thoroughly reviewed. Specific attention should be paid to the definition of the GE junction, dissection of the fat pad, preservation of the vagi, closure of the crura, size of the bougie, and the technical details of construction of the fundoplication itself. To summarize, as detailed above, our work-up generally includes EGD and barium swallow. We also obtain manometry and pH monitoring when feasible. In patients requiring redo surgery, a gastric emptying study is also typically obtained to evaluate the functional integrity of the vagus nerves.

Operative Technique

The operative technique described here is based on the technique described by Pearson and colleagues. An epidural catheter is placed to optimize postoperative pain control and a left thoracotomy is planned. The patient is intubated with a double-lumen endotracheal tube. Adequate venous access is established, and a Foley catheter and an arterial line are also placed. An on-table EGD is then performed by the surgeon. Specific note is made of hiatal hernia, esophageal length, Barrett's esophagus, esophagitis, and any other sequelae of reflux disease. The patient is placed in the right lateral decubitus position, pressure points are padded, and sequential compression devices are placed on the lower extremities. A posterolateral thoracotomy is then performed and the chest is entered through the seventh intercostal space. The latissimus is divided while the serratus is preserved. We excise a 1 cm segment of the eighth rib posteriorly. After placement of the rib spreader, the ribs are separated to a minimal extent as a 5 cm opening is usually adequate for exposure.

With isolation of the left lung, the lower lobe of the lung is retracted superiorly and the inferior pulmonary ligament is divided. The paraesophageal hernia contents enclosed by the sac are clearly visible. The mediastinal pleural overlying the esophagus is opened starting from the level of the inferior pulmonary veins and extending inferiorly. The vagi are identified here and preserved for the remainder of the dissection. The esophagus is dissected superiorly away from the sac and encircled with a Penrose drain, which is placed after circumferential mobilization, encompassing the esophagus along with the vagus nerves and this drain can be used for traction as the esophagus is mobilized (Fig. 35.14). The mobilization of the esophagus can be extended superiorly to the aortic arch. The next step is the dissection of the sac. In long-standing hernias, there may be dense adhesions between the sac and the pericardium and the lung. The sac is dissected and the adhesions between the sac and the pericardium are divided. Care must be exercised while retracting the pericardium, and hemodynamics should be monitored closely. It is best not to grasp the pericardium but use gentle traction during this dissection. The hernia sac is dissected away from the pericardium and the contralateral hemithorax, with care to avoid entry into the right pleural space. The hernia sac dissection is carried inferiorly around the hiatus and the hiatal defect is defined. Inferiorly, the esophagus is dissected between the hiatus and the inferior pulmonary vein by freeing up the mediastinal attachments. It is important to mobilize the esophagus along with vagi, with special care taken not to injure the vagi. Similarly, when dissecting the sac, one has to be careful not to injure the vagi.

After the dissection of the sac, the sac may be entered near the hiatus, and adhesions between the sac and the stomach

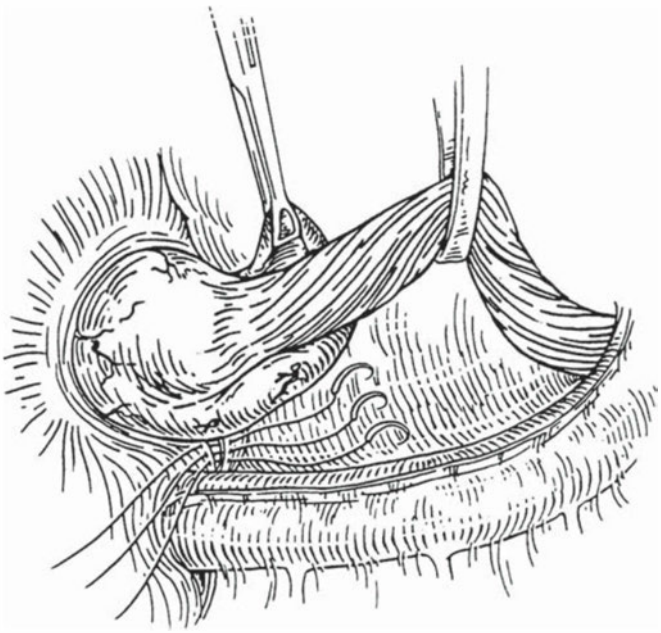


Fig. 35.14. All transthoracic fundoplications are preceded by the placement of crural stitches, which are left untied until the fundoplication wrap is completed. The fundus of the stomach is brought posterior to the esophagus in preparation for a Nissen fundoplication.

divided. The sac is opened in a circumferential manner anteriorly to posteriorly around the hiatus, adhesions within the sac divided, and the sac dissected completely off the stomach and GE junction. It is again important to emphasize the careful preservation of the vagus nerves during the dissection of the sac. In a moderate-to-large hernia, after complete dissection of the sac, the sac is excised.

The hiatus is then defined, and enlarged if needed, so as to be able to reduce the stomach along with the planned fundoplication into the abdomen. The esophagus is then freed up from the diaphragmatic hiatus to the inferior pulmonary vein. If there is tension noted when the stomach and the GE junction are reduced into the abdomen and a shortened esophagus is suspected, the esophagus should be mobilized to the level of the aortic arch. This requires ligation or cauterization of aorta–esophageal branches as the esophagus is fully mobilized. It is important to have a tension-free intra-abdominal segment of the esophagus (at least 2 to 3 cm) to assure a successful and durable repair. If there is persistent esophageal foreshortening despite complete mobilization of the esophagus, a Collis gastroplasty is performed prior to the fundoplication. In large series, the type of fundoplication described when using a transthoracic approach include a Belsey partial fundoplication (Pearson) and a transthoracic Nissen fundoplication (Orringer).

Defining the Gastroesophageal Junction, Mobilization of the Gastroesophageal Junction, and Mobilization of the Stomach

In moderate-to-large hernia, the sac is excised. The retroperitoneal attachments are then divided to mobilize the GE junction. An inconstant artery ("Belsey's Artery") communicating between the left gastric and the inferior phrenic arteries should be divided. This is typically encountered in the posteromedial dissection, and it is important to control these vessels before division. The crural decussation and crural defect are then clearly exposed.

The GE junction fat pad is then dissected along with left vagus nerve and mobilized anteriorly, and the GE junction is defined. The superior short gastric vessels are then divided to mobilize the gastric fundus, particularly if a Nissen fundoplication is planned. It is important to adequately mobilize the fundus so as to prevent any tension during construction of the fundoplication. At this stage, it is also important to make an assessment of esophageal length and assess whether a tension-free fundoplication can be performed. After reduction of the hernia, there should be a minimum of at least 2 to 3 cm of intra-abdominal esophagus without any tension. As mentioned above, if there is any concern for esophageal shortening, complete mobilization of the esophagus should be performed; a Collis gastroplasty should be performed if there is noted a tendency for the GE junction to migrate back into the chest after complete mobilization.

Esophageal Shortening and Collis Gastroplasty

If there is esophageal shortening despite esophageal mobilization, a Collis gastroplasty is performed. This is performed after careful placement of an esophageal bougie, with the bougie traversing the GE junction extending into the stomach. The greater curvature of the stomach is then grasped with a Babcock instrument and subsequently an endo-GIA stapler is applied and fired, immediately adjacent to the bougie tight to the lesser curvature of the stomach (Figs. 35.15 and 35.16). The staple line is then oversewn with continuous absorbable 3-0 sutures along the esophageal side without inverting the staple line so that the esophageal lumen is not compromised; the staple line is inverted with continuous sutures on the gastric side of the staples.

Placement of the Crural Sutures

The crural edges are clearly defined. The retraction of the stomach and the GE junction facilitates crural exposure. Generally, 0 silk nonabsorbable sutures are placed 1 cm apart in the crura. Typically, three to four sutures are placed posteriorly but not tied at this time. When there is a large hiatal defect, a couple of sutures may be placed anteriorly, so as to avoid excessive angulation of the esophagus by the placement of several posterior sutures. These crural sutures are all placed, clamped with hemostats, but not tied (Fig. 35.14).

Fundoplication

A Belsey partial fundoplication or a Nissen fundoplication is then performed. While Pearson and colleagues prefer a Belsey fundoplication, Orringer and colleagues have preferred a Nissen fundoplication.

For the Belsey fundoplication, the stomach is mobilized posteriorly into the chest, and a 54F bougie is then placed in the esophagus across the GE junction; this should be placed by the surgeon carefully under direct visualization. A 240- to 270-degree wrap is then performed as below (Figs. 35.17–35.19). The first row of mattress sutures is placed with 2-0 silk on an atraumatic needle. A total of three mattress sutures are placed in each row,

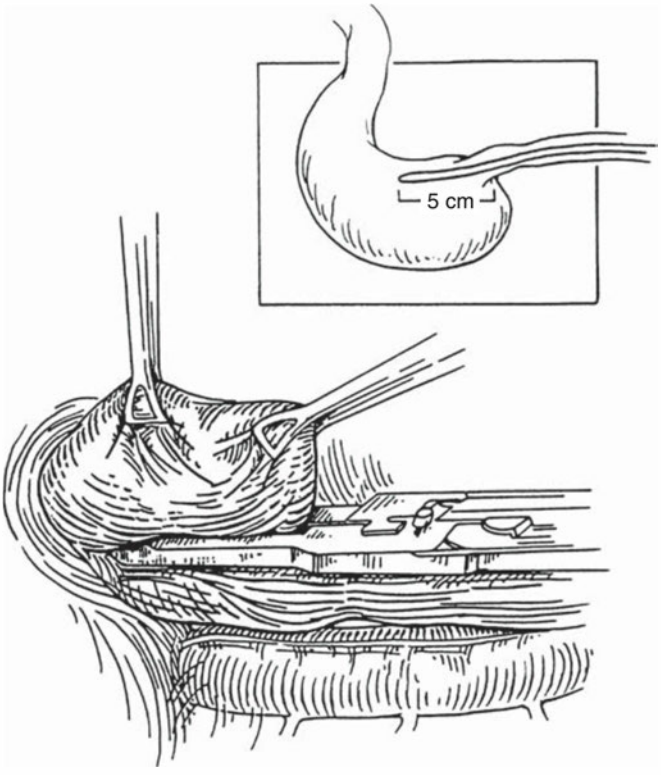


Fig. 35.15. A Collis gastroplasty is created by firing a stapler parallel to the lesser gastric curvature, snugly placed against a bougie that has been positioned across the esophagogastric junction. 4-5 cm of neoesophagus is typically created by the gastroplasty (*Inset*).

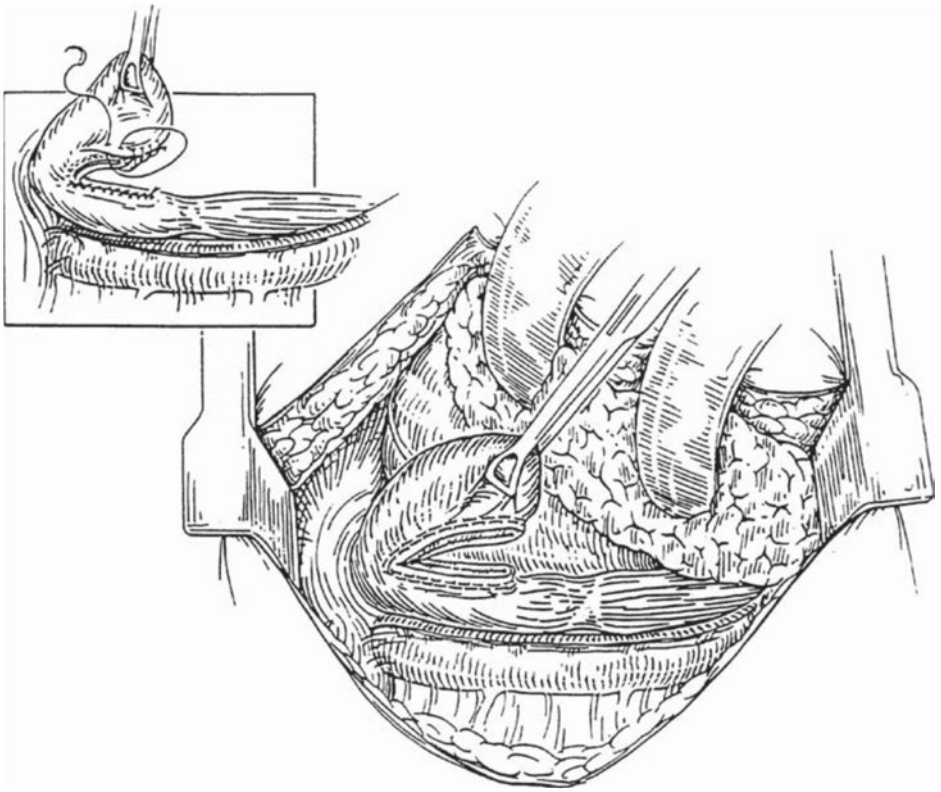


Fig. 35.16. The staple line of the gastroplasty is oversewn as described in the text, creating an extended esophagogastric tube and enlarging the gastric fundus.

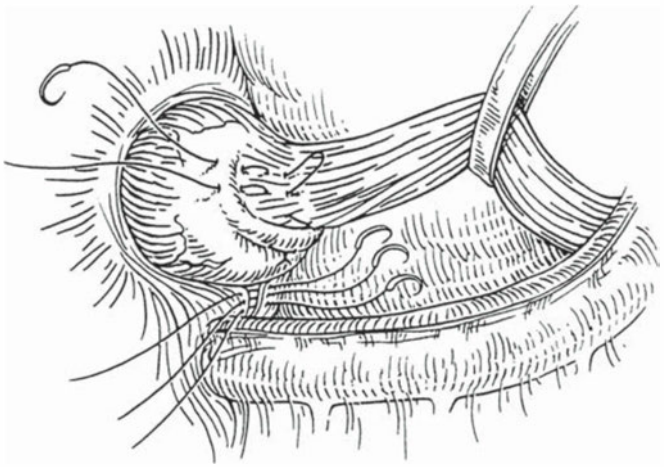


Fig. 35.17. A Belsey fundoplication is begun by mobilization of the upper stomach into the chest. The initial row of three mattress sutures incorporates 1 cm of stomach and 1 cm of esophagus and is evenly spaced between the vagus nerves. The untied previously placed crural sutures are also shown.

each about 1 cm apart. The first row of sutures is placed about 1 cm cephalad to the GE junction in the esophagus and 1 cm distal to the GE junction in the stomach, care being taken not to injure the vagus nerves. The most posterior stitch is anterior to the left vagus nerve and the most medial stitch is placed adjacent and just anterior to the right vagus nerve. It is critical to incorporate the submucosa of the esophagus and the stomach with each suture bite. These sutures should traverse the esophagus for about a 5 mm distance incorporating the submucosa, and the second arm of the first suture is placed about 0.5 cm from the first. Similarly, the second suture is placed in the midline approximately 135 degrees from the first. The third suture is then placed in a similar manner 135 degrees from the previous suture; the second arm of this suture is placed just anterior to the right vagus nerve. Once all these sutures of the first row are placed, they are tied. It is important to avoid making the sutures too tight, and this should be done more by vision than by feel.

The second row of mattress sutures is then placed incorporating the esophagus, the stomach, and the diaphragm. These sutures are again placed in a similar manner, about 1 to 1.5 cm proximal and distal to the first row in the esophagus and the stomach. It is important to avoid placing the sutures in the fundus too far apart. A modified teaspoon is then placed through the hiatus to protect the abdominal viscera, and the suture is passed through the diaphragm. These sutures are placed but left untied. The partial Belsey wrap is then gently reduced into the abdomen, the sutures are pulled up tight, removing any redundancy in the sutures, and then tied. A modification of this technique is to tie the second row of sutures after the placement of the esophageal and gastric sutures, but leave the suture ends long with the needle intact (or alternately the suture can be rethreaded into a needle). The fundoplication is then gently reduced back into the abdomen manually. The suture needles are then passed through the diaphragm with a rim of 1 to 2 cm, with a modified teaspoon in place to protect the viscera, and this suture is then tied. If a Collis gastroplasty is performed, three rows of sutures are required for the Belsey fundoplication. In this case, the third row of sutures is used to anchor the fundoplication to the diaphragm (Fig. 35.20).

If a Nissen fundoplication is planned, the fully mobilized gastric fundus, after the division of the short gastric vessels, is delivered into the chest. The decision on the use of the Collis gastroplasty is made as detailed previously. A 54F esophageal bougie is placed by the surgeon so that the bougie traverses the GE junction and the tip is positioned in the stomach. The fundus is then passed posteriorly, and a lack of tension is assured. The fundoplication is then performed with three sutures, 1 cm apart to form a 2 cm fundoplication. These sutures incorporate the fundus on the left of the esophagus, the esophageal wall, and the fundus on the opposite side (right side) of the esophagus. All these sutures are placed and then tied (Fig. 35.21). The fundoplication is then reduced into the abdomen prior to tying the crural sutures. The Nissen–Collis repair is shown in Figure 35.22.

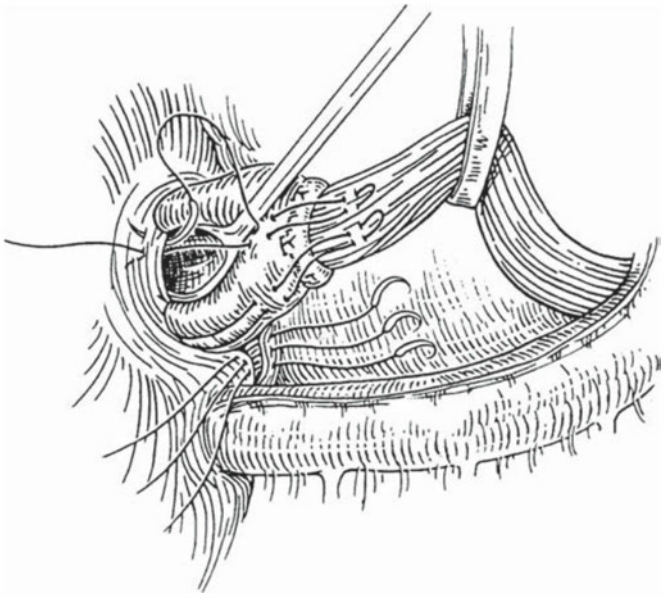


Fig. 35.18. A second row of sutures for the Belsey fundoplication is placed 1 cm from the first and incorporates the diaphragm at the beginning and at the end.

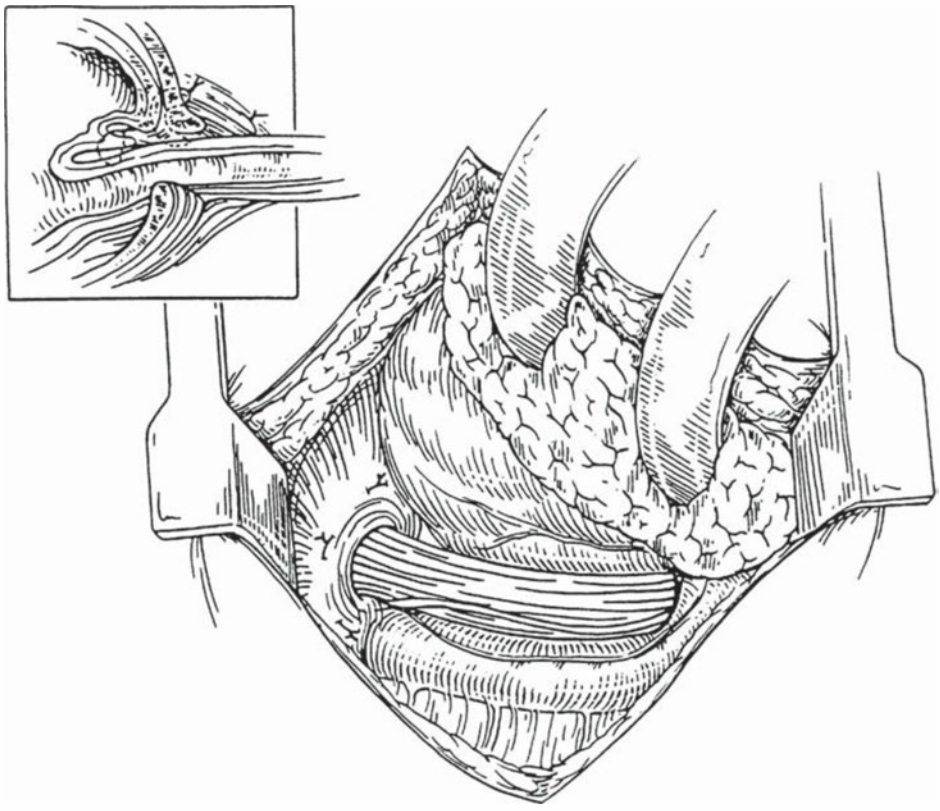


Fig. 35.19. The completed fundoplication showing the wrap anchored below the diaphragm by the second row of sutures.

Approximation of the Crura

The crural sutures are then tied, starting with the most posterior suture. Care must be taken to avoid making the repair too tight and this should be calibrated. After the crural sutures are tied, the index finger should be able to slide easily along the posteromedial aspect of the crus. A nasogastric tube is then placed and guided into the stomach by the surgeon. A 28F thoracostomy tube is placed to drain the pleural space. The thoracotomy incision is then approximated in layers.

It is important to prevent retching or vomiting in the postoperative period and to that end antiemetics should be given prophylactically. The nasogastric tube should be functional to avoid postoperative gastric distension. The patency and function of the nasogastric tube should be assured by the staff. We obtain a follow-up barium swallow and if no abnormalities are seen, and the nasogastric tube drainage is low, the nasogastric tube is removed. The patient is subsequently started on clear liquids and typically discharged on a soft diet. Stool softeners are prescribed while the patient is taking pain medications. Patients are specifically instructed not to lift weights >10 lb or strain for a minimum of 2 months.

RESULTS OF LAPAROSCOPIC REPAIR

Although laparoscopic Nissen fundoplication has become a well-established procedure in the treatment of GE reflux disease (GERD), laparoscopic management of GPEHs has been somewhat controversial. Although early studies of laparoscopic repair of GPEH were promising, concerns have been raised about the safety and efficacy of a minimally invasive approach in handling this complex problem. Dahlberg reported the Mayo clinic experience in 37 patients undergoing laparoscopic repair of GPEH. Successful repair was accomplished laparoscopically in 35 of the 37 patients. Intraoperative complications included two splenic injuries and one crural tear. Postoperatively, there were two cases of esophageal leak and one small-bowel obstruction at a port site. Mortality was 5.4%. In short-term follow-up, 4 of 37 patients (12.9%) developed recurrent paraesophageal herniation. It was concluded that laparoscopic repair of GPEH is a technically challenging operation associated with significant morbidity and mortality. Similar results were obtained in a series of 60 patients by Weichman. In this series, reoperation for recurrent herniation was required in 5.5% of patients, with an associated 1.9% mortality. This study also concluded that laparoscopic repair of GPEH is a technically challenging procedure, but that with increasing experience and technical refinements, results could approximate those of the open approach.

As surgeons have gained experience with advanced laparoscopic techniques, improved results after laparoscopic repair of GPEHs have been observed. We initially reported the University of Pittsburgh experience with laparoscopic repair of 203 consecutive GPEHs. Median follow-up was 18 months. The most common preoperative

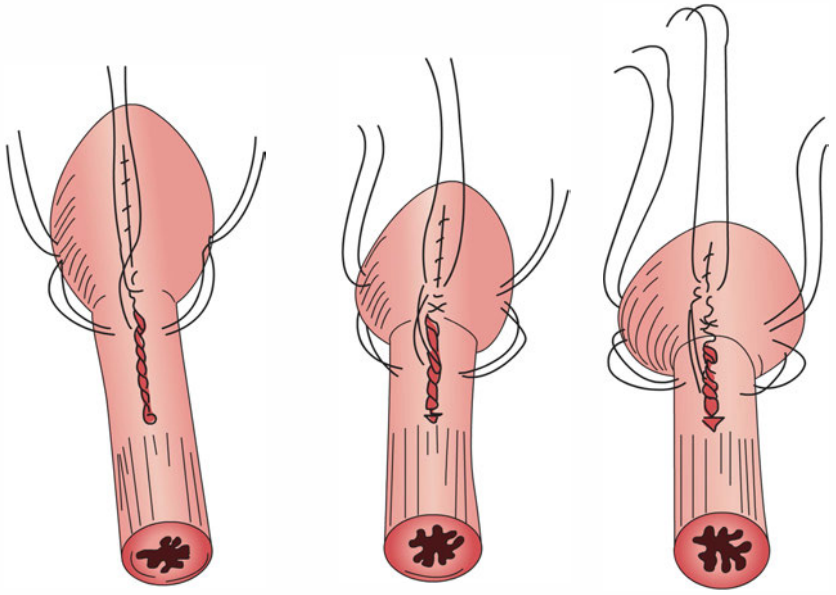


Fig. 35.20. The sequence of suturing for the 270-degree Belsey fundoplication with a Collis gastroplasty. The Belsey fundoplication is performed after construction of the Collis gastroplasty. In all, three tiers of fundoplication sutures are placed; each tier is positioned about 1.5 cm above the other (Adapted from Pearson FG. Open gastroplasty. In: Pearson FG, Patterson GA, eds. Pearson's Thoracic and Esophageal Surgery. 3rd ed. Philadelphia, PA: Churchill Livingstone/Elsevier; 2008:326-336).

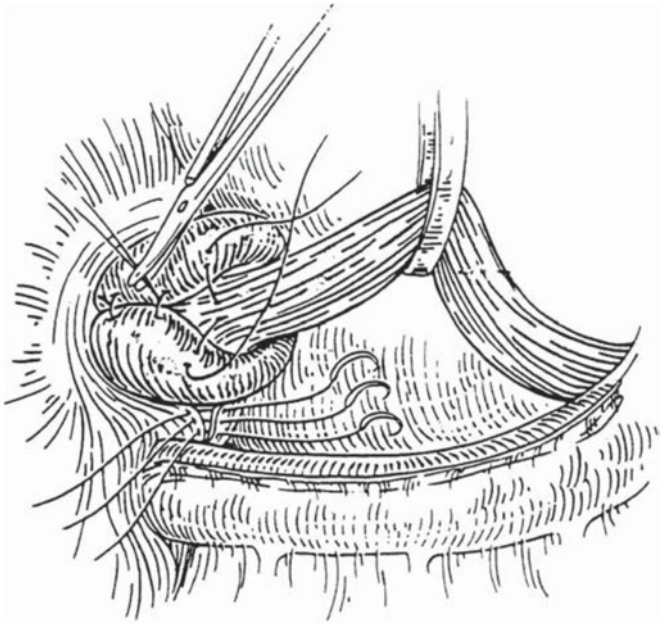


Fig. 35.21. The transthoracic Nissen fundoplication is completed using three sutures spaced 1 cm apart, extending from the stomach, through the esophagus, to the wrapped fundic tip.

symptoms include heartburn (47%), dysphagia (35%), epigastric pain (26%), and vomiting (23%). Laparoscopic procedures performed included 69 Nissens, 112 Collis–Nissens, and 19 other procedures. Only three patients were converted to an open procedure, all nonurgently secondary to adhesions. Median length of stay was 3 days. Complications (minor or major) occurred in 57 of 203 patients (28%). There were six postoperative esophageal leaks (3%) and only one death. Five patients (2.5%) required reoperation for recurrent hiatal hernia. Good to excellent results were obtained in 92% of patients based on a postoperative questionnaire. The mean postoperative Gastroesophageal Reflux Disease Health-Related Quality-of-Life (GERD-HRQoL) score was 2.4 (scale 0 to 45; 0 = no symptoms, 45 = worst).

More recently, we published our results of laparoscopic repair of GPEH in 662 patients over a 12-yea r period, and this represents the largest series published to date (Luketich et al). The median patient age was 70 years, with a median percentage of herniated stomach of 70%. In this series, a Collis gastroplasty was performed in 63% of patients. The operative mortality was 1.7%. Postoperative GERD-HRQoL scores were available for 489 patients (30-month median follow-up), with good to excellent results in 90% (438/489). Reoperation was required in 3.2%, which is comparable to results reported in series where laparotomy or thoracotomy was performed.

In another study, we evaluated the long-term outcomes after laparoscopic repair of GPEHs in a subset of 187 patients, with a median clinical follow-up of 77 months. We evaluated clinical outcomes, barium esophagram, and QoL measures. A Collis gastroplasty was performed in a majority of patients (86%). A barium esophagram obtained in 82% of patients, demonstrated a radiographic recurrence in 15% of patients, though many recurrences were small and asymptomatic. A total of seven reoperations (4%) were required for symptomatic recurrence. The GERD-HRQoL scores were excellent to good in 87%. These results evaluating long-term follow-up show that there is a high degree of satisfaction among patients, and the rates of reoperation for symptomatic recurrence are low after laparoscopic repair in centers with extensive experience with open and minimally invasive esophageal surgery.

As the experience with antireflux surgery in the setting of GPEH has increased, we have come to recognize the important role of esophageal shortening in this condition. As described earlier, 75% to 90% of patients with true GPEH will have a GE junction located well above the diaphragmatic hiatus, and it is important to evaluate for a short esophagus. The proper assessment of esophageal length can be tricky because there is a tendency to overestimate the intra-abdominal length of the esophagus because of the diaphragmatic elevation caused by the pneumoperitoneum during the laparoscopy as well as the downward traction that is applied to the stomach. In addition, we have also gained experience with extensive mediastinal mobilization of the esophagus, when required. Once a short esophagus is recognized, complete mobilization of the esophagus in the mediastinum is important, prior to deciding whether to perform a Collis gastroplasty. We have found that with more extensive experience, and better mobilization of the esophagus, the need for a Collis gastroplasty may be reduced. An esophageal lengthening procedure should, however, be strongly considered in

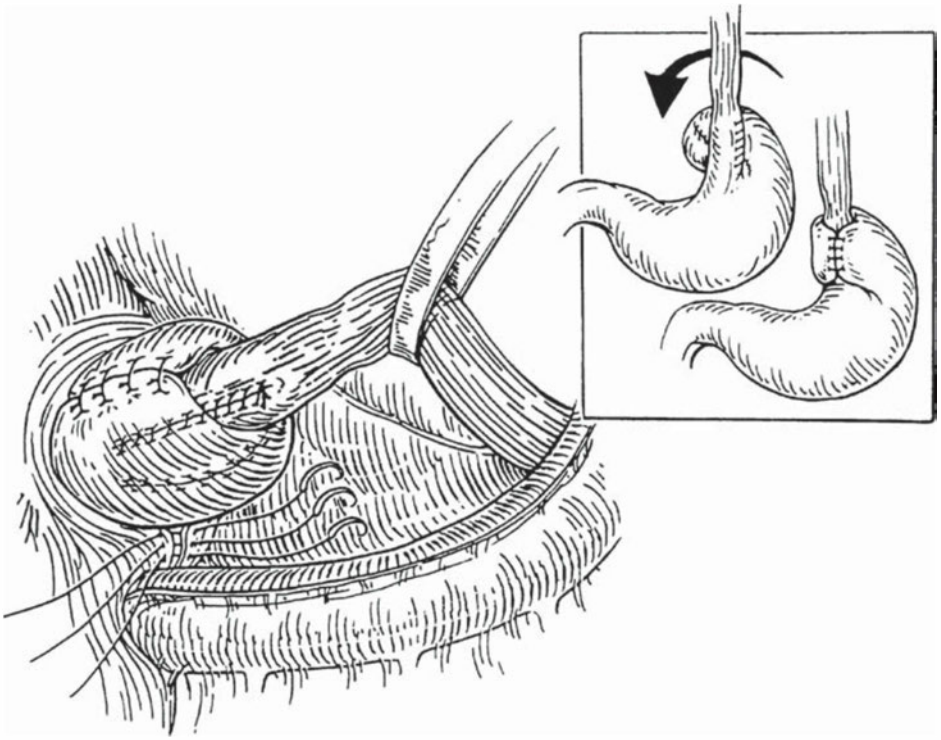


Fig. 35.22. A Nissen fundoplication is performed after completing Collis gastroplasty. Posterior wrapping of the fundus around the neoesophagus is indicated by the black arrow.

the presence of a short esophagus despite adequate mediastinal mobilization.

In addition to an esophageal lengthening procedure, proper reconstruction of the hiatal defect is also critical to the long-term success of the repair. It is critical to preserve the crural integrity, while fully mobilizing the crura. In our experience, in the majority of patients the crura can be approximated primarily without undue tension, and we rarely have used mesh as an adjunct to the repair. In a review of the literature, the radiographic recurrence rate has been reported to be higher following a laparoscopic approach, ranging from 23% to 42%, depending on the duration of follow-up. Crural breakdown and wrap migration have been implicated in up to two-thirds of these failures. Failure rates are particularly prominent when the hiatal defect exceeds 5 cm since the crura may be quite attenuated and may not hold sutures well. Despite complete mobilization of the sac and crura, if the crura cannot be reapproximated without tension, the induced pneumothorax described above may allow a viable, good integrity diaphragm to be approximated without tension. If this is absolutely not possible, the surgeon has several options. Synthetic mesh has the propensity to erode into the esophagus. Thus, we try to avoid synthetic mesh if at all possible in this location, but there will be rare occasions when this may be considered to avoid leaving an obvious large defect. Some surgeons have described the use of a relaxing incision in the diaphragm that reduces the tension at the level of the hiatus. The relaxing incision is then closed with a polytetrafluoroethylene patch. Another approach, advocated by some authors, is bioprosthetic mesh cruroplasty. Biological mesh may be used to buttress the crural muscle so that sutures do not tear through or to actually serve as a patch to achieve closure of the hiatus.

In one randomized study evaluating mesh versus no mesh in laparoscopic repair of paraesophageal hernia, the early results favored placement of mesh; however, the long-term results showed no significant difference in the rate of recurrent hernia (Oelschlager). In this randomized study, primary diaphragm repair (PR) was compared with primary repair buttressed with a biologic prosthesis (small intestinal submucosa [SIS]). With early follow-up, the rate of radiologic hiatal hernia recurrence was higher with PR (24%) than with SIS buttressed repair (9%) after 6 months. The long-term results were recently reported. At a median follow-up of 58 months, there was significant recurrence of the hernia (>50%) in both the groups with 20 patients (59%) in the PR group and 14 patients (54%) with recurrent HH in the SIS group with no difference between the groups ($P = 0.7$). The repair of large paraesophageal hernia is a complex operation and there are several elements critical for the successful repair of a GPEH. This randomized trial addresses one element, the crural repair, that shows a very high recurrence rate at long-term follow-up with or without the addition of mesh.

It is critically important that several important aspects of the repair in this complex operation be considered and addressed. These include complete dissection of the hernia sac and removal from the chest, complete reduction of the hernia contents into the abdomen, preservation of the vagi, dissection of the GE fat pad and determination of the location of the GE junction, evaluation of intra-abdominal esophageal length, extensive mediastinal mobilization of the esophagus, the addition of a Collis gastroplasty if there is a short esophagus despite mediastinal mobilization, proper construction of the fundoplication, preservation of crural integrity, mobilization of the crura and a tension-free crural repair, and the use of a buttress in selected circumstances when a tension-free repair is not possible.

RESULTS OF TRANSTHORACIC REPAIR

The results of transthoracic repair of paraesophageal hernia with thoracotomy is well described with classical series of Pearson, Orringer, and Skinner. In the classic paper of Pearson (Maziak, et al.), the results of 94 patients with GPEHs repaired with an open approach were described. The preoperative symptoms included postprandial pain, dysphagia, iron deficiency anemia, and a history of symptomatic reflux. An open transthoracic approach was used in 97% of patients. A Belsey Mark IV repair was performed and a Collis gastroplasty was added in 80% of patients ($n = 75$) due to a short esophagus. The mean follow-up in this cohort of patients was 94 months and represents one of the longest reported in the literature. Good to excellent results were obtained in 93% of patients, and fair results were obtained in 4%. Two patients had a poor result, and both required reoperation with the addition of gastroplasty for a short esophagus.

In another series (Patel et al.), the results in 240 patients who underwent primary transthoracic repair of paraesophageal hiatal hernia were reported. Presenting complaints included reflux (69%), pain (67%), dysphagia (36%), and bleeding or anemia (33%). Preoperative esophageal function testing showed abnormal reflux in 86%. Hernia types were combined (type III) in 92% and type IV in 8%. All patients had reduction of the hernia and a Nissen fundoplication; an esophageal lengthening

Collis gastroplasty was performed in 96% of patients. The perioperative mortality was 1.7%. The mean follow-up in 226 patients was 42 months (median 27.8 months). Satisfactory results were obtained in 86% of patients. Of 19 patients with an anatomic recurrence, 4 (2%) had required reoperation. Postoperative esophageal function testing, obtained in 28% of the patients, showed abnormal GE reflux in two. The authors concluded that an open transthoracic repair of paraesophageal hiatal hernia provides good to excellent long-term control of both the hernia and GE reflux with relatively low early morbidity.

Some authors have questioned the need for a Collis gastroplasty, in light of the extensive transthoracic mobilization of the esophagus that is possible during thoracotomy. In another series (Altorki, Skinner), 47 patients with GPEH underwent transthoracic repair ($n = 46$) with reduction of the hernia and an antireflux procedure (Belsey in 28; Nissen in 19) without a Collis gastroplasty. The thoracic esophagus was extensively mobilized to the level of the aortic arch. An excellent to good result was reported in 90% of patients at a median follow-up of 45 months.

CONCLUSIONS

In summary, the repair of large paraesophageal hernia is a complex operation with several elements that are critical to achieving a successful repair. As shown by Pearson and colleagues and others, the transthoracic approach with a thoracotomy is a well-established technique for the repair of these hernia but increasingly the laparoscopic repair of GPEH is being utilized and is feasible, safe, and with lower morbidity, in centers with extensive experience in open and minimally invasive esophageal surgery. This has become the authors' preferred approach.

The laparoscopic approach should adhere to the principles established during open operations: these include complete dissection of the hernia sac and removal from the chest, reduction of the stomach and herniated intra-abdominal contents to the abdomen, preservation of the vagi, dissection of the GE fat pad and determination of the location of the GE junction, careful assessment for esophageal shortening, the appropriate use of the Collis gastroplasty in patients with a short esophagus despite extensive mobilization of the esophagus, proper construction of the fundoplication, preservation of the integrity of the crura, mobilization of the crura and a tension-free crural repair, and the use of a buttress in selected circumstances when a tension-free repair is not possible. All these components of the operation are important and careful attention to all these elements of repair may reduce the incidence of recurrent herniation and improve long-term functional results.

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EDITOR'S COMMENTS

Pennathur and colleagues have the world's most extensive experience with the laparoscopic approach for the repair of giant paraesophageal hernias and have shown that in experienced hands results are equivalent, if not superior, to those achieved with an open procedure. The major issue remains the recurrence rate and thus far no procedure, when followed for an extended period, has been able to achieve a zero recurrence rate. Many of the patients who present with these hernias are elderly and have additional comorbidities so there is great appeal for a minimally invasive approach if the results are equivalent or better than those achieved via a standard open procedure.

Recognizing the low probability of an asymptomatic hernia causing problems the question as to whether to electively repair these remains an open one. One can make the case for continued observation while recognizing the very high potential mortality if strangulation should occur. The argument for elective repair of the asymptomatic patient becomes much more compelling if the operation can be done via the laparoscopic approach that the authors advocate. The caveat, however, is that these are complex procedures and should likely be done only by the most experienced laparoscopic surgeons who also have significant experience repairing these hernias via an open procedure. It is imperative that strict attention be paid to the technical details spelled out by the authors if a high success rate for these procedures is to be achieved. The authors make a convincing argument for the addition of an antireflux procedure to accompany the hernia repair, noting that it is likely that the appropriate dissection significantly disrupts the LES mechanism. The type of fundoplication should be determined based both on the surgeon's experience but perhaps more importantly on the preoperative physiologic findings and should be individualized.

It seems that a significant factor involved in recurrence following hernia repair is the failed recognition of a foreshortened esophagus. Recognizing that the assessment of esophageal shortening is difficult at the time of operation, there should be a low threshold to perform a Collis-type gastroplasty if there is any tendency for the reduced viscera to migrate back into the chest. It is likely that the

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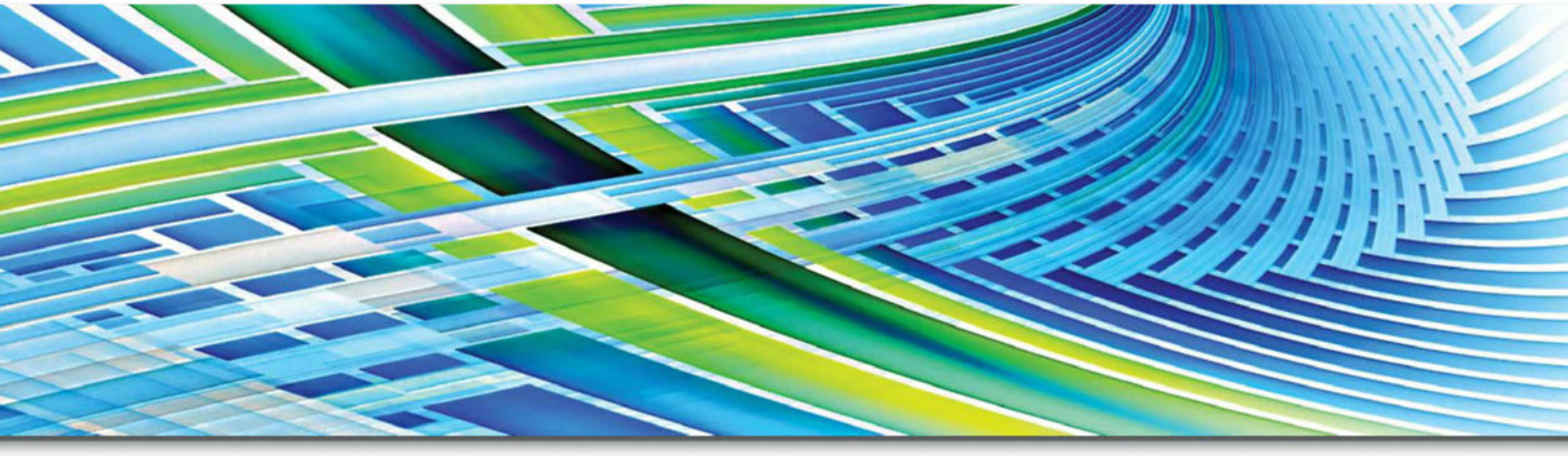
EDITOR'S COMMENTS (*continued*)

majority of patients, especially those with chronic reflux symptoms, have a shortened esophagus and it is probably a good practice to error on the side of performing a gastroplasty though recognizing the possibility of slightly greater morbidity. The potential for a slightly higher incidence of morbidity needs to be weighed against the higher likelihood of recurrence. Once again the experience of the surgeon is key here in making this assessment.

With laparoscopy having become such an integral part of general surgery and of surgical training, it is probably safe to say that the thoracic approach to these lesions, and especially the Belsey-type repair done through the chest, is rarely done and likely will disappear altogether. Very few thoracic training programs provide enough experience with this procedure and the thoracic trainees all come with significant laparoscopic experience. It is highly likely that the Belsey Mark IV repair will be of historical interest only if that is not already the case (with apologies to Griff Pearson and the late Mr. Belsey).

LRK

Adult Cardiac Surgery II



A General Considerations

36 Cardiopulmonary Bypass

Ivan K. Crosby and Richard K. Zacour

INTRODUCTION

The incidence of technical complications associated with the use of cardiopulmonary bypass (CPB) has decreased greatly over the years, aided by the standardization of the equipment and techniques. The standardization of cannulation, cannulae, heart–lung machine, and myocardial protection has resulted in fewer iatrogenic complications such as aortic dissection, air embolism, or end-organ malperfusion. The use of such a standardized protocol is especially important for surgeons learning to use CPB, as well as for the other members of the operative team (anesthesiologists, nurses, and perfusionists), who will anticipate and prepare for each "next step" in the lengthy sequence of important steps in each operative procedure. The omission of one step (e.g., forgetting to give the heparin or forgetting to de-air the heart) can have devastating results in an otherwise well-performed operation. To facilitate the implementation of a standard protocol, all anesthesiologists, surgeons, and perfusionists should agree to standardization of their respective parts of the operation.

It goes without saying that surgeons-in-training should understand the perfusion circuitry, the priming solutions, maximum and minimum flow rates, safe levels of hemodilution during CPB, and the technical details of the system of myocardial protection being used. There should be an understanding of the chain of command, an appreciation of the areas of responsibility of each team member, and the need for open and concise communication among all team members during the course of the operative procedure.

Prior to going to the operating room the surgeon should have a plan for the operation, the expected pathology to be encountered, the type of incision best suited for this operation, the cannulation technique, the level of hypothermia, and the details of the myocardial protection to be used.

DEVELOPING A PLAN

Before each operation the surgeon must develop a plan for conducting the operation, especially the use and style of CPB. Although it is unnecessary for the anesthesiologist and perfusionists to know every technical detail of the planned procedure, it is critical that they understand the planned incisions, method of cannulating the heart and great vessels, the systemic and myocardial temperatures desired, the possible need for low flow or circulatory arrest, and any anticipated pathologic or anatomic variations that may require alterations in the plan of the procedure. It is advantageous for the surgeon to "think through" the entire operation before arriving in the operating room. This intellectual exercise begins by considering the critical elements of the planned operative procedure, which will determine the anatomic exposure required. For example, repair or replacement of the mitral valve requires maximum exposure of the right lateral aspect of the heart and the interatrial groove. Although neither the ventricles nor the left side of the heart need to be elevated, they must be able to be displaced into the left side of the pericardial cavity, or into the left pleural cavity. On the other hand, aortic valve replacement can usually be performed with simple aortic and right atrial cannulation "in situ," and minimal manipulation or displacement of the ventricles. In general, coronary artery bypass grafting (CABG) requires exposure of all surfaces of the ventricles. Adequate exposure for the planned operation will be achieved by "suspending" or elevating the edges of the pericardium to make the relevant parts of the heart closer to the surface of the mediastinum, such that cannulation for CPB is easier, and the areas of the heart to be incised are closer to the surface. The operative plan will include the procedural steps of the operation, the technical details of the planned myocardial protection, and an estimate of the myocardial and systemic temperatures required for the specific operation.

EXPOSURE OF THE HEART

For CABG the best exposure of the heart is via a median sternotomy. For aortic valve procedures, either a full sternotomy or an upper hemisternotomy can be used. The full sternotomy makes it easier to insert a left ventricular (LV) vent through the right superior pulmonary vein, and to insert the retrograde cardioplegia cannula via the right atrium into the coronary sinus. A young surgeon should use the full sternotomy for all aortic valve procedures until he or she feels comfortable using the smaller "mini-valve" incision. Mitral valve operations require a different exposure. A large left atrium, as in long-standing mitral stenosis or mitral regurgitation, is easily approached by decompressing the heart using a vent and excellent venous drainage, using superior and inferior caval cannulae, and retrograde and antegrade cardioplegia. A vertical incision in the left atrium close to the interatrial groove is made, and using a mitral valve retractor the surgeon has a great view of the mitral valve. When the left atrium is small, as in acute bacterial endocarditis or severe mitral regurgitation following acute myocardial infarction, it

may be necessary to use caval snares, enter the right atrium, and incise the interatrial septum to expose the mitral valve. Again, although the surgeon might anticipate a "standard" valve or CABG procedure, he or she must always prepare for the unexpected: the valve that is irreparable; the infected aortic valve with a root abscess; the CABG patient with a shortage of suitable conduits; or the left internal mammary that is damaged during take-down, can it be salvaged and be a quality graft for the left anterior descending? Meticulous attention to technical details should prevent such calamitous iatrogenic problems such as sternal saw injuries to the right ventricle, ascending aorta or innominate artery, or unrecognized "sheathing" of the aortic cannula in the wall of the ascending aorta, causing fatal total aortic dissection. The time-honored adage "chance favors the prepared mind" is very appropriate for all surgeons performing cardiac surgery.

The careful communication between perfusionists, anesthesiologist, surgeon, and nurse is essential in preventing and dealing with crises during the operation.

PREPARATION

A member of the surgical team should be present in the operating room as soon as the patient arrives. The anesthesiologist and associates are responsible for the induction of anesthesia, endotracheal intubation, as well as placement of most monitoring devices. The induction of anesthesia should not commence until good intravenous lines have been inserted and stable EKG monitoring established. If a patient is hemodynamically unstable an arterial line and pulmonary artery catheter should be inserted prior to the induction of anesthesia.

Positioning the Patient

After the insertion of all monitoring lines and the administration of anesthesia, the patient is positioned for the appropriate surgical incisions. All pressure points are padded to prevent pressure necrosis of the skin or nerve damage. All monitoring cables, infusion lines, and EKG cables are secured to prevent displacement or disconnection during the operation. Most cardiac surgical procedures utilize a median sternotomy. This is best done when the sternum of the patient is parallel to the operating room floor. This is achieved by placing a padded roll transversely beneath the patient's shoulders and a padded ring placed under the head. In coronary revascularization procedures both legs should be "frogged" to facilitate harvest of the saphenous vein conduits. If a radial artery graft is planned, the left arm should be on an arm board abducted from the left side of the table. When a submammary right thoracotomy is planned the right chest is elevated 30 degrees on a "bump" under the right scapula and another one is placed under the right hip. The patient's right arm is at the side on the table.

Skin Preparation and Draping

After the anesthetized patient has been properly positioned, a protective screen is placed at the head of the table just superior to the manubrium of the patient. This serves two purposes: (a) it supports the weight of the sterile drapes; (b) it separates the anesthesia team from the sterile field. It is important that the framework of this "ether screen"—traditional name for it—allows easy access to the endotracheal tube and neck lines by the anesthesia team. Antibacterial skin preparation needs to commence prior to the patient's arrival in the operating room. For at least 1 day prior to surgery the patient should have one or more showers and hair wash with chlorhexidine, and gentle scrubbing of the chest, abdomen, and groins with a soft foam scrubber. After positioning in the operating room, the lower neck, chest, abdomen, both groins, and for multivessel coronary revascularization procedures, both lower extremities are painted with a two-solution antibacterial skin prep. When the skin is dry an antibacterial adhesive plastic drape is applied covering all the areas that need to be accessible for surgical incisions. The rest of the traditional sterile drapes complete the draping of the patient.

For every open-heart operation it is important to prep and drape both groins to facilitate rapid institution of CPB before the sternum is opened, should it become necessary, or to permit insertion of femoral arterial monitoring lines or an intra-aortic balloon pump.

The height of the operating room table is usually a compromise between the height desired by the surgeon and that desired by the first assistant. Some surgeons feel that the table height is optimal when the surgeon's wrists are 1-3 cm below the elbow height when operating. The assistants adjust their position and elevation to accommodate the surgeon. The operating room lights are positioned initially to give the best exposure for the sternal incision. Subsequent adjustments are made to illuminate the appropriate area of the heart being treated. For mitral valve operations one light is positioned behind the surgeon, midway between the head and right shoulder such that it shines directly on the mitral valve.

The heart–lung machine and cell-saving equipment are brought into position and the pump lines and suckers are passed off the field to the perfusionists and circulating nurse. The pump lines are secured to the drapes on the side of the table near the patient's right hip in such a way that the operative field is not compromised and the surgeons are unhampered. The lines should be secured to the drapes such that even excessive force cannot displace them. Inexperienced members of the team are instructed not to touch or compress the lines.

Incisions

The most common incision for cardiac surgical procedures is the median sternotomy. The length of the incision requires consideration of safety, effective exposure of the appropriate parts of the heart, and cosmesis. Anatomic and pathologic variations require careful planning to avoid disaster. A large ascending aortic aneurysm pressing against the back of the sternum, severe pectus excavatum, a patent internal mammary graft, or a redo operation all require the sternotomy incision to be performed with great caution. The length of the sternotomy incision can be shortened for cosmetic reasons. The superior extent of the skin incision might be midway between the top of the manubrium and the manubriosternal junction. Inferiorly, the incision can be stopped at the superior aspect of the xiphisternum. Experienced surgeons will elevate the skin of the inferior margin of the sternotomy incision and divide the xiphoid process longitudinally along with the superior aspect of the linea alba, to allow effective exposure of the heart, but limiting the length of the skin incision. It is important for less-experienced surgeons not to compromise safety for cosmesis. When the sternal edges are spread using the sternal retractor the anterior aspect of the pericardium is clearly visible. The anterior layer of the pericardium is grasped with hemostats and the pericardium is incised from the acute angle of the heart inferiorly up to the pericardial reflection on the ascending aorta.

There are two basic techniques for elevating the heart closer to the sternal surface to facilitate exposure of the cardiac structures. One approach utilizes #1 silk traction sutures where the pericardium is stitched to the fascia at the edge of the sternal incision, or to the retractor itself. In the second technique two Kocher clamps grasp each side of the incised pericardium, the sternal retractor is loosened, and the edges of the pericardium in the Kocher

clamps are pulled up between the blades of the sternal retractor and the sternal edge. As the retractor is opened further the whole heart is elevated anteriorly giving excellent exposure. Caution should be used in spreading the sternal retractor too wide, as this can cause traction on the brachial plexus causing pain and numbness in the fourth and fifth fingers of the left hand postoperatively, or acute hypotension. If the length of the sternotomy skin incision is limited inferiorly, the pericardial incision should be carried down onto the diaphragm to give better exposure.

Table 36.1 Anticoagulation Management Protocol

Add 4 U of beef lung heparin per milliliter of the pump prime to the cardiopulmonary bypass circuit.

The surgeon injects 300 U of beef lung heparin per kilogram of patient body weight into the right atrium.

An uncontaminated whole-blood sample is drawn 3–5 min after heparin injection for an activated clotting time (ACT) measurement.

The ACT must be >2.5 times the baseline ACT before initiating cardiopulmonary bypass, and must be maintained at >480 seconds during cardiopulmonary bypass.

The ACT should be monitored every 20 min during normothermic cardiopulmonary bypass, every 30 min during hypothermic cardiopulmonary bypass, and more frequently if the patient shows heparin resistance.

Heparin

Once the aortic and atrial pursestring sutures are in place, heparin should be administered. In some centers the heparin is given by the anesthesiologist into a central line. In other centers the surgeon will inject the heparin directly into the right atrial appendage so that there is no doubt that all the heparin is circulating. If the heparin is given through a peripheral IV line tragedy will ensue if that IV has infiltrated. The normal heparinizing dose is 300 units/kg of patient bodyweight (Table 36.1). Additionally, 4 U porcine heparin/ml of pump prime is injected directly into the CPB circuit. However, heparin activity is patient specific and may vary if underlying coagulation defects are present. Confirmation of systemic anticoagulation is achieved by performing an activated clotting time (ACT). An uncontaminated whole blood sample is sent for ACT measurement 3 to 5 minutes after the heparin has been given. This test measures heparin activity, which plays a more significant role in anticoagulation management than heparin concentration. The optimal ACT range for CPB has not been defined. Our protocol requires the ACT be >2.5 times the baseline ACT before initiating CPB, and maintained at >480 seconds during CPB. We prefer to maintain ACTs between 400 and 600 seconds because ACTs <180 seconds are dangerous and ACTs >600 seconds are unnecessary (Table 36.1). We monitor the ACT every 20 minutes during normothermic CPB, every 30 minutes for hypothermic CPB, and more frequently if the patient shows heparin resistance (Table 36.1).

Heparin Substitute

In situations where heparin cannot be used we go to bivalirudin (Angiomax). Our protocol is as follows:

1. 50 mg in pump prime
2. 1 mg/kg bolus 20 minutes prior to CPB
3. 2.5 mg/kg/h continuous infusion following initial bolus
4. 0.1 to 0.5 mg/kg bolus to maintain desirable ACT of 500 seconds
5. Check ACT every 10 minutes while on bypass
6. Discontinue bivalirudin following termination of CPB
7. DO NOT use blood cardioplegia. Use crystalloid cardioplegia (Plegisol)
8. Flush CPB circuit with clear prime fluid into cell-saver once terminated
9. Process CPB circuit volume via cell-saver as soon as possible.

Cannulation

All cannulation pursestring sutures MUST be inserted before the heparin is given (Fig. 36.1). The full heparin dose must be given and have circulated BEFORE any cannulae are inserted. In its simplest form CPB requires a cannula in the ascending aorta and a cannula in the right side of the heart. The type of procedure to be performed will dictate where the cannula is placed in the ascending aorta and what type of cannulation is performed on the right side of the heart. If a coronary bypass procedure is being performed, the cannula in the ascending aorta will be placed close to the innominate artery or even in the arch area. This will allow a large area of the ascending aorta for the placement of proximal graft anastomoses. Remember: PURSESTRING SUTURES first, then HEPARIN, then CANNULATION!

Aortic Cannulation

Trainee cardiac surgeons traditionally learn cannulation techniques from their immediate superiors or from their consultants or

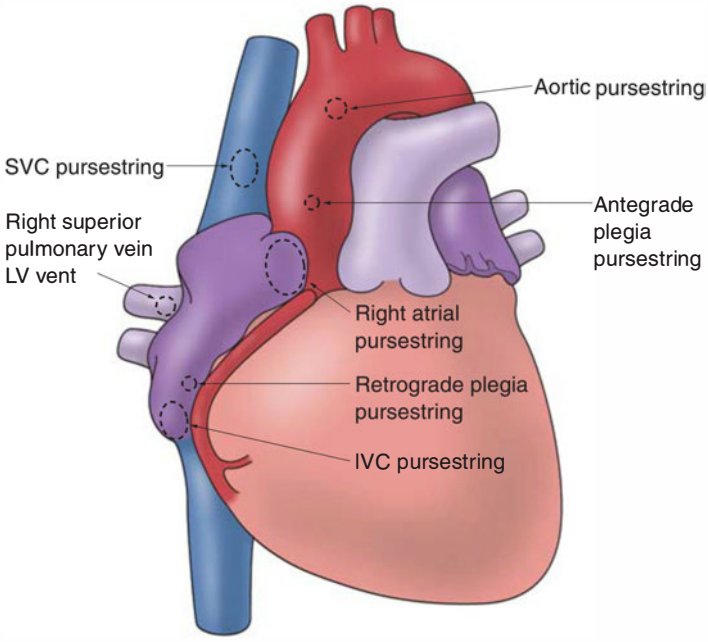


Fig. 36.1. Sites on the heart and great vessels for cannulation sutures. IVC, inferior vena cava; LV, left ventricle; SVC, superior vena cava.

professors. Before any aortic pursestrings are inserted, however, the surgeon should perform gentle digital evaluation of the entire ascending aorta, innominate artery, and aortic arch. In choosing a cannulation site he or she needs to avoid areas of calcification or atherosclerosis in the aorta. If there is any question, the aorta should be evaluated with an epiaortic probe. Of prime importance is the need for the cannula tip to lie in the lumen of the aorta (Fig. 36.2), not against or penetrating the wall of the aorta ("sheathing" the cannula, causing a type A aortic dissection). Also it should not be inserted so far that the tip of the cannula goes up the innominate artery or the left carotid artery. The jet from the cannula tip should not act like a "fire hose" directed at atherosclerotic plaque and shower emboli to the brain. Many surgeons prefer that the length of the cannula that penetrates the aortic wall should be no more than 2 to 3 cm (Fig. 36.2). Although some surgeons prefer a much longer cannula penetration into the aorta such that the tip of the cannula lies in the distal aortic arch, avoiding the innominate, the left carotid, and the left subclavian artery orifices, we feel that it is hard for trainee surgeons to be sure that the tip placement is perfect using this latter technique. The aortic pursestring is usually a 3-0 monofilament or braided suture. The diameter of the pursestring is usually 1 to 1/3 times the diameter of the aortic cannula being used. There is usually an inner pursestring and an outer pursestring and some surgeons prefer to use pledgets with these pursestrings (Figs. 36.3, 36.4, 36.5). The needle bites of the double-armed sutures should penetrate the adventitia and into the media of the aortic wall, but not into the lumen of the aorta. After completing the pursestrings, the needles are removed and both ends of the suture drawn through a tourniquet or snare. There are thus two tourniquets to secure the aortic cannula when it is inserted.

There are three commonly used techniques for inserting the aortic cannula in the aorta:

(a) Using a #11 blade, and taking care not to cut the pursestring sutures, the surgeon makes a stab incision through the aortic wall inside the pursestrings circle (Fig. 36.5). Removing the blade, the left index finger quickly tamponades the incision to prevent hemorrhage. The "bumper" on the aortic cannula has already been adjusted to limit the depth of penetration of the cannula into the aorta. As the left index finger moves from the stab incision in the aorta the right hand gently but quickly pushes the aortic cannula into the aortic lumen. The snares or tourniquets are tightened down and then tied to the cannula above the bumper to prevent the cannula from backing out of the aorta. The clamp on the aortic cannula is briefly released to remove all air from the cannula and the clamp reapplied (Fig. 36.2).

(b) Some surgeons prefer to use a #15 blade and make a vertical incision within the pursestrings, layer by layer of the aorta, until only the intimal layer remains. Holding the preadjusted cannula in the right hand the cannula is carefully pushed through the intima into the lumen of the aorta. The aortic cannula is deaired as above and secured (Fig. 36.2).

(c) Ambidextrous surgeons can make the incision into the aorta inside the pursestring circle with the left hand using a #15 blade, and keeping the body of the #15 blade inside the aorta, they hold the cannula in their right hand and slide the tip of the cannula off the #15 blade into the aortic lumen (a "shoe-horn" maneuver). The cannula is then secured and deaired as above.

No matter which technique is used to insert the aortic cannula into the aorta, the surgeon must meticulously remove all air and debris from the cannula as it is connected to the arterial line from the heart–lung machine, and the clamp is removed from the aortic cannula.

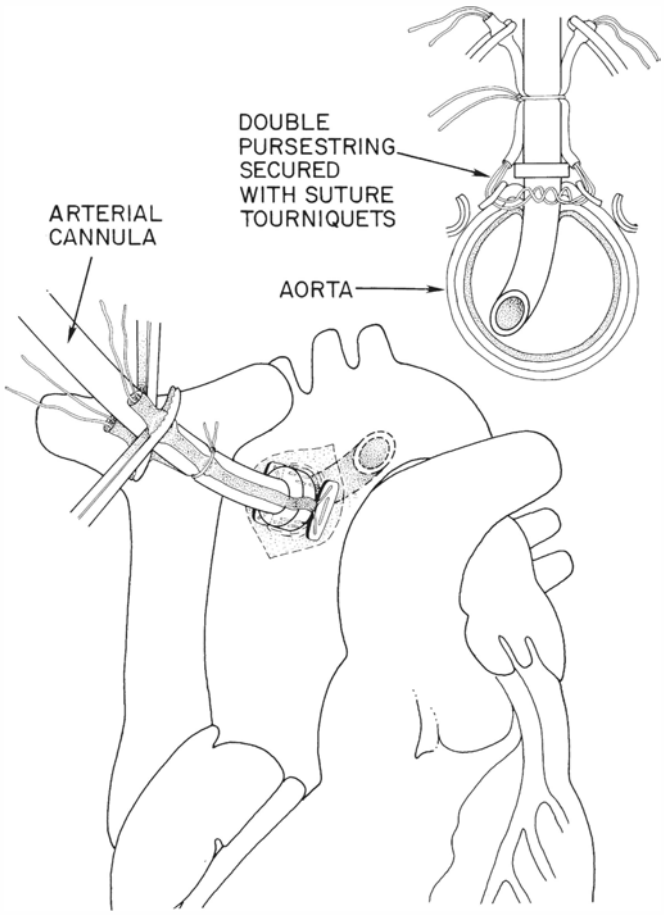


Fig. 36.2. The final position of the aortic cannula in an adult.

Alternative Cannulation Sites

Arterial cannulation of the distal ascending aorta or aortic arch may not be desirable or possible if there is extensive calcification of the entire ascending aorta and arch, or if there is an acute aortic dissection involving the ascending aorta and arch (type A), or a large aneurysm involving the arch. In such circumstances there are two alternative cannulation sites to consider: the right axillary artery or the femoral artery.

The right axillary is exposed using a small subclavicular incision. The pectoralis muscle is split or divided, exposing the axillary artery. Gentle mobilization is necessary to avoid injury to the adjacent brachial plexus, or causing spasm of the axillary artery itself. After heparinization of the patient a segment of the artery is isolated

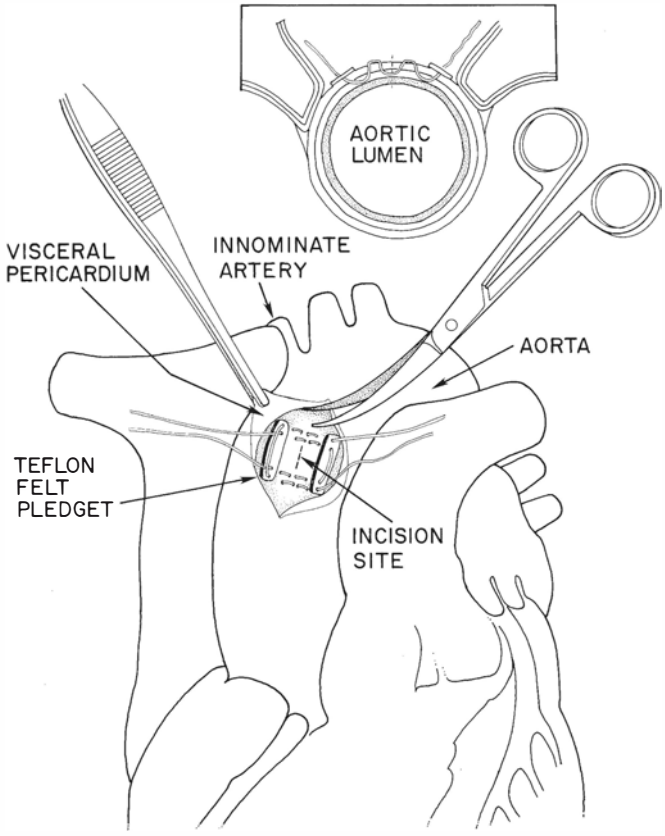


Fig. 36.3. Preparing an adult ascending aorta for cannulation.

between gentle vascular clamps, and an 8 mm longitudinal incision is made on the anterior aspect of the artery. A 10 cm length of 8 mm diameter prosthetic graft is sewn end-to-side to the 8 mm arteriotomy using a 5-0 polypropylene running suture. The arterial cannula is inserted into this graft and secured with a heavy silk suture ligature.

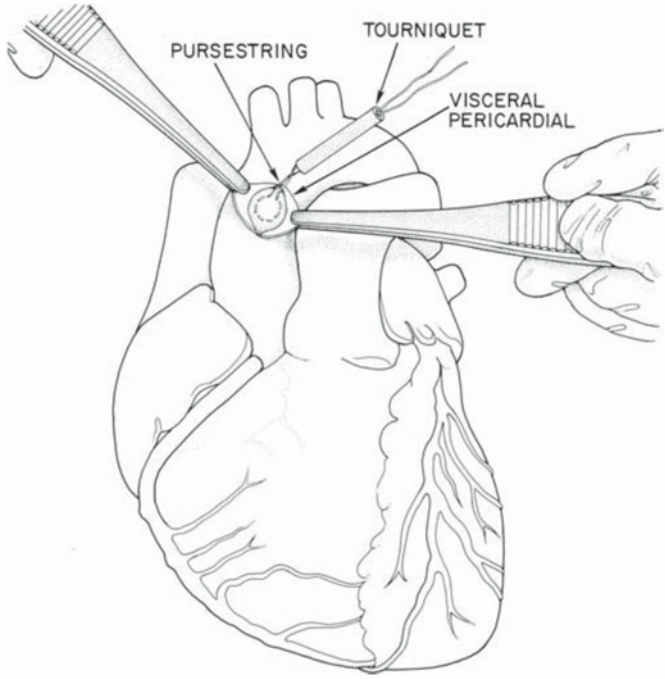


Fig. 36.4. Preparing an infant or pediatric aorta for cannulation.

The tip of the arterial cannula should be 2 to 3 cm from the axillary artery. After the CPB is discontinued and the patient is quite stable, the cannula is removed and the prosthetic graft is transected, leaving a 5 mm cuff which is oversewn with a running 5-0 suture.

Femoral cannulation is not desirable if the patient has aortoiliac occlusive disease with severely diminished or absent femoral pulses. When used it requires a short vertical skin incision directly over the common femoral artery. Using electrocautery for hemostasis, the common, superficial, and profunda arteries are encircled for control. An umbilical tape is passed around the common, and the ends drawn through a tourniquet. With the patient fully heparinized, the distal vessels are occluded, a vascular clamp occludes the common femoral, and a transverse arteriotomy is made in a soft part of the common femoral on its anterior surface, not longer than 50% of the circumference of the artery. The arterial (femoral) cannula is advanced proximally up the common femoral artery and the umbilical tape tourniquet is tightened just enough for hemostasis, and is then secured to the cannula with a heavy silk ligature. On completion of CPB the cannula is removed, and the arteriotomy closed with a continuous 5-0 suture.

Percutaneous femoral cannulation can be performed as an alternative to open exposure of the femoral artery or vein (or both) using the Seldinger technique.

Atrial Cannulation

When the right atrium is opened for repair of septal defects or tricuspid valve surgery two venous cannulae will be placed—one in the superior vena cava (SVC) and the second in the inferior vena cava (IVC) (Figs. 36.6 and 36.7). Caval "snares" or tourniquets are then placed around the superior and inferior cavae, such that when the tourniquets are tightened around the superior and inferior caval cannulae, the patient is then on "total" CPB, and the right atrium can be opened with a dry operative field. When mitral valve procedures are being performed, two venous cannulae are frequently used to facilitate exposure of the mitral valve. The mitral valve retractor must not compromise the venous return. For many cardiac operations, such as aortic valve replacement or coronary bypass surgery, a "two-stage" single venous cannula is inserted via the right atrial appendage (Fig. 36.8).

Bicaval Cannulation

When two caval cannulae are used to facilitate the venous drainage of the heart, a

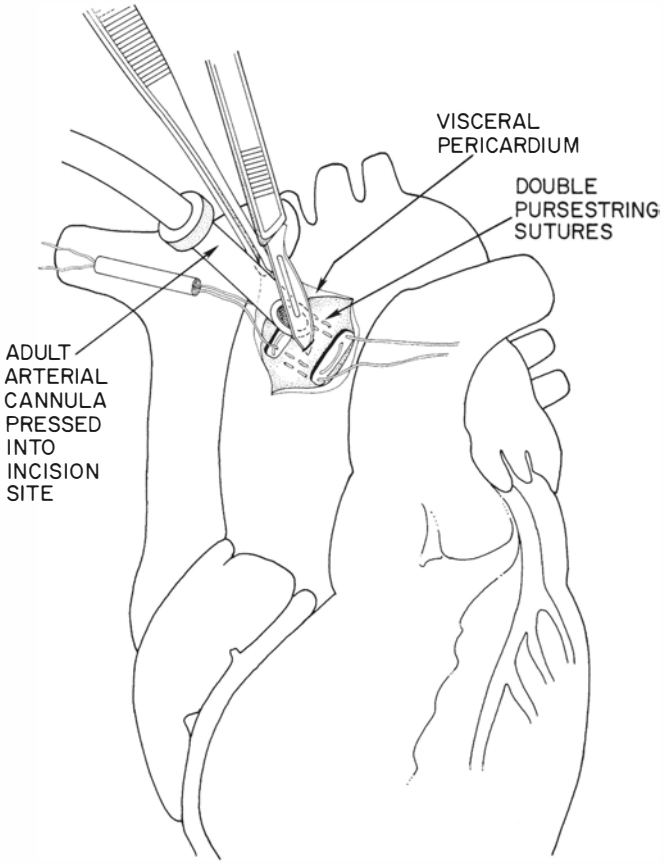


Fig. 36.5. Incision of an adult ascending aorta and insertion of a soft aortic cannula.

separate pursestring suture is inserted for each caval cannula (Fig. 36.1). The superior caval pursestring encircles the right atrial appendage or less commonly is placed directly in the superior cava itself. These areas are easily accessed through the sternotomy incision. The insertion of the inferior caval pursestring is more difficult, and should be inserted first. The diameter of the atrial pursestring sutures should be 1 to 1/3 times the diameter of the cannula. The inferior pursestring should be placed on the inferior aspect of the right atrium near the junction with the IVC (Fig. 36.1). With a 4×4 sponge on the heart at the junction of the right atrium and the right ventricle the assistant retracts the heart gently to expose the area for the insertion of the inferior pursestring suture. The pursestring for the superior caval cannula is placed around the right atrial appendage (Fig. 36.1).

To insert the inferior caval cannula, a vertical or cruciate incision is made within the pursestring with a #11 blade. The assistant tamponades this incision with a finger briefly, and then the surgeon places the cannula into the right atrium through this incision, and directs it down the IVC and the tourniquet is snared. As the tubing clamp near the top of the cannula is released briefly, the surgeon's right hand presses on the right upper quadrant of the patient's abdomen to fill the cannula with blood, the tubing clamp is reapplied, and the venous cannula connected to the venous return line to the heart–lung machine. Insertion of the superior caval cannula is often done by placing a vascular clamp across the base of the atrial appendage that is encircled by the superior pursestring. The tip of the appendage is amputated, and as the vascular clamp is removed, the superior caval cannula enters the right atrium, is advanced into the SVC, and secured with the tourniquet. Connection of the two caval cannulae to the venous return line is accomplished in a meticulous way. Both venous cannulae should be filled with blood. A bulb syringe filled with saline fills the connector on the venous line and the venous line itself with saline. Both caval cannulae are then connected to the venous line Y-connector. The tubing clamps are then reapplied keeping air out of the heart (Figs. 36.6 and 36.7).

Two-Stage Venous Cannula

The two-stage venous cannula, with or without suction, provides excellent

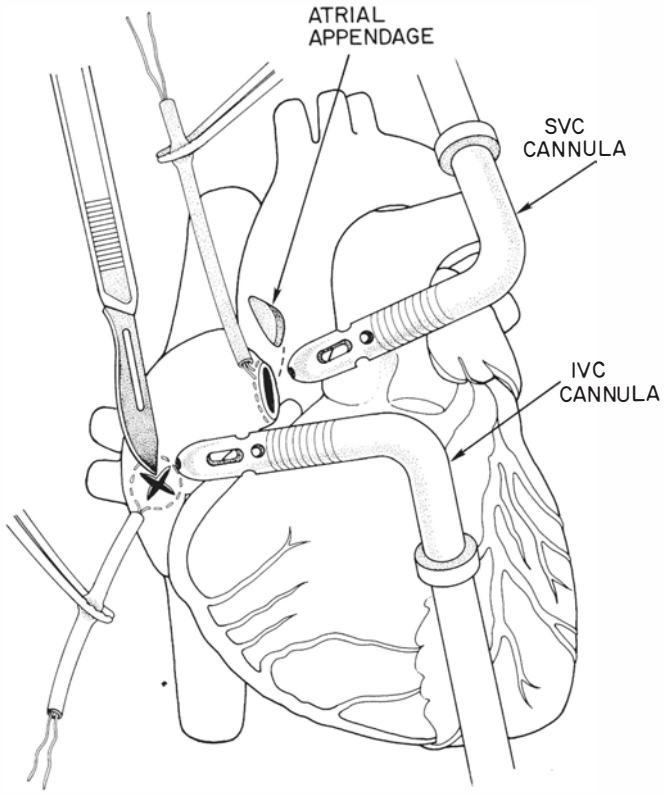


Fig. 36.6. Double venous cannulation. IVC, inferior vena cava; SVC, superior vena cava.

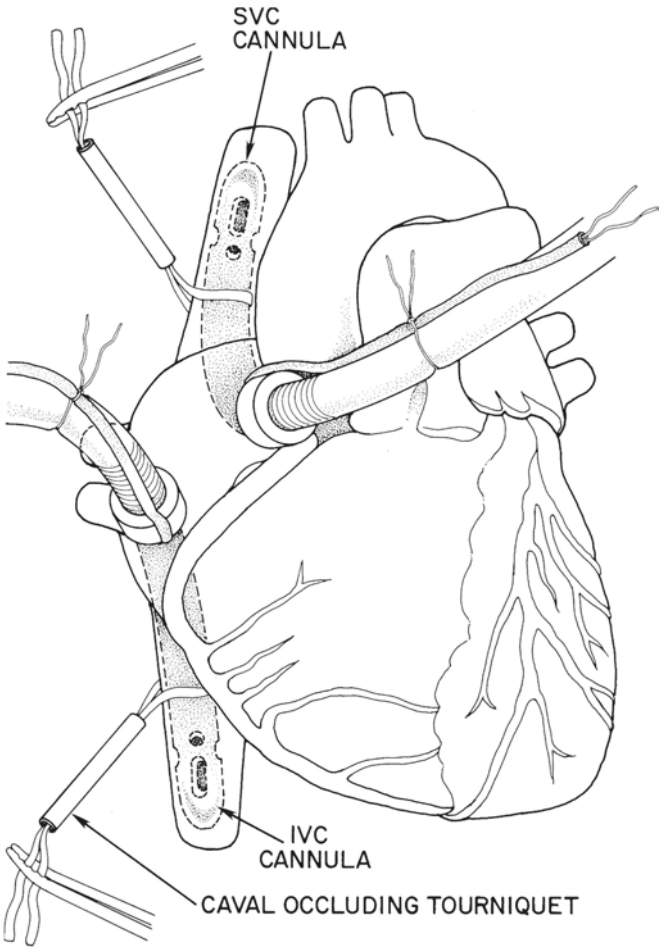


Fig. 36.7. The final position for double cannulation. Note that the side holes must be distal to the vena caval tourniquets. IVC, inferior vena cava; SVC, superior vena cava.

drainage of the heart. It is used routinely in aortic valve procedures, coronary bypass procedures, and AVR+CABG procedures. In all these operations the right side of the heart is not opened. Thus this venous drainage system is used more frequently than bicaval drainage. A single pursestring suture is inserted around the base of the right atrial appendage (Fig. 36.1). With a noncrushing vascular clamp across the base of the right atrial appendage, the tip of the appendage is amputated. The clamp is removed and the two-stage cannula placed into the right atrium and directed down the IVC. The cannula is backfilled and connected to the venous return line (Fig. 36.8).

Retrograde Cardioplegia Cannula

Before the heparin is administered, a 4-0 polypropylene pursestring suture in the lower right atrial area near the IVC is placed for the retrograde cardioplegia cannula (Fig. 36.1). After the heparin has circulated, using a #11 blade, a stab incision is made inside the pursestring and the retrograde cannula inserted through the right atrium into the coronary sinus. This is best done before the two-stage or inferior caval cannula is inserted. It is harder to thread the retrograde cardioplegia catheter into the coronary sinus with a venous cannula nearby. The accurate positioning of this cannula is facilitated by the surgeon placing his right hand between the right ventricle and the diaphragm. The third and fourth fingers of the right hand help manipulate the cannula into the coronary sinus.

Antegrade Cardioplegia Cannula

A #4-0 polypropylene pledgeted pursestring suture is placed in the ascending aorta above the aortic valve but proximal to where the aortic cross clamp will be applied (Fig. 36.1). In CABG procedures this cannula is positioned such that this cannulation site in the aorta is used for one of the proximal anastomoses. This allows insertion of the antegrade cardioplegia/aortic vent cannula into the aorta. When the cannula is back flushed and filled with blood the cardioplegia line is connected, while a vent suction line is connected to the other tube of the cardioplegia/vent cannula, which is then occluded.

Indications for Left Ventricular Venting

Venting of the left side of the heart is most important in different circumstances:

(a) Severe aortic valve regurgitation. The institution of CPB in a patient with severe aortic valvular insufficiency causes immediate LV distension. Unless this is corrected immediately (by cross clamping the aorta and venting the LV), it will result in severe damage to the LV.

(b) Antegrade cardioplegia. If there is any degree of aortic valve regurgitation when antegrade cardioplegia is being administered, the LV will become distended with cardioplegia. To prevent damage to LV function the LV needs to be vented.

(c) Incomplete drainage of the venous return. If there is inadequate drainage of the right side of the heart or kinking/obstruction of the venous return line, both sides of the heart will become distended. If severe, this can cause a dramatic fall-off in cardiac contractility. An LV vent can obviate this problem, together with improved drainage of the right side of the heart.

(d) Evacuation of air from the heart. For such operations as aortic valve replacement, mitral valve repair or replacement, LV aneurysmectomy, atrial or ventricular septal defect procedures, after completion of the definitive procedure, a considerable amount of air will be present in the left atrium and LV. While the heart is arrested and motionless this is not a problem. However, unless all this air is evacuated before the heart starts beating, there is a serious risk that some of this air will be pumped from the heart into the arterial circulation. This can cause cerebral obtundation or stroke, air down the coronary arteries resulting in infarction or LV power failure, end organ or peripheral ischemia.

To prevent air embolism most surgeons use a LV vent and an aortic vent. The LV is decompressed using a catheter inserted through the right superior pulmonary vein (Fig. 36.1) and advanced into the LV chamber. Air is evacuated from the ascending aorta using the antegrade cardioplegia/aortic vent cannula.

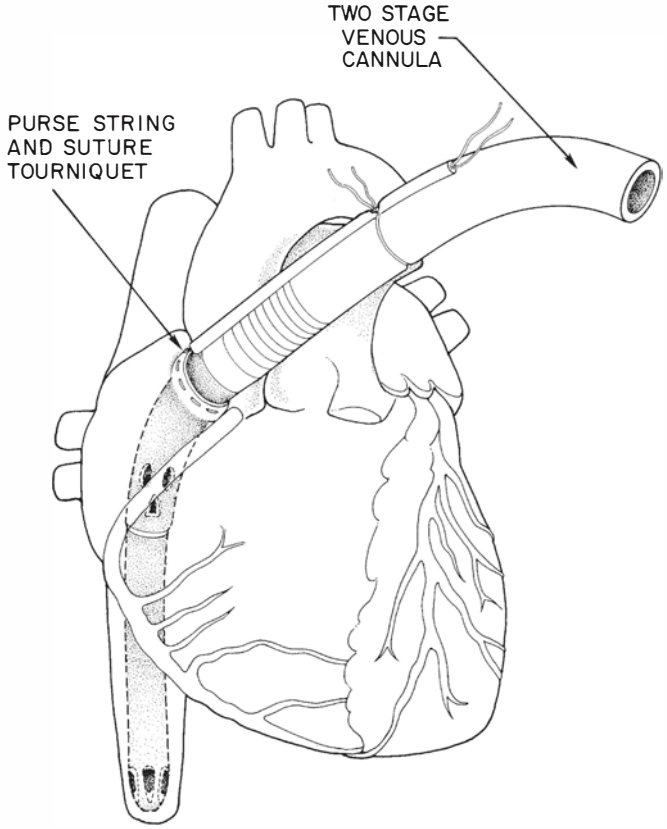


Fig. 36.8. Single venous cannulation. The "two-stage" construction places drainage ports in the right atrium and the inferior vena cava.

cut edges of the aorta together to prevent bleeding. When the surgeon is ready, the forceps are parted, and the aorta is raised to give counter pressure as the surgeon inserts the cannula, entering the aorta at a right angle to the direction of blood flow and tilting the external cannula inferiorly as he or she advances it the equivalent of 1 to 1/3 times the aortic diameter (Fig. 36.9). The position of the cannula is confirmed by palpation to be sure it has not entered the innominate artery and that the tip lies on the inner curve of the aortic arch just distal to the pericardial reflection. The pursestring suture is drawn tight in its tourniquet and secured to the cannula with a heavy ligature. After the aortic cannula has been de-aired and connected to the arterial pump line, it is secured to the drapes to avoid undue tension.

In infants and young children, venous cannulation is performed by modifying commercially available thin-walled, wire-wound cannulas. For double-caval cannulation, the unwired portion of the tip is trimmed to provide two proximal side holes and an oblique bevel. Insertion techniques are similar to those for adults, but often a right-angled cannula with a metal or plastic tip is inserted directly into the SVC, rather than into the right atrium,

PEDIATRIC CARDIOPULMONARY BYPASS

Pediatric patients are not small adult patients, and require different management strategies from the adult. Pediatric patients have a smaller blood volume, higher oxygen consumption, compensatory shunting, significant levels of hemodilution, reduced clotting factors and plasma proteins, and require a wider fluctuation of perfusion flow rates and temperatures per procedure. These differences in the pediatric population mandate great attention to detail, and the ability to adapt and improvise to a wide variety of circumstances.

In infants and young children, the aorta is cannulated with a wire-wound, thin-walled cannula (Fig. 36.9). The aorta within the pursestring is cleared of its visceral pericardium to expose the aortic adventitia (Fig. 36.4), and the assistant, using two fine-tipped forceps, grasps the visceral pericardium on each side of the pursestring. The surgeon, using a scalpel with a #11 blade, makes an incision equal in length to the diameter of the cannula. As the scalpel is withdrawn, the assistant presses the

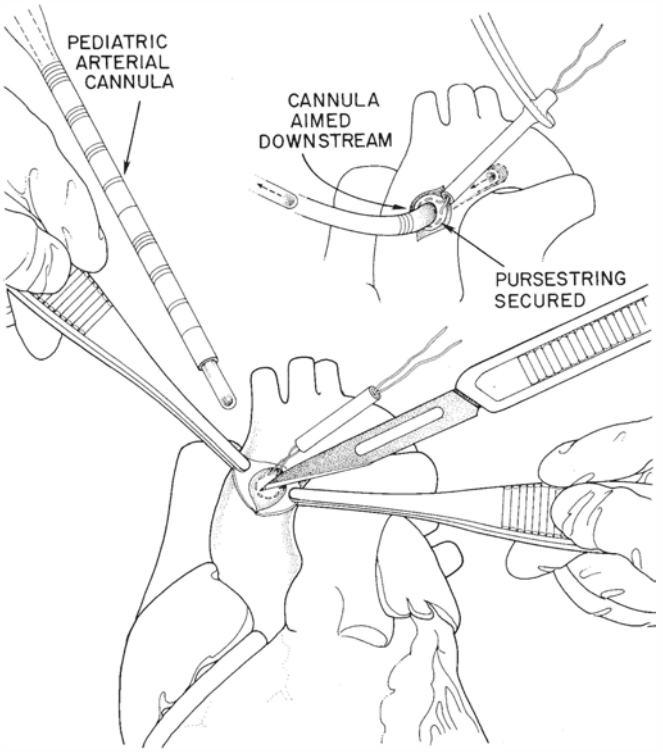


Fig. 36.9. A wire-wound pediatric aortic cannula and obturator. This is an 8F cannula. Because of the thin wall, it will permit flow rates of 800 ml/min and is adequate for patients weighing as much as 5 kg.

when using two venous cannulae. When a single venous cannula is to be used in infants, it is modified to leave four side holes and is inserted through the right atrial appendage, so that only the tip lies in the orifice of the IVC to provide free drainage of the right atrium. In neonates a single 12F or 14F cannula is used.

Left Ventricular Venting Technique

If an LV vent is needed, it is inserted before commencing CPB. The anesthesiologist performs a Valsalva maneuver to elevate the left atrial pressure. An incision is made in the pursestring suture at the juncture of the right superior pulmonary vein and left atrium (Fig. 36.1), and the fluid-filled cannula is immediately inserted. The cannula should have a curve that forms nearly a right angle and should be aimed with its tip hugging the anterior atrial surface and directed toward the LV apex. Except in mitral stenosis, it will pass immediately into the LV in its appropriate position, which can be confirmed by signs of pulsatility within the cannula, or by the surgeon's hand palpating in the oblique sinus. If the vent cannula is stiff, caution must be used to limit the length of the cannula inserted through the pursestring into the heart, so that the tip of the stiff cannula does not puncture or perforate the apex of the LV.

Some surgeons prefer to insert the LV vent while on CPB. The perfusionist fills the heart by occluding the venous line, translocating blood volume from the bypass circuit to elevate the left atrial pressure. When the stab incision is made within the pursestring, blood under pressure flows from the left atrium (preventing the ingress of air into the left atrium), while the LV vent catheter is advanced into the LV.

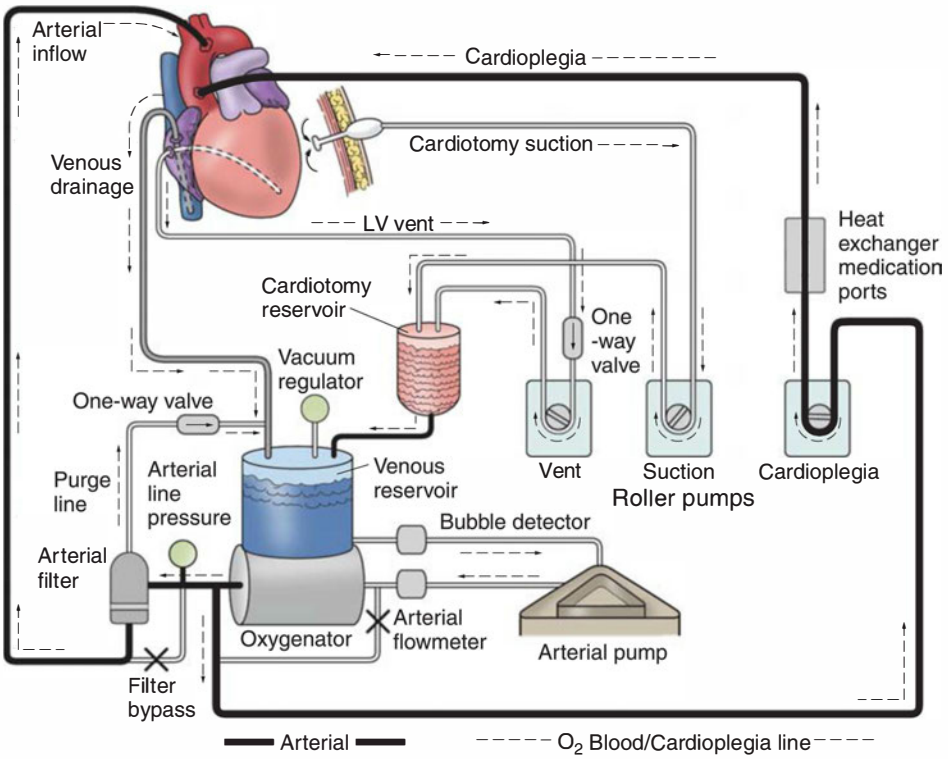


Fig. 36.10. Example of a complete cardiopulmonary bypass circuit. LV, left ventricle; P, pressure transducer.

Table 36.2 Two Methods for Calculating "Full" Blood Flow Rates

Age (y)	1. Blood flow rate (BFR; L/min/m²) from body surface area (BSA; m²)	Body weight (kg)	2. Blood flow rate (ml/min/kg) from body weight (in kg)
0–2	BSA × 2.6	0–5	150
2–4	BSA × 2.5	6–10	125
4–6	BSA × 2.4	11–15	100
6–9	BSA × 2.3	16–25	90
>9	BSA × 2.2	>25	70

Cardiopulmonary Bypass Circuit

The foremost principle of CPB management is to minimize the patient's metabolic adaptation to a nonphysiologic state, a state of "controlled shock" and nonpulsatile perfusion. To accomplish this, the perfusionist must meticulously prepare the extracorporeal circuit and manage the dynamic characteristics of CPB (Fig. 36.10). The circuit design and the selection of the arterial and venous cannula are based on the calculated blood flow rates necessary to provide adequate hemodynamic support and gas exchange (Table 36.2). The cannula tip is the narrowest component of the extracorporeal circuit and creates an environment of high-resistance, high-pressure gradients and turbulence. The cannulae are described by either French size or in millimeters, which reflects only the external dimensions, without regard to the internal diameter or performance characteristics. Although flow characteristic and pressure drop can be calculated for straight tubing, cannula performance characteristics complicate these calculations due to varying lengths, the presence of side holes or curves, and irregular diameters. These factors, along with ratio of internal diameter to external diameter (ID/OD) need to be considered when selecting a cannula (Table 36.3). Performance characteristics and the pressure-flow relationship of any given cannula should be reviewed and a pressure gradient not exceeding 100 mmHg at "full flows" is recommended for an arterial cannula.

The performance characteristics of venous cannula are even more vital than those of the arterial cannula since blood flow through the extracorporeal circuit is dependent upon the venous return, which may be either passive or "assisted." Passive venous return is the more traditional method of providing extracorporeal circulation, and is dependent upon gravity, the level of the heart above the venous reservoir (40 to 70 cm), and large-bore tubing.

Table 36.3 Required Inside Tubing Diameter for Different Patient Weights

Patient weight (kg)	Arterial line (in.)	Venous line (in.)	Vacuum-assisted venous line (in.)
<8	1/4	1/4	1/4
>8–17	1/4	3/8	3/8
17–50	3/8	3/8	3/8
>50	3/8	1/2	3/8

"Assisted" venous return has been used in some centers in the United States for 5 to 6 years, and is achieved with the aid of vacuum being applied to the venous line or reservoir. Assisted venous return provides some advantages over the traditional venous drainage, such as permitting smaller-diameter venous lines and cannulae, smaller incisions, and lowering the extracorporeal circuit priming volume. However, potential disadvantages of assisted venous return include an increase in gaseous microemboli if the vacuum is too great and the reservoir volume is too low to allow proper dissociation of gaseous microemboli from the blood, and an increase of hemolysis if the vacuum rate is too great. Because of these concerns, we prefer to maintain a negative pressure similar to gravity drainage, limit the amount of vacuum to less than –60 mmHg, and prefer to maintain a venous reservoir volume sufficient enough to provide a 10-second reaction time. Currently in the United States about 55% of heart surgery teams use passive venous return, and 45% of teams use assisted venous return. Regardless of the method utilized, venous return will be reduced if the side ports of an oversized venous cannula are obstructed by an overstretched cava, or if the internal diameter of the venous line and/or cannula is too small to accommodate the expected venous return. This results in overdistension of the right heart and/or flooding of the operative field. An appropriate venous cannula will have a pressure gradient of less than 30 mmHg at the rated blood flow, and will not allow an excessive negative pressure to develop (the ideal venous pressure should range between 0 to 5 mmHg). The patient's size determines the design of the extracorporeal circuit and the priming volume. The blood volume of all patients, from pediatric to adult, can be calculated from Table 36.4. The degree of hemodilution may be calculated before bypass is initiated, and if the expected priming volume would cause an "unacceptable" anemia, occasionally packed red blood cells may be added to the extracorporeal circuit before commencing CPB (Tables 36.5 and 36.6).

Table 36.4 Patient Blood Volume

Age	Blood volume (ml/kg)
Premature	100
Newborns	90
1–12 mo	80–85
1–10 y	75–80
Adult	70–75

Hemodilution provides an advantageous effect for perfusion by decreasing fluid viscosity and by augmenting blood flow. One deleterious side effect of hemodilution is the reduction of oncotic pressure, resulting in tissue edema. There is an inverse relationship between tissue edema and colloid oncotic pressure, and when albumin or mannitol is added to the extracorporeal circuit to obtain an oncotic pressure of 16 mmHg, extracellular fluid accumulation is reduced. Although blood flow reflects the interaction of many influences, hemodilution aids in negating those inherent effects by diminishing blood's viscosity and resistance to flow, and promotes increased microcirculatory flow and tissue perfusion. Hypothermia also influences blood rheology and vascular geometry. A decrease in temperature provokes direct vasoconstriction and increases viscosity, creating sludging and stasis at the capillary level, resulting in reduced blood flow. These effects are counteracted by hemodilution. Is there an "acceptable" degree of hemodilution? It is common to see hematocrits of 18% to 21% during CPB, and even extreme hemodilutional hematocrits of <15% are well tolerated when used for circulatory arrest cases or for patients who are Jehovah's Witnesses. A general rule of thumb is that the hematocrit in percent should not exceed the desired level of hypothermia in degrees Celsius. Currently our threshold for adding packed cells to the pump is a hematocrit of <18%.

We use a blood conservation strategy as follows:

1. A transfusion threshold at a hematocrit of less than 18% unless the patient has a history of cerebrovascular disease, carotid stenosis, or diabetes
2. The patient's clinical condition mandates a transfusion trigger of 21% to 24% (advanced age, severity of illness, cardiac function, end-organ ischemia, massive or active blood loss)
3. Routine use of cell-saver, except in cases of infection and/or malignancy
4. Utilizing low-prime and mini-extracorporeal bypass circuits (Table 36.5).

Table 36.5 Priming Volume per Length of Tubing of Given Diameter

Tubing diameter (in.)	Priming volume (ml/ft)
3/16	5.00
1/4	9.65
3/8	21.71
1/2	38.61

Instituting Cardiopulmonary Bypass and Decompressing the Left Heart

CPB is begun at the instruction of the surgeon. Visual inspection of the field, monitors, and bypass lines as the perfusionist initiates CPB will provide an immediate assessment of effectiveness of the conversion to artificial cardiopulmonary support. After confirming the surgeon's instruction, the perfusionist initiates CPB by releasing the arterial line clamp and slowly transfusing the patient with volume from the heart–lung machine (Fig. 36.10). The arterial blood flow in the

Table 36.6

$$\text{Hemodilutional hematocrit (hct) (\%)} = \frac{\text{Patient blood volume} \times \text{wk (kg)} \times \text{hct (\%)}}{(\text{Patient blood volume} \times \text{wt [kg]}) + \text{priming volume (ml)}}$$

extracorporeal circuit should be free flowing and exhibit a reasonable extracorporeal line pressure. A sudden spike in the extracorporeal line pressure may indicate an occluded arterial line, a malpositioned aortic cannula, or an aortic dissection. Should this occur, CPB is terminated immediately and the cause identified and corrected. As soon as it is obvious the arterial flow is unobstructed, the perfusionist releases the venous clamp, diverting the patient's venous return blood into the CPB circuit (Fig. 36.10). The right heart should be decompressed and the central venous pressure should be <5 mmHg. A high central venous pressure and poor venous drainage at the initiation of CPB may indicate a malpositioned venous cannula, a kinked venous line, an "air lock," venous cannulae that are too large or too small, an inappropriate height between the operating table and the venous reservoir, an inappropriate amount of vacuum, or a vacuum leak. During this transition period of 1 to 2 minutes, the perfusionist gradually increases the rate of arterial flow, the ventricles receive less blood, and the pulsatile arterial waveform diminishes and becomes "flat-lined." Once total bypass is achieved, a continued pulsatile arterial waveform signifies the LV is receiving unwanted blood from aortic valvular insufficiency, excessive bronchial venous return, or incomplete drainage of the systemic venous return.

An acute, transient state of systemic arterial hypotension, resulting from hemodilution and vasoactive substance release, is common on the initiation of CPB. Treatment with alpha agonists is usually not necessary to overcome this acute state because the mean arterial pressure will generally increase with the initiation of systemic cooling (due to its induced vasoconstriction) and increased levels of endogenous catecholamines and angiotensin. The arterial pressure during total CPB is a mean or "flat-line" pressure, unless an artificial means is used to generate pulsatile flow. The mean pressure should be viewed as an index of the relationship between blood flow, volume, and arteriolar resistance, and not as a measure of perfusion adequacy.

The body surface area and weight of the patient determine the desired flow through the heart–lung machine while on CPB (Table 36.2). Although the subject of much debate, an "acceptable" mean arterial pressure ranges from 35 to 90 mmHg. However, in the presence of known coronary artery stenosis or ventricular hypertrophy, a perfusion pressure of 60 to 80 mmHg should be maintained. In patients with known carotid artery stenosis, we recommend keeping the perfusion pressure between 80 to 100 mmHg. In general, if the systemic vascular resistance index and mixed venous blood gases are normal, the resultant mean arterial pressure is "acceptable." In patients with severe aortic regurgitation, the surgeon should be ready to cross-clamp the ascending aorta if the heart becomes distended or if ventricular fibrillation occurs. If the heart continues to beat, the left atrium should not be entered until the aorta has been cross-clamped to prevent embolization of air to the brain. In children and infants with congenital heart disease, the right heart should not be opened until the aorta is cross-clamped or ventricular fibrillation has occurred. However, decompression of the LV is best achieved in infants and small children by tightening the vena cava tapes, opening the right atrium, and passing a small suction line through the patent foramen ovale into the LV. If the foramen is not patent, a small stab wound will permit this approach.

Cooling the Patient

The perfusionist can begin cooling the perfusate to induce hypothermia once full flow and adequate decompression are established. The primary advantage of hypothermic CPB is the reduced metabolic rate and oxygen consumption; although not linear, this approximates 5% to 7% per degree Celsius that the body temperature is lowered. In addition, hypothermia sustains intracellular reservoirs of high-energy phosphates (essential for cellular integrity) and preserves high intracellular pH and electrochemical neutrality (a constant OH^-/H^+ ratio). As a result of these associated interactions, hypothermic patients can survive periods of circulatory arrest of up to 1 hour without suffering from the effects of anoxia (Table 36.7).

Hypothermia may be induced by surface cooling using cooling blankets and ice packs applied directly to the patient, or by core cooling with cold perfusate from the extracorporeal circuit. Because tissues and organs have varying blood flows, systemic cooling is not a uniform process. To minimize this, we combine the two methods, maintain high perfusion flow rates of 2.2 to 2.5 L/min/m^2, and limit the rate of the cooling to <1 degree Celsius/minute until the desired temperature is reached. Thereafter, perfusion flow rates are adjusted to maintain "normal" mixed venous blood gases. Bladder and nasopharyngeal temperatures are monitored to ensure uniform temperatures.

Rewarming

As the intracardiac procedure is being completed, systemic rewarming is instituted by gradually increasing the perfusate temperature. Rewarming is slower than cooling because of the maximum 10 degrees Celsius permissible temperature gradient between perfusate and nasopharyngeal temperatures, the maximum allowable blood temperature of 42 degrees Celsius, and the reduced thermal exchange as the temperature gradient between the patient and perfusate narrows. During this state, the warming blanket is set to 40 degrees Celsius, the perfusion flow rates are increased to 2.5 to 3.0 L/min/m^2 (Table 36.2), and pressure permitting, pharmacologic vasodilation is used. When the bladder temperature reaches 32 degrees Celsius, the patient begins to vasodilate spontaneously and the pharmacologic vasodilator may be terminated.

Removal of Air from the Heart

With the aorta cross-clamped, the patient is placed in a 30-degree head-down position, the caval tourniquets are loosened, and the perfusionist restricts venous return to the pump. The right heart begins to fill, and the anesthesiologist ventilates the lungs. The heart is gently massaged while the vent in the LV continues to drain. The antegrade cardioplegic cannula is placed on suction and, as more blood is massaged through the left heart, some air is removed via the antegrade cardioplegic cannula. Once all air appears to have been evacuated, the balloon of the

Table 36.7 Definition of Levels of Hypothermia and Approximate "Safe" Circulatory Arrest Times

Hypothermia level	Patient temperature (°C)	Circulatory arrest times (min)
Mild	37–32	5–10
Moderate	32–28	10–15
Deep	28–18	15–60
Profound	>18	60–90

retrograde cardioplegic cannula is deflated, pump flow is reduced to one-half of calculated flow, arterial pressure is reduced to 50 mmHg, and the aortic cross-clamp is removed while suction is maintained on the antegrade cardioplegic cannula. Transesophageal echocardiography is used to determine if there is residual air within the heart. If there is, the operating table is rocked from side to side, the left atrial appendage is inverted, and gentle massage of the heart is continued, and the sternum and sternal retractor can be shaken by the surgeon to dislodge air bubbles within the heart. The heart is defibrillated when all air has been evacuated. Venous return is restricted intermittently to allow the heart to eject. Suction on the intracardiac vent is discontinued, and suction is maintained on the aortic cardioplegia/vent cannula as the heart continues to eject. When echocardiography confirms that the left heart is free of air, the operating table is restored to a level position and the aortic cardioplegia/vent cannula, the retrograde cardioplegia cannula, and the LV vent are removed, and the pursestrings tied.

Appropriate postoperative monitoring lines are inserted by the anesthesia team prior to the sternotomy and, if indicated, temporary pacing wires are sutured to the right ventricle and the right atrium. Rewarming is continued until the nasopharyngeal temperature is 37 degrees Celsius and the bladder temperature is >36 degrees Celsius.

This rewarming usually takes 3 to 5 minutes for each 1 degree Celsius increase.

Discontinuing Cardiopulmonary Bypass

Especially if the patient had compromised ventricular function preoperatively, or had a lengthy or complex repair, termination of CPB is performed gradually, with constant communication between surgeon, perfusionist, and anesthesiologist. The surgeon will always check to be sure the LUNGS are being VENTILATED before weaning the patient from CPB. Because the heart cannot generate a cardiac output until the central blood volume is restored, the perfusionist progressively occludes the venous return line, translocating blood volume from the venous reservoir into the patient's vascular system. The patient is now on "partial" CPB, with blood flowing through the heart and pulmonary circulation. When the blood volume in the heart reaches an adequate level, the aortic valve begins to open with each heartbeat, and a measurable cardiac output and arterial waveform will be observed with each cardiac contraction. The translocation of volume by the perfusionist is continued until the arterial systolic pressure reaches 100 mmHg. The perfusionist then terminates CPB by completely occluding the venous and arterial lines. Thereafter, the perfusionist transfuses volume to the patient to maintain a systolic blood pressure of 100 mmHg unless the heart becomes distended.

If the heart does not function effectively when CPB is terminated, bypass is reinstituted to prevent overdistention of the LV or hypoxia, and inotropic support will be started. When good cardiac function and hemodynamic stability have been achieved, CPB is discontinued, and decannulation can begin. If two venous cannulas were used, the inferior caval cannula is removed, and the pursestring suture is tied. The superior caval cannula can be repositioned in the right atrium in case CPB needs to be reinstituted. With stable cardiac function the superior cannula is removed, and the tourniquet is drawn tight. A few minutes are allowed to confirm hemodynamic stability and adequate cardiac function. The ventilation, blood gases, cardiac output, and blood pressure are watched carefully over the next 5 to 10 minutes to ensure stable hemodynamic function of the heart, and good oxygenation by the lungs. Transesophageal echocardiography will confirm that the repair is complete and myocardial function is adequate.

Reversal of Heparin

With stable cardiac function, now is the time to reverse the anticoagulation by administering protamine. All pump suckers are turned off, and the anesthesiologist administers a test dose (50 mg) of protamine. If there is no allergic reaction within a few minutes, the anesthesiologist administers one-third to one-half of the total calculated dose of protamine. Under no circumstances should shed blood be returned to the extracorporeal circuit once protamine is introduced into the patient's circulation for fear of clotting the CPB equipment. If there has been no hemodynamic instability from this dose of protamine after 5 minutes, decannulation can begin, and the rest of the protamine is given slowly over 15 minutes. If the full dose of protamine is given rapidly over 5 to 10 minutes, a "protamine reaction" with vasodilatation and profound hypotension may ensue.

Removal of the Arterial Cannula

In adult patients with two pursestring sutures, the tourniquets are slipped off the pursestring sutures. The surgeon holds the arterial line while the assistant places the first overhand throw in the pursestring suture on his or her side. The arterial cannula is removed, and the assistant's pursestring is cinched down and held in position for hemostasis. The surgeon takes the pursestring on his or her side and ties this firmly. The assistant then completes tying the other pursestring suture. Once all protamine has been administered and the patient is stable, the pursestring suture on the right atrial appendage is tied and, if necessary, reinforced with a second suture. Thereafter, final hemostasis and surgical closure of the wound are performed according to protocol.

Sternal Closure

The pericardial cavity, and frequently the left pleural cavity (if the left chest has been opened with take-down of the mammary artery), is drained with two or three chest tubes. Our preference is for a flat tube to be placed under the right ventricle and into the posterior part of the pericardial cavity; a round Blake tube drains the anterior retrosternal space, and another Blake drain is placed in the left chest. These chest tubes are brought out through 10 to 12 mm incisions just below the bottom end of the sternotomy incision, on either side of it. The left chest tube can be brought out through an intercostal incision in the anterior axillary line just above the abdomen (draining the bottom of the left hemithorax), or through a small incision just lateral to the mediastinal tube incisions.

Closure of the sternum can be accomplished in a variety of ways. We favor a very secure closure, as if a patient sneezes; it is always very disappointing if he/she dehisces his/her sternotomy closure. Our skin incisions are closed with a subcuticular closure.

PUMP-ASSIST CORONARY ARTERY BYPASS GRAFTING

In off-pump CABG the coronary revascularization is usually performed without using CPB. However in a small percentage of cases the support of CPB makes the operation safer. If a patient has had a recent transmural myocardial infarct, or has an ejection fraction of 5% to 10% some surgeons use "pump assist." Normothermic CPB is used without arresting the heart,

and the coronary bypasses performed in the usual off-pump way.

FEMOROFEMORAL BYPASS

Certain complex problems can be managed more safely using femorofemoral bypass, rather than traditional CPB. A mycotic aneurysm of the ascending aorta that is right up against the back of the sternum, and is bulging out over the top of the sternum would pose a major risk of massive hemorrhage with a standard sternotomy. Again, a redo CABG operation where there is a patent internal mammary graft in the retrosternal area, or a redo operation when the right ventricle is stuck to the posterior table of the sternum might also benefit from femorofemoral bypass. Finally, rapid rewarming of a patient who has undergone a hypothermic cardiac arrest can easily be achieved by this form of full CPB support. Again, this can be performed using percutaneous cannulation or open cannulation techniques. Especially if there is vacuum-assisted venous return, full CPB can be established using femorofemoral bypass, with core-cooling and rewarming.

LEFT HEART BYPASS

Patients with descending thoracic or large thoracoabdominal aortic aneurysms have a lower incidence of spinal cord complications if left heart bypass improves distal organ perfusion during the aortic repair. This support system is not truly CPB as there is no oxygenation of the blood, but simple pumping of oxygenated blood from the left atrium to the arterial vessels distal to the clamped-off aneurysm. For this type of bypass a cannula is passed through a pursestring suture in the left inferior pulmonary vein, and advanced into the left atrium. All air is removed by back-bleeding and then clamping the atrial cannula. The distal cannula is placed through two pursestrings in the distal thoracic aorta below the aneurysm, or into the femoral artery. During this left heart bypass the flows range from 1 to 3 L/min. After completion of the aneurysm repair the cannulae are removed and the pursestrings tied.

BIG PICTURE

Modern CPB, incorporating excellent drainage of the right side of the heart, antegrade and retrograde myocardial protection, aortic and if necessary LV venting, allows the performance of complex reconstructive procedures with great safety. However, it requires many technical procedural steps, and makes for a very busy operative field (Figs. 36.10 and 36.11).

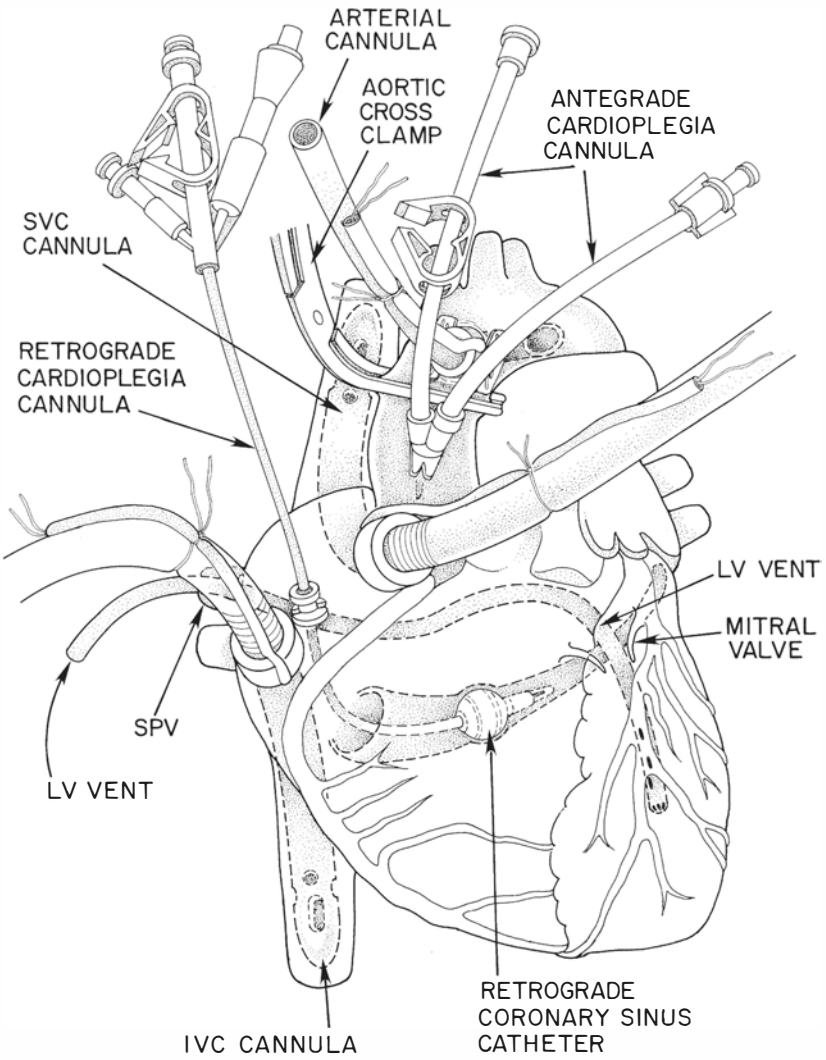


Fig. 36.11. Completed cannulation for total cardiopulmonary bypass. The left ventricular, venting catheter has been inserted via the right superior pulmonary vein. The retrograde cardioplegia catheter is in the coronary sinus. The antegrade cardioplegia catheter is in the ascending aorta proximal to the aortic occluding clamp. (IVC, inferior vena cava; LV, left ventricle; SVC, superior vena cava.)

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EDITOR'S COMMENTS

The authors of this chapter have done a very careful analysis of the basics of cardiopulmonary bypass. They have emphasized familiarity with the devices, standardized protocols, and most importantly teamwork. Cardiac surgery is a team sport and this cannot be overemphasized. Discussions at the beginning of each surgery will certainly clarify any issues between the various groups involved in the care of these very important patients. I think the techniques for cannulation as well as the approaches to avoid the disasters that are inherently present in our specialty are very well done. I would like to emphasize the importance of the discussion on venous drainage particularly assisted venous drainage. Keeping the right heart empty markedly protects the heart by avoiding distention and induced fibrillation as blood enters the coronaries in a retrograde fashion. This is an area that I truly emphasize in my practice and it is well described in this chapter.

ILK

37 Myocardial Protection

Richard J. Shemin and Yousef M. Odeh

This chapter reviews current concepts regarding myocardial protection during cardiac procedures including minimally invasive procedures with or without robotic assistance. In addition, myocardial protection in orthotropic heart transplant will be discussed. We provide the rationale of our strategy and describe the methodology of our approach in detail.

The strategies available for myocardial protection have led to adversarial positions regarding warm versus cold blood cardioplegia, as well as antegrade versus retrograde delivery. This creates confusion and potentially deprives the patient of the benefits of each method. In principle, cardioplegia markedly reduces oxygen demand in the arrested heart and must be delivered in sufficient quantity to all regions to match myocardial demand (Fig. 37.1). This has led to our use of antegrade and adjunct retrograde delivery, cold methods to reduce the demands allowing bloodless visualization, and warm reperfusion for resuscitation.

Our "integrated method" is a strategy of myocardial protection that combines the advantages of different concepts, allowing the operation to be conducted without interruptions.

The principle goals of cardioplegia are:

(1) Providing and maintaining electromechanical arrest of the myocardium.
(2) Sustained cooling of the myocardium, while flushing out unwanted toxins and warm blood.
(3) Limiting myocardial edema and providing buffering capacity.
(4) Limiting ischemic and reperfusion injury to the myocardium.

Electromechanical activity during cardiac arrest raises oxygen demand during ischemia. Hypothermia is a crucial component of myocardial protection because it reduces myocardial oxygen demand (Fig. 37.1). Blood cardioplegia is now preferred by most surgeons because of its versatility. It maintains oncotic pressure, is a natural buffering agent, has advantageous rheologic properties, and is a free-radical scavenger. Blood cardioplegia also limits reperfusion injury and reverses ischemia/reperfusion changes in the acutely ischemic myocardium. These beneficial features may not be possible with crystalloid cardioplegia.

Noncardioplegia methods have been applied in special situations. For example, when a severely calcified aorta is encountered, the preferred technique is deep hypothermic arrest (approximately 20°C) without aortic clamping.

COLD BLOOD CARDIOPLEGIA

Cold blood cardioplegia allows complete myocardial recovery after 4 hours of ischemia in normal hearts. Hypothermia lowers myocardial oxygen demand and ischemic damage when coronary flow is interrupted in the course of the revascularization procedure, provided the cardioplegic perfusate is distributed adequately with reinfusions. Hypothermia alone, however, does not avoid injury in chronically "energy-depleted" (ischemic) hearts (Fig. 37.2). Furthermore, hypothermic crystalloid cardioplegia adds some disadvantages, such as shifting the oxyhemoglobin association curve leftward, retarding Na^+/K^+ adenosine triphosphatase to produce edema activation of platelets, and creating free radicals. Blood cardioplegia consists of four parts of blood to one part of crystalloid solution. This limits the hemodilution seen with crystalloid cardioplegia during repeated infusions.

WARM BLOOD CARDIOPLEGIA

Warm blood cardioplegia (37°C) given initially (induction) limits reperfusion damage in ischemic hearts. It enhances metabolic repair by channeling aerobic adenosine triphosphate production to reparative processes. Other cardioplegic components, citrate phosphate dextrose (CPD) and buffers (THAM [tromethamine; tris-hydroxymethylaminomethane]), limit calcium influx and acidosis. Clinical studies confirm experimental findings and show that warm blood cardioplegia ("hot shot") after ischemia improves recovery. Warm cardioplegic induction and reperfusion solutions are augmented with the amino acids glutamate and aspartate to replenish key Krebs-cycle intermediates depleted by ischemia. These additions enhance the reparative processes after a period of myocardial ischemia (Fig. 37.2).

Warm (37°C) cardioplegia may also be used after aortic unclamping to restore rhythm and improve hemodynamics in the unstable patient following cardiac surgery. When used for impaired hemodynamics, the left ventricle requires venting to reduce distention and myocardial stress. The antegrade flow is given for 5 to 10 minutes.

ANTEGRADE/RETROGRADE PERFUSION: INTERMITTENT OR SIMULTANEOUS?

A cardioplegic perfusate requires even distribution. Adding retrograde perfusion improves subendocardial perfusion, avoids ostial cannulation during aortic valve procedures, limits removal of retractors during mitral procedures, and permits flushing of air and atheroma during coronary reoperations. Transatrial coronary sinus cannulation allows safe and rapid retroperfusion into the coronary sinus and has become widely adopted. Experimentally, right ventricular nutritive flow appears limited, thus hypothermia is essential to lower oxygen demand and provide effective right ventricular protection. Clinical studies show that switching from antegrade to retrograde perfusion increases oxygen uptake and lactate washout, indicating that each mode perfuses different areas. Therefore, both antegrade and retrograde perfusions are required (Fig. 37.3).

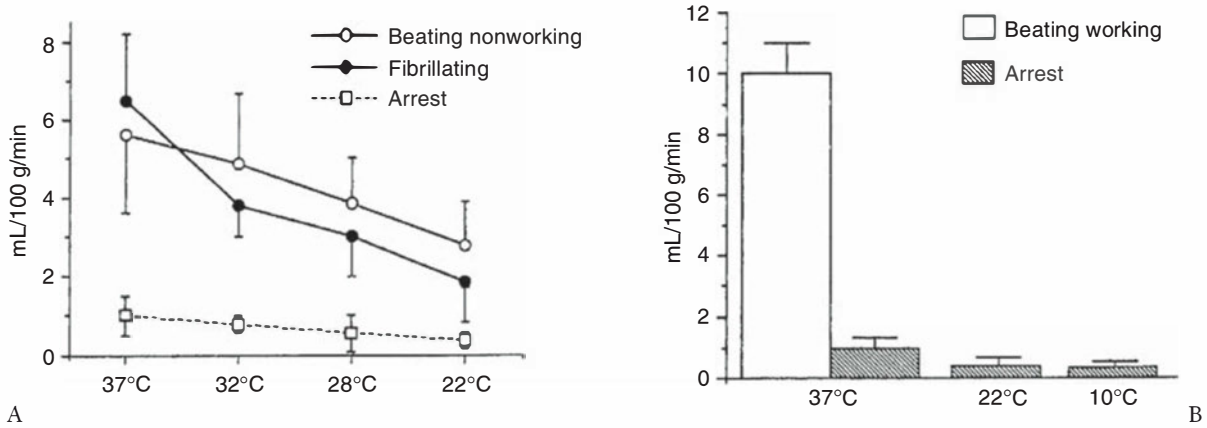


Fig. 37.1. (**A**) Left ventricular oxygen requirements of beating nonworking, fibrillating, and arrested hearts from 37°C to 22°C. Note the lowest requirements during arrest. (**B**) The left ventricular oxygen requirements of a beating working and an arrested heart at 37°C, 22°C, and 10°C. Note in panel A the low oxygen demands of arrest and in panel B the negligible change between 22°C and 10°C as the heart rewarms from collateral flow. Panel B shows higher demands than panel A if electromechanical activity recurs when systemic perfusate washes out the cardioplegic solution.

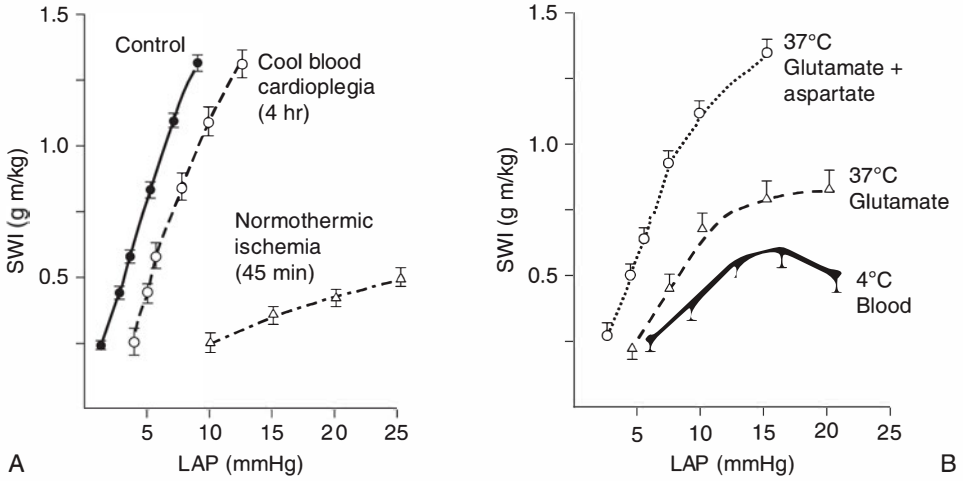


Fig. 37.2. (**A**) Left ventricular function in normal hearts subjected to 4 hours of aortic clamping with cardioplegia with blood every 20 minutes compared with depressed function after 45 minutes of normothermic arrest without cardioplegia. The type of protection is more important than the duration of aortic clamping. (**B**) Left ventricular function when jeopardized hearts undergoing 45 minutes of normothermic ischemia are subjected to two more hours of aortic clamping. Note (1) no further improvement when only cold cardioplegic perfusate is given over the 45-minute arrest period and (2) progressively increased recovery when the cardioplegic solution is supplemented with warm glutamate and aspartate during induction of cardioplegia and reperfusion with intermittent cold doses of blood every 20 minutes of supplemental aortic clamping. These data suggest the value of amino acid enrichment in jeopardized hearts. LAP, left atrial pressure; SWI, stroke work index.

The limitations of antegrade and retrograde delivery were first overcome by *using* both antegrade and retrograde perfusions intermittently. It is possible to *simultaneously* deliver retrograde cardioplegia via the coronary sinus and antegrade cardioplegia via direct ostial or vein graft infusion. Myocardial venous hypertension is prevented by drainage through the Thebesian veins. Studies document the safety of simultaneous vein graft and coronary sinus perfusion.

INTERMITTENT/CONTINUOUS INFUSION

Continuous cardioplegic perfusion has been advocated to avoid ischemia by antegrade or retrograde delivery, but adequate protection may not be achieved at usual flow rates and vision becomes obscured during infusion. A quiet/dry field requires "intentional" ischemia by intermittently stopping the flow of cardioplegic solution. Intermittent replenishment restores hypothermia, maintains electromechanical arrest, flushes accumulated metabolites, and counteracts acidosis and edema. Initially, the heart is arrested with high-dose potassium (20 to 40 mEq/L) blood cardioplegia (Table 37.1), and low-dose cold potassium (8 to 10 mEq/L) blood cardioplegia (Table 37.2) is used for intermittent repeat doses. Such reinfusions (every 15 to 20 minutes) are often retrograde, directly into the aorta or, via the each coronary ostium. Cold blood with a hematocrit of

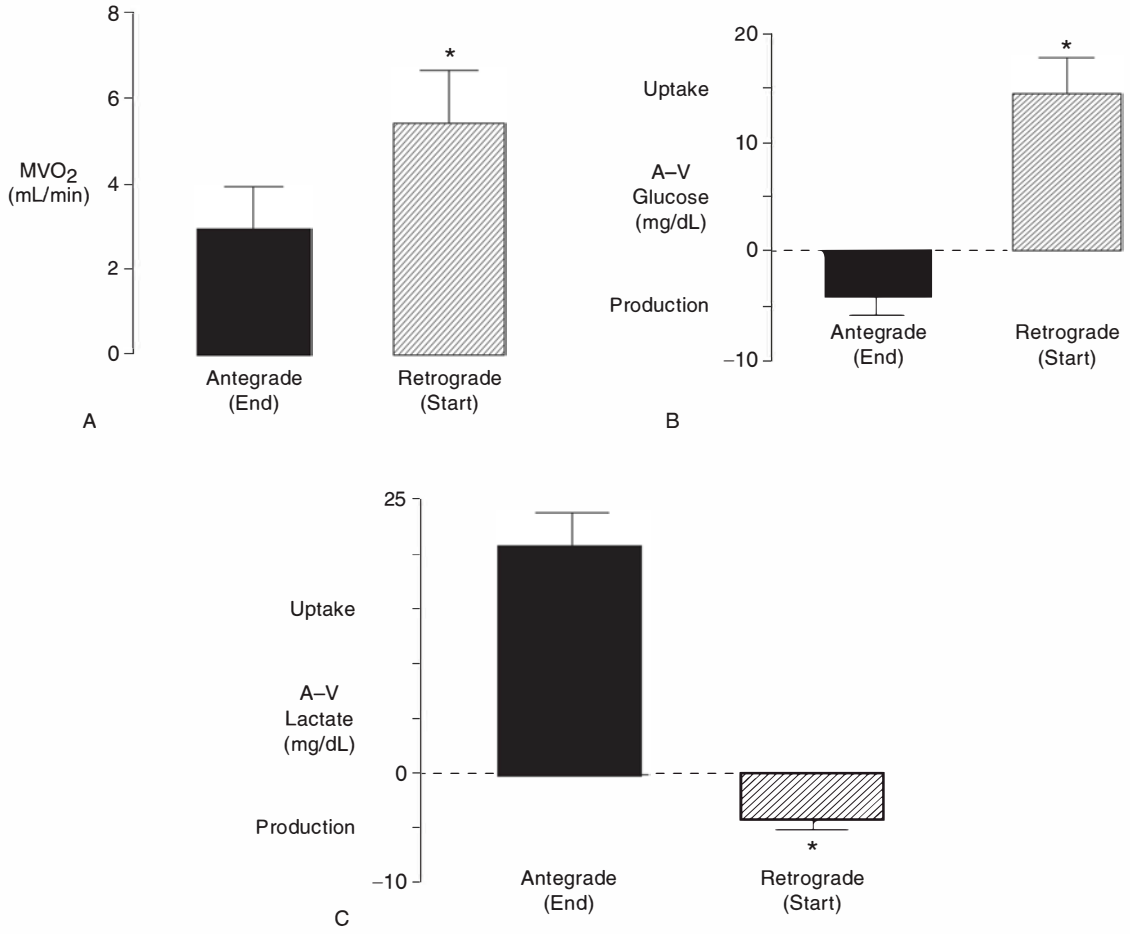


Fig. 37.3. Myocardial metabolic changes in coronary patients when administration of the cardioplegic induction solution is converted from antegrade to retrograde. Note the increase in myocardial oxygen uptake, glucose uptake, and lactate production, suggesting different areas of perfusion by the antegrade and retrograde methods of delivery. This implies an advantage to both methods. A–V, arteriovenous.

Table 37.1 Cold Blood Cardioplegia Solution

Cardioplegia additive	Volume added (ml)	Component modified	Concentration delivered[a]
Potassium chloride (2 mEq/ml)	31	Potassium ion	26–28 mEq/L
$NaHCO_3$ (mEq)	5	pH	pH 7.6–7.8
2.5% Dextrose in 0.45% NS	500	Osmolality	280–300 mOsm

[a]When mixed in a 4:1 ratio with blood.

Table 37.2 Cold Multidose Blood Cardioplegia Solution

Cardioplegia additive	Volume added (ml)	Component modified	Concentration delivered[a]
Potassium chloride (2 mEq/ml)	7.5	Potassium ion	8–10 mEq/L
$NaHCO_3$ (mEq)	5	pH	pH 7.6–7.8
2.5% dextrose in 0.45%NS	500	Osmolarity	280–300 mOsm

[a]When mixed in a 4:1 ratio with blood.

approximately 20% is infused at a pressure of <40 mmHg and infused for a duration of 2 minutes.

At the completion of the cardiac procedure, controlled reperfusion is achieved by administering a dose of warm cardioplegia ("hot shot"), administered either antegrade, retrograde, or both. This can be followed by a continuous infusion of warm blood into the aortic root while the cross-clamp is still in place at a flow of 300 to 350 ml/min with a pressure of ≤80 mmHg. The aortic clamp is released within 5 minutes after adequate contractility is observed.

CANNULAS AND DEVICES FOR DELIVERING AND MONITORING ANTEGRADE/RETROGRADE AND SIMULTANEOUS ANTEGRADE/RETROGRADE CARDIOPLEGIA

All operations include the use of a cardioplegic heat exchanger for cold and warm perfusion (Fig. 37.4) cannulas for

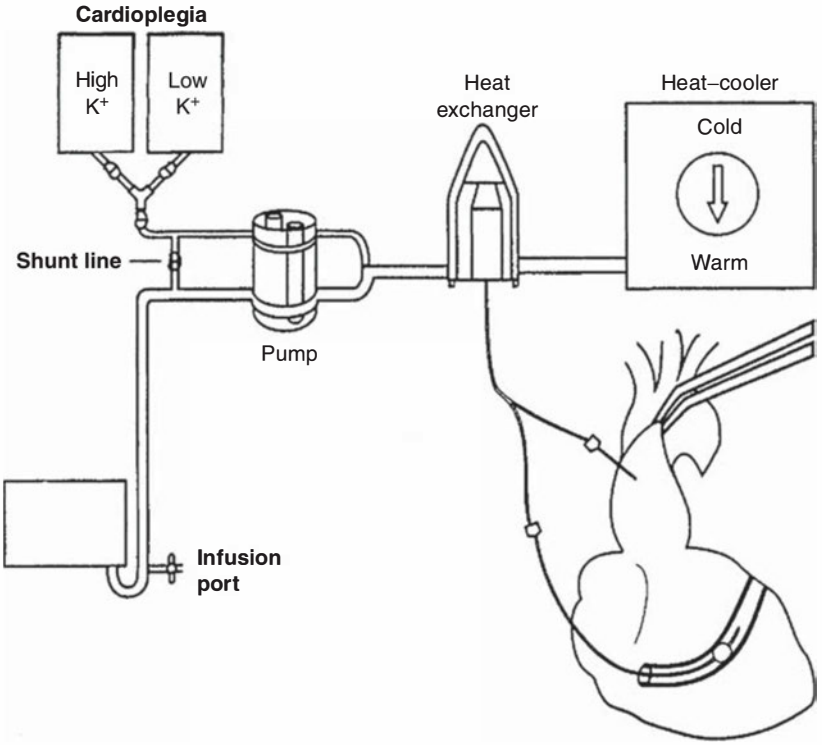


Fig. 37.4. Blood cardioplegic delivery system, whereby the cardioplegic solution contains high-dose and low-dose potassium bottles. These are mixed 4:1 with blood from the oxygenator. The shunt line allows delivery of terminal regular blood through the roller head of the pump after the warm reperfusate is given.

antegrade and retrograde delivery and a monitoring-infusion system. The system we use is shown in Figure 37.5. This has been found to be the simple, yet effective way for delivering and monitoring cardioplegia in conventional cardiac cases.

The main cardioplegia line is connected to a three-way stopcock. From here, the cardioplegia is set with branches allowing either antegrade flow or retrograde flow. The retrograde catheter has a port to transducer pressure allowing measurement and display of the waveform. The stopcock allows alternating between antegrade and retrograde flows.

Antegrade Cannula

An antegrade cannula is placed high in the ascending aorta below the proposed site of the aortic cross-clamp. This site is not used as a proximal graft site. A 4-0 purse-string suture with a tourniquet is used to secure the cannula and eventually tied at the end of the procedure, and oversewn to ensure hemostasis. After commencement of cardiopulmonary bypass, the heart should empty, as indicated by the collapse of the pulmonary artery, indicating good systemic venous drainage. The aorta is clamped, and antegrade cardioplegia is delivered for 2 minutes at a rate of 200 ml/min. In coronary bypass procedures, additional antegrade cardioplegia is hand delivered through the saphenous vein graft. The proximal vein graft proximal anastomosis is completed. Therefore, subsequent antegrade doses are delivered via the completed graft(s). All proximal grafts are performed during the cross-clamp period.

Retrograde Cannula

Transatrial approaches permit coronary sinus cannulation without right heart isolation. Cannulation is best performed before venous cannulation to avoid the venous cannula obstructing the path the retrograde cannula traverses to easily access the coronary sinus ostia. This is usually performed under ECHO guidance. We use a malleable stylet and inflating balloon cannula. A high atrial 4-0 purse-string suture is placed from the right side of the operating table (Fig. 37.6). The cannula should always enter the coronary sinus easily. The cannula is advanced 2 to 3 cm within the coronary sinus. The position is confirmed with ECHO images, the presence of dark blood emerging from the cannula, and the appropriate

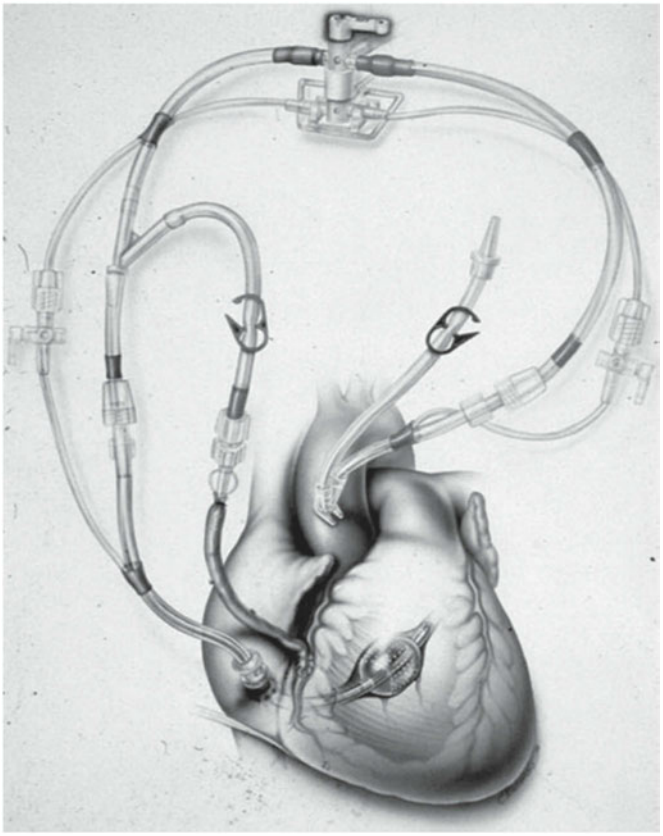


Fig. 37.5. Tubing use for antegrade, retrograde, and vein perfusion connected to the retrograde arm. Note the stopcock that allows simultaneous delivery of cardioplegic perfusate and monitoring of arterial or coronary sinus pressure. The arm is connected to the retrograde limb, allowing simultaneous retrograde/antegrade perfusion down the vein graft or right coronary ostium during aortic valve replacement.

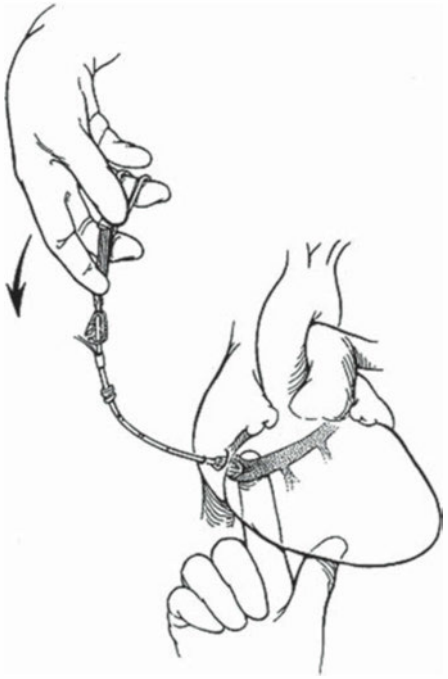


Fig. 37.6. Method for introducing the retrograde cannula from the right side of the operating table. Note the direct course at 45 degrees from the coronary sinus to the junction of the coronary sinus and the left atrium.

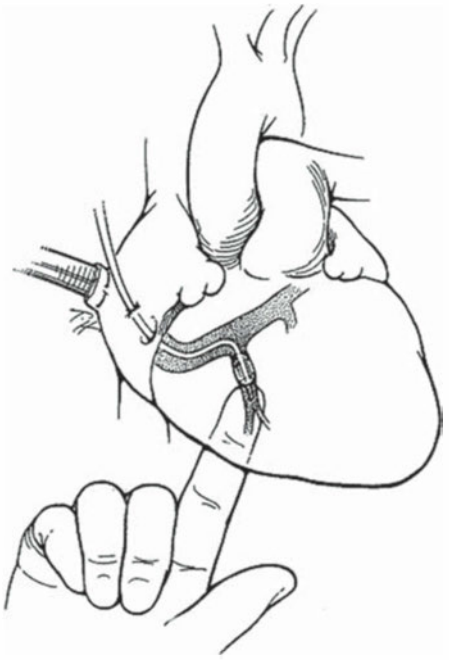


Fig. 37.7. Palpation of the cannula tip without deep palpation. This means the cannula is in the posterior descending vein and it should be withdrawn and repositioned so that it can be palpated far posteriorly.

"ventricularization" waveform that is transduced from the retrograde pressure measuring line.

Cannulation on partial bypass is done with the right atrium slightly distended to keep the coronary ostium open. Either ECHO-guided techniques or finger palpation can be employed to guide the retrograde cannula into position. Failure to intubate the coronary sinus is rare (1% to 2%) and indicates a fenestrated Thebesian valve or a flap over the ostium. Occasionally, the cannula fails to advance because it has gone into the posterior descending vein (Fig. 37.7). Alternatively, bicaval cannulation can be used. The right atrium is opened with a small incision and cannulation is performed directly after the flap is retracted or the Thebesian valve is opened. If the right atrium is open, a purse-string suture around the coronary sinus prevents reflux. This direct approach is preferred when the right atrium is opened for tricuspid repair/replacement; transseptal approach to the mitral valve and biatrial MAZE procedures. The cannula is pulled back to the purse string to allow perfusion of proximal veins entering the sinus and allow cryothermy to the mitral annulus and coronary sinus without the cannula interfering.

During retrograde infusions, the posterior descending vein fills readily with oxygenated blood, which confirms an extensive myocardial venovenous collateral network. The cannula is withdrawn slightly if there is failure to fill the posterior descending vein or if there is no egress of blood from the right coronary ostium or open right artery during aortic or coronary operations. This dark blood signifies nutritive flow despite some shunting into the right atrium with inflating balloons. The blood becomes red, which reflects adequate perfusion. This is especially important if normothermic continuous cardioplegia is delivered.

Coronary Sinus Injury

The coronary sinus can be injured due to forceful cannulation or continued administration of cardioplegia when coronary sinus pressure exceeds 50 to 60 mmHg. This can occur while the heart is elevated during circumflex artery grafting. The perfusionist notes high pressure, and then low pressure as a consequence of acute perforation, or the surgeon sees red blood accumulation within the pericardial well during infusions. Perforation can be repaired directly with 6-0 prolene sutures or with a pericardial patch if there is concern that the repair will constrict the coronary sinus. Hematomas can sometimes be left unattended and observed because low venous pressure allows self-containment after heparin reversal.

Monitoring Pressure

Measuring infusion pressure avoids edema and endothelial damage. During retrograde infusion, measurement of pressure verifies correct placement of the coronary sinus cannula.

Retrograde Pressure

Coronary sinus pressure ranges from 30 to 40 mmHg at an infusion rate of 200 to 250 ml/min. A coronary sinus pressure of >50 mmHg means improper positioning or cardiac retraction that kinks the venous system. This is treated by reducing the flow rate immediately, repositioning the catheter, and resuming flow. Balloons should not be moved during infusion because of coronary sinus wall traction and possible injury. Coronary sinus pressure of <20 mmHg implies that the balloon is not inflated or not occluding the coronary sinus. The cannula tip and balloon should then be palpated and repositioned. Other causes of low pressure are (1) persistent left superior vena cava (SVC), (2) failure of balloon inflation, and (3) rare left atrial unroofing of the coronary sinus noted during mitral procedures. The presence of a left SVC is usually determined before cardiopulmonary bypass, and the vessel is occluded with a tourniquet only if an intact innominate vein is present. If the innominate vein is absent, only antegrade cardioplegia is used, to avoid myocardial under perfusion.

CARDIOPLEGIC SOLUTIONS

Two important basic principles of blood cardioplegia are: (1) blood optimizes the rate of aerobic metabolism and (2) oxygen uptake occurs over time, so that infusion duration is more important than dose. Delivery at a fixed pressure for too short a time (especially with warm infusions) deprives the myocardium of the benefits of a full dose. The setup should include flow clamps to direct antegrade or retrograde flow of the cardioplegic solutions for induction, maintenance, and reperfusion. In addition, the option to flow the cardioplegia through an "inline" white blood cell filter to add leukodepletion to the cardioprotective strategy should be considered.

The following guidelines are used for flow rates in normal hearts and are increased by 50 to 100 ml/min for hypertrophied hearts (Table 37.3). Many formulations

Table 37.3 Delivery of Cardioplegic Perfusate

Induction	Antegrade perfusion	Retrograde perfusion
Cold	300 ml/min × 2 min until arrest	150 ml/min × 2.5 min
Maintenance	200 ml/min × 1 min	200 ml/min × 1 min
Reperfusion	150 ml/min × 2 min	150 ml/min × 2 min
Noncardioplegic blood	200 ml/min	200 ml/min

with various additives are used with excellent results. High-dose potassium (20 to 30 mEq/L) solution that contains components has been described previously. It arrests the heart promptly during either warm or cold induction and remains available if electromechanical activity recurs. Enrichment with amino acids (glutamate/aspartate) is useful in high-risk patients (Table 37.1), those with low output, ischemia, or hypertrophy. It is switched to the low-dose, nonenriched potassium solution immediately after arrest with cold induction to limit hyperkalemia. The low-dose potassium (10 to 15 mEq/L) solution (Table 37.2) is used for maintenance doses during cold cardioplegic infusions and does not contain amino acids. At the completion of all cardiac repairs, all patients receive a terminal warm cardioplegic perfusate ("hot shot"). This solution is substrate enhanced in patients with poor ventricular function or long cross-clamp times in complex procedures. In transplant patients, an additional 3 to 5 minutes of noncardioplegic warm normal blood (leukocyte depleted) with the aorta clamped is given. During infusion of the warm reperfusate, it is not uncommon to observe a mild degree of transient hypotension due to vasodilation.

Normocalcemic blood from the extracorporeal circuit is added to the cardioplegia solution. In adult patients, if citrated solutions (CPD) are used; calcium should be added to the cardiopulmonary bypass circuit prime prior to weaning from bypass. This avoids unintentional hypocalcemia, which can damage the membranes of the sarcolemma. In pediatric patients, systemic hypocalcemia occurs when the pump is primed with citrated blood. Consequently, no calcium is added until the end of the procedure, when the calcium concentration is normalized before bypass is stopped.

CORONARY BYPASS PROCEDURE

During coronary bypass, the body's core temperature is cooled and kept between 32°C and 34°C. The heart is arrested with high-potassium *cold* blood cardioplegia in elective low-risk operations with good ventricular function. In cases where acute ischemia is present or in cases of severely depressed systolic function, high-potassium substrate enriched *warm* blood cardioplegia ("warm induction") is used initially to arrest the heart. The administration of substrates in this situation has been proven to aid in the recovery of function after surgical repair. After the warm induction, low-potassium cold blood cardioplegia is infused into the aortic root and/or retrograde into the coronary sinus. Intermittent low-potassium cold blood cardioplegia is used during the course of the grafting (Table 37.3). The ventricle is palpated; short axis view of the transesophageal echocardiography (TEE) and PA pressure are observed because cardiac distortion or aortic insufficiency (low aortic root pressure) may harmfully distend the ventricle. Similarly, retraction of the heart during retrograde delivery raises coronary sinus pressure by kinking the venous system. If pressure exceeds 50 mmHg, flow is reduced.

Following cardioplegic arrest, all anastomoses are constructed in a dry operative field using distal/proximal grafting in sequence. The right coronary graft is constructed first because retrograde nutritive perfusion is limited. Retrograde perfusion (200 ml/min for 2 minutes) is started after the last suture is placed in the first distal anastomosis with the suture line patulous to allow de-airing of the coronary artery with blood. The flow clamp connected to the graft is opened to evacuate air from the graft and to provide simultaneous graft antegrade and whole-heart retrograde cardioplegic perfusion while the suture is tied. The antegrade injection can be given by hand injection to remove air from the graft, test for anastomotic leaks, and determine resistance to runoff.

The vent is closed to distend the aortic root. The graft length is facilitated by holding the distended vein or arterial graft adjacent to the pericardium taking into account pulmonary artery distention when the heart will be full. A no. 11 blade is used to make a small incision where the proximal will be attached to the aorta, and a 4.0- to 4.5-mm punch is inserted into the small aortotomy and deployed. The proximal anastomosis is performed. Then, antegrade cardioplegia is administered allowing the root to fill with blood, evacuate any air, and the suture is then tied. Newly constructed grafts perfuse the heart after each proximal connection. The next distal graft is then constructed in a dry operative field while the aorta is vented. The sequence is repeated for each distal and proximal anastomoses, leaving one proximal graft to be connected to the aorta after the final distal left internal mammary artery (LIMA) anastomosis is constructed. Systemic rewarming is started while the LIMA anastomosis is begun. After the LIMA is anastomosed to the anterior descending artery, antegrade warm cardioplegia is delivered for 2 minutes initially and then retrograde.

After cross-clamp removal, the aortic root is vented and the heart usually begins to resume contraction. Clamps on the LIMA graft is removed just before starting the warm cardioplegia. When antegrade flow is started to de-air the aortic root, the grafts are de-aired with a fine needle vent hole to prevent air embolus.

In combined valve/coronary procedures, distal grafts are first placed. The valve is then replaced or repaired. Every 15 to 20 minutes, maintenance cold blood is delivered through retrograde catheter. All the proximal anastomoses are performed lastly with the aorta still cross-clamped. The warm (2 to 4 minutes) cardioplegic reperfusion is started. After the high-potassium cardioplegia is washed out, the heart begins to contract slowly.

The antegrade infusion of warm cardioplegia or warm blood is then infused into the aortic root with the cross-clamp in place at a flow of 250 to 300 ml/min. The heart resumes vigorous contractility within a couple of minutes. The aortic clamp is then released, and the aorta is vented.

REPEAT CARDIAC SURGERY

The integrated cardioplegia method has been found to be particularly useful in repeat operations. In coronary bypass operations, a generally accepted rule is that old grafts should be left undisturbed. This "no-touch" technique prevents embolization of friable atherosclerotic debris into the distal vasculature.

After sternotomy, only the aorta and the right atrium are dissected free of surrounding adhesions. After cannulation and institution of cardiopulmonary bypass, the retrograde cannula is inserted. The aorta is clamped and flow through a patent

LIMA is occluded. Induction cardioplegia or cold blood cardioplegia is administered into the aortic root to arrest the heart. Retrograde cardioplegia follows or is instituted early if there is ventricular distention.

After patent atherosclerotic vein grafts are divided proximally, the heart can be safely freed of surrounding pericardial adhesions. This avoids compression of old vein grafts and embolization. The added time of ischemia is only about 10 minutes, and the hemodynamics is not compromised by avoiding mobilization by dissection of the beating heart before arrest. Distal and proximal grafts are constructed in sequence as with all coronary operations. The single aortic cross-clamping technique avoids compression of old grafts and ischemia that can be induced by the side-biting method, in which graft flow is temporarily interrupted.

Clinical outcomes with this method in repeat coronary bypass grafting have demonstrated no increased mortality, even in patients with markedly reduced systolic function preoperatively.

MYOCARDIAL PROTECTION DURING MINIMALLY INVASIVE CARDIAC PROCEDURES

Although more cardiac procedures are being offered through minimally invasive incisions, it is imperative that the operation be as safe and more importantly have the same outcomes as traditional sternotomy approaches.

In this section, we will describe cardioplegia set-up and techniques for minimally invasive aortic valve procedures as well as for mitral valve and tricuspid valve operations.

With minimally invasive aortic valve operations, I choose to use a small (3 inch) upper sternotomy incision. The sternal incision extends from the sternal notch and ends at the level of the right third/fourth intercostal space. This exposes the entire ascending aorta and aortic root. Through this same incision, I have performed ascending and arch aortic aneurysms as well as aortic root operations, including valve-sparing root procedures.

With regard to aortic cannulation, the placement of the antegrade cardioplegia catheter is unchanged from my sternotomy approaches. The retrograde cardioplegia catheter is placed through a purse-string suture in the right atrium just below the appendage cannulation.

The catheter is then placed under ECHO guidance into the coronary sinus. Once cannula placement is confirmed with ECHO, waveform tracing, and dark blood return, the purse string is snared and the cannula position is secured. After the retrograde catheter is positioned, the venous cannula is placed. It is important to place the retrograde cannula first to avoid the pathway to the coronary sinus becoming obstructed by the venous cannula.

If the right atrium is not visible due to the upper sternotomy incision or that cannulating the right atrium is difficult because of exposure, then I cannulate the femoral vein percutaneously with a long venous 25F cannula. If there is not a contraindication, I prefer to cannulate the right femoral vein. The cannulation is performed using the Seldinger technique and assistance is gained from ECHO that verifies the wire placement and the advancement of the cannula over the wire into the right atrium. As with central venous cannulation, having the retrograde catheter in place prior to placement of the long venous catheter eases placement and avoids dislodgment of the retrograde catheter.

Shortly after the initiation of cardiopulmonary bypass, the aortic cross-clamp is applied and the TEE transgastric short axis view of the heart is monitored for ventricular distention. After 500 cc of pressure-controlled antegrade cardioplegia and cardiac arrest, the stopcock is turned and retrograde cardioplegia with a sinus pressure of approximately 40 mmHg is administered for a total dose of 1.0 to 1.2 L.

Throughout the case, retrograde cardioplegia is delivered at least every 20 minutes or at optimal times to ensure a dry field. While the aortotomy is being closed, the perfusionist prepares the warm reperfusion dose. Prior to tying the sutures, the retrograde followed by antegrade warm cardioplegia is given. Air is allowed to evacuate the aorta and the sutures are secured. After the warm cardioplegia is administered, the aortic root is vented with the head of the patient dependent. The cross-clamp is removed and the heart is allowed to return to sinus rhythm.

MYOCARDIAL PROTECTION DURING MINIMALLY INVASIVE (ROBOTIC-ASSISTED) MITRAL VALVE REPAIR/REPLACEMENT: WITH OR WITHOUT TRICUSPID REPAIR

For minimally invasive mitral and/or tricuspid valve operations, I use a submammary right mini thoracotomy incision. The chest is entered in the fourth intercostal space.

The femoral vein and artery are prepared for cannulation through a small infrainguinal transverse incision. Purse-string sutures and direct Seldinger technique allow cannula insertion with TEE guidance and position confirmation.

A SVC cannula is inserted into the jugular vein with TEE confirmation of the cannula's position. An antegrade cardioplegia/aortic root venting cannula is placed directly into the ascending aorta. A retrograde cardioplegia cannula is only used in patients with aortic insufficiency. This is placed through a purse-string suture in the right atrium. Under the guidance of TEE, the cannula is placed in the coronary sinus. Confirmation of placement is obtained with TEE, waveform with the balloon inflated, and the return of dark blood into the retrograde catheter. A percutaneous coronary sinus catheter as well as a combination of endoballoon catheter with antegrade cardioplegia and venting ports is commercially available. The principles of cold blood cardioplegia delivery and warm reperfusion cardioplegia are similar to full sternotomy approaches. The TEE and EKG are essential monitors of successful arrest and ventricular distention.

MYOCARDIAL PROTECTION DURING ORTHOTOPIC HEART TRANSPLANT

The donor heart is arrested with cold blood cardioplegia solution. The heart is sterilely wrapped for cold storage transplant. Reliable myocardial protection for 4 to 5 hours can be expected.

During implantation, topical ice slush with an insulating pad protecting the left phrenic nerve provides myocardial protection. After completing the anastomosis of the left atrium, a 10F catheter is guided from the right superior pulmonary vein across the body of the atrium and into the left ventricle. Cold (4°C) plasmalyte is allowed to infuse to cool the endocardium. The plasmalyte solution (Table 37.4) will exit from the donor aorta and is continued during the pulmonary artery and aortic anastomosis.

The pulmonary artery sutures are not tied to allow venting of the right ventricle during weaning from cardiopulmonary bypass. After the SVC and IVC anastomoses are completed, warm substrate-enhanced antegrade cardioplegia is given at a controlled reperfusion pressure of less then 80 mmHg for 4 minutes followed by warm blood (leukocyte depleted) for 3 to 4 minutes. The aortic cross-clamp is removed and the aortic root is vented.

TEE of the cardiac chambers ensure complete air removal. The pulmonary

Table 37.4 Constitution of Plasmalyte A (per 100 cc)

Sodium chloride	526 mg
Sodium gluconate	502 mg
Sodium acetate trihydrate	368 mg
Potassium chloride	37 mg
Magnesium chloride	30 mg
Osmolarity	294 mOsmol/L
pH	6.5–8.0
Sodium	140 mEq/L
Potassium	5 mEq/L
Acetate	27 mEq/L
Gluconate	23 mEq/L

artery suture line is tied as the wean from CPB is completed. A left atrial pressure line is inserted to monitor directly the left atrial pressure. The TEE is essential to monitor both left and right ventricular functions.

MYOCARDIAL PROTECTION DURING AORTIC ROOT REPLACEMENT

The myocardium must be carefully protected during extensive and long operations on the aortic root, as encountered with ascending aortic aneurysm or dissection. Here, the coronary ostia are often isolated for reimplantation into a graft. Intermittent cardioplegia can be administered directly into these ostia buttons, but care must be taken to avoid injury, particularly if operation is indicated because of dissection. Occasionally, when dissection extends down the right coronary or torsion cannot be prevented by extensive right coronary mobilization, grafting the right coronary artery with saphenous vein is helpful.

Cardioplegia should be administered intermittently retrograde and antegrade. This permits maximal distribution throughout the myocardium and does not interfere with the operation. The aortic root and ascending aorta can be repaired with ease in a dry field. After the root has been reconstructed and the left coronary ostia reimplanted into the graft, the graft is clamped distally and antegrade cardioplegia is infused. This allows left main cardioplegia to be given, checks for anastomotic leaks, graft orientation for the distal anastomosis and the site for implantation of the right coronary button. A final retrograde and antegrade dose of warm substrate-enhanced cardioplegia is administered for 2 to 4 minutes.

MYOCARDIAL PROTECTION FOR VENTRICULAR RESTORATION

Ventricular remodeling is an approach used in rebuilding the ventricular chamber in congestive heart failure patients. These patients are in the highest risk category with low EF and normally undergo coronary grafting, and approximately 25% need mitral valve repair. Both coronary artery bypass grafting (CABG) and mitral repair are accomplished with the previously described antegrade and retrograde methods of warm induction, cold maintenance, and substrate-enhanced reperfusion.

After removal of the aortic clamp, the reconstruction of the spherical left ventricular chamber into an oblique shape is performed. The method of protection is the "beating open" technique. Competence of the aortic valve together with ventricular venting via the left superior pulmonary vein avoids ventricular filling during repair.

Aortic perfusion pressure is kept at >75 mmHg because both experimentally and clinically, this higher pressure improves subendocardial perfusion: An improvement in the force of twisting is usually observed when perfusion pressure is increased. Restoration, with continuous perfusion, is *always* done *after* coronary grafting is completed to take advantage of the improved flow provided by the CABG procedure. Ventricular palpation with the beating method is very useful in the thick-walled akinetic heart, in which visual distinction between scar and viable muscle is difficult, especially if there is trabecular scar, or in the early phases after infarction when there is no evidence of scarring. Other advantages include (1) limiting ischemia by allowing ongoing coronary perfusion in severely dilated hearts during early learning curve, when the time period of restoration may be prolonged, (2) continuous perfusion to repay the "ischemic debt" after prolonged aortic clamping with cardioplegic protection was needed during the CABG and mitral repair phases of the procedure, and (3) more rapid discontinuation of bypass as the reperfused heart undergoes a long period of nutritive flow during restoration. Consequently, the beating method offsets the interval of "resting the heart" by prolonging the end of the procedure.

Determination of preoperative aortic insufficiency by ventriculogram or intraoperatively by ECHO is critical because only a small amount (i.e., 500 ml/min) will obscure the field. We normally use the beating technique. If aortic regurgitation is anticipated, the aortic clamp is left in place; proximal grafts are perfused via a Y connector rather than connected to the aorta, the internal mammary artery graft is opened, and antegrade normal blood perfusion is delivered to the beating heart during repair as shown in Figure 37.8.

CONCLUSION

Myocardial management as described is useful in all adult cardiac procedures by allowing them to proceed smoothly and more rapidly than usual because there is minimal interruption or delay for infusion of cardioplegic perfusate or noncardioplegic blood reperfusate. The duration of cardiopulmonary bypass is actually shortened while one simultaneously takes advantage of the benefits of different methods that compliment effective and efficient myocardial protection strategies. Ongoing studies continue to explore the incorporation of additional cardioprotective methods such as preconditioning agents, white blood cell filters, oxygen radical scavengers, endothelium-enhancing agents, and molecular factors that will further improve the safety of ischemic intervals and limit reperfusion damage.

The myocardial protection methods presented reflects the author's current approach with over 35 years of research and clinical experience. However, there is no one unique technique of myocardial protection. The individual must understand the basic science and principles that have evolved over the past several decades. The individual surgeon must have a consistent and adaptable approach. The techniques must be flexible to allow for intraoperative flexibility to deal with specific clinical situations. Team communication with the perfusionist, the operating room nurses, the surgical assistants, and the anesthesiologist is essential. However, even the best techniques of myocardial protection do not make up for a normally perfused heart. No matter what technique is used, time is of the essence. Time management and a coordinated cardioplegia approach that compliments and does not delay the operative flow must be the goal of the operating surgeon.

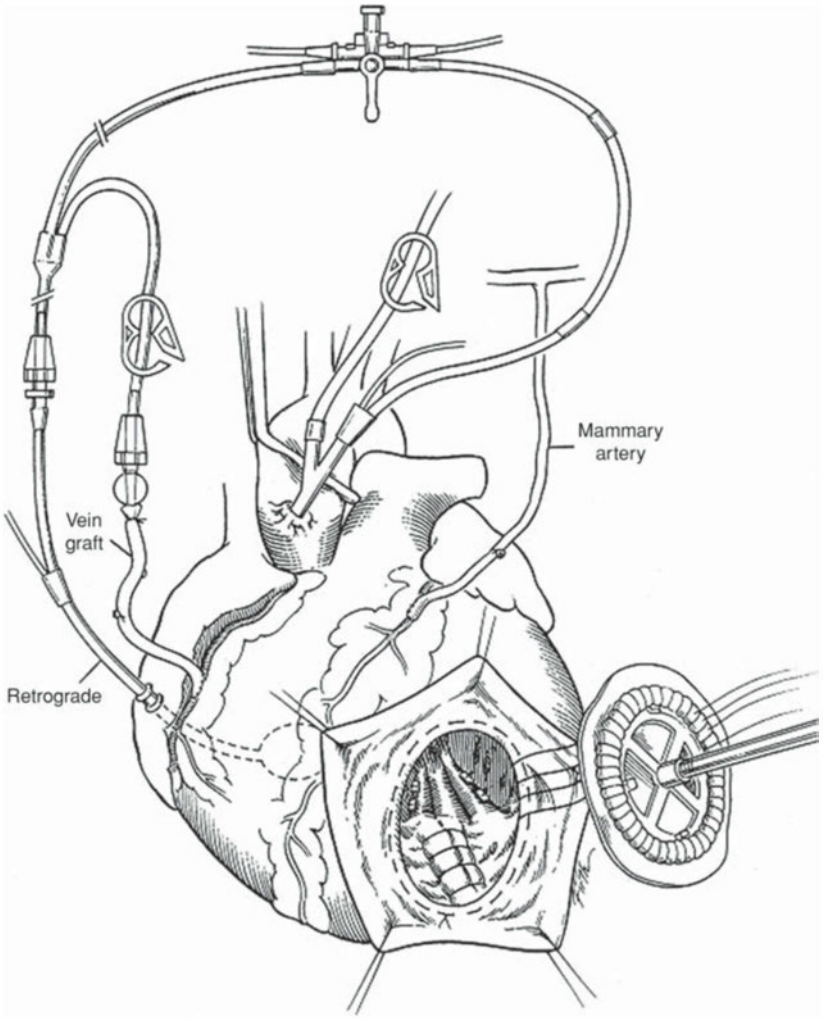


Fig. 37.8. Beating heart myocardial protection during ventricular respiration in a patient with aortic insufficiency. Note the clamped aorta, with perfusion via the internal mammary artery, vein grafts, and retrograde via the coronary sinus. This method is used with either anterior or inferior repairs.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Dr. Odeh and Dr. Shemin have done an outstanding job presenting a thorough approach to myocardial protection. I believe their descriptions are comprehensive and their overall approach will allow for very safe cardiac surgery. It is extraordinary how things have developed over the last 20 years. It was not unusual at one time that at least 10% of the patients would have required an intra-aortic balloon pump to come off bypass. This almost never occurs presently in elective or either urgent cases due to our myocardial protection strategies. I believe one should go by the book and have a careful orchestered approach to myocardial protection as the authors have described.

Our group has two slight differences from the techniques discussed here. The authors have discussed using antegrade and retrograde cardioplegic simultaneously. They mentioned that there are studies suggesting this could be done safely. I have at least theoretic concerns about this and we have avoided doing this at our institution. I do not think there is any issue with giving antegrade followed by retrograde.

The second issue relates to redo surgeries. The authors describe controlling the patent internal mammary artery graft to allow for better protection. I think this is okay but we have demonstrated that leaving an IMA open in patients requiring valve surgery is a nonissue. We have noted that there is less incidence of injury to the internal mammary artery using this technique, and we have found no issues with protecting the heart even with the IMA perfusing. In fact, it may provide better protection overall because of continuous blood being given to the left anterior descending distribution.

ILK

38 Database

Ashok N. Babu, A. Laurie Shroyer, and Frederick L. Grover

Quality improvement endeavors within the field of cardiac surgical care have made substantial progress over the last 40 years. A key aspect that has promoted improvement in the processes, structures, and outcomes in cardiac surgical care is the development of multi-institutional databases to monitor clinical risk-adjusted outcomes following cardiothoracic surgery. Quality improvement database reports offer several inherent advantages to a cardiothoracic surgeon in clinical practice. Using statistical prediction models, reports generated from complete and high-quality databases offer a unique assessment of the interactive effects of multiple risk factors on outcome.

The majority of these cardiac surgery database initiatives began by focusing reports upon relatively crude measures of unadjusted operative (30 days) mortality as their primary outcome criteria. Since the mid-1990s, there has been a progressive increase in the level of sophistication in the databases. For example, risk models can be constructed from a list of preoperative variables, and these models can be used to predict the likelihood of adverse outcomes. Whereas database outcomes initially focused on mortality, more recent efforts expand the focus for risk-adjusted outcomes to also include major morbidity as well as risk-adjusted efficiency outcomes such as length of stay and cost. Databases offer the opportunity for both feedback and information exchange; facilitating the comparison of institutions, providing data to local clinical care teams to self-assess performance, and coordinating future improvement strategies. This exchange offers, in turn, the potential to further improve the processes, structures, and outcomes of care in cardiac surgery.

HISTORICAL CONSIDERATIONS

Local databases have been used by many institutions for more than 40 years. For example, hospital-based databases have been used by Duke University, the Cleveland Clinic, the Mayo Clinic, and others to monitor volume and mortality statistics within their institutions. The first large, multi-institutional monitoring of cardiac surgery outcomes began in the Department of Veterans Affairs (VA) with the establishment of the Cardiac Surgery Consultants Committee (CSCC) in 1972. Under a congressional mandate, the VA semiannual reports were combined with ongoing surveillance work toward the goal of assuring the quality of cardiac surgical care in the VA system. Until 1988, the CSCC used unadjusted mortality and volume as the main parameters to judge outcomes. Takaro and colleagues analyzed the VA experience from 1975 to 1984 and noted that the annual volume of cardiopulmonary bypass surgery rose from 3,000 to more than 6,400 procedures. Correspondingly, the observed mortality rate for coronary artery bypass procedures dropped from 4.7% to 3.6%. They noted that patient-related comorbidities accounted for most of the operative mortality, but advances in surgical and medical techniques also may have played an important role in this mortality rate reduction.

On March 12, 1986, the Department of Health and Human Services Health Care Financing Administration (HCFA) released to the public raw mortality data for coronary artery bypass grafting (CABG). This report actually listed hospitals that had mortality rates for Medicare patients undergoing CABG that were in excess of predicted mortality rates. Although these data were to be used originally for state peer-review organizations as a quality improvement tool, HCFA was forced to release these data through the Freedom of Information Act. These mortality rates were unfortunately not risk adjusted for clinical patient risk factors. In addition, diagnostic categories were grouped together without comprehensive clinical patient-specific risk adjustments related to severity of disease and comorbidities. Furthermore, the public release of these data prompted widespread concern that nonrisk-adjusted mortality could not offer an accurate reflection of the caseload mix, and relative outcomes within an individual program. Without appropriate patient-specific clinical risk stratification, accurate comparison of cardiac surgery outcomes between programs was widely perceived not to be useful for patient care, program management, or policy decisions. This action by HCFA served as a strong stimulus for the Society of Thoracic Surgeons (STS) to initiate its own cardiac surgical database with risk-adjusted methods.

In 1987, the VA Cardiac Surgery Advisory Group, then called the VA Cardiac Surgery Consultants Committee, was also concerned with the use of unadjusted operative mortality and volume as major quality indicators for the approximately 45 VA cardiac surgery programs. Using the logistic regression method approach developed by the Collaborative Study in Coronary Artery Surgery, Drs. Hammermeister and Grover developed a multivariable risk model using clinical and angiographic predictors of 30-day operative mortality separately for coronary artery bypass procedures and valve plus other operations. The method included capturing data on all cardiac surgery procedures performed using a single-page data sheet (originally of 52 data elements) including patient-specific clinical risk factors, cardiac catheterization assessment data, operative details, and a set of both mortality and major morbidity outcome variables. These initial efforts proved successful in the attempt to "level the playing field" by accounting for the individual patient risk factors that could play a role in determining outcomes after cardiac surgical procedures (according to the first VA report). Based on these early events, two large national databases, the STS National Adult Cardiac Surgery Database and the VA Cardiac Surgery Database, emerged in a complementary and synergistic manner to provide routine reports of cardiac surgical practices for their use in their own local quality improvement endeavors. Similarly,

large regional databases also evolved, including (but not limited to) the Northern New England Cardiovascular Study Group (NNE), the New York State databases, and more recently state databases in California, Massachusetts, and Pennsylvania.

DATABASE CONSTRUCTION

The factors that may influence the differences in outcomes include variations in patient risk factors, in the processes and/or structures for the clinical care provided, as well as random chance. Conceptually, there are different categories of patient risk characteristics that may influence cardiac surgical outcomes including patient demographic factors, socioeconomic factors, severity of cardiac disease, comorbidities, and patient lifestyle/health behaviors. As a general rule, patient risk factors should be assessed as close to the starting time frame for the cardiac surgical procedure as possible to most accurately and reliably evaluate the patient's inherent risk. Using statistical modeling approaches, risk stratification for a given procedure holds constant the patient-specific characteristics that may influence the likelihood of specific patient outcomes. Risk-adjusted outcomes provide indirect measures to begin exploring the potential differences for quality of care.

Virtually all of the national, regional, and local cardiac surgical databases provide predictive risk stratification—that is, an estimation for a patient with a prespecified risk profile what is the risk of having an adverse cardiac surgical event occur. Surgeons must, however, be aware of the limitations and factors pertaining to the design, construction, and management of a particular database if it is to be optimally utilized. Of primary importance are the completeness, accuracy, reliability, timeliness, sensitivity to change, and integrity of the data entered. "Garbage in = garbage out" is operative in considering any analysis of a database. Ideally, all patients receiving a cardiac surgical procedure should be entered for analysis and the adverse outcomes (such as 30-day operative mortality) appropriately validated. For any given patient record, the data submitted must also be internally consistent. For example, patients entered into the database as needing emergent operations should have firm clinical data supporting the need for such a classification. Both intrafield and interfield edit checks may be used to evaluate data quality discrepancies. Finally, standardized definitions (with training for the data capture team) are important for all data elements. Common inconsistencies in application of definitions have been demonstrated for patient-related outcomes, such as perioperative morbidities, where the use of different diagnostic test assessments is not consistently performed across all settings. Surgeons should be aware of the important aspects of the management/operation of any given database, including the following:

1. Which outcomes are examined, and are definitions for the measured variables standardized? Mortality, for example, is the most widely used outcome because of its importance and it is readily available from hospital records. Such a dichotomous variable would seem relatively standard. However, some databases measure in-hospital mortality, whereas others track 30-day mortality after CABG (including both in-hospital deaths and deaths that occur within 30 days postsurgery). Other important outcome variables to monitor commonly include the occurrence of major complications (infection, stroke, prolonged ventilation, renal failure, etc.), cost, and length of hospital stay. These are more difficult to measure because complications may not be uniformly recorded across sites. Standardized definitions for these major morbid events are rigorously defined by both the STS and VA cardiac databases.
2. What is the intent of the data analysis? The database may be designed to provide a predicted outcome for a given patient or to compare surgeon-specific or program-specific outcomes. In addition, outcomes analysis may be used to drive national policy decisions or managed care accreditation. Discussions have been initiated also about using risk-adjusted outcomes for physician profiling and credentialing.
3. How are the data collected? Input for databases may be gathered in a retrospective, concurrent, or prospective manner. Depending on the database, data acquisition may be mandatory (VA database) or voluntary (STS database). Data acquisition and entry may be collected by members of the patient care team or by independent data collectors. For example, in the VA database, data are collected by nurse investigators who are independent of the surgical team. It is believed that independent collection of such data may result in more thorough and unbiased reporting (e.g., self-reporting for perioperative complications may not be consistently assessed and/or uniformly recorded across surgeons).
4. How are the missing data treated? Any large database will have incomplete data elements. Missing data happens more commonly in databases where the data are gathered and/or entered retrospectively. If the outcome of interest is missing, then the records with missing outcome data must be discarded because no reasonable method exists that is well accepted to impute outcomes. For all other nonoutcome variables, a variety of techniques may be used to estimate the missing data. Occasionally, there are enough missing data elements within any specific record to justify dropping this specific case. Common imputation techniques derive a value to be inserted for the missing variable based on values for other data fields for that same patient, in context of the data gathered for other patients. As the completeness rates for cardiac surgery databases have increased, however, the need for complex imputation techniques has begun to diminish.
5. How did the predictive model perform that was generated from the database? Construction of a predictive model typically involves several steps. First, a conceptual model is generated that identifies the clinically relevant variables (e.g., risk factors) that are thought to be associated with a particular outcome (e.g., mortality). In context of the previous literature published within the field, these clinical models are constructed by a panel of experts. After these risk factors have been identified, rigorous definitions must be used to standardize the data collection. After all database completeness, accuracy, and reliability verifications (including any required data record updates), both univariate and multivariate analyses may be performed for a procedure, for a selected population of patients, and for prespecified outcomes to determine which of these risk factors are statistically associated with adverse patient outcomes. As part of the univariate screening, the outcome rates for patients with a given risk factor are compared with the outcome rates for patients without the risk factor. The number of patients required for such an analysis varies based on the nature and frequency of the outcome studied, the number of patient risk variables planned for evaluation, and the statistical approach planned. Based on the rule that in logistic regression analysis, there should be no more than 1 risk factor assessed per 10 to 15 adverse

events, if ~10 risk variables are used, >5,000 cases are required to develop a logistic regression model for CABG-only procedure, assuming an average mortality rate of <3%.

After univariate screening analyses have been performed, a multivariable predictive model is next generated. In the majority of the cardiac surgical literature, the endpoints analyzed for risk models have included risk-adjusted mortality or presence/absence of major perioperative morbidity. For any dichotomous modeling endpoint, the analytical technique originally used by the STS to generate a predictive equation was initially based on Bayesian theory to address the inherent challenges in a voluntary database related to missing data. However, the STS switched to multivariate logistic regression in 1997 when the data record completeness and quality improved such that the performance of the logistic regression approach was determined to be more robust. For dichotomous endpoints (e.g., 30-day operative mortality), the Northern New England Cardiovascular Consortium, VA, New York State, California State, and Massachusetts State databases have also used multivariate logistic regression as the most common risk modeling approach.

Multivariate logistic regression is a statistical technique that can be used for either explanation (to understand associations between risk factors and outcomes) or prediction (to estimate risk for new and different populations). To apply the logistic regression equation for prediction purposes, risk factors are normally entered into the mathematical equation for a given patient. Mathematically, application of the logistic regression equation to a specific patient's data captured can yield the statistical probability for the adverse outcome of interest.

Before being actively used for quality assessment activities, the performance for any statistical modeling approach(es) employed should be thoroughly evaluated. For logistic regression models, the "predictive power" (or *c*-index) of the model describes the concordance of the outcome predictions in comparison to the actual outcomes observed. The *c*-index describes the area under the receiver-operating characteristic curve related to the true-positive rate (sensitivity) and the false-positive rate (1—specificity). For example, a *c*-statistic of 0.5 would indicate that the risk model has no better chance than flipping a coin of identifying "at risk" patients for adverse outcome events. Ideally, a predictive risk model will have a *c*-index of >0.70, meaning that on average 70% of the time that the outcome predicted by the model is accurate, leaving the remaining 30% subject to the unmeasured risk factors, the nature of the processes and structures of care rendered, as well as random chance.

Statistical risk models are best for predictions performed on large volumes of patient records, where the confidence intervals around the predictions may be relatively small, as compared with predictions made for a single patient where the confidence interval around the point estimate for risk prediction may be large. Given that it is virtually impossible for a single risk model to incorporate comprehensively all of the risk factors that are relevant to all patients, there commonly may be unmeasured risk variables (specifically, rare patient risk factors) that may predispose a given patient to an adverse event. The final predictive variables included in a logistic regression model, therefore, are generally those factors that have the largest impact and occur reasonably frequently. For example, a patient may have had chest radiation in the past, which can increase their risk of adverse perioperative outcomes. As chest radiation occurs infrequently, however, this risk factor may not be commonly placed upon the list of prediction variables that determine estimated operative morbidity and mortality. For this reason, statistical risk models may either underpredict or overpredict the risk for an individual patient, so that physicians need to take this into account when diagnosing unusual patient risk factors when deciding on appropriate therapy and discussing this with a given patient.

For logistic regression models, another key performance metric to be evaluated is the model's calibration. Model calibration describes how well the model performs over the full range of risk estimates, as compared with the observed outcome rates. Models may work well for high-risk or for low-risk situations but not uniformly work well across all ranges of risk. A common metric used to assess logistic regression model performance related to calibration is the Hosmer–Lemeshow [H-L] test for "goodness of fit." The H-L test is adapted from the Chi-square test, evaluating to what degree there is "no difference" in the predicted versus observed outcome rates across ordered risk categories (such as ordered deciles of risk). To be able to state that there is not a calibration challenge, therefore, the H-L test statistic's *P*-value should be ideally >0.05—stating that there was no difference in calibration across the risk categories that were observed.

In addition to multivariable logistic regression for single endpoints, more complex regression approaches for multiple endpoints have recently been implemented by the STS. One notable example is the development of a comprehensive composite metric that assesses multiple aspects of quality rather than focusing only on risk-adjusted mortality. This acknowledges the fact that not all patients who survive CABG surgery have had equivalent quality procedures. The STS Database team and its Quality Measurement Task Force under the guidance of Dr. Shahian has developed a meaningful, NQF endorsed CABG Composite Score to assess the overall quality of care rendered. The results of this methodology are presented as both numerical scores and, for ease of consumer interpretation, as a "star" rating system. In the latter, programs whose results are estimated to have at least a 99% Bayesian probability of being superior to the STS average are awarded three stars (about 10% to 15% of all STS participant programs), whereas those with a similar probability of being worse than the STS average (again, about 10% to 15%) are awarded one star. The remaining or average quality programs are classified as two stars.

To calculate the STS CABG Composite Score for each hospital, the following four metrics were combined:

1. freedom from 30-day operative mortality (both any in-hospital death regardless of timing and any death within 30 days regardless of venue; risk-adjusted);
2. freedom from major morbidity (defined as the risk-adjusted occurrence of any one or more of the following endpoints: renal failure, reoperation, stroke, sternal wound infection, prolonged ventilation);
3. use of at least one internal thoracic arterial bypass graft conduit (considered as a "best surgical practice"); and
4. use of all recommended perioperative medications (preoperative beta blockade; discharge antiplatelet drugs, lipid-lowering agents, and beta blockade) (an "all-or-none" endpoint; considered as a "best medical practice").

The above metrics were analyzed together in a hierarchical model in order to estimate success rates for each of the four metrics by a participant. A participant's final composite score was obtained by averaging their four success rates. Albeit more complex analytically, this innovative

quality metric is multidimensional (evaluating simultaneously mortality, morbidity, and reflects compliance with what is considered the optimal CABG surgical and ischemic heart disease medical therapy practice) and fully risk adjusted. This intellectually elegant and clinically relevant composite metric provides an assessment of the quality metrics that hospitals can control (specifically their hospital's compliance with best practices) while adjusting for the factors that hospitals cannot control (specifically, the risk characteristics of their patient population).

CURRENT ADULT CARDIAC SURGERY DATABASES

New York State Database

In response to the HCFA release of cardiac surgery data and as much as fivefold variations in mortality between hospitals, New York State organized a Cardiac Surgery Advisory Committee, which was charged with investigating issues related to quality assurance for cardiovascular disease. A clinical registry for cardiac surgery began collecting data in 1989 and one for percutaneous coronary intervention (PCI) began in 1992. This group developed a patient-specific cardiac surgical report form. This form reported demographic data, surgical procedures, admission and discharge dates, and information on risk factors, complications, and discharge status. Data forms are completed at each surgical program and forwarded to the Department of Health for analysis. Data reporting is mandatory.

The first report on these data was published in 1990 and described hospital mortality rates for 28 hospitals, 4 of which had significantly higher than expected mortality rates. The overall statewide mortality rate for cardiac surgery was 4.87%. Identifying these poor performing centers allowed site visits to occur to identify the quality-of-care issues. For example, Winthrop Hospital in New York had one of the highest risk-adjusted mortalities with the initial data. They were placed on probation that allowed them to hire a new chief of cardiac surgery, centralize all patients in one area, hire nurse specialists and physician assistants dedicated to cardiac surgery, review all cases preoperatively, and hire a dedicated team of cardiac anesthesiologists. These interventions allowed risk-adjusted mortality to fall from 9.2% to 4.6% to 2.3% between 1989 and 1991.

The surgeon-specific outcomes reporting has also had an effect on the cardiac surgery climate in New York state. A more recent study by Jha and Epstein covering the years of 1989 to 1999 reported that 20% of the surgeons in the lowest performing quartile abandoned cardiac surgery or relocated within 2 years of the report, whereas only 5% of surgeons in the top performing quartile did so. Interestingly, in the same study, the authors found that a higher risk-adjusted hospital mortality did not affect that hospital's market share negatively.

A study comparing PCI to CABG in multivessel CAD was published in the New England Journal of Medicine in 2008. They compared the 18-month outcomes of patients meeting their inclusion criteria from 2003 to 2004. They demonstrated a statistically significant lower mortality and rate of repeat revascularization in the CABG group at 18 months.

Some studies have also been performed to answer the question of whether high-risk cases were turned away due to fear of excess mortality. A study by Omoigui et al. demonstrated an increase from 61.4 patients per year to 96.2 patients per year being treated at the Cleveland Clinic in the 10-year period before and after 1989. These patients transferred in the later decade were also sicker on average than those treated in New York State. Another study compared the preprocedural severity of illness of patients undergoing PCI in New York versus Michigan. They found lower rates of acute MI, cardiogenic shock, and congestive heart failure. As one would expect, the overall hospital mortality was much lower in New York, and risk-adjusted mortality was not significantly different. Both studies have their weaknesses and this remains a controversial and unresolved issue.

Despite these types of criticisms, the overall mortality in New York State continues to decrease. The New York State Database remains active and continues to identify significant independent risk factors for cardiac surgery and avenues to improve outcomes.

Northern New England Cardiovascular Disease Study Group

The Northern New England Cardiovascular Disease Study Group was organized in 1987 to help improve the quality, safety, and cost of cardiovascular interventions in the region. This group comprises representatives from 11 institutions performing cardiac surgery in Maine, New Hampshire, and Vermont. This group of institutions voluntarily reports outcomes on patients undergoing coronary bypass operations, valve operations, and PCI. At last report, more than 190,000 patients had been registered with this database. This database was originally founded as a regional prospective study to examine variations in outcome following CAB in this region. The initial report from this database in 1991 evaluated 3,055 patients undergoing isolated CABG operations. This study noted that crude, unadjusted mortality rates provided insufficient data to gauge performance. Although the study documented a range for in-hospital mortality from 3.1% to 6.3%, these regional data also suggested that differences observed in in-hospital mortality rate could not be explained solely by patient risk factors. Rather, the data suggested that differences in isolated CAB outcomes may have resulted from unmeasured differences in care.

This group then explored a regional strategy to learn "best practices" from each other. This strategy employed four components: regular feedback of outcomes data, efforts to determine causes of mortality, structured round-robin visits between organizations, and determinations of cause-specific mortality. This innovative sharing of information resulted in a subsequent reduction in the CAB mortality rate for the region by 24% over an 18-month period despite a higher risk population being operated on during this period.

In 2000, the group published data demonstrating 44% relative risk reduction over a 5-year period from 1992 to 1997 for aortic valve replacement and a 53% relative risk reduction for mitral valve replacement, potentially associated with the group's quality improvement efforts. A randomized trial comparing neurocognitive outcomes from on-pump versus off-pump coronary bypass was published in 2007 demonstrating a lack of difference in this outcome. This group has over 100 peer-reviewed publications from the database covering a myriad of topics focused on risk factors and quality improvement. The NNE cardiovascular group continues to collect data and explore and try to alter risk factors for negative outcomes in cardiac surgery as well as PCI.

Michigan Regional Collaborative Improvement Programs

As we have learned from the New York state database review, public reporting of hospital and surgeon outcomes paradoxically does not negatively affect market share of poor performing programs. Taking that into consideration, regional collaborative programs like the one in Northern New England may be able to identify problem

areas and correct them to improve quality. The Michigan Society of Thoracic and Cardiovascular Surgeons (MSTCVS) began analyzing STS data for half the cardiac surgery programs in Michigan to implement a quality initiative in the late 1990s. Subsequently, Blue Cross/Blue Shield of Michigan (BC/BS), which covers 47% of the state's population, began funding nine regional collaborative quality improvement programs in the mid-2000s, including a cardiac surgery program in 2006. The cardiac surgery collaborative was based on the pre-existing efforts of the MSTCVS. This program was to include all cardiac surgery programs in the state and BC/BS would provide funding for personnel and data collection. BC/BS also funded required quarterly meetings to review data. The program uses STS data registry with some additional variables and provides hospital and physician-specific outcomes data and statewide and national benchmark data to aid in local quality improvement. In the first 3 years following inception, this collaborative was able to identify significant variance relative to internal mammary artery use and prolonged ventilation and significantly improved both variables at outlier centers. The Michigan program is yet another example of a successful regional quality improvement program. What makes it unique is its private payer funding and collaboration that results in net benefit to the patients, hospitals, surgeons, and payer in the form of reduced costs.

Virginia Cardiac Surgery Quality Initiative

The Virginia Cardiac Surgery Quality Initiative (VCSQI) was formed in 1996 as a voluntary consortium of cardiac surgery programs and now includes 17 hospitals and 13 surgical groups that perform all of the cardiac surgeries in Virginia except for the Veterans Administration. The mission of this group is to improve clinical performance among all cardiac surgery programs by using outcomes analysis and process improvement. The database includes STS registry data (all hospitals report to the STS) and also links these cases to the costs as reported by the hospital. The meetings include physicians, nurses, administrators, data managers, and others to create a multidisciplinary atmosphere. As with other local collaborative, VCSQI has sought to identify outlying outcomes and review them to make improvement. Additionally, they have implemented several best practice initiatives based on their own data to reduce perioperative transfusion, atrial fibrillation, prolonged ventilation, and hyperglycemia. The unique combination of outcomes data with cost data has allowed VCSQI to estimate costs associated with complications and hence to estimate cost reduction by quality improvement. This has led to their pioneering efforts in pay for performance models and quality-based payments.

California Cardiac Surgery and Intervention Project

California first entered into public reporting in 1996 with the California CABG Mortality Reporting Program. Reporting was voluntary and many hospitals, perhaps preferentially those with suboptimal results, did not report data. Public reporting in California began from this but the data were too limited to make solid conclusions from. A law supported by the California Society of Thoracic Surgeons (CASTS) was passed in 2001 by the California State Senate to require public reporting of all surgical outcomes, beginning with CABG in 2003. The project was begun by the CASTS and the California Cardiac Surgery and Intervention Project (CCSIP) was formed to help enhance the database with a focus on quality improvement. The state uses STS data elements, with slight modifications and definitions, and analyzes the data and issues surgeon-specific risk updated mortality reports for CABG publically.

The first paper in 2005 compared California's outcomes in CABG and PCI with the well-established New York database. Surprisingly, CABG mortality was 33% higher in California and PCI mortality was double that of New York. However, programs performing >300 CABG per year had equivalent mortality to New York (2.45%). Excess mortality only occurred at the lower volume programs, and this relationship did not change with risk adjustment. The PCI mortality had no volume–outcome relationship.

A follow-up study was published in 2009 after several years of public reporting and CCSIP intervention. Total CABG volume decreased from 28,495 cases in 1997 to 15,520 cases in 2006. On the other hand, PCI volume increased from 38,098 to 53,703 in the same period. Even with the decrease in CABG volume, risk-adjusted mortality decreased from 4.7% to 2.1% for CABG and from 3.4% to 1.9% for PCI. Interestingly, combined rates of death, MI, or repeat revascularization were double in the PCI group. Additionally, the volume–outcome relationship was no longer statistically significant, though the trend still existed. This suggests that quality improvement initiatives did alleviate the quality issues at some low volume centers. This is yet another example of successful quality intervention.

Massachusetts

As in many other states, Massachusetts cardiac surgeons and legislators began thinking about establishing a cardiac surgery database along with public reporting in the late 1990s. Massachusetts was somewhat unique in that there were only 11 cardiac surgery programs due to strict "determination of need" certification on the part of the state. However, community hospitals that had banded together strongly requested to be allowed to perform cardiac surgery, and it was this debate that accelerated the need for outcomes measurement and quality control. The cardiothoracic surgeons of Massachusetts succeeded in having the STS national cardiac database used as the acquisition tool for the data. The data are reported both to the STS NCD as well as Massachusetts Data Analysis Center (MDAC) to allow for state-specific report generation. It was decided that hospital-level public reporting would occur, whereas surgeon-specific data would be protected and analyzed privately.

Data collection began January 1, 2002 and the first report card was published October 19, 2004 demonstrating no significant differences among cardiac surgery programs in Massachusetts. In the subsequent years, this database has spawned multiple clinical research projects and resulting publications identifying effectiveness of specific interventions and risk factors for poor outcomes.

Veterans Administration Database

The Cardiac Surgery Consultants Committee was organized under a congressional mandate to review the surgical results of the 43 VA medical centers performing cardiac surgery beginning in 1972. In 1987, the committee acknowledged the limitations of using only raw unadjusted mortality data to determine whether the appropriate level of surgical care was being delivered. This group sought and received authorization and funding from the VA to incorporate patient risk factors for the assessment of cardiac surgical results. Dr. Karl Hammermeister, one of the members of the committee, had experience with risk adjustment for cardiac surgery through his involvement with the Coronary Artery Surgery Study (CASS). He, therefore, took this knowledge

to the VA to help develop the risk-adjusted methodology for measuring outcomes in cardiac surgery for the purpose of quality improvement. He worked with Dr. Grover to initiate this in the Veterans Administration (VA) Cardiac Surgery Database, the first national risk-adjusted cardiac surgery database. A similar initiative was undertaken 2 years later by Dr. Shukri Khuri in the area of general surgery and other surgical subspecialties with the development of the VA National Surgery Quality Improvement Program (NSQIP).

The initial report of the VA's National Risk Adjusted Cardiac Database occurred at the STS meeting in 1989. The VA Cardiac Surgery Database vastly improved in 1991, when 44 nurse data managers were assigned to the hospitals performing cardiac surgery to collect data for both cardiac and general surgery programs. This allowed for virtually 99.9% complete capture of data points, a crucial factor in having valid data. Data entry from each surgical program is mandatory. The data were distributed back to the local hospitals, including the cardiac surgical team, cardiologists, and hospital administrators on a 6-monthly basis, with the observed and expected mortality rates for each of hospital blinded expected except for one's own hospital. Over a 20-year period, there has been a significant reduction in risk-adjusted mortality for VA cardiac surgery (Fig. 38.1). The expected mortality rate for each cardiac surgery program is computed by calculating the expected probability of the 30-day mortality for each patient and then averaging the probabilities for all patients having cardiac surgery at the given hospital. A ratio of the observed mortality rate to the expected mortality rate is then calculated. This analysis also includes major morbidities that are also risk adjusted.

This information is now reviewed by the VA Surgical Quality Improvement Program (VASQIP) Executive Committee and the Cardiothoracic Surgery Advisory Board on a quarterly basis. VA cardiac surgery is one of numerous surgical subspecialties that are monitored by the VA and the VASQIP Executive Committee and each subspecialty has their own Surgery Advisory Board (SAB). It is noteworthy that 59 publications have occurred from the cardiac surgical database since its inception. One of the most important studies was the prospective randomized ROOBY Trial, which demonstrated better 1 year outcomes with on-pump coronary bypass as compared with off-pump.

Within the VA system, the risk-adjusted surgical results are used for two purposes—to prompt oversight review by the committees and the individual cardiac surgery programs and feedback to the individual cardiac surgery programs to provide comparison data with the other VA cardiac surgical programs to stimulate their attention to quality improvement.

History of the Development of the Society of Thoracic Surgeons National Database

The STS began initial meetings to develop databases in 1988, including adult cardiac surgery, congenital heart surgery, and general thoracic surgery. The initial emphasis was in the adult cardiac surgery area because of the public release of hospital CABG data in 1986 by HCFA (Healthcare Financing Administration, now CMS) and subsequently in New York State by the New York State Cardiac Surgery Database. By 1994, over 1,500 surgeons operating in 706 hospitals in 49 states had entered more than 500,000 patients into the database with a decrease in coronary bypass operative mortality from 3.7% to 3.1%. In 1996, efforts were made to expand the goals of the STS database, leading to the hiring of the first national database coordinator, Mary Eiken. In 1999, a contract with Duke Clinical Research Institute (DCRI) was signed for warehousing and analysis of the data and multiple software vendors were contracted after approval for data entry and transmission of data to DCRI. This marked a new era for the STS Databases and the STS

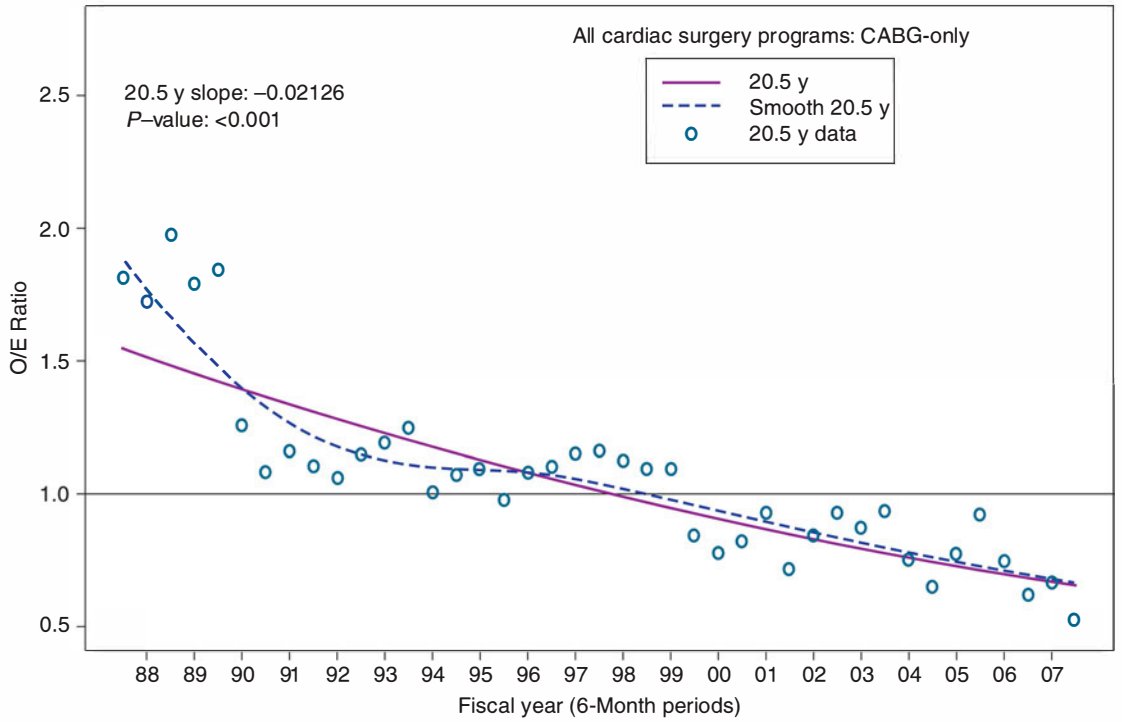


Fig. 38.1. This graph demonstrates a very significant decrease in risk-adjusted operative mortality following coronary artery bypass grafting over a 20-year period in the National VA Cardiac Surgery Programs. (Shroyer ALW, et al. Improving quality of care in cardiac surgery: evaluating risk factors, processes of care, structures. Semin Cardiothorac Vasc Anesth 2008;12:140.)

Adult Cardiac Database progressed in its maturity and the sophistication of its analysis and research, and much progress was made in the development of more mature general thoracic and congenital cardiac databases. In 2004, Cynthia Shewan was recruited to be the Staff Director of Quality Research and Patient Safety. **The STS Databases are now the largest registries of clinical cardiothoracic surgery data in existence**. The STS Database is now chaired by Dr. David Shahian and it has taken on many functions. However, the highest priority remains quality improvement in the care of cardiothoracic surgical patients.

Society of Thoracic Surgeons Adult Cardiac Surgery Database

The STS Adult Cardiac Surgery Database now has 1,067 groups participating, which is well over 90% penetration of all cardiac surgical programs in the United States. It contains over 4.8 million records including 1.8 million isolated CABG records from 2000 to 2011. Data auditing was put in place in 2009 and an increasing number of sites are audited each year, forty in the past year with 96% overall agreement. Performance measures have been endorsed by the National Quality Forum and the AQA are used by many hospital systems and payor systems in their quality programs. The data are now distributed back to cardiac surgery programs, their hospitals, and cardiologists on a quarterly basis using a three-star composite score, with three stars outstanding, two stars satisfactory, and one star needing improvement (Fig. 38.2). The details of this were discussed earlier in the chapter.

Over the past two decades, as in the VA program, there have been significant reductions in risk-adjusted operative mortality for cardiac surgery patients (approximately 60%) as demonstrated by the reduction of coronary bypass grafting operative mortality in the decades of the 1990s and the first decade of the 21st century (Fig. 38.3 A, and 38.3 B). The coronary bypass data are now being publically reported through the Consumers Union Website as well as the STS' Web site. Currently, approximately 40% of groups in the United States are voluntarily reporting their data publically and this percentage continues to modestly increase each year. Risk models are updated every several years and new ones are created with the adult cardiac surgery database with risk models for CABG only, aortic and mitral valves only, and CABG/valve procedures. Plans are also underway for publically reporting outcomes for other adult cardiac procedures.

The Society of Thoracic Surgeons General Thoracic Surgery Database

The General Thoracic Surgery Database now receives data from over 229 groups (785 surgeons) participating with 312,000 operations entered since 2002. The data are harvested twice a year, with the spring 2012 harvest being the fourteenth. Data are distributed back to the surgeons and their local hospitals. The first risk models have been completed for prolonged length of stay for lobectomy and for morbidity and mortality for esophagectomy. Audits were initiated in 2010 and 10 sites were audited in 2011 and will increase to 10% of sites in 2012. Seven general thoracic measures have been endorsed by the National Quality Forum. Eight publications have been published from this database since 2008.

The Society of Thoracic Surgeons Congenital Cardiac Database

There are currently 103 participants using the STS Congenital Cardiac Surgery Database, from an estimated 125 U.S. pediatric cardiac surgery programs, with 213,416 total operations in the database. The STS-EACTS scores are utilized for risk adjustment. Audits began in 2007 with five occurring in 2011 and 10% of sites for 2013. The 20th harvest took place in the spring of 2012. Eighteen publications have come from this database since 1995, nine in the last 3 years, including a paper analyzing the complex relationships between surgical case volumes and mortality (Fig. 38.3B).

RECENT ADVANCES AND PROJECTS

There has been major activity in linking the STS Database with other databases, both administrative and clinical, including CMS, the American College of Cardiology (ACC) interventional database, ACC pediatric cardiac database, the National Death Index, the Society Security Death Index, the Pediatric Cardiac Intensive Care Society database, the Extracorporeal Life Support Organization registry, hospital billing systems, and private payor systems. Through these linkages, it is possible to follow long-term outcomes as well as to obtain cost data, which can then be used to evaluate the early and late value of the procedures that should be useful for value-based purchasing of health care, which is currently being initiated. Research using the STS databases has resulted in 121 peer-reviewed publications.

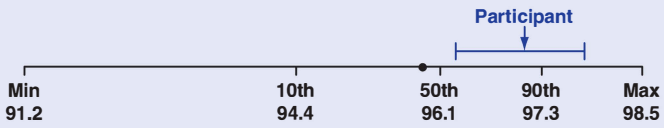
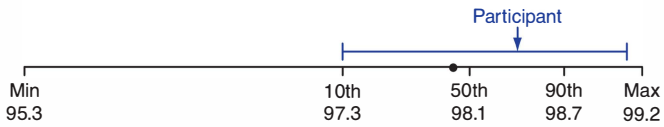
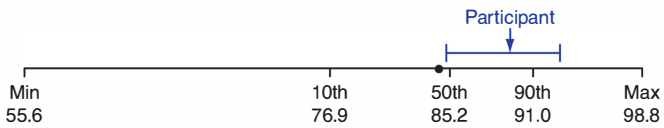
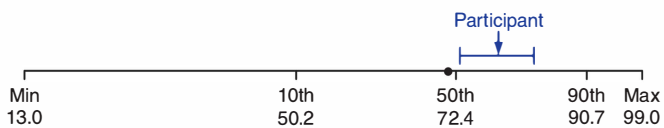
Because of the increasing interest and activity in research, an STS Research Center was established in 2012, with Fred Edwards serving as the Director of the Center and Cynthia Shewan as the Director of Research and Scientific Affairs. DeLaine Schmitz was hired at this time as Director of Quality.

The STS also has received numerous grants from external funding sources utilizing the databases. The most prominent grant thus far is the NIH Go Grant as part of the President's Stimulus Program, which funded the STS and the ACC and DCRI to perform a comparative effectiveness study of PCI versus CABG linking the STS and ACC databases with the CMS administrative database (ASCERT Trial). This study was published recently in the New England Journal of Medicine and there are several sub-studies of that data now underway.

This study demonstrated that during the first year, there was a lower risk of mortality in the PCI group, but after 1 year there was progressively favorable survival with coronary bypass as compared with PCI for two- and three-vessel coronary artery diseases (Fig. 38.4). The study like any other retrospective observational study may have unknown and/or uncaptured confounding variables but has demonstrated results similar to the New York State Database observational studies and the prospective randomized Syntax Trial.

HEALTH POLICY AND COLLABORATION WITH THE GOVERNMENT AND OTHER ENTITIES

Because of the high penetration of the STS Databases in practicing programs and their reputation for accuracy and sound scientific methodology, these databases have been very effective in building strong relationships between the cardiothoracic surgical community, health policy groups, and the government including the legislative and executive branches, and regulatory agencies. Recently, a landmark decision has been made by the FDA and CMS to invite the STS and the American College of Cardiology to be involved with the criteria and measurement for the efficacy and safety of transcatheter aortic valve replacements. Working with FDA and CMS, criteria were developed to determine which patients are initially eligible for these valves, what the program standards are for both cardiology and cardiac surgery, and very importantly, establishing that data will be captured on all patients receiving these valves in

Quality domain	Participant score (98% CI)	STS mean participant score	Participant rating[1]	Distribution of participant scores • = STS mean
Jan 2008–Dec 2008 Overall	**97.1% (96.3, 97.8)**	**95.9%**	★★★	
Jan 2008–Dec 2008 Avoidance of Mortality	98.1% (97.3, 99.1)	98.0%	★★	
Jan 2008–Dec 2008 Avoidance of Morbidity[2]	89.4% (84.9, 92.9)	84.4%	★★★	
Jan 2008–Dec 2008 Use of IMA[3]	96.1% (93.2, 98.0)	93.9%	★★	
Jan 2008–Dec 2008 Medications[4]	78.5% (73.1, 83.5)	71.3%	★★★	

[1]*, Participant performance is significantly lower than the STS mean based on 99% Bayesian probability; **, participant performance is not significantly different than the STS mean based on 99% Bayesian probability; ***, participant performance is significantly higher than the STS mean based on 99% Bayesian probability.
[2]Includes reoperations, renal failure, deep sternal wound infection, prolonged ventilation, and CVA.
[3]Excludes patients with prior CABG surgery.
[4]Includes preoperative beta blockade, discharge beta blockade, discharge anti-lipids, and discharge anti-platelets. Excludes v2.61 contraindicated/not indicated records.

Fig. 38.2. This is an example of the Society of Thoracic Surgeons reports that individual cardiac surgical groups receive on their performance and includes the three-star rating with three stars being above average, two stars average, and one star below average.

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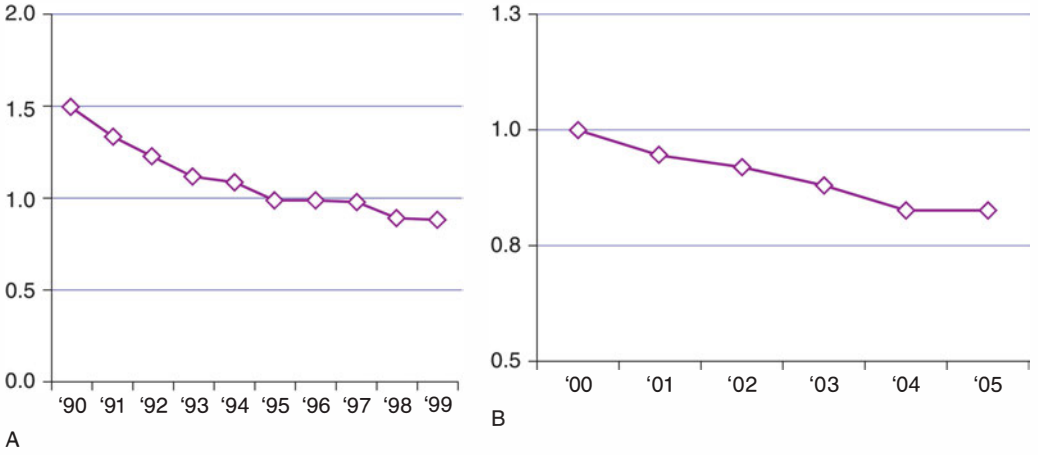
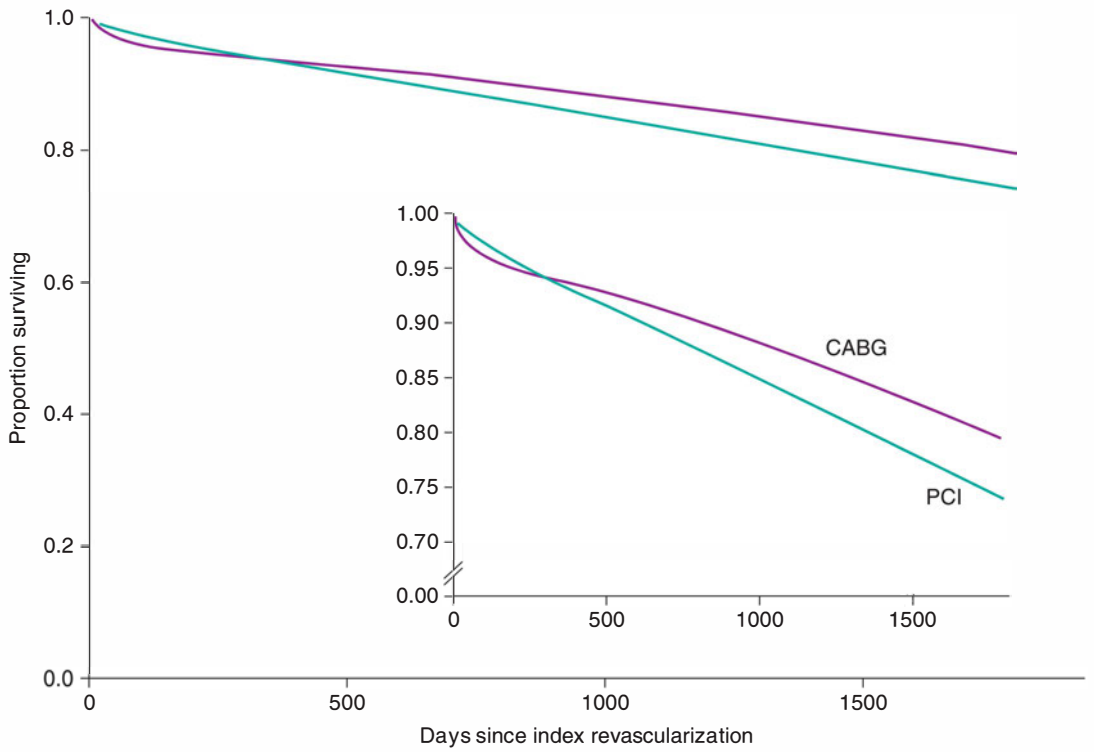


Fig. 38.3. These Society of Thoracic Surgeons graphs demonstrate an approximately 60% reduction in the observed to expected mortality rates for coronary artery bypass patients for the decades of the 1990s (**A**) and for the first 5 years of the 21st century (<0.0001) (**B**). (From: Grover FL. The bright future of cardiothoracic surgery in the era of changing health care delivery: an update. Ann Th orac Surg 2008;85:8-24, doi:10.1016/J.ATHORACSUR.2007.10.100, with permission.)



	30 and D	1 and Y	2 and Y	3 and Y	4 and Y
Mortality after CABG, % (95% CI)	2.25 (2.09–2.41)	6.24 (5.97–6.50)	8.98 (8.68–9.29)	12.4 (12.0–12.8)	16.4 (15.9–16.9)
Mortality after PCI, % (95% CI)	1.31 (1.21–1.41)	6.55 (6.35–6.76)	11.3 (11.0–11.6)	15.9 (15.6–16.3)	20.8 (20.4–21.2)
Relative risk with CABG (95% CI)	1.72 (1.52–1.89)	0.95 (0.90–1.00)	0.79 (0.76–0.83)	0.78 (0.75–0.81)	0.79 (0.76–0.82)

Fig. 38.4. This is a graph of early and late mortality with inverse probability weighting for patients receiving percutaneous coronary intervention versus coronary artery bypass grafting for two- and three-vessel coronary artery disease. You will note that after the first year, survival is progressively greater with the coronary bypass group with a 21% less relative risk of death. CABG, coronary artery bypass grafting; PCI, percutaneous coronary intervention. (Modified from Weintraub WS, Grau-Sepulveda MV, et al. Comparative effectiveness of revascularization strategies. N Engl J Med 2012;366(16):1467-1475., doi:10.1056/NEJMOA1110717.)

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the United States. This database is being housed in the Duke Clinical Research Institute, with the data being collected through the ACC on the front end and the STS and ACC along with DCRI participating in the analysis and reporting of the data to the FDA, CMS, and industry and participating surgeons and cardiologists and their hospitals. This is extremely important because it involves physicians participating in the process of the "rational dispersion" of new technology. Hopefully, this process will lead to the development of appropriate use criteria and minimizing inappropriate use of these valves until data are out showing not only their short-term but also their long-term efficacy. This relationship between the above organizations will hopefully lead to similar applications for other new cardiothoracic and cardiology technologies.

As another example of collaboration with the government and other groups, the STS and DCRI have recently been asked by CMS to work with the Yale Group to develop a method of measuring and evaluating hospital readmission rates following CABG using STS clinical data and Yale administrative data (obtained from CMS).

SUMMARY

The cardiothoracic surgery, cardiology, and general surgery specialties have set an example for other physicians to fulfill their professional obligations to society by developing robust quality improvement programs based on risk-adjusted clinical data. These databases have given our cardiothoracic specialty credibility with Congress, the Executive Branch, including CMS and the FDA, payors, purchasers, and the major quality improvement organizations, such as the National Quality Forum. The databases offer real-world research potential and the opportunity for prospective randomized trials as well. In addition, they offer opportunities for short- and long-term comparative effectiveness studies of various treatments and procedures. They are and will be utilized more in measuring efficiency and value going forward. They can be used for public reporting so that our specialties are seen by the public, the health policy groups, the government, and the purchasers as being transparent, which is very important in our culture. They have also been helpful in reimbursement negotiations demonstrating not only the quality and services rendered, but the workload involved. They are also a valuable tool for recertification and maintenance of certification for all physicians. Finally, databases are emerging as a tool that can be utilized to develop specialty society–government collaboration with organizations such as CMS and FDA and with industry in partnering in device monitoring and the dispersion of new technology.

The database acquisition of risk-adjusted clinical data through professional societies databases including the STS, the American College of Cardiology, the American College of Surgeons, and the VA databases has been integral in improving patient care by allowing each surgeon and physician to compare their results with national and regional peer groups to identify areas for improvement.

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EDITOR'S COMM NTS

Dr. Grover and his colleagues have true expertise in the field of databases and this is the reason I asked them to do this chapter. Very few developments have had more impact on the practice of cardiac surgery than those of national databases. This chapter goes through the history of the STS database, which is the measurement of results for our entire field. I think new practitioners need to understand that they will be judged by the results of databases and they need to carefully measure their own outcomes. In addition, they really have to be cognizant of the administrative databases that included billing data. These are public record and we truly have to understand the nuances of what they mean and do. They are not perfectly accurate yet they are relevant.

Though databases have many positive aspects, there is the issue discussed by the authors whether cases are turned down to improve results. Certainly, every experienced practitioner knows that they can determine their own mortality for coronary artery bypass grafting. There have been studies that the authors have quoted reviewing whether in fact the turn down rate is higher in places where there is public reporting. Eventually, there needs to be some system that studies both patients who are operated on the patients that are treated at a given institution. One must know numerator and denominator. However, this is not possible at this point.

ILK

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39 Prevention of Neurologic Injury after Coronary Artery Bypass

George J. Arnaoutakis and William A. Baumgartner

INTRODUCTION

Coronary artery bypass grafting (CABG) increases long-term survival compared with medical therapy for patients with significant coronary artery disease. Improvements in anesthesia, surgical technique, and myocardial protection have led to improved operative mortality rates. Despite these improvements, neurologic injury remains a significant risk for patients undergoing CABG. Aside from being a leading cause of morbidity in CABG patients, neurologic sequelae also account for escalating medical costs in the form of increased length of hospital stay and subsequent rehabilitation.

The spectrum of neurologic injury ranges from clinical stroke to more prevalent, yet subtle neurocognitive changes. According to a recent Society of Thoracic Surgeons (STS) report clinically evident stroke occurs in 1.4% of patients undergoing CABG and is associated with significantly worse early and long-term survival after CABG. Delirium occurs in up to 10% of patients >65 years of age and is also associated with worse long-term mortality. Other early postoperative neurocognitive abnormalities occur in up to 60% of patients and manifest as mild deficits in memory, attention, concentration, and language. Many of these subtle neurocognitive changes are transient, however, and are known to resolve at 3 months post-CABG. The three main etiologic factors involved in neurologic injury are atherosclerotic emboli, cerebral hypoperfusion, and generalized perioperative inflammation. Identifying the mechanism of injury and potentially modifiable risk factors for unfavorable neurologic outcomes is an active area of research, and many potential neuroprotective strategies have emerged.

Preoperative comorbidities predisposing to neurologic complications after cardiac surgery include advanced age, prior neurologic events, hypertension, diabetes, renal failure, atrial fibrillation, peripheral vascular disease, and carotid stenosis. These risk factors identify individuals likely to have diffuse cerebrovascular disease, impaired cerebral blood flow (CBF), or increased susceptibility to thromboembolic events. To screen for patients with asymptomatic carotid stenosis, carotid duplex ultrasonography should be performed in patients with audible bruits or in elderly patients at high risk for cerebrovascular disease.

Studies have also identified intraoperative predictors for perioperative neurologic events. These independent risk factors include the presence of significant aortic arch atherosclerosis, longer duration of cardiopulmonary bypass (CPB), and CABG with concomitant carotid endarterectomy. The preoperative recognition and assessment of risk factors is an important step in reducing the morbidity and mortality associated with perioperative neurologic injury.

MECHANISM OF NEUROLOGIC INJURY

The development of potential therapeutic strategies depends on our understanding of the mechanism of neuronal cell injury following cardiac surgery. "Glutamate excitotoxicity" is a major mechanism of neuronal injury. Glutamate is the major excitatory amino acid neurotransmitter in the central nervous system (CNS) and can cause neuronal hyperactivity and death during periods of metabolic stress such as hypoxia or ischemia (Fig. 39.1). Glutamate triggers a cascade of events via binding to an *N*-methyl-D-aspartate (NMDA) receptor, ultimately resulting in neuronal necrosis or apoptosis. Histologically, the brain regions most significantly affected are those in which NMDA receptors are prominent: the hippocampus, cerebellum, and basal ganglia. Pharmacologic blockade of this receptor also confers some degree of neurologic protection in animal models, thus reinforcing this mechanism of injury.

Nitric oxide (NO) is a ubiquitous molecule that also acts as a neurotoxin. Induction of neuronal nitric oxide synthase (nNOS) via a hypoxic/ischemic insult leads to diffuse NO formation in the brain. NO and its metabolite peroxynitrite impart toxic effects on the neuronal mitochondria, resulting in free radical proliferation and DNA fragmentation (Fig. 39.2). Mitochondrial energy failure is considered to play a central role in neuronal cell death. Investigations using a canine model of hypothermic circulatory arrest (HCA) showed that inhibition of nNOS results in decreased NO production and superior neurologic function compared with untreated HCA canines. These studies demonstrated that pharmacologic intervention at specific points in the injury cascade might potentially mitigate the neurologic deficits that can result from cardiac surgery.

CEREBRAL PROTECTION TECHNIQUES

Minimizing the Systemic Inflammatory State

CPB is associated with an intense inflammatory response due to contact of blood with the artificial bypass surfaces, conversion to laminar, nonpulsatile flow, and leukocyte and endothelial cell activation following ischemia/reperfusion. The systemic inflammatory response is characterized by activation of the complement, fibrinolytic, and cytokine cascades. Complement activation occurs immediately after the blood comes into contact with the foreign surfaces of the bypass circuit. This leads to leukocyte activation and increased inflammatory cytokine production, such as interleukin-6 and tumor necrosis factor. The leukocyte–endothelial cell interactions result in microvascular occlusion and end-organ ischemia. This inflammatory response is the suspected mechanism underlying the global cerebral edema seen on post-CPB magnetic resonance imaging (MRI).

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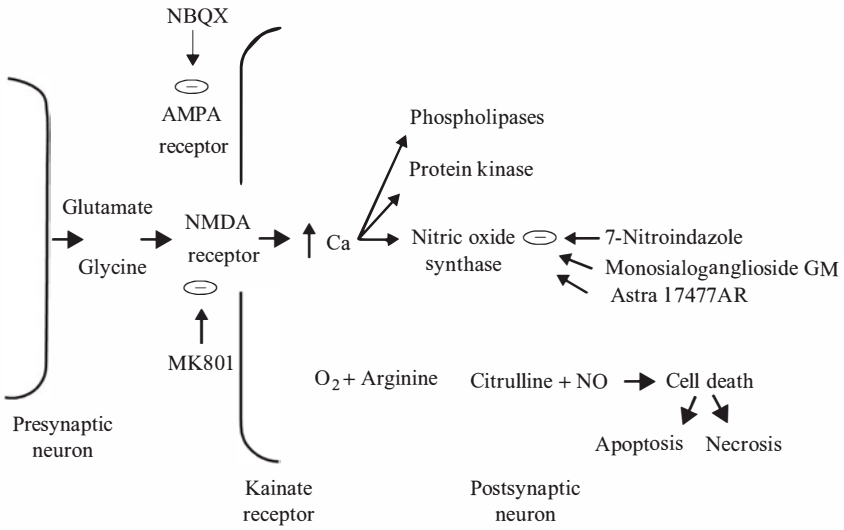


Fig. 39.1. The excitotoxicity mechanism of neuronal cell injury and points of intervention. AMPA, α-amino-3-hydroxy-5-methylisoyazole-4-propionic acid; NMDA, *N*-methyl-D-aspartate. (Reprinted with permission from Baumgartner WA. Neurologic injury after cardiopulmonary bypass surgery. J Neurosurg Anesthesiol 2004;16:102.)

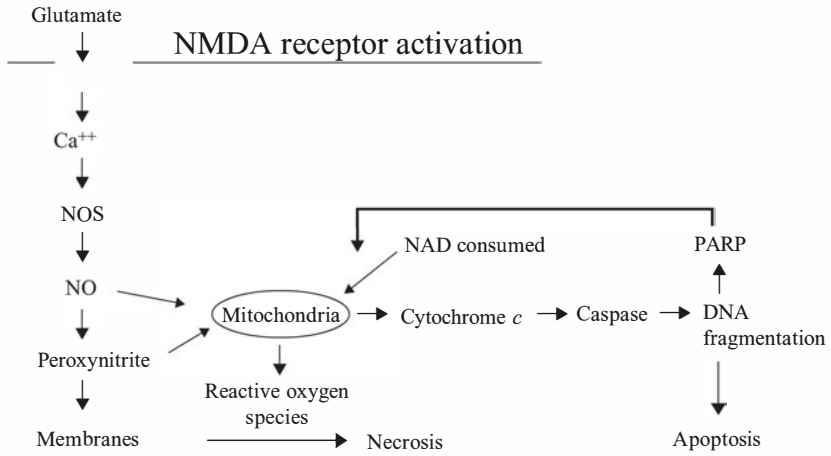


Fig. 39.2. The role of the mitochondria in neuronal cell injury. NAD, nicotinamide adenine dinucleotide; NMDA, *N*-methyl-D-aspartate; NOS, nitric oxide synthase; PARP, poly ADP-ribose polymerase. (Reprinted with permission from Baumgartner WA. Neurologic injury after cardiopulmonary bypass surgery. J Neurosurg Anesthesiol 2004;16:102.)

Many therapeutic strategies have been used in an attempt to mitigate the CPB-induced inflammation. These include both pharmacologic and mechanical therapeutic modalities. Corticosteroids have been shown to reduce CPB-induced inflammation by decreasing complement activation and levels of circulating cytokines. However, a large Cochrane Database review of over 3,600 patients revealed that the routine use of preoperative corticosteroids in adults did not confer any clinical benefit, including no improvement in neurologic complications. The serine protease inhibitor aprotinin was shown to reduce the inflammatory response post-CPB and possibly decrease postoperative stroke risk. However, due to increased risk of fatal myocardial infarction and renal failure, this drug is no longer available. Experimental evidence in animal models suggests decreased post-CBP inflammatory response in subjects pretreated with statin therapy; however, a large cohort study failed to demonstrate a clinical benefit in neurologic outcomes.

Mechanical strategies to reduce the release of proinflammatory cytokines include leukocyte and hemoconcentration filters, which wash the cells before returning them to the bypass circuit. However, despite several studies documenting reductions in measured cytokine concentrations, significant clinical benefit has not been shown. In an attempt to increase the biocompatibility of CPB, heparin-coated circuits also reduce complement activation, proinflammatory cytokine levels, and neutrophil adhesion. A prospective, randomized trial of heparin-coated circuits demonstrated a significant improvement in postoperative neuropsychometric tests as compared with conventional circuits.

Maintenance of Cerebral Perfusion

Under normal physiologic conditions, cerebral autoregulation maintains a constant CBF over a wide range of systemic arterial pressures (50 to 120 mmHg). Below 50 mmHg, cerebral oxygen delivery (CDO_2) becomes pressure-dependent. This can be compensated for by an increase in cerebral oxygen extraction. Therefore, CDO_2 remains relatively constant even at systemic pressures as low as 30 mmHg during moderate hypothermia. However, because of the burden of comorbidities prevalent in CABG patients, including hypertension, diabetes, and cerebrovascular disease, the ischemic tolerance and autoregulatory capacity of the brain are diminished. For this reason, higher minimal perfusion pressures may be required to support adequate cerebral oxygenation. Thus, flow rates during CPB are adjusted to preserve end-organ and cerebral perfusion above an ischemic threshold.

However, the watershed areas of the brain still remain at greatest risk for neurologic injury. A prospective, randomized trial of 248 elective CABG patients showed that those maintained at lower perfusion pressures (50 to 60 mmHg) had significantly higher mortality and stroke rates than those maintained at a higher perfusion pressure (80 to 100 mmHg). This study demonstrated that maintaining patients on CPB at a higher perfusion pressure was technically safe and effectively improved outcomes after cardiac surgery. The current perfusion practices, which err on the side of higher perfusion pressures, result in excellent outcomes in the vast majority of patients, and there is little evidence to suggest that altering these practices will have a positive influence on CBF.

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Embolization Reduction

There are three main types of embolic phenomena that may occur during cardiac surgery and result in postoperative neurologic dysfunction: particulate, gaseous, and lipid embolization. Perhaps, the most significant of these is the particulate emboli resulting from atheromatous debris present in the diseased aorta and great vessels. The platelet–fibrin aggregates and debris generated by the CPB circuit itself also have a causative role. To limit particulate embolization, surgical manipulation of the heart and great vessels must be minimized and care should be taken in the placement of cannulae and clamps on atherosclerotic vessels. In addition to intraoperative palpation, transesophageal echocardiography (TEE) and epiaortic ultrasonography are commonly used to assess the degree of aortic atherosclerosis and to identify more favorable areas for cannulation and clamping. In a recent study, patients undergoing epiaortic scanning showed a lower incidence of cognitive dysfunction than patients evaluated only with aortic palpation.

Cross-clamping of the atherosclerotic aorta can result in dislodging of plaque material and cerebral embolization. A number of recent studies evaluated the use of a single-clamp technique (SCT) versus a double-clamp technique (DCT) for cerebral protection during CABG. In the DCT, the cross-clamp is removed before the proximal aortic anastomoses, which are then performed using a partial occluding clamp. This technique reduces the overall CPB and myocardial ischemic time but results in increased aortic manipulation and risk of embolization. A retrospective study of 189 SCT versus 272 DCT patients showed DCT to be an independent risk factor for neurologic injury on multivariate analysis. A recent prospective trial randomized 268 patients undergoing CABG to either SCT or DCT. The DCT group included two patients (1.5%) with a postoperative stroke and two patients (1.5%) with postoperative confusion. The SCT patients demonstrated improved cerebral protection, with no postoperative neurologic complications, and no adverse effect on myocardial protection or postoperative outcome.

Novel alternatives have also evolved to decrease aortic manipulation and subsequent embolic risk. Endovascular balloon occlusion of the ascending aorta has been described as one method for avoiding the potential injury caused by cross-clamping of the aorta. Intra-aortic filtration systems (Embol-X; Embol-X, Inc., Mountain View, CA) have also been developed for deployment before cross-clamp release in an effort to reduce the embolic burden. A large, randomized trial of >1,200 patients demonstrated the filtration system to be both safe and effective with a filter embolic capture rate of 96.8%. However, postoperative event rates including mortality, stroke, transient ischemic attack, and renal insufficiency showed no significant differences between the control and filter arms of the study. When specifically examining a high-risk subgroup, there did appear to be a clinical benefit with respect to a composite outcome of mortality or major morbidity. Aortic manipulation can also be minimized with the use of automated aortic connectors for the construction of sutureless proximal saphenous vein graft–aortic anastomoses. Again, a recent prospective trial of 77 primary CABG patients randomized to either the automated anastomotic device or conventional handsewn anastomosis demonstrated no significant differences in neurocognitive deficits, stroke, or mortality.

Cerebral air embolization is another potential source of postoperative neurologic dysfunction. Air can be entrained into the heart from the surgical field. However, this is more pertinent to open-chambered procedures such as valve operations than with routine CABG. Flooding the surgical field with CO_2 is one method used to decrease the embolic burden. The CPB circuit can introduce air into the patient, but this is theoretically avoided by the venous reservoir design and arterial line filter. A dynamic bubble trap has been used to reduce gaseous microemboli, and in a prospective study was found to improve neuropsychological function at 3 months postoperatively. The number of perfusionist interventions, defined as the administration of drugs or the injection of blood into the venous reservoir, has also been correlated with the incidence of air microemboli. A prospective study of 83 CABG patients demonstrated that those with increased perfusionist interventions, and therefore increased gaseous microemboli, had significantly worse performance on neurocognitive testing. This study further implicates the CPB circuit as a potential source of clinically significant microemboli. TEE is routinely used to assess the amount of intracardiac air and can assist in de-airing procedures.

Fat embolization into the cerebral circulation is a common occurrence due to aspiration of mediastinal and cardiac fat into the cardiotomy suction and CPB circuit. This lipid material is often small and deformable, and so can easily pass through arterial line filters. Lipid emboli in the cerebral circulation are an extremely common occurrence in post-cardiac surgery patients, as determined postmortem. These emboli are associated with an upregulation of inflammatory cytokines in the brain, which may result in the systemic inflammatory response seen in some patients. Therefore, the current practice at our institution is to avoid the use of cardiotomy suction to prevent the return of lipid debris to the pump. Cell-saver or blood-salvage devices, however, can decrease the amount of particulate or lipid debris returned to the CPB circuit. When comparing cell-saver suction to standard cardiotomy suction, in a randomized trial of 226 patients, cell-saver was associated with less cognitive dysfunction. The main drawback to cell-saver devices is the washing away of platelet and coagulation factors that also occurs. Further technological refinements may circumvent this issue in the future.

Temperature Management

The issue of hypothermic versus normothermic CPB is an ongoing matter of debate. The potential benefit of hypothermic CPB is a decreased cerebral metabolic rate, which has been shown to decrease the neuronal injury associated with ischemia. Mild hypothermia also allows for lower flow rates, which can lower the risk of embolization. This risk, however, is greatest during aortic cannulation, cross-clamping and unclamping, and initiation of CPB. Unfortunately, the brain remains normothermic during these critical time periods due to the timing of hypothermia after the onset of CPB. In addition, hypothermia requires rewarming, which lengthens CPB and overall operative time. The rewarming process also harbors some potential to cause moderate hyperthermia if performed too rapidly, which has been associated with worsened neurologic outcomes. It is therefore important that during the rewarming phase the bladder temperature does not exceed 37°C to 37.5°C. Studies examining the effect of hypothermic versus normothermic CPB have produced conflicting results. Some studies suggest a decreased stroke rate and improved neuropsychological outcomes associated with hypothermic CPB, whereas others report no difference in neurologic complications. Overall, common practice is to induce a mild state of

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hypothermia (32°C to 34°C), which is generally protective of all organs during CPB.

Acid–Base Management

The increased solubility of carbon dioxide in blood at lower temperatures has resulted in two different methods of acid–base management during hypothermic CPB. The pH-stat management technique uses a temperature-corrected arterial blood gas and maintains the pH at 7.40. This technique can result in a loss of cerebral autoregulation and an increased risk of neurologic injury. The α-stat management technique, however, does not use temperature-corrected arterial blood gases and therefore maintains the autoregulation of CBF. Studies comparing these two management strategies have had varied results. Some animal studies have advocated the use of the pH-stat method due to improved functional outcomes and less severe neuronal injury. However, other studies report a higher incidence of postoperative neurologic dysfunction in patients treated with the pH-stat method. This may be due to increased CBF associated with the loss of autoregulation, which can lead to an increased delivery of microemboli to the cerebral circulation. Despite conflicting opinions, the majority of adult practices today utilize the α-stat management technique.

Pharmacologic Intervention

Many different pharmacologic agents have been explored for their potential neuroprotective effects in both experimental and clinical models. There are no approved therapies for the specific prevention of CABG-associated neurologic events. However, a multi-institutional prospective observational study revealed that early initiation of aspirin in the post-CABG period improved numerous outcomes, including a 50% reduction in stroke rates.

In addition, various anesthetic agents have been used in conjunction with electroencephalography in an effort to suppress cerebral oxygen metabolism. Anti-inflammatory agents have been tried to prevent the adverse inflammatory sequelae of cerebral ischemia. However, the limited success observed with these agents may not be due to a direct neuroprotective effect, but rather by an indirect effect on cerebral emboli. As the mechanism of injury is further elucidated, the potential targets for intervention continue to increase. Our understanding of glutamate excitotoxicity with increased NO production and neuronal cell death has led to the rational exploration of glutamate-receptor blockers and NO synthase inhibitors. These agents have shown beneficial neurologic effects in our experimental canine model of HCA. HCA is considered to carry the greatest risk of neurologic injury and is therefore a practical model for pharmacologic investigations.

The inhibition of mitochondrial energy failure is another strategy for preventing the ischemia-induced neuronal injury in our canine HCA model. Ischemic preconditioning (IPC) is a paradoxical form of protection against lethal ischemia by exposure to brief episodes of ischemia prior to the primary insult. Although initially described in cardiac myocytes, a similar mechanism was also found in neurons and may confer mitochondrial protection. One potential mechanism of IPC relies on the opening of ATP-dependent K^+ channels on the inner mitochondrial membrane. This effect can be achieved pharmacologically by using a variety of different agents. Diazoxide, an antihypertensive medication no longer in clinical use, is one such ATP-dependent K^+-channel agonist. Pretreatment with diazoxide before neurologic insult has been correlated with improved functional outcomes and histopathology. This has been further substantiated by the antagonistic effect of glibenclamide, an ATP-dependent, K^+-channel antagonist. In the current age of molecular-targeted drug design, further success in pharmacologic neuroprotection will become increasingly feasible as our mechanistic understanding continues to improve. There is also evidence in our canine model that the anticonvulsant medication valproic acid may offer neuroprotection; however, concerns regarding metabolic acidosis have limited broader application in the clinical setting.

Hematocrit Management on Cardiopulmonary Bypass

Hemodilutional anemia has been used during hypothermic CPB to reduce blood viscosity to allow maintenance of baseline CBF. This technique is perceived to reduce the risks of adverse outcomes due to arterial hypertension such as aortic dissection and collateral blood flow to the coronary arteries during cross-clamping of the aorta. Although a goal hematocrit level of approximately 21% was traditionally common practice, multiple recent studies have contradicted the appropriateness of this technique. In a randomized trial of 147 infants, Jonas and colleagues randomly assigned patients to a lower hematocrit strategy (21.5%) or a higher hematocrit strategy (27.8%) during hypothermic CPB. The lower hematocrit group had worse perioperative outcomes with significantly lower scores on the Psychomotor Development Index at 1 year of age. Another prospective observational study of 10,000 patients found that each 1% decrease in nadir hematocrit on CPB was independently associated with a 10% increase in the odds of postoperative stroke. At our institution, we similarly observed an independent association between low post-CPB hemoglobin levels and increased stroke risk. These studies suggest that inadequate oxygen delivery due to anemia may be the mechanism of cerebral injury. While adverse neurologic outcomes have been observed with low hematocrit levels on CPB, prospective randomized studies specifically addressing this issue are needed to better guide the optimal target hematocrit during CABG surgery.

Glucose Control

Disruption of cerebral energy metabolism plays a causative role in the neurologic injury that occurs following cerebral ischemia. Episodes of brain ischemia initiate anaerobic glycolysis with subsequent intracellular lactic acidosis, which causes cellular toxicity. An increase in blood glucose concentration at the time of ischemia provides more substrate for anaerobic metabolism with exacerbation of the acidosis. Studies show that hyperglycemia at the onset of cerebral ischemia worsens postischemic neurologic function and histopathologic injury. In a primate model of complete cerebral ischemia, animals receiving a glucose infusion immediately before ischemic insult had significantly worse neurologic function at 96 hours than control animals receiving a crystalloid infusion. Large clinical studies have shown that diabetic CABG patients with poorly controlled perioperative hyperglycemia had higher rates of sternal wound infections and worse long-term survival. However, because of concern of serious hypoglycemia, there has been a paradigm shift away from strict glycemic control (90 to 120 mg/dl) toward more moderate glycemic control (120 to 180 mg/dl) in CABG patients. The appropriate intraoperative management of hyperglycemia in patients undergoing CABG continues to evolve, but currently most centers strive to maintain glucose levels in the 120 to 180 mg/dl range, in concert with current STS recommendations.

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Intraoperative Cerebral Monitoring

Intraoperative neurophysiologic monitoring reduces the incidence of neurologic complications after CPB. Several different modalities exist, including near-infrared cerebral oximetry, jugular bulb oximetry, EEG, somatosensory-evoked potentials (SEPs), and transcranial Doppler (TCD) ultrasound. These multimodality techniques can help detect cerebral hypoperfusion and impaired oxygenation, which contribute to brain injury during cardiac surgery. Cerebral oximetry is a simple method of measuring cerebral oxygenation. The level of cerebral oxygenated hemoglobin has been shown to correlate with high-energy phosphates and can predict histologic brain injury in an animal model. In addition, near-infrared spectroscopy can be used to continuously monitor CBF autoregulation in CABG patients. Real-time assessment of cerebral perfusion autoregulation may be useful in tailoring hemodynamic goals to an individual patient, thereby minimizing injurious hypotension during CPB.

Jugular bulb oximetry provides similar information regarding oxygen imbalance. However, it is more invasive due to the need for jugular vein cannulation. EEG and SEP monitoring have also been used as measures of cerebral metabolic activity. EEG silence and the disappearance of SEPs can be used as markers of adequate cerebral protection and can potentially shorten the cooling period during CPB. These two techniques, however, are more commonly utilized in patients undergoing HCA. TCD ultrasound can detect sudden changes in either blood flow or vascular resistance, as well as aid in the identification of embolic phenomena. In addition to detecting abnormalities, these modalities can be used to make adjustments in perfusion, oxygenation, or anesthetic administration. When used effectively, intraoperative cerebral monitoring shortens hospital stay, decreases costs, and prevents neurologic complications.

Off-Pump Coronary Artery Bypass

With the development of cardiac immobilization techniques that enable complete revascularization of the beating heart, there was speculation that by avoiding use of CPB off-pump CAB (OPCAB) would reduce neurologic complications. CPB is associated with an intense inflammatory response secondary to the conversion to laminar flow, blood contact with the artificial bypass surface, cold cardiac ischemia, and reperfusion. In addition, other CPB-related factors such as hemodilution and aortic cross-clamping play a significant role in postoperative neurologic dysfunction. OPCAB has the potential advantage of avoiding the inflammatory response and micro/macroemboli that may result from the CPB circuit and manipulation of the atheromatous aorta. Some retrospective studies suggest a decreased early stroke rate with OPCAB, although delayed stroke rates were equivalent. However, numerous prospective, randomized studies have failed to demonstrate significant neurologic benefit over CABG with traditional CPB. Although OPCAB may offer some theoretical advantages, its ability to reduce the risk of neurologic complications is presently unproven. The growing recognition that OPCAB does not appear to improve neurologic outcomes has led to a shift in focus toward patient-related factors, such as arteriosclerotic burden, that predispose patients to neurologic morbidity.

ASSESSING NEUROLOGIC INJURY

Neuropsychological Testing

Traditional studies in the area of postcardiac surgery neurologic outcomes focused on clinically obvious neurologic and psychological dysfunction such as stroke, disorientation, and depression. However, with more widespread use of neuropsychometric testing, more subtle injury patterns can be observed. This testing typically includes measures of language, memory, attention, concentration, and psychomotor performance. In a prospective study of 127 patients undergoing CABG, patients were given a battery of cognitive tests preoperatively, 1 month postoperatively, and 1 year postoperatively. This study established that the incidence of cognitive decline varies among the different domains of cortical function tested. More importantly, however, these patients must be followed longitudinally because initial deficits may eventually show either improvement or further decline. We recently performed a prospective study examining the longitudinal neuropsychological performance of CABG patients versus nonsurgical controls with coronary artery disease. The cognitive testing results at 3 months and 1 year after baseline examination were comparable between the two groups, suggesting that the cognitive decline during the early post-CABG period may be transient and reversible. Therefore, any long-term cognitive dysfunction may be related to age and other comorbidities such as vascular dementia, as opposed to directly attributable to CPB exposure. With further research into neurobehavioral assessment techniques, it will be possible to evaluate the effectiveness of surgical and pharmacologic strategies for mitigating neurologic injury after CAB surgery.

Molecular Markers

Various proteins released by the injured brain have been used to measure cerebral injury. Neuronal, glial, and endothelial cells of the CNS elaborate these substances in response to ischemia/reperfusion injury. Assays for these biochemical markers provide a relatively noninvasive means of quantifying the extent of neuronal injury. If these assays can demonstrate a direct correlation with clinical outcomes, these biomarker assays may offer a simpler way of assessing postoperative cognitive dysfunction. A recently investigated protein is ubiquitin C-terminal esterase-L1 (UCHL1), which is specific to neurons, is expressed in high concentrations in the CNS, and functions in the degradation of misfolded proteins. This serum biomarker correlates with neuronal injury in animal and human studies of subarachnoid hemorrhage and traumatic brain injury. In our laboratory, we detected serum elevations in this biomarker in animals subjected to neurologic injury after being exposed to a prolonged period of HCA. Current studies are underway to examine the usefulness of this biomarker in human patients undergoing CABG.

S100β is another candidate biomarker, which normally promotes axonal growth, glial proliferation, and neuronal differentiation. This protein is released by injured glial cells and also demonstrates increased blood concentration after cardiac surgery. It has been shown to correlate with intraoperative cerebral microemboli, quantified using TCD sonography. Furthermore, a significant correlation was detected with neuropsychological deficits assessed at 6 months postcardiac surgery in a small study of 16 patients. Many of the other previously studied CNS biomarkers, such as tissue-specific enolase, are limited by poor specificity or late detection. In addition to UCHL1 and S100β, recent experimental evidence suggests that spectrin breakdown products and glial fibrillary acidic protein have promise as useful biomarkers in the clinical setting of patients undergoing CABG. Although no CNS biomarker is used routinely at the present time, a reliable, noninvasive biochemical test for detecting early neurologic deficits would provide a great benefit to the postoperative care of CABG patients.

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Magnetic Resonance Imaging

MRI has demonstrated cerebral edema as early as 1 hour after CABG. This is often clinically insignificant and resolves by 1 week. Diffusion-weighted MRI can also help to differentiate acute from chronic brain ischemia; however, it has been difficult to correlate the presence of lesions on MRI with clinical cognitive dysfunction. Magnetic resonance spectroscopy (MRS) is another useful adjunct, which may have better predictive value for cognitive dysfunction. We have used MRS in our laboratory canine HCA model, which has allowed us to detect and quantify subcellular metabolic changes within the brain after HCA. The most significant finding on MRS was a decline in the ratio of *N*-acetyl-aspartate to choline (NAA:Cho) 24 hours post-HCA, which is a known marker of neuronal mitochondrial dysfunction. MRS also provided a way of assessing the efficacy of different pharmacologic agents. Pretreatment with diazoxide limited neurologic injury versus controls, which was reflected in a preserved NAA:Cho ratio. These results could be correlated with the degree of clinical neurologic injury observed. Similarly, MRI/MRS and neurocognitive testing were performed serially during the pre- and postoperative period in a clinical study of 35 CABG patients. Transient changes were detected in the ratio of NAA to creatinine (NAA:Cr), with the degree of decline closely correlating with deterioration in neurocognitive function, increased patient age, and longer CPB times. Normalization of the NAA:Cr ratio also accompanied the recovery of neurocognitive performance over time. These studies clearly demonstrate that MRS may offer an early, noninvasive means of assessing neurologic injury and the effectiveness of neuroprotective agents.

CONCLUSION

Neurologic injury remains a significant complication after CABG. This is especially true given the continual improvements in surgical and anesthetic techniques that have enabled cardiac surgeons to operate on older and higher acuity patients today. Therefore, it is important that the individual risk profile of each patient be taken into consideration. The various neuroprotective strategies available to the cardiac surgeon continue to expand. Pharmacologic neuroprotection is an attractive option for preventing neurologic injury and therefore is a very active area of research. Although we have had success in some laboratory and small clinical studies, no single agent is available for routine use in CABG patients. However, as our fundamental understanding of the mechanisms of brain injury after cardiac surgery continues to improve, we will be better equipped to design enhanced neuroprotective strategies in the future.

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EDITOR'S COMMENTS

These authors have presented the mechanisms of neurologic injury and potentially preventive strategies for prevention. There is no doubt in my mind that this is the most difficult area of comorbidity now facing cardiac surgery. Neurologic injury is a difficult area and clearly affects the present and future of our patients. I entirely agree with the authors about the avoidance of embolization as well as some of the preventive measures they have proposed. Dr. Baumgartner has studied the single cross-clamp technique for coronary bypass surgery and there is no doubt this is the way on-pump bypass surgery should be done. This is the technique we presently employ. The authors have also demonstrated that off-pump coronary surgery will not completely prevent neurologic injury.

I agree with the authors that there will be pharmacologic adjuncts that will help us in more complex cases including circulatory arrest. The Cardio thoracic Surgery Network sponsored by the NHLBI is reviewing several protocols that will potentially examine such agents. I believe we will have a totally different approach in the future to prevent these complications.

ILK

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40 Patient Safety

Thoralf M. Sundt

The term "patient safety" is widely—and sometimes loosely—used today in a variety of contexts and it would convey a variety of meanings. The suggestion made over a decade ago that patients were unsafe in our hospitals was initially greeted with skepticism if not hostility but now has given way to acceptance as the medical community has come to recognize that there are indeed instances of avoidable harm. With a turn in focus to reducing "adverse events" and "errors," a veritable "movement" has arisen within the medical community.

The scope of the topic is enormously broad, encompassing equipment design to make their use more intuitive through legible and proximity-compatible information delivery and ergonomic structure, to process design to make them simpler and more resilient. Less obvious may be issues of teamwork and communication. While all aspects of medical error impact the entire medical community, for practitioners of diverse specialties the emphasis may vary. For pharmacists or medical oncologists, the area of greatest risk and hence greatest focus may be avoiding medication error. Surgeons have focused for many years on aspects of technical error. As the complexity of surgical care has increased, however, errors in nontechnical skills such as cognitive errors, communication errors, and errors in situational awareness have become more important. We will focus here on these domains in the interest of developing a basis for understanding the interventions proposed for the surgical arena. The potential impact of such interventions is powerful, as disruptions in teamwork and communication have been shown repeatedly to be strongly correlated with errors, outcomes, and even legal actions. Furthermore, the interventions required to improve teamwork and communication are almost entirely within our own personal control.

It is also worthwhile carefully defining our terms. "Error" as defined in the Institute of Medicine Report is "the failure of a planned action to be completed as intended or the use of the wrong plan to achieve an aim." As such, there is no blame implied nor is the occurrence of an adverse event requisite. Regardless of the clinical consequences, an error stands as an error just the same. Accordingly, its pathogenesis can be examined, as can any recovery efforts successful or not. Indeed many would argue that the most information one can gather is by examining "near misses" as they are more plentiful (thankfully) than adverse events, and that successful error capture and recovery has more to teach about resilient teams than uncorrected ones.

The meaning of the term "adverse event" is self-explanatory; an injury or complication occurring during medial treatment. Adverse events may be avoidable or not, and they may be related to an error or not. The focus on adverse events resulting from the patient's underlying condition has long been a staple of the medical literature, such as the incidence of dialysis-dependent renal failure and its association with comorbidities such as long-standing hypertension or diabetes. To be sure "patient safety" can be improved via interventions to mitigate that risk as well; however, more often when reference is made to patient safety efforts, it is in the context of adverse events occurring as a consequence of medical error, particularly those that might be preventable. Of course, not all adverse events lead to lasting or significant harm, and the definition of "preventability" is thorny as well. A narrow perspective may rest with the legal definition of negligence, which addresses whether or not care provided meets the standard of care that can be reasonably expected of an average physician in those same circumstances. On the contrary, a more liberal definition of preventability will likely lead to more progress in improving outcomes.

There has been significant argument over whether it is more productive to focus on errors or adverse events. Argument in favor of the former identifies them as the ultimate precursors and belies the presumption that prevention of error will reduce adverse events and preventable harm. The difficulty is that errors are so common that it can be overwhelming just keeping track of them. Those who argue in favor of a focus on adverse events suggest that such an approach with concentrate attention on the most serious errors worthy of attention. There really can be no resolution to this argument as both sides have merit. It is worth noting, however, that one must be clear about definitions at the outset. Improved safety will be the inevitable product of reducing all of these.

Although it was the Institute of Medicine report "To Err is Human" published in 2000 that raised the topic most prominently to our collective consciousness, the origins of this alternate perspective can be traced back at least as far as Ernest Amory Codman who challenged the medical community in Boston to adopt what de Leval has called a "forensic" approach to surgical outcomes in contrast to the more traditional "statistical" approach. Codman insisted on reporting of 1-year outcomes including the reasons why perfection had not been attained. He classified errors and adverse events and queried the role of human, organizational, and equipment factors. This approach was not welcomed in his home town or nationally. The same theme, however, was taken up by de Leval, initially surrounding the arterial switch operation in his own hands and subsequently in the United Kingdom broadly. The impact of even minor errors on outcomes was highlighted in this study, reported to the American Association for Thoracic Surgery in 1999, as was the role of "Human Factors" in general. He had begun his journey many years previously as told in this *Lancet* article "Human Factors in Cardiac Surgery: A Cartesian Dream," which represented a summary of his Mannheimer lecture focused on his own approach to technical errors after the death of a child early in his career due to a technical error. In it, he outlines the lessons to be learned from other disciplines and his journey into systems thinking, recognition of the impact of many factors beyond patient risk factors, and surgical skill on outcome. Importantly, such a perspective provided him and us a window into other opportunities to impact those factors and improve outcomes.

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LEARNING FROM OTHERS

Among the fields de Leval encourages us to explore is that of human factors science. This field incorporates knowledge from cognitive psychology, engineering and industrial design, and ergonomics to understand human capabilities and limitations, and their impact on performance of tasks be they medical or otherwise. This understanding helps in the design of systems or technologies to reduce the chances of human error. While the origins of process design can be found in 19th century "time and motion studies" conducted to improve factory efficiency, human factors came into its own as a recognized field in World War II when aircraft became so remarkably complex that their operation exceeded the cognitive capacity of a single human mind. The result was an effort to design the plane to fit the human rather than vice versa. For example, Alphose Chapanis demonstrated that shape coding aircraft cockpit controls for the flaps and landing gear reduced landing accidents. It is because of the dramatic accomplishments in the field of aircraft engineering that the analogy between medicine and aviation is cited so frequently, sometimes to the chagrin of physicians. The analogy holds, however, as fundamentally the issue is human performance in complex environments. Closely related is the field of cognitive psychology including biases, heuristics, and intuitive decision-making.

The relevance of these fields to surgery resides in *practical* solutions to *common* human challenges in *complex* environments that have been explored and developed elsewhere but can be modified and applied in medical care in general and surgical care in particular. The cognitive aspects are particularly applicable to surgery and in particular cardiothoracic surgery, as our field demands a large number of decisions be made on the basis of incomplete information on a short timeline with profound implications for our patients. When things go wrong, we can apply the discipline of accident analysis to understand the potential interventions that might prevent the same in future. The applicability of the Human Factors Analysis Classification System, which was employed productively to reduce accidents among military pilots, to cardiac surgery has been demonstrated.

A nontrivial barrier to the application of forensic analysis and human factors science to health care is the fear of retribution either from peers or the legal system. The former is within our control even if the latter is not. In order to maximize our ability to learn from errors, it is necessary that we develop a "blame-free" culture in which errors can be openly discussed. This does not imply disappearance of accountability, but rather a reframing. Individuals are still accountable for their own actions such as willing rule violations or reckless behavior, whereas the accountability for systems issues is shared among those responsible for the systems themselves. In some instances, individual accountability is replaced by collective accountability—not disappearance of accountability. This concept has been termed " Just Culture."

COMPLEXITY

It is reasonable to ask "why change from the way we have done things in the past?" Apart from concern about "political correctness" what is so different about medical care today? Why is a conscious focus on medical error is now necessary?

Just as occurred with rapid technological developments in the military during WWII, the complexity of our environment has increased exponentially in recent years. The body of surgical knowledge, the complexity of our technology, the comorbidities of our patients, and the organizational complexity of medical institutions including interdisciplinary collaborations have made medicine a much different field today that only a few years ago. The consequences of this increased complexity are profound.

Complexity itself is now a field of scientific endeavor. Complexity theory and systems science focus on the properties of complex systems. Complex, interconnected systems behave as more than the sum of their parts. Complex systems are, by their very nature, nonlinear, sensitive to initial conditions, and self-organizing. The interactions among individual elements lead to unpredictability and unintended consequences. Perrow has argued convincingly on the basis of study of nuclear power, petrochemical plants, mines, deep-sea oil drilling operations, and marine commerce that accidents are inevitable—they are "Normal" —in highly complex environments. The risks to the system as a whole posed by normal accidents depend in significant measure on the degree to which the interactions among system components are "tightly or loosely coupled" and our ability to capture them on whether the relationships are "linear or complex." Ironically, technological "solutions" such as safety systems layered on top of these complex systems may simply make them even more complex and incomprehensible.

The implication if we wish to continue to make progress, then, is that we must develop means of dealing with such accidents. The so-called "High Reliability Organizations" (HROs) such as the nuclear power industry or flight-deck operations on aircraft carriers have done so. HROs are defined as organizations that must function reliably in highly complex environments in which the consequences of error are catastrophic. In such organizations, the focus is on error management—not just prevention—and anticipation of the unexpected; they focus on error identification, capture and recovery as well as the critical importance of resilience. Complexity is fundamentally best dealt with by decentralized control, and HROs demonstrate this in the manner in which they are structured.

HUMAN COGNITION

Recent years have seen marked advancements in our understanding of human cognition and an understanding of the strengths and limitations of the same. Metacognition—thinking about thinking—has become a topic of popular books and Nobel Prizes (Kahneman). An understanding of those cognitive processes and limitations are critical for surgeons called upon to make decisions quickly and under pressure on a regular basis. Uncertainty, time pressure, and the gravity of the decisions increase our vulnerability to error. For these reasons, it is important to understand how our brains work and, perhaps more importantly, when we are at greatest risk of making an error and means of mitigating this risk. Again, this is an area of opportunity for us to improve outcomes.

In order to deal with our complex environment and what would otherwise be an overwhelming number of sensory inputs, human perception demands application of filters; the consequence is systematic and predictable biases. Equally, to make decisions efficiently, we employ cognitive shortcuts or "heuristics." Kahneman has identified two complementary cognitive systems—one fast and one slow—called system 1 and system 2 or more descriptively the automatic and contemplative or reflective. The former is more efficient in complex environments, particularly when time is of the essence. Unfortunately, it is also susceptible to pitfalls. Many of the interventions proposed to improve patient

Table 40.1 Some Common Pitfalls in Perception and Decision-Making

Hindsight bias: "I knew it all along" or "Monday morning quarterback"—refers to the tendency to view what has already happened as obvious. This is unfortunately the basis of many legal actions.

Confirmation bias: Refers to the tendency to dismiss disconfirmatory or contradictory evidence and cling to a theory or interpretation. When manifest among a collective, this phenomenon is referred to as "group think."

Representativeness bias: Refers to the tendency to overestimate the likelihood of an explanation or diagnosis based on its similarity to a memory of the same rather than the actual likelihood of the explanation (classically ignoring base rates). It derives from a reflex tendency to look for similarities.

Valence effect: The tendency to think positive outcomes to be more likely than negative ones. This is responsible in part for the Framing Effect (see below).

Certainty effect: The impact of a reduction in the probability of an outcome is greater if it can be eliminated completely.

Biases secondary to context

Contrast effect: Contrasted stimuli that are similar to each other tend to alter perceptions of each other. A circle surrounded by smaller circles "looks" larger that the same-sizeed circle surrounded by larger ones. Remarkably, it remains true even when one knows that the circles are of the same size!

Primacy effect: Perceptions tend to be overinfluenced by impressions made early, particularly when one is comparing opposite sides of an argument. This is why there is a subtle advantage to leading off in a debate. This effect is particularly apparent if the subject is queried soon after the impressions are made.

Recency effect: In contrast, in other instances, it is the final event that has the greatest impression. In this instance, one may best remember the last presentation in a debate more clearly. This is more apparent if the subject is queried late after the event. The same phenomenon accounts for the tendency to consider the same diagnosis today as one saw yesterday, even if less likely than another.

The Halo effect: The tendency to attribute additional characteristics to a person or phenomenon based on other characteristics (e.g., good looking and intelligent, etc.)

Framing effect: Responses will depend on the way in which the questions themselves are stated or framed, for example, closed versus open questions, the number of responses (many/few, odd/even), and losses versus gains such as mortality/survival.

Certainty effect: An opportunity to reduce the probability of an outcome is more impactful if the outcome was initially thought certain.

The endowment effect: Endowment effect or "loss aversion" refers to our tendency to overvalue that which we already possess (be it an object or an idea).

Regret theory: When making a decision we mentally simulate the possible outcomes and then compare the way we feel about the loss of either possible positive scenario.

safety are, in fact, defenses against just these pitfalls. Some common pitfalls are presented in Table 40.1.

Perceptions are the critical first element in decision-making, and preferences impact perceptions (Fig. 40.1). Humans typically use different criteria depending upon whether we are subconsciously or consciously motivated to prove or disprove a concept. All judgments are influenced by selective perception and the innate drive to cognitive consistency. The former is impacted by motivational factors (what we want to be the case) as well as expectations. Expectations in turn are impacted by personal experience and memory (availability bias). And the drive to cognitive consistency derives from our amygdale; we are literally driven to reduce inconsistency and make sense of a situation as the alternative is emotionally unpleasant. Note too that an implication of this is that once committed to an action or course, we are inherently motivated to feel positively about that course of action. It is easy to say that "an average surgeon sees what he/she believes, but a great one believes what they see" but this caters to the heroic superman view. The truth is that humans all see what they believe to some degree.

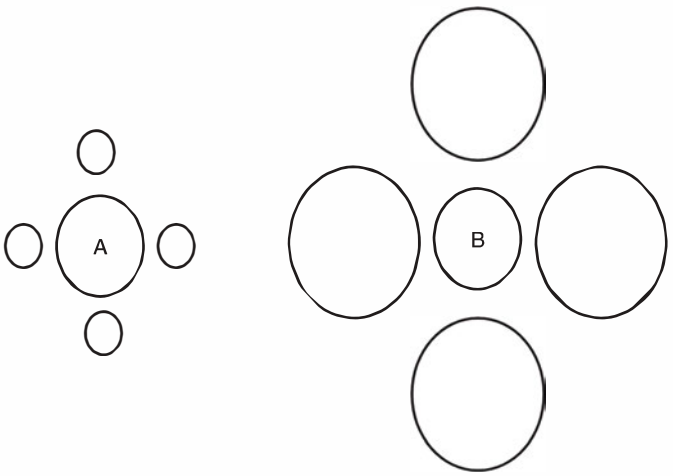


Fig. 40.1. This example of the contrast effect, in which contrasted stimuli altered perceptions of the other, demonstrates the remarkable power of such illusions even when one is aware of the facts. Although the center circles are exactly the same size, the circle surrounded by larger ones looks smaller than its counterpart surrounded by smaller circles. The illusions persist even though one knows that the circles are in fact the same size.

Finally, again along the lines of understanding our own cognition, there is a growing body of knowledge about decision-making itself. Some of the biases mentioned

in Table 40.1 directly impact decision-making. Beyond that, however, are theories of "expert decision-making" that again have direct relevance to surgery.

Expert or naturalistic decision-making stand in contrast to the means whereby novices make judgments and decisions. While novices think algorithmically, relying on "slow thinking" the way we were taught our craft, experts rely on intuition and "fast thinking" to increase speed and efficiency. This is how an expert comes to a diagnosis or course of action so quickly given a patient or situation similar to but not exactly like one they have encountered before. Rather than deductive, logical thinking including analysis of probabilities and use of statistical methods, in natural settings we look for analogous experiences as prototypes in a manner termed "recognition-primed decision-making" to size up a situation and mentally model possible solutions (and their pitfalls). These modes of thinking are particularly evident when experienced decision-makers are under time pressure to make high stakes decisions with inadequate information under dynamic conditions. This is surgery.

While efficient, this nonalgorithmic thinking has its weaknesses; it does not permit evaluation of alternate courses of action side-by-side because it does not list a series of options. It depends upon mental simulation of the course of action asking only "will it work." The alternative strategy—the "rational choice" strategy employed by novices is time inefficient. Mental simulations can run awry, however, as we are prone to explain away contradictory information (confirmation bias) until the explanations themselves become so complicated that we loose faith and reexamine. Sensemaking can go wrong if we rely too much on our intuitions and expertise as our own mindset may make us blind to other explanations. This explains how we can be amazed by the medical student that twigs to the correct answer when all of the "experts" have been following the wrong path, or why we can be mystified at mortality and morbidity conference by our own blindness to what we "really going on." It is also why listening to the input of "nonexperts" can be so valuable.

Added to these challenges is the biology of the sense of certainty. Unfortunately, perhaps, our confidence in decisions tends to increase as the difficulty of those decisions increases and, consequently, as the accuracy of those decisions approaches chance. Importantly, this discrepancy between accuracy and confidence is unrelated to intelligence. The involuntary "feeling of knowing" derives from the limbic system. Furthermore, the more uncertain the situation—due to missing information, incorrect information, conflicting information, noisy information, or confusing information—the more the decisions are based on emotion, instinct, and heuristics.

There are also important implications of this understanding for training: if we wish to train clinical intuition and expert decision-making, students need experience in actually making decisions and managing difficult cases. In years past, this was accomplished by long hours in the hospital. Today it may be possible to accomplish by developing training programs with intensive exercises and realistic scenarios demanding rapid action. Deliberate practice in decision-making with immediate feedback can actually improve this skill.

Note also that an understanding one's own expertise is important when communicating to others. Many things that are obvious to experts are completely invisible to novices. One must take the time to explain how one has come to a conclusion or the patterns one sees that simplify the situation cognitively for you.

An additional cognitive challenge in identifying problems or pitfalls for surgeons in particular is termed "stance." This term refers to your approach when carrying out the work. Are you focused on the performance of task itself or on identifying potential problems? It is difficult to do both, which is why the individual running a cardiac arrest code should not be the one actually performing cardiopulmonary resuscitation. And yet it is the nature of what we do that the operating surgeon must do both. Furthermore, it has been demonstrated that if continually interrupted one will be even more challenged in identifying problems. To some degree, this is an inescapable fact of life. It is also an argument again for engaging the rest of the team cognitively and leveraging all of the minds in the room.

The practical utility of this metacognition is in developing awareness of when and how one may misperceive a situation or make an erroneous decision and develop defenses. Checklists are one such defense, but only one, and they are useful for a particular type of cognitive error. They are useful for preventing memory lapses during the conduct of a sequence of steps that are the same every time and must be conducted in the correct order. They are particularly useful for the human–machine interface. They are decidedly not, however, a universal solution—they address only memory lapses. They do not address biases in perception or cognitive traps. The broadest solution is collective intelligence. This, in turn, demands effective communication and teamwork.

HIGH RELIABILITY

Other high risk/high consequence endeavors have adopted strategies to manage these same challenges via teamwork and collective intelligence. The so-called "High Reliability Organizations" can be defined as "Organizations that consistently avoid catastrophes in environments where normal accidents can be expected due to complexity." They share a fundamental character: they are organized to expect, identify, and contain the unexpected. They exhibit resilience. Their aim is error management, not just error prevention. To accomplish this, they are organized around five principles:

- **Preoccupation with failure:** Lapses are taken seriously as a symptom that there is something wrong with the system.
- **Reluctance to simplify interactions:** Deliberate steps are taken to constantly update interpretations to create a complete and nuanced picture.
- **Sensitive to operations:** Recognition that unexpected events originate in latent failures that are revealed during normal operations on the front line.
- **Commitment to resilience:** Deliberate development of capabilities to detect, contain, and recover from *inevitable* errors.
- **Deference to expertise:** Decisions need to be made where the expertise is—on the front line.

Managing the *unexpected* demands an understanding of the *expected* and how expectations are shaped and, in turn, how to manage those expectations mindfully. As noted above, cognitive filters based on expectations are necessary to deal with any complex environment. Importantly, mindfulness demands more than situational awareness—it demands ongoing questioning of expectations and a willingness to accept new interpretations. The focus is on emerging threats and potential means of dealing with them. We see this in the experienced surgeon who asks, "What is the danger at this point in the operation and what options do you have to deal with it if it occurs?" The key cultural components are a reporting culture, a just culture, a flexible culture, and a learning culture.

Dealing with the unexpected demands resilience—an ability to remain in control despite external or internal challenges. Resilience demands an ability to (1) absorb strain, (2) to bounce back rather than collapse, and (3) to learn from the experience. These can be properties of both individuals and organizations alike. Importantly, accomplishing these aims demands flexibility. Unfortunately, at an institutional level, all too often the response is to implement rules and regulations that reduce flexibility. Worse, they may actually increase the complexity of getting the work done and absorb more of the cognitive capacity needed for adaptive thinking. The most powerful tool is teamwork.

TEAMWORK

The most powerful countermeasure to pitfalls in perception and decision-making is effective teamwork. Collective cognition—harnessing the diversity of expectations, biases, and perspectives—enables effective error management and resilience. High-level teamwork, in turn demands effective communication, cooperation, and coordination. This is the contribution of complex sociotechnical systems theory as applied to the problem of medical error. The interactions and interfaces are the key to high reliability and improved outcomes as suggested by de Leval in his Mannheimer lecture.

Miscommunication—be it due to ineffective "encoding" of the message because of vague, ambiguous, or incomplete language; inaccurate decoding; or misdirection to the wrong individual—obviously undermines teamwork. It stands in the way of establishing "shared mental models" or "common ground" for sense making. The problems of language have, perhaps unsurprisingly, been particularly studied in aviation where recordings of communications among pilots and between pilots and air traffic controllers provide a unique window. The parallels to the operating room are clear; problems of ambiguity (such as when a surgeon says "OK" meaning "go on bypass"), homophony (an increasing problem among staff for whom English is a second language—"on" and "off" can sound a lot alike), punctuation and intonation as well as problems of reference (does "vent on" refer to the LV vent or the root vent?), and problems of inference (either false or absent as in the case when an assistant suggests "are you sure that you did not leave a sponge behind?" meaning "you left a sponge behind the heart") are encountered daily. This is the basis of emphasis on structured communication in aviation and the military. Even if language is clear, and situational awareness is common, the presence of hierarchy may present a barrier to "speaking up."

Formal programs in team training and communications abound; while they have proven effective in aviation, and initially so in health care, their impact in the latter has proven difficult to sustain. TeamSTEPS is a program developed by the Department of Defense in cooperation with the Agency for Healthcare Research and Quality and is freely available but appears to demand continued coaching. Surgeons and human factor scientists in the United Kingdom developed the Nontechnical Skills for Surgeons program, but its roll-out to U.S. sites has been limited.

An alternative approach, favored by the author, has been the institution of structured, participatory briefings. Their implementation is low-technology and time-efficient, and the impact appears durable as long as the prescribed behaviors are sustained. Participatory preoperative briefings establish multidirectional dialog, such that if and when problems arise, team members are more likely to speak up. They have been shown to reduce flow disruptions and waste even when instituted among familiar teams. The participatory aspect is enhanced if the order of speaking is in reverse order of hierarchy or empowerment. The effectiveness of this approach during briefings or, indeed, during floor rounds, is remarkable. In addition, a conscious effort can be made to invite and celebrate the mindful engagement of the entire team in the care of the patient. Suggestions should be welcomed even if not taken.

ACCOUNTABILITY

Finally, a word about accountability; the movement toward a "system perspective " and focus on teamwork itself presents unique challenges, with regard to accountability. The "system perspective," recognizes that the operation of a system is critically influenced by the nature of the interactions among the components of the system. Therefore, understanding of the function and the faults demands a holistic approach. In this context, accountability for those functions and for those faults may well lie within the system itself. Furthermore, identification of those system issues is most effective in a "blame-free culture" in which errors are readily identified or confessed without fear of retribution. The individual should not be blamed for systems problems. Who then is accountable? Is the individual ever accountable? Is no one accountable?

The balance must be struck between appropriate accountability and absence of accountability. In an era before systems factors were acknowledged, it was clear who was "accountable" even if they were not to blame! With a systems focus there is the risk that "nobody is to blame" but the system. The alternative is collective accountability of the team.

A related concept is that of "Just Culture" in which we can satisfy the demands for accountability while also allowing for reporting and learning. It distinguishes between unintended errors and negligence, recklessness, and knowing rule violations. In another construct from James Reason's work, errors may be skill based (such as accidental slips or lapses) or mistakes (either rule-based or knowledge-based). More simply still, Charles Bosk distinguishes between technical and normative errors, the latter being regarded much more negatively in surgical culture. In practice, one may ask the question whether another well-intended, unimpaired individual under the same circumstances may have committed the same action—if so the individual should not be blamed.

SUMMARY

Patient safety can most productively be considered as the product of a reduction in preventable adverse events leading to patient harm. Not all adverse events are the consequence of errors, but many if not all preventable events are likely so. Accordingly, a focus on errors of all types can be expected to improve patient safety. Errors may be due to organizational/systems issues, device design issues, process design, or individual human performance including both technical and nontechnical matters. As leaders within our institutions, we should each contribute productively to improvements in systems of care. As innovators, we may be able to contribute to device design in collaboration with our industry partners.

Most immediately, however, each and every one of us can improve the performance of our teams. We are all humans and as such are subject to recognized human fallibilities. Our brains have evolved to accomplish remarkable tasks, and at the same time demonstrate remarkably consistent vulnerabilities. These limitations have been recognized in other industries and countermeasures developed,

which may be considered for modification and application to health care in general and surgery in particular. Optimal safety demands high team performance, which in turn depends upon effective leadership skills. These nontechnical skills can be learned. Social intelligence and emotional intelligence should be recognized as important skills and developed. Optimizing the performance of your team requires active effort and is a critical part of our jobs as leaders and care providers. We will provide the best outcomes to our patients through effective teamwork. This is the most direct path to transforming surgical practice into a high reliability endeavor.

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EDITOR'S COMMENTS

This chapter on Patient Safety by Dr. Sundt is a new addition to this book. We as surgeons have tended to base our results on the last best or worst case. Clearly, this is an inaccurate and inappropriate technique for improving performance. We have done better studying databases and looking at collective results.

Dr. Sundt has demonstrated much more expert approaches to patient safety and avoidance of error. In particular, he emphasizes a blame-free environment and a team approach to the patient. This is easier said than done. He discusses the approach of briefing and a discussion process that includes all members of the team. In addition, he presents the example of the person who attends a cardiac arrest and runs a code. One must be able to step away from the emotion and physical resuscitative actions to best reduce error and improve outcome. To say the least, this is a new area for all of us. We are being judged on patient safety by groups such as Leap Frog and Consumer Report. There is no doubt this will continue to be emphasized and improve as we learn the skill sets to truly deal with this.

ILK

41 Reoperative Cardiac Surgery—General Principles

Damien J. LaPar and Irving L. Kron

CARDIAC REOPERATION

Reoperations for cardiac surgery following prior sternotomy are associated with elevated morbidity and mortality. Major adverse events occurring in the intraoperative and perioperative setting are often predictable but may still occur despite careful attention and surgical detail. As cardiac reoperations are fundamentally different from routine, nonreoperative cases, the general approach to patient selection, preoperative assessment, and surgical technique should be modified from that commonly employed in the routine, nonreoperative setting. In fact, certain advantages may be derived from a protocol-driven approach to reoperations to improve surgical technique and patient outcomes. This chapter reviews general principles that apply to the preoperative and intraoperative management of patients undergoing cardiac reoperations following prior sternotomy.

INCIDENCE AND POSTOPERATIVE OUTCOMES OF REOPERATION

Performance of cardiac operations after prior sternotomy continues to confer an increased risk of operative mortality and morbidity. Recent trends indicate that the performance of cardiac reoperations has been affected by an increase in patient life expectancy and by improved patient outcomes following primary cardiac operations. While evidence demonstrates that the number of patients requiring reoperations for valvular disease has increased, those requiring reoperations for coronary disease has decreased due to improved secondary medical management and evolving surgical techniques. Mortality rates, nonetheless, remain elevated for a variety of cardiac reoperations, including those with prior coronary artery bypass grafting (CABG) and cardiac valve procedures. In a retrospective review of 946 cardiac reoperations performed at our institution from 1995 to 2010, the overall operative mortality was 8.2%. According to recent estimates from the Society of Thoracic Surgeons and other single institutions, mortality for reoperative aortic valve replacements (AVRs) range from 5% to 14%; however, we have recently demonstrated that improved outcomes for reoperative AVRs can be achieved and may, in fact, be comparable to that of primary operations (2.0% vs. 3.5%; $P = 0.65$). We believe that the improved outcomes for cardiac reoperations observed over time are due to enhanced approaches to patient assessment and selection as well as intraoperative management.

PREOPERATIVE PLANNING

Patient selection and assessment for cardiac surgery following previous sternotomy begins with a comprehensive history and physical exam. A thorough medical history should be conducted to include not only an extensive cardiovascular history but also a detailed history related to prior cardiac surgical interventions or surgical procedures within the thorax and mediastinum. Specifically, special attention should focus upon the type, date, and nature of prior cardiac operations, prior incisions and thoracic approach, and postoperative complications following prior cardiac operations, including respiratory failure, tracheostomy, and sternal wound infections. In addition, a prior history of mediastinal infections, previous chest irradiation, number of cardiotomies, prior pericardiectomy, number, conduit type, and location of previously performed CABGs, and prior implanted valve size and type should be considered. Previous operative notes must be carefully reviewed.

A complete physical exam should be performed. Particular attention should be paid to important findings related to a prior cardiac procedure. In addition to routine physical exam techniques, prior surgical scars on the chest should be assessed for location and extent of healing. Whether a prior cardiac procedure was performed through standard full or partial median sternotomy, thoracotomy, or other minimally invasive approaches often provides valuable insight into the degree of underlying scar tissue that may be encountered upon resternotomy and which may alter existing mediastinal anatomy. Degree of prior wound healing and overall appearance of prior scars may provide additional insight into the setting of prior wound infection, breakdown, or deep sternal wound infection. Potential graft conduit sites should also be considered during the performance of the physical exam for reoperative cases requiring myocardial revascularization. This assessment includes evaluation of both upper and lower extremity venous and arterial conduit sites, including the performance of bilateral Allen tests to assess for collateral flow in patients who may be candidates for radial artery CABGs. Similarly, peripheral vascular access sites for potential femoral or axially cardiopulmonary bypass cannulation should be assessed for patency, prior scars, and ease of access.

Cardiac Catheterization

Standard preoperative evaluation with routine catheterization is fundamental to success for cardiac reoperations. In addition to routine preoperative laboratory analysis, chest roentography, cardiac electrocardiography, and echocardiography, all patients undergoing cardiac reoperations in the setting of prior CABG should undergo coronary angiography within 3 to 6 months of surgery to assess for graft orientation, to determine graft patency, and to establish the contribution of each graft to myocardial perfusion. Coronary angiography also helps to determine the location and proximity of previously placed

bypass grafts in cases of graft adherence to the sternum. This information is crucial to avoid injury and to develop a preoperative plan for myocardial revascularization in the event of intraoperative injury. Timing of preoperative angiography is an issue that affects patient morbidity and mortality. Recent series have demonstrated that contrast loads provided within 24 hours of operation result in significantly higher rates of postoperative renal failure, translating into higher mortality rates and increased hospital lengths of stay and resource utilization. At our institution, we favor cardiac catheterization ≥48 hours prior to operation in order to limit renal dysfunction in the postoperative setting.

Radiographic Evaluation

Preoperative computed tomography (CT) should be performed as part of the evaluation of all patients undergoing cardiac reoperations. More specifically, computed tomography angiography (CTA) provides a more complete radiographic assessment of mediastinal anatomy, architecture, and location and status of prior CABG. CTA has many advantages over other imaging modalities due to its ability to provide cross-sectional imaging, which can often be used to provide three-dimensional reconstructions of the heart and mediastinum (Fig. 41.1). Preoperative information that can be obtained from CTA includes the proximity of sternal incision to various mediastinal structures such as the right ventricle, aorta, pulmonary artery, CABG, and other vascular structures crossing the mediastinum. Details related to aortic size and degree of aortic calcification also aid in cannulation and cross-clamping strategies

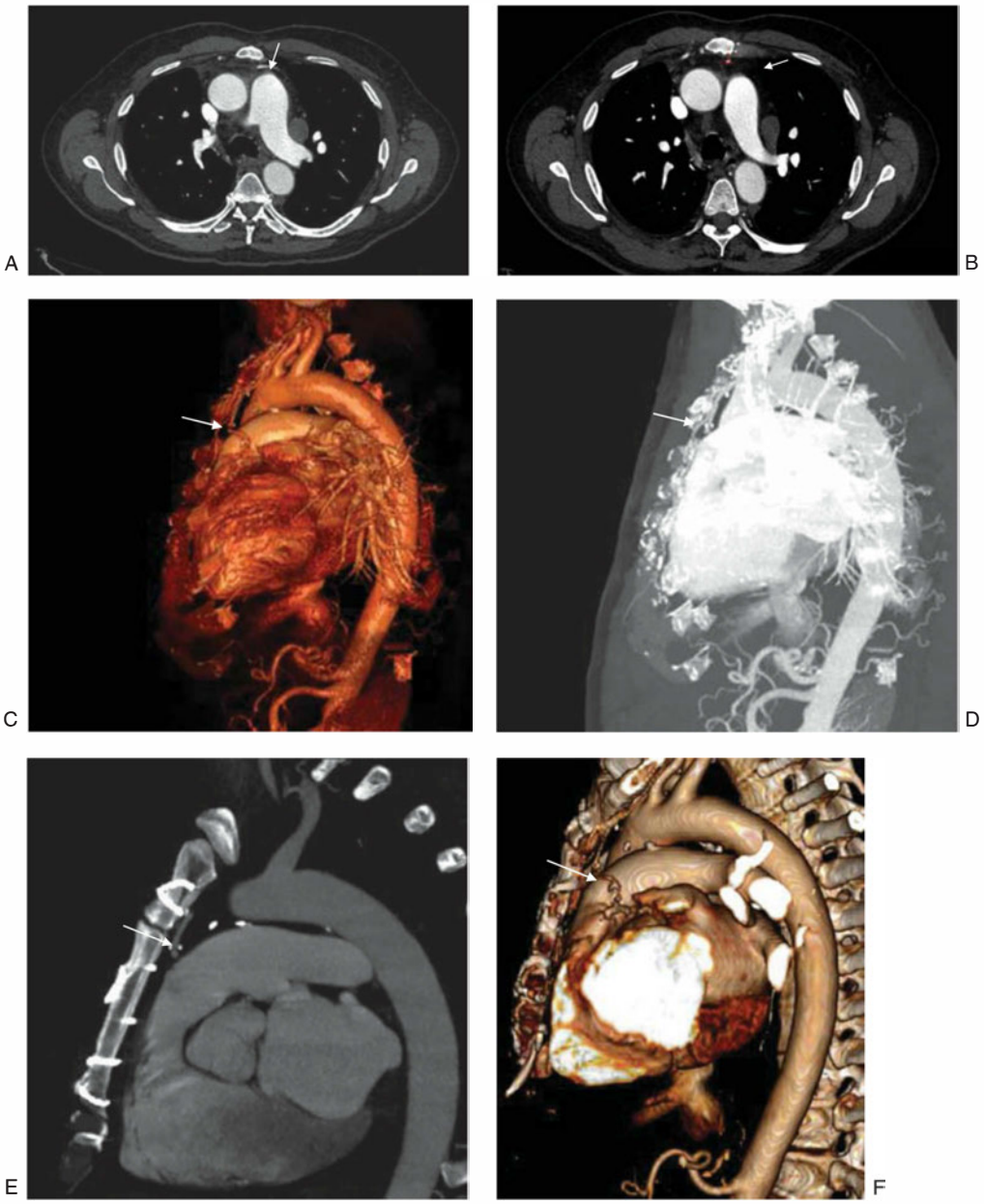


Fig. 41.1. Representative preoperative computed tomography angiography cross-sectional (**A** and **B**) imaging with three-dimensional reconstructions (**C**–**F**), demonstrating a patent left internal mammary artery coronary artery bypass graft (*arrows*) that is in close proximity (4 mm) to the retrosternum and is at high-risk for reentry injury upon resternotomy.

prior to incision. The use of prior synthetic materials or felt pledgets may also be suggested by the presence of dense adhesions visible on preoperative imaging that may ultimately distort retrosternal anatomy and place cardiac and vascular structures at increased risk for reentry or intraoperative injury. Alternatively, for patients with baseline renal insufficiency of allergies to intravenous contrast, noncontrast CT imaging or magnetic resonance imaging can be performed.

The utilization and benefits derived from preoperative CTA or CT for cardiac reoperations appears to be increasing. Cremer and colleagues demonstrated a 98% safe reentry rate and low injury rate (2%) in their study of patients undergoing reoperative CABG following assessment with preoperative CTA. Other series have reported on the role of preoperative CT imaging to develop alternate surgical strategies in up to 80% of cardiac reoperations. In one of the largest reported series to date, the benefits of establishing cardiopulmonary bypass via peripheral cannulation prior to sternotomy was demonstrated in the setting of dense retrosternal adhesions identified by preoperative CT. Most importantly, the use of preoperative CT imaging has been shown to reduce postoperative morbidity including reentry injury and postoperative stroke rates, and at our institution it remains routine practice to obtain and review preoperative CT imaging on all patients undergoing reoperations following prior sternotomy.

Other Considerations

Several other important considerations exist for patients undergoing reoperative cardiac surgery. Adequate peripheral intravenous and arterial access is required prior to induction of anesthesia. This often requires placement of two central venous catheters and two peripheral arterial lines for adequate monitoring and resuscitation in the event of a life-threatening reoperative injury. Choice of arterial line placement is important. If the radial artery is a potential CABG conduit, the contralateral radial artery should not be accessed for arterial line placement. Patients undergoing reoperations also have a higher prevalence of prior pacemaker and automatic implantable cadioverter-defibrillator devices (AICDs). Consequently, the placement of external defibrillator pads prior to induction is recommended, and the function and type of all implanted devices should be reviewed and interrogated prior to surgery. Finally, the treatment mode of all AICDs must be turned off prior to induction.

INTRAOPERATIVE CONSIDERATIONS

Intraoperative Patient Preparation

Intraoperative preparation of the reoperative cardiac surgery patient begins immediately upon the patient's arrival to the operating room. A quick review of preoperative imaging, studies, and cannulation strategy is paramount to operative success. We recommend that all the available preoperative cardiac catheterization and cross-sectional radiographic imaging are displayed in the operating room upon patient arrival. Next a reevaluation of all prior scars including primary incisions, prior drain sites, and graft conduit sites should be performed. Potential access sites for extrathoracic, alternative cardiopulmonary bypass cannulation (femoral and axillary) should be marked. At our institution, placement of femoral arterial and venous guidewires is performed for all cardiac reoperations in the event that alternative cannulation is required for cardiopulmonary bypass support. This is most commonly performed percutaneously using the Seldinger technique and soft-tipped wires. The wires are then secured appropriately to prevent migration or accidental removal. The patient should then be prepped and draped in the usual sterile fashion to the extent that all alternative, extrathoracic cannulation and potential graft conduit sites are included in the surgical field. At this point, all cardiopulmonary bypass lines should be brought to the surgical field, and the bypass pump should be primed and ready prior to sternotomy in the event that it becomes necessary to emergently initiate cardiopulmonary bypass.

Incision and Dissection

The choice of incision for reoperation is most appropriately left to the discretion and experience of the operating surgeon. For the majority of cardiac reoperations, standard median resternotomy is most appropriate. Alternatively, a thoracotomy may be used; however, this has been associated with higher risk of postoperative stroke and may limit the ability for myocardial revascularization. For resternotomy, the sternal skin incision and dissection of the sternum are carried out in the standard surgical manner. Prior sternal wires are then removed. We do not routinely recommend leaving wires in place to protect the mediastinum during division of the anterior and posterior tables of the sternum as this not only limits the ability to control an injury upon sternal reentry but also can result in multiple wire fragments that can prove injurious to underlying mediastinal structures.

Careful mediastinal dissection is fundamental to success during cardiac reoperations. The primary goal of mediastinal dissection is to liberate available cardiopulmonary bypass cannulation and appropriate aortic cross-clamp sites. While preoperative radiography often provides enough detail to develop an adequate cross-clamping strategy, occasionally intraoperative surveying of aortic architecture is required to minimize the disruption and embolization of aortic plaque and calcifications. Minimal manipulation and dissection of the aorta also reduces the likelihood of subadventitial dissection. Overall, the "golden rule" of reoperative cardiac dissection is that only areas necessary for the planned procedure should be dissected. During reoperation, it is customary at our institution to perform minimal mediastinal dissection, to practice a no-touch operative technique for prior CABGs, and to not routinely clamp the internal thoracic artery for cases following a prior CABG. Furthermore, for patients undergoing reoperations for isolated valve repair or replacements, it is not necessary to dissect out the left side of the heart, and it is acceptable to leave adherent pericardium to the right atrium to avoid injury if extensive adhesions are encountered. Other considerations include the use of sharp dissection around prior conduits and vessels to avoid thermal injury from electrocautery. Electrocautery can predispose to ventricular arrhythmias during mediastinal dissection. For difficult, high-risk reoperations, initiation of early cardiopulmonary bypass can also aid mediastinal dissection by decompressing the heart.

Cardiopulmonary Bypass and Cannulation

Cardiopulmonary bypass can be initiated either prior to or following resternotomy or thoracotomy, and the choice for either peripheral or central cardiopulmonary bypass cannulation is often dictated by the surgical risk of the case and the quality of vasculature available. While central bypass cannulation is often preferred, extensive calcification of the ascending aorta, a high number of prior central cannulations,

multiple patent bypass graft anastomoses to the aorta, presence of significant adhesions to the right atrium, or patients at high risk for catastrophic injury to mediastinal structures may require alternative cannulation strategies. In these situations, either axillary or femoral cannulation may be performed. For axillary cannulation, the axillary artery can be easily accessed via vascular cut down performed in the deltoid crease. Alternatively, the femoral artery may be accessed percutaneously using a Seldinger technique or through a femoral cut down technique in the inguinal crease. The choice of axillary versus femoral cannulation is often dictated by the preoperative assessment of individual patient vasculature. Axillary artery cannulation has been associated with reduced risk of limb ischemia and cerebrovascular complications compared with femoral access, and the femoral artery usually carries a higher atherosclerotic burden. Complications of axillary cannulations include brachial plexus injuries, and the most serious immediate complication of femoral access includes the risk of vascular perforation and retroperitoneal hemorrhage. Regardless of choice in peripheral arterial cannulation site, however, it is important to serially assess peripheral pulses following decannulation in order to address early limb ischemia and the potential for evolving vascular dissections and pseudoaneurysms.

The initiation of cardiopulmonary bypass should be dictated by operative risk. For most cardiac reoperations, cardiopulmonary bypass should commence following completion of all mediastinal dissection in order to reduce the overall pump times and avoid any added morbidity associated with prolonged cardiopulmonary bypass. However, in certain high-risk cases, the early initiation of cardiopulmonary bypass has important advantages and implications. For cases with anticipated difficult mediastinal dissections or those carrying a high risk of reentry cardiac or aortic injury, the initiation of bypass prior to resternotomy or thoracotomy is advised. In these scenarios, bypass not only decompresses the heart to aid resternotomy and dissection, but it also reduces the impact of myocardial ischemia resulting from injury to or division of prior patent coronary bypass grafts as well as the risk of significant bleeding from reentry injuries. Thus, both intraoperative and postoperative blood product transfusion requirements are often minimized and total cardiopulmonary bypass times may be unaffected, or in some cases even reduced, by the avoidance of additive surgical repair of reentry injuries or myocardial revascularization. It is our practice to initiate cardiopulmonary bypass prior to sternotomy for cases with a high potential for aortic injury.

A PROTOCOL-DRIVEN APPROACH TO CARDIAC REOPERATIONS

A routine protocol-driven approach to preoperative assessment and intraoperative management of patients undergoing cardiac reoperations can prove useful to reduce patient morbidity and mortality. At our institution, we have incorporated a protocolized approach to preoperative planning and operative techniques employed for all cardiac reoperations. Our protocol utilizes routine preoperative CTA or noncontrast CT to evaluate mediastinal anatomy and location and status of bypass grafts, the placement of soft-tipped arterial and venous femoral guidewires in all reoperative patients prior to induction of anesthesia, minimal mediastinal and cardiac dissection, no dissection of prior internal mammary artery or other bypass grafts, and early initiation of cardiopulmonary bypass prior to sternotomy for cases with a high potential for resternotomy or aortic injury. As a result, among 946 patients undergoing reoperations from 1995 through 2010 at our institution, the adoption of this routine, protocol-driven approach has resulted in a significantly decreased incidence of re-entry injuries (2.6% vs. 9.6%; $P < 0.001$) and lower operative mortality (5.8% vs. 9.6%; $P = 0.04$) compared with reoperations performed without the routine performance of these measures. In an era of increasingly complex cardiac reoperations, we suggest the adoption of a similar protocolized approach to the preoperative and intraoperative management of patients undergoing reoperative cardiac surgery.

CONCLUSION

Cardiac reoperations following prior sternotomy are becoming increasingly performed within the United States and worldwide as patient life expectancy increases and patient outcomes following cardiac surgery continue to improve. General principles guiding the approach to cardiac reoperations should incorporate many unique measures for adequate preoperative patient assessment and intraoperative management. Adoption of a routine protocol-driven approach to cardiac reoperations should be considered to reduce intraoperative injury and postoperative outcomes.

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EDITOR'S COMMENTS

Drs. LaPar and Kron have given a comprehensive review of the general principles to be considered in reoperative adult cardiac surgery. Emphasis is placed, as it should be, on proper preoperative evaluation and planning to include a CT angiogram of the chest and coronary angiogram to determine the location and status of important structures such as bypass conduits. The strategies employed at their institution are those in common practice.

The authors emphasize the increased mortality rate for reoperative surgery, which should be discussed with the patient and family. The operative strategy they employ is to place arterial and femoral lines in case of the need for emergent peripheral bypass in the event of cardiac injury on reentry. In cases at high risk for such injury (such as a patent internal mammary artery graft in a precarious position), consideration should be given to surgical exposure of the femoral artery and vein prior to sternotomy to decrease the time required to initiate cardiopulmonary bypass. In low-risk cases, surgical exposure of the femoral vessels may be avoided. Compatible blood for transfusion should be available and in the operating room at the time of sternal reentry in the event of serious bleeding. The cardiopulmonary bypass machine should be primed and lines in place on the sterile field.

The authors describe the removal of the sternal wires in order to eliminate the risk of fragments injuring surrounding structures and reduce the time needed for a more emergent reentry. Our group and many others follow this practice but many groups also leave the wires in place as a safety measure to prevent entry of the saw into the right ventricle. There are groups that use a standard reciprocating sternal saw for redo-sternotomy, although the vast majority use an oscillating bone saw. Having the "feel" of the location of the posterior plate is easier with the hands in a relaxed neutral position and the use of lower saw speed. Additionally, heavy scissors may be used to divide the posterior table under direct vision.

Once the sternum is reopened, the emphasis should be on formulating a clear strategy, usually starting with the goals of aortic and right atrial cannulation for initiation of cardiopulmonary bypass. The obtuse margin of the right heart is often free of adhesions once the initial top layer is entered. One should not randomly dissect the heart but instead have a clear series of stepwise goals in mind: right atrium clear enough to cannulate, aorta able to cannulated, aortic cross-clamp able to be placed, cardioplegia and vent lines able to be placed, caval tapes when needed, and so on.

Reoperative cardiac surgery requires a "bag of tricks" to fit various patients. Whether it be alternative cannulation sites (i.e., axillary cannulation), alternative cardioplegia cannulation (percutaneous retrograde cardioplegia coronary sinus catheter placement by anesthesia), or even circulatory arrest prior to sternotomy as in the case of large arterial pseudoaneurysms, these procedures should be performed by surgeons with significant experience in reoperative procedures.

There is no substitute for careful and thoughtful preoperative strategic planning for these operations.

TSG and LRK

42 Mitral Valve Repair

Lawrence H. Cohn and R. Scott McClure

Mitral valve repair is one of the most important technical advances in cardiac surgery over the past 35 years and is the treatment of choice for most conditions of the mitral valve, most notably the prolapsed valve with myxomatous degeneration. This chapter outlines the current concepts and reparative techniques for the myxomatous degenerated mitral valve. Elements of the pathoanatomy and principles of repair will be reviewed followed by a detailed explanation of the surgical techniques used and refined at the Brigham and Women's Hospital by the lead author over a 40-year career. Select adjunct techniques popularized at other major centers throughout the world are also discussed.

DEVELOPMENT OF TECHNIQUES OF MITRAL VALVE REPAIR

Elliott Carr Cutler at the Peter Bent Brigham Hospital did the first successful operative repair of the mitral valve in 1923. The surgery was performed on a 12-year-old girl diagnosed with severe rheumatic mitral stenosis. Using a neurosurgical tenotomy knife via a median sternotomy, Cutler, boldly and ingeniously inserted the knife transapically into the left ventricle of the beating heart, incising each fibrotic commissure in a blinded manner. Isolated successes with this operation were rare. It was not until after World War II, when Harken in Boston and Bailey in Philadelphia popularized the method of closed mitral commissurotomy for mitral stenosis using the finger fracture technique, that reasonable surgical successes were achieved for the management of what was inevitably a terminal disease. For the next 25 years, until the early 1970s, the closed mitral commissurotomy procedure was one of the most widely utilized techniques to treat and relieve mitral stenosis.

Similar to severe mitral stenosis, severe mitral regurgitation was also recognized early on to incur adverse outcomes prompting similar though more sporadic attempts at surgical correction. Yet unlike the surgical successes garnered with mitral stenosis, mitral regurgitation generated little in the way of innovative progress or reproducible surgical techniques until the development of the heart–lung machine in the mid-1950s. In the late 1950s and early 1960s, several suture annuloplasty techniques were developed to relieve mitral regurgitation with a modicum of success. One of the more successful attempts to relieve mitral regurgitation at the time was a technique developed by Harold Kay in Los Angeles who obliterated the commissures using a sequence of mattress sutures. A circular technique developed by Paneth and DeVega, involving a circumferential suture around the annulus, also aided in the early attempts at mitral valve repair. In the early 1960s, Dwight McGoon proposed a reparative technique for the degenerative prolapsed mitral valve with ruptured chordae that involved resecting a small segment of the mitral leaflet from the mitral valve. He rationalized that small resections of the mitral leaflet would prevent overall distortion of the valve and in the few cases where it was trialed the technique proved successful. However, with the advent of prosthetic devices to replace the mitral valve in the early 1960s, efforts to repair the mitral valve quickly waned with mitral valve replacement emerging as the preferred treatment choice for mitral regurgitation at the majority of institutions.

One of the ironies of the early 1960s was that most surgeons of the era believed it prudent to cut the papillary muscle and all chordal attachments to the mitral leaflets to ensure an anatomically secure mitral valve replacement. It is now recognized that utilizing chordal sparing techniques to preserve the subvalvular apparatus and annular-papillary continuity during mitral valve replacement retains left ventricular geometry. This portends to improve left ventricular mechanics and left ventricular function. Late cases of severe left ventricular decompensation and cardiomyopathy began to emerge in patients having undergone corrective surgery for mitral regurgitation where the chordal attachments were excised. Poor outcomes in these early experiences with corrective surgery for mitral regurgitation caused many surgeons to retreat from such operations. And cardiologists for their part became reluctant to refer such patients to surgery as a treatment option. A lack of appreciation for the importance of the subvalvular integrity when performing mitral surgery, which resulted in suboptimal outcomes, in addition to the prevailing yet erroneous "pop off" valve theory of the time, sustained the reluctance to intervene on the regurgitant mitral valve for many years. The "pop off" theory was the misconception that if a patient had progressed to having left ventricular dysfunction secondary to mitral regurgitation, the regurgitant valve was now a necessary evil, as it became a "pop off" valve into the left atrium that relieved stress on the failing heart. Any effort to correct the mitral valve in this situation was presumed to impart further stress on the already failing ventricle and thus cause deleterious effects. These two key misconceptions, removal of the subvalvular apparatus and the fallacious "pop off" construct, stymied advancements in surgery to treat mitral regurgitation well into the 1980s.

The late 1970s and early 1980s heralded a new development in mitral valve reconstructive surgery with the invention of prosthetic remodeling annuloplasty rings. The works of Alain Carpentier and Carlos Duran, pioneering surgeons independently credited with first implanting prosthetic rings during mitral valve reconstructive operations to refashion and remodel the mitral annulus, were seminal developments. The concept of implanting a prosthetic remodeling annuloplasty ring enabled mitral valve reparative procedures to be performed in a reproducible manner by surgeons worldwide. Along with the introduction of prosthetic rings to aid in annular remodeling, these two surgeons are credited with outlining the basic principles of mitral valve repair for the generations of cardiac surgeons to follow. In 1983, as an honored lecturer of the American Association for Thoracic Surgeons, Dr. Carpentier gave the now classic lecture entitled, "The French Correction," outlining the basic principles of repair for the prolapsed mitral valve and emphasized the importance of an annuloplasty ring. This lecture helped to stimulate the thought process of the cardiac surgery community regarding the surgical treatment of patients with mitral regurgitation. A number of surgeons have subsequently developed rings of different caliber, consistency, and shape. Though the multitude of rings now available each profess their advantages over one another, the basic fundamental principles to mitral valve repair remain faithful to the initial tenets outlined by Carpentier and Duran, with a key tenet being the integral importance of an annuloplasty ring to remodel the annulus for long-term success. A commitment to annuloplasty ring implantation, more so than the type of ring implanted, is a far more important factor for successful outcomes.

PRINCIPLES OF MITRAL VALVE REPAIR

The principles of repair for the mitral valve have evolved with experience for each of the four etiologies of mitral regurgitation: rheumatic, endocarditic, ischemic, and myxomatous. Regardless of the etiology, the principles are basically the same: (1) create apposition of the anterior and posterior leaflets during systole, (2) increase valve mobility, (3) prevent valve stenosis, (4) reduce annular dilatation, (5) remodel the annulus, (6) prevent systolic anterior motion (SAM) of the mitral valve into the left ventricular outflow tract causing obstruction. Whereas the focus of this chapter is primarily the myxomatous mitral valve, other chapters of this textbook very explicitly outline reparative techniques for rheumatic, ischemic, and endocarditic mitral valve disease.

REPARATIVE TECHNIQUES FOR THE MYXOMATOUS DEGENERATED MITRAL VALVE

The following principles of mitral valve repair for degenerative mitral valve disease are now commonly accepted: (1) apposition of the posterior and anterior leaflets during systole, (2) reduced height of the posterior leaflet such that the prolapsing segment of the posterior leaflet is obliterated or reduced causing the overall height of the leaflet to be shortened, (3) stabilization of the anterior leaflet by repairing and replacing ruptured chords or shortening very elongated chords, (4) remodeling the annulus, which is distorted and distended using an annuloplasty ring. The annuloplasty ring may be a full encircling ring or a partial C-band, either rigid or flexible.

Conventional terminology used to describe the normal segments of the posterior and anterior mitral valve leaflets are depicted in Figure 42.1A. The surgical anatomy of the mitral valve and adjacent structures are illustrated in Figure 42.1B.

Apposition of the anterior and posterior leaflets in systole is fundamental to providing a durable mitral valve repair. The optimum level at which the leaflet apposition creates a tight coaptation point preventing any leakage across the valve can be determined by the "Carpentier" reference point in unaffected areas of the posterior leaflet in comparison to the anterior leaflet. Once this reference point has been identified, adjacent segments of the mitral leaflets where the reference point has not been adhered to, denotes those areas of the valve

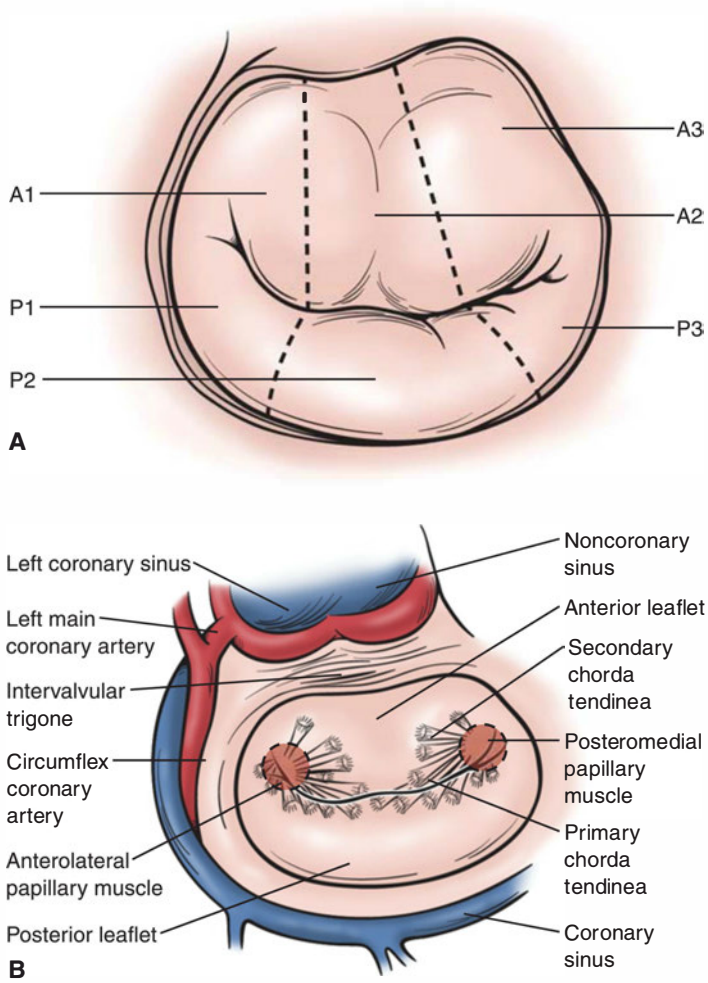


Fig. 42.1. (**A**) Anterior and posterior segments of the mitral valve. The normal partitions of the anterior and posterior leaflets of the mitral valve are illustrated. The leaflets appear in their closed position obscuring the valve orifice. Conventional terminology, used to describe and measure optimal points of repair, designates the anterior leaflet "A" and the posterior leaflet "P." The leaflets are then divided into three segments designated as 1, 2, and 3 moving *left* to *right* from the anterolateral commissure toward the posteriomedial commissure. (**B**) Surgical anatomy of the mitral valve. Thorough familiarity with the surgical anatomy of the mitral valve is key to avoiding injury to the coronary vasculature and other surrounding structures during reparative procedures.

where alterations to the height of the leaflets need to occur so as to restore a single uniform plane of apposition. Typically, the surgeon will reduce the height of the posterior leaflet letting it serve as a "doorstop" for the anterior leaflet; the "height" refers to the distance from the base of the leaflet at the annulus to the leading edge of the leaflet at the free margin. Re-creating a uniform plane of apposition across the entire mitral valve eradicates the persistent leak and results in successful repair.

MITRAL VALVE REPAIR TECHNIQUES FOR ISOLATED POSTERIOR LEAFLET PROLAPSE

In patients with myxomatous disease of the mitral valve, more than 80% of the patients have pathoanatomy requiring repair of the posterior leaflet, predominantly P2, having either markedly elongated chords or actual rupture and flail chordae (Fig. 42.2A). There are many different techniques to repair this defect. Our preference is a limited resection of the flail segment, removing the minimal number of adjacent chordae and only as much of the supporting structures of the posterior leaflet as is necessary. A silk suture is placed at the middle of the offending segment and the area is excised in a trapezoidal shape, with the narrowest portion of the trapezoid at the annulus, to ensure maximum preservation of adjacent chordae. Once the flail segment is excised, there is a "drawing down" of the adjacent parts of P2 with two running 4-0 monofilament sutures, bringing the two remaining parts of the leaflet together (Fig. 42.2B–E). This is a "modified" leaflet advancement technique promoted by the lead author. The classic leaflet advancement technique popularized by Carpentier is shown in Figure 42.3. The drawback here is that one must incise the posterior leaflet completely off the annulus and then re-suture the leaflet back onto the annulus. As the two segments of the posterior leaflet are re-sutured, they are advanced medially toward the midpoint of the posterior annulus such that the two leaflet segments meet in the middle, reducing leaflet height in the process. Complete removal of the leaflet from the posterior annulus can be a daunting task for the less experienced surgeon. The "modified" advancement technique achieves the same goal as the classic Carpentier technique yet is more simplistic, less time-consuming, and more easily taught and adopted by cardiac surgeons with a more modest mitral valve case volume.

With either of these techniques, again, approximate the residual posterior leaflet segments to the annulus using two running 4-0 monofilament sutures advanced medially toward the center of the annulus. Then, with these sutures held on tension approximate the two residual leaflet segments to one another. With a double-armed 4-0-monofilament suture, begin at the leading edge of the leaflets and tie four knots such that equal lengths of the suture reside on each side of the knot. The two lengths of the suture are then run back toward the annulus suturing the leaflet edges together and tying a knot with the adjacent advancement sutures previously left untied and held on tension. The adjacent leaflet segments are brought together with a double over-and-over technique. From the leading edge of the leaflet, the first suture is a running mattress and the second suture is a simple over-and-over running stitch to prevent any chance of dehiscence. We do not use pledgets of any kind on the mitral valve leaflets, as they tend to produce scarring and may provide a site for potential thromboemboli. Once the posterior leaflet is refashioned, the valve is then tested for competency. Even without a supporting ring the valve is usually competent in 90% of patients at this point. An encircling annuloplasty ring or partial C-band is then fixated to the annulus to complete the repair. Of note, though we advocate the use of the "modified" advancement technique for its simplicity, in the case of true Barlow's syndrome where all segments of the posterior leaflet are elongated, the classic Carpentier technique is required (Fig. 42.3), as the "drawing down" of P2 will not adequately reduce the height of the posterior leaflet sufficiently in this situation.

"Leaflet resection" is the time-honored traditional approach to mitral valve repair with isolated myxomatous posterior leaflet pathology. More recently, however, select surgical groups have endorsed a philosophy of "leaflet preservation." This alternative approach to repairing the myxomatous prolapsing posterior leaflet implores maneuvers to preserve the excess tissue as opposed to resecting it. Polytetrafluoroethylene chordae, measured to the appropriate length, are anchored to the papillary muscle and sutured to the posterior leaflet. These strategically placed "neochords" reduce the height of the posterior leaflet and restore the line of apposition with the anterior leaflet, heeding to the same basic principles followed for "leaflet resection" techniques. Historically, the biggest challenge to using neochords has been constructing chords of the appropriate length. The work of Friedrich Mohr in Leipzig, Germany, with the description of the Loop technique, using an intraoperative caliper to facilitate the construction of looped chords of precise length, has removed much of this guesswork and is credited for the increased popularity of this approach to posterior leaflet repair (Fig. 42.4).

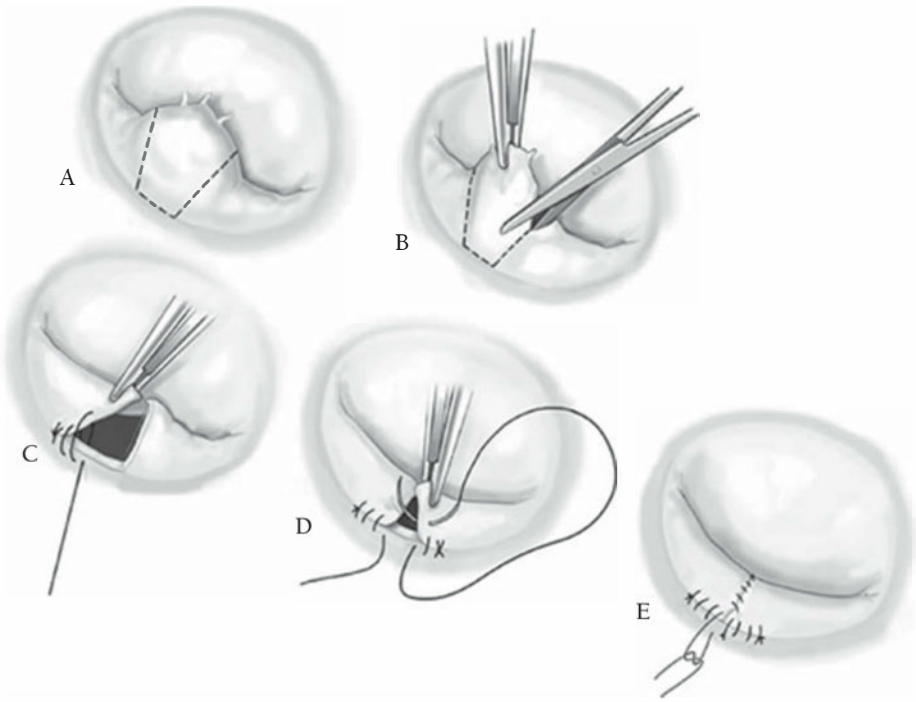


Fig. 42.2. (**A–E**) Repair of the myxomatous mitral valve using the modified leaflet advancement technique. The figure illustrates the repair of a valve with ruptured chordae underlying the middle segment (P2) of the posterior leaflet.

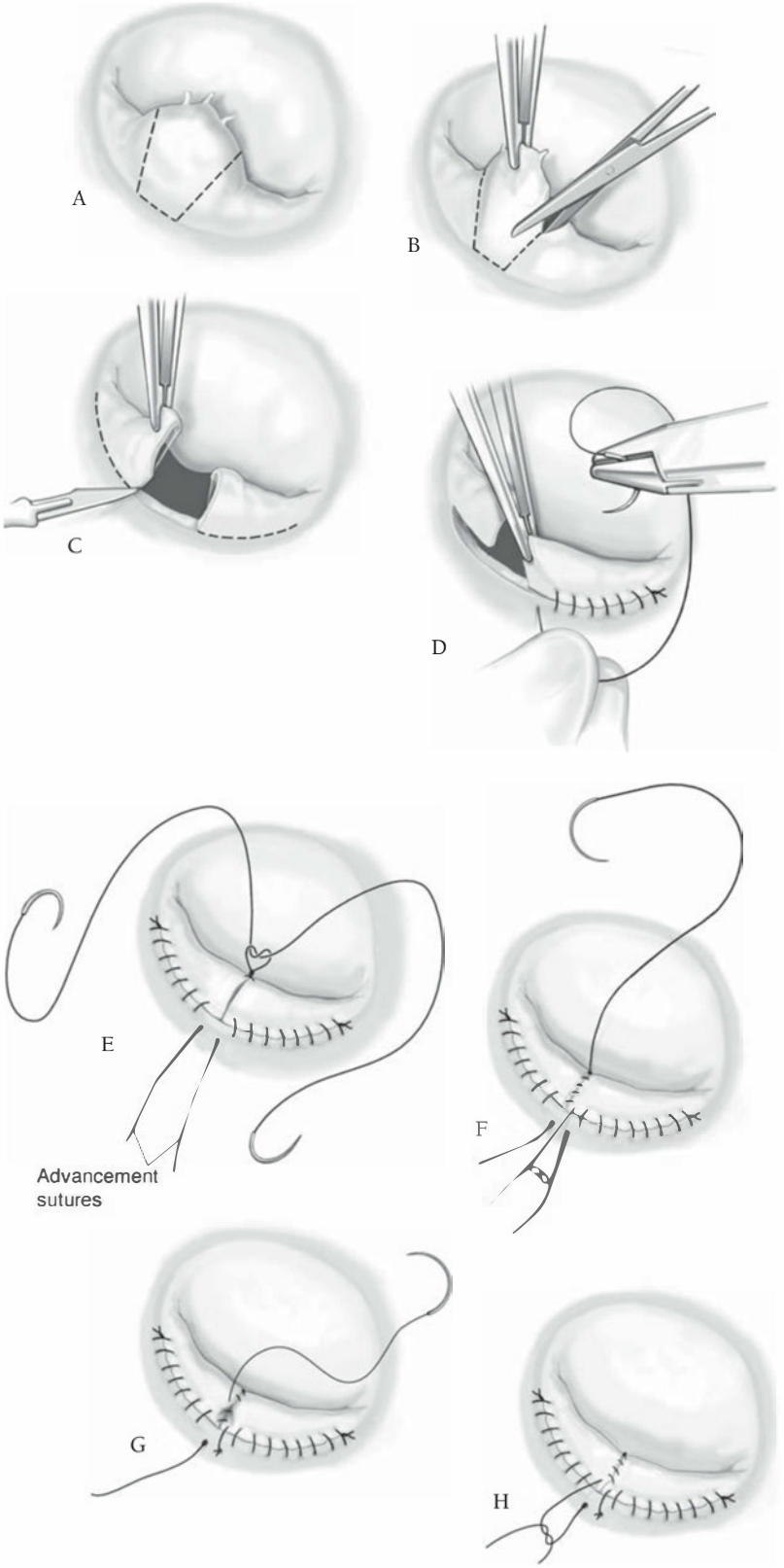


Fig. 42.3. (**A–H**) Mitral valve repair by means of the classic leaflet advancement technique described by Carpentier. This technique is required to repair extreme cases of Barlow syndrome.

An additional "leaflet preservation" technique advocated by the lead author and reserved for posterior leaflet prolapse *without ruptured chordae* is to simply fold over the redundant section of the posterior leaflet. A large needle double-arm 4-0 monofilament stitch inserted at the leading edge of the elongated segment is brought underneath the leaflet and directed back toward the annulus. The suture emerges at the annular attachment site and is securely tied in place. This simple technique, known as a "folding" or foldoplasty (Fig. 42.5), reduces the height of the posterior mitral leaflet by half. A single stitch is often all that is required; however, an additional two or three stitches placed in a similar manner may be necessary. An annuloplasty ring is then placed and a competent valve is achieved without any leaflet resection following a rather quick operation. This technique has been validated with published results to document its medium and long-term efficacy. Regardless of which technique one chooses, several simplified approaches are now available to reduce the posterior leaflet to normal height.

SIZING AND FIXATION OF THE PROSTHETIC ANNULOPLASTY RING

As previously mentioned, despite the apparent integrity of the valve after repairing the posterior mitral leaflet, because of the presence of annular distortion that will eventually lead to mitral valve leakage, the long-term competency of the valve repair is significantly enhanced with the placement of an annuloplasty ring. Sizing, placement, and fixation of the ring are key elements to remodeling the mitral annulus. As noted earlier, though there are many different rings, in our experience the most important aspect of the ring implantation is to find the size and shape that accomplishes the physiologic purposes of stabilizing the annulus. The keys to appropriate ring selection are shown in Figure 42.6. The importance of annuloplasty ring size and annuloplasty suture placement to properly fixate the ring cannot be overstated. An important aspect of ring placement is the identification of the trigone of the mitral valve anterior leaflet, which inextricably is the underside of the noncoronary leaflet of the aortic valve. This intertrigonal distance is extremely important to secure ring fixation. Whatever the type of the ring used, it *must be anchored to the trigonal area, not to the commissural area.* A C-band such as the Cosgrove–Edwards (Edwards Lifesciences Irvine, CA), Medtronic Future GC (Medtronic, Inc., Minneapolis, MN), or the partial Duran ring (Medtronic, Inc., Minneapolis, MN), have all been acceptable choices in our practice provided the anchoring sutures are properly placed in the trigonal area as shown. The commissural areas are mistaken for the trigones and used inappropriately as the anchor point for ring fixation in a surprising number of cases, resulting in early referral for reoperation.

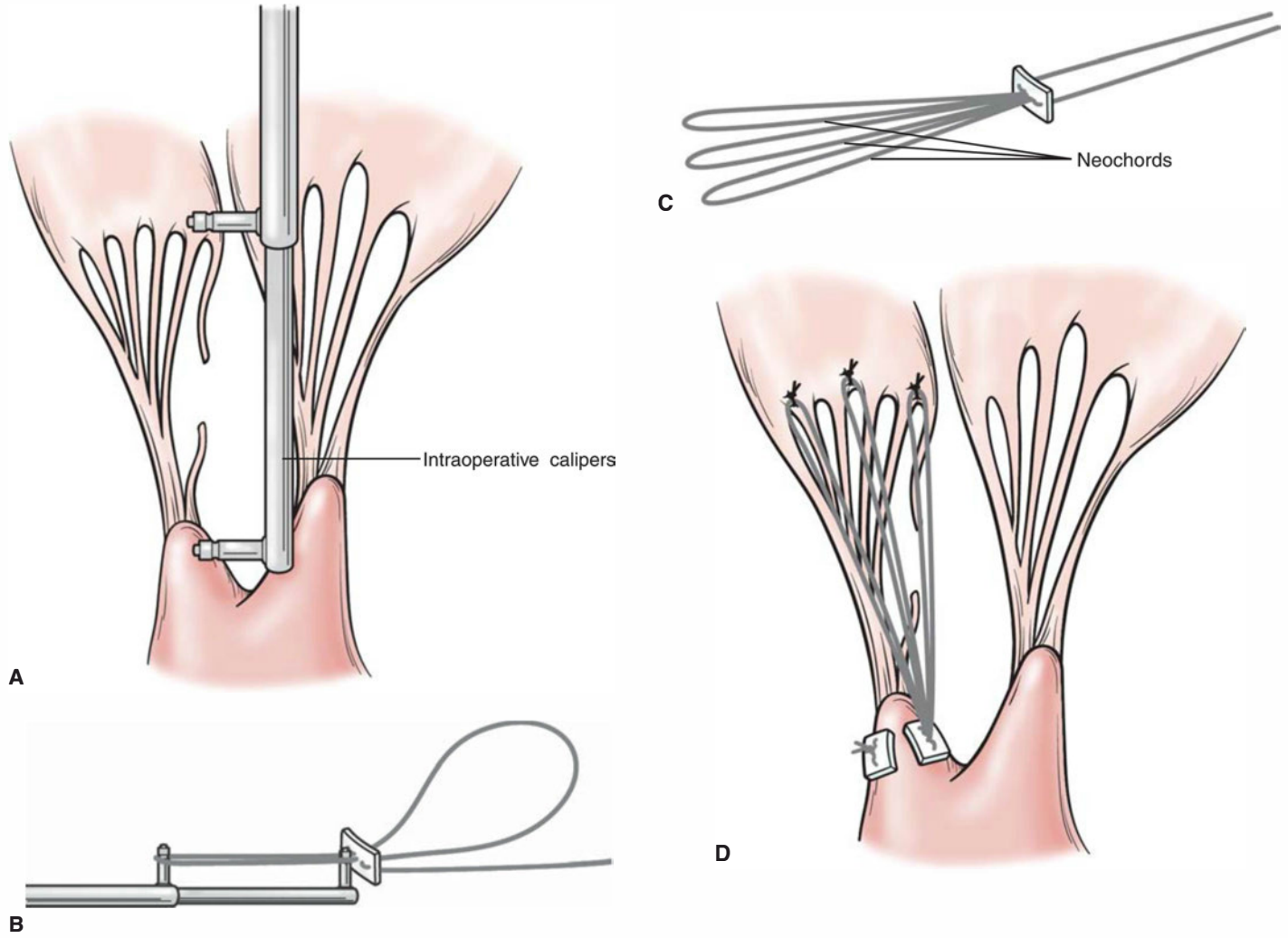


Fig. 42.4. (**A–D**) Polytetrafluoroethylene chord insertion for mitral valve repair employing the Loop technique. Intraoperative calipers facilitate the construction of "premeasured" chords of precise length. The neochords are then sutured to the papillary muscles and affixed to the free edge of the prolapsing leaflet of interest.

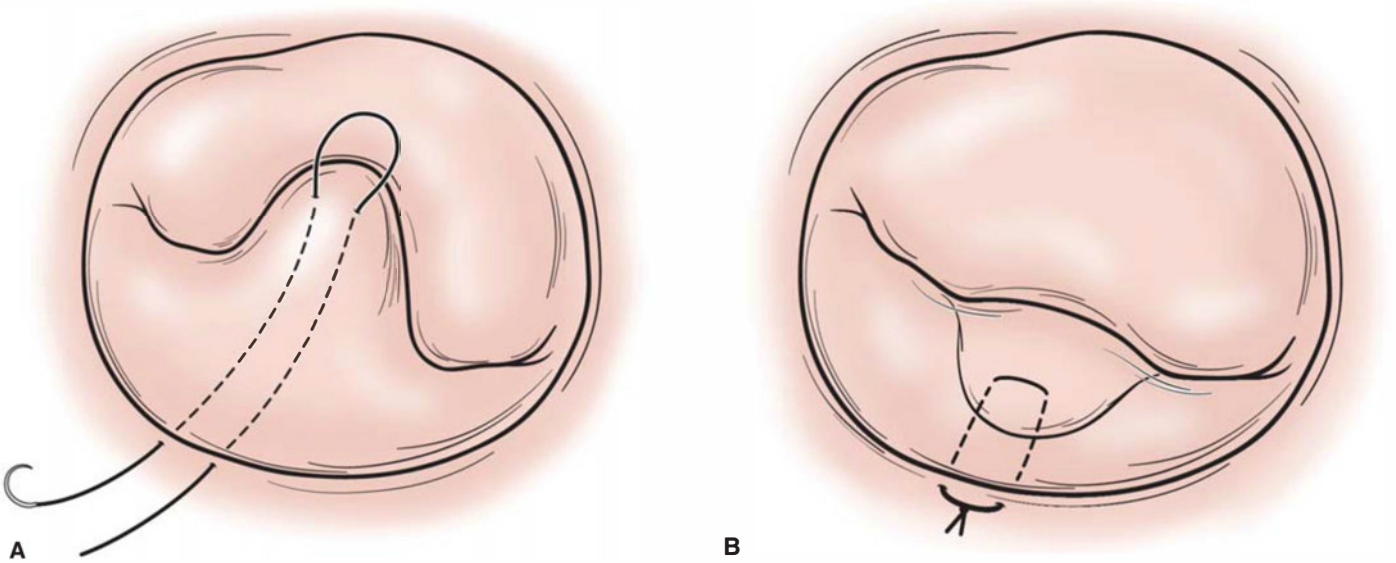


Fig. 42.5. (**A** and **B**) Foldoplasty. A simple reparative technique that can be used when there is a prolapsing segment of redundant posterior leaflet and no ruptured chordae. A double-arm 4-0 monofilament suture is passed through the leading edge of the area in question and brought back underneath the leaflet to re-emerge at the annulus. The suture is then tied in a simple mattress configuration.

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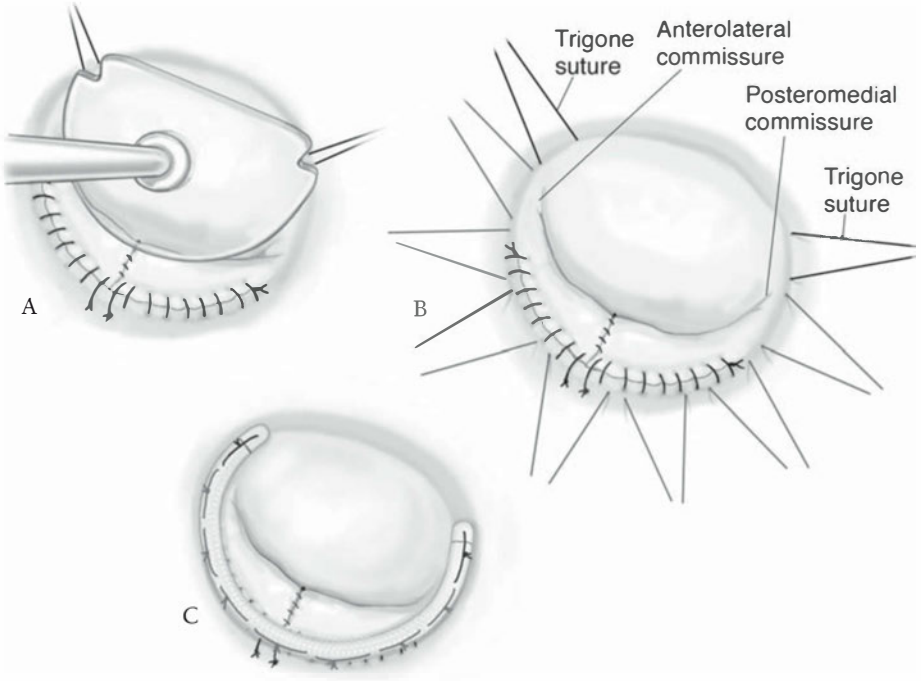


Fig. 42.6. Sizing and placement of the prosthetic annular ring. (**A**) A ring sizer is used to mark suture placement within the intertrigonal space. Nine to 12 sutures are generally sufficient to encircle even the most dilated annular ring. (**B**) The intertrigonal space is not prone to consequential degrees of dilation, which makes either a full encircling or partial C-ring suitable for repair. (**C**) U stitches are placed at the ends of the partial C-ring well in the trigonal area.

We place U stitches in the ends of the C-ring in the trigonal area well above the commissures as shown in Figure 42.6C. Rings can be implanted with running sutures; however, we prefer to use interrupted multifilament sutures with an emphasis on deep annular stitches that go into the ventricle and out again. In general, 9 to 12 sutures are sufficient to encircle ones prosthesis of choice for even the most dilated annulus. The sizing of the ring has been a source of confusion for many surgeons. The sizing is accomplished by measuring the length of the anterior leaflet. The intertrigonal distance can be very misleading, particularly in some patients in whom the anterior leaflet is extremely long but the intertrigonal distance is relatively short. Several reoperative referrals from other centers to the lead author have stemmed from the fact that the original rings were placed on the basis of the intertrigonal distance. This inevitably causes marked SAM because the ring is too small to accommodate the excess anterior leaflet tissue.

With the aid of transesophageal echocardiography, the length of the anterior leaflet can be measured and the ring size approximated before a surgical incision has even occurred. The measurement for the large myxomatous valve is usually at least 34 mm and in many situations will be as large as 45 mm. This is to emphasize the point that the larger sizes of the different annuloplasty rings are likely indicated when presented with myxomatous degenerated mitral valve in need of repair. Moreover, if for whatever reason one is unsure as to what size of ring to implant, it is generally better to upsize the ring rather than downsize the ring, as again, this will minimize the possibility of SAM of the anterior leaflet. Once the ring has been properly sized, sutures are placed in a mattress manner to fixate the ring to the annulus. The ring is then lowered to the annulus and the sutures are tied and cut. Importantly, many rings come with mandrels, which should be removed before the final three or four sutures are tied near one commissure or the other. This prevents unnecessary tension on the final sutures and protects against tissue tearing in the trigonal area. The valve is then tested. If the valve is competent the left atrium is closed, intracardiac air is evacuated with the intraoperative assistance of transesophageal echocardiography, and the patient is weaned from cardiopulmonary bypass.

VARIATIONS ON A THEME

There are a number of variations to the classic mitral valve repair techniques that should be in every surgeon's armamentarium when considering mitral valve repair—specifically, (1) commissural prolapse, (2) cleft closures, (3) sub-leaflet calcification, and (4) intra-leaflet calcification.

Commissural prolapse with or without ruptured chords is one of the more simplistic defects to repair (Fig. 42.7). A leaflet segment with a ruptured chord at either the anterior lateral or posterior medial commissure can simply be obliterated without resection. The annulus of these patients is always so enlarged that a simple 4-0 monofilament mattress suture will obliterate the commissure and correct any leak resulting from the prolapsing chords at the site of interest. No further reparative techniques are necessary except for the annuloplasty ring. The area within the valve is so large that this obliteration makes little difference. It is a very quick repair and one that can be done with complete alacrity. In addition, a cleft in the posterior leaflet may sometimes be exaggerated when a ring is placed in a valve that is hugely dilated, some being as large as 60 mm. Testing of the valve will show a leak at such clefts. A simple 4-0 suture to elevate this area and keep it at the same plane as the adjacent segment is more than enough to extirpate this problem. This is usually accomplished after the ring is in place and valve competency has been tested.

Subannular calcium of the posterior leaflet can be a vexing problem. Carpentier has advocated a rather radical approach to this issue recommending complete removal of the calcified bar. This can lead to disassociation between the left atrium and the left ventricle requiring the difficult undertaking of reconstructing the atrioventricular groove. In the majority of cases, we would contend that this approach is unnecessarily aggressive and potentially dangerous. With careful debridement, one can remove an appropriate amount of calcium to allow a flexible repair. We have found subtotal excision of calcium to be satisfactory for leaflet coaptation in the majority of situations. In very rare circumstances, one may encounter a patient with ruptured chords of the posterior medial leaflet in the setting of an exorbitantly diffuse degree of calcification encasing the entire posterior annulus. A corrective repair is still feasible in this setting; however, the extensive calcium may prohibit insertion of an annuloplasty ring. Yet, if the calcium is so rigid as to prevent the insertion of an annuloplasty ring, it has inherently become a fixated structure that is unlikely to succumb to any further annular dilatation. This makes the role of a ring less vital to successful repair in this unique situation.

Finally, if there is calcium within the integrity of the mitral valve leaflets themselves, we believe that this is not likely to be repaired satisfactorily unless one can

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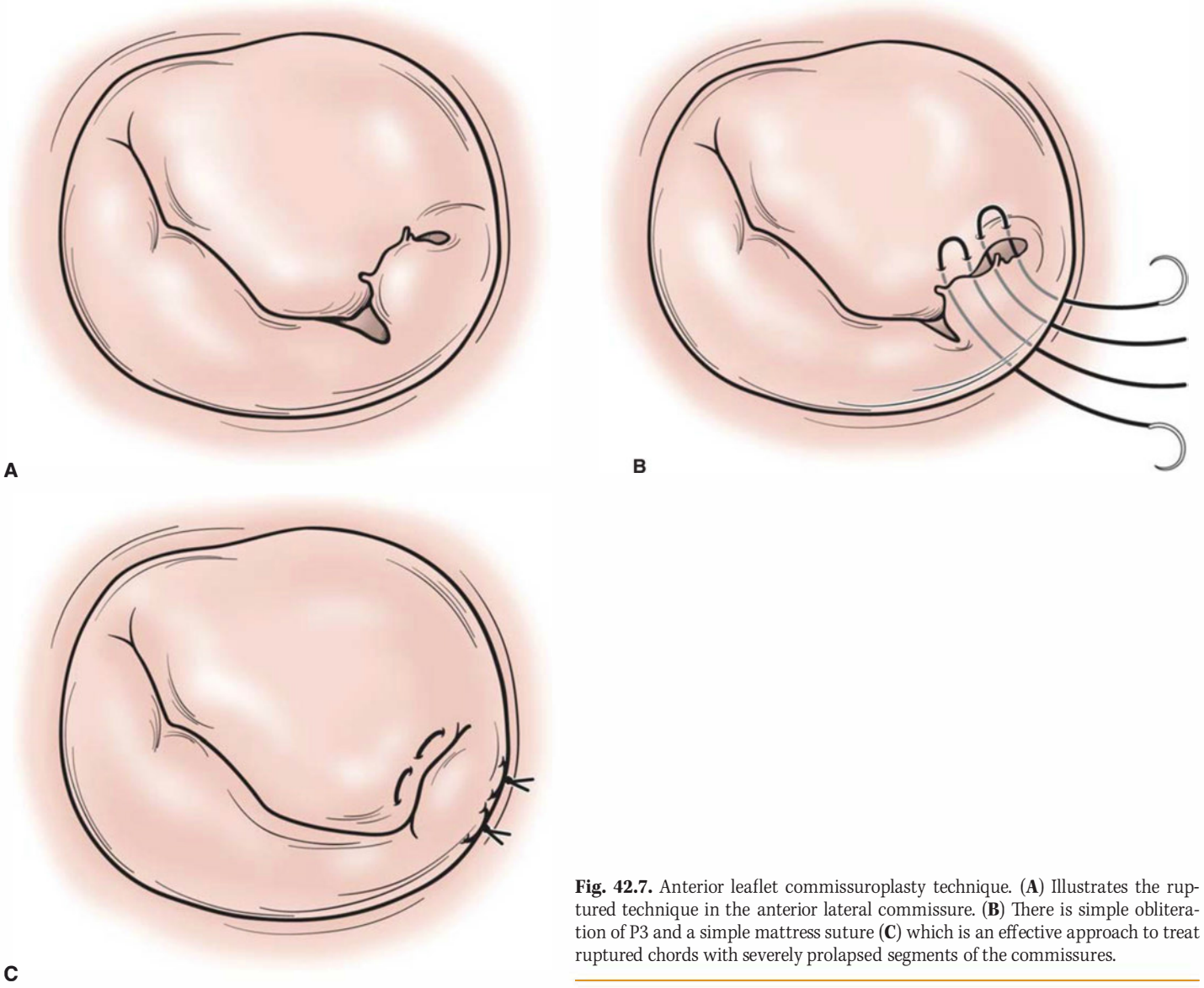


Fig. 42.7. Anterior leaflet commissuroplasty technique. (**A**) Illustrates the ruptured technique in the anterior lateral commissure. (**B**) There is simple obliteration of P3 and a simple mattress suture (**C**) which is an effective approach to treat ruptured chords with severely prolapsed segments of the commissures.

debride the calcium from the anterior leaflet. Calcified leaflets do not hold up in the long-term and are often the cause of reoperations. At times, a history of endocarditis will present as small residual calcific nodules embedded within the leaflets. These nodules are an exception to the rule as they are generally debrided with ease and leaflet flexibility restored. Otherwise, if leaflet flexibility cannot be retained it is our experience that achieving a long-lasting competent mitral valve repair will be difficult.

MITRAL VALVE REPAIR TECHNIQUES IN THE PRESENCE OF ANTERIOR LEAFLET PATHOLOGY

Many patients are referred to the surgeon with the diagnosis of "bileaflet prolapse" raising concern about reparability. There is no question that true anterior leaflet pathology makes a repair more complicated, but, in actual fact, many experienced mitral valve surgeons have noted that once a good posterior leaflet repair is done and the remodeling ring applied, "prolapse of the anterior leaflet" is not a factor. This can be determined by looking at chordal length upon examination of the valve. Thus, our approach to anterior leaflet pathology, unless ruptured chords of the anterior leaflet are present, is to perform the posterior leaflet repair, insert the appropriate sized annuloplasty ring, and only then see what else must be done. The expression "less is more" is never better attributed to an operation than here because in the vast majority of patients, the prolapse in the anterior leaflet is rather modest and it is the posterior leaflet pathology that is the predominant cause for the regurgitant valve.

When it is absolutely required to lower the height of the anterior leaflet or there are ruptured anterior leaflet chordae, it can be accomplished in a few relatively straightforward ways. The first option is to insert polytetrafluoroethylene chords into the anterior leaflet in the same manner as was discussed above for posterior leaflet pathology. Though the use of neochords has gained increased popularity for posterior pathology in recent years due to the simplification of neochord measurements, neochords were first introduced more than 20 years ago by Tirone David in Toronto and Robert Frater in New York. To be precise, in the original descriptions, neochord placement was primarily promoted as a corrective technique for anterior leaflet pathology. The more traditional approach

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to neochord placement is to determine neochord length "after" attachment of the neochord to the papillary muscle. Once the neochord is secured to the papillary muscle with a double-arm suture, the two ends of the suture are brought up through the underside of the leaflet at the leading edge left untied. This is different than the Loop technique, where neochord length has been predetermined and constructed at a side table "before" attachment to the papillary muscle based on caliper measurements. With the traditional technique, the neochord is set to the appropriate length by tying the sutures at the leading edge of the leaflet with the left ventricle fully distended with saline solution. The surgeon then determines the height at which to secure the knot at the leading edge, which then sets neochord height, so that the knot occurs at the exact point where coaptation with the posterior leaflet is achieved (Fig. 42.8). Another technique first popularized by Duran is the "flip over" technique (Fig. 42.9). Here the surgeon incises a noninvolved segment of posterior leaflet with normal chordae and brings this across to the anterior leaflet of the mitral valve. This healthy posterior mitral leaflet segment is "flipped over" onto the anterior leaflet and becomes the supporting chordae for the affected area of the anterior leaflet. This can work well in experienced hands but we have not used this technique because we prefer not to disfigure a normal posterior leaflet. A final option that may be considered is triangular resection of the anterior leaflet (Fig. 42.10). Though we do not use this technique, as it is our general contention that anterior leaflet resection should be avoided, heeding the advice of Dr. Carpentier who professed that anterior leaflet resection can distort the leaflet and impede coaptation, we acknowledge the work of Irving Kron, coeditor of this textbook. Dr. Kron has utilized triangular resection for both anterior and posterior pathologies, and in his experience results have been excellent with this technique.

SYSTOLIC ANTERIOR MOTION

SAM of the mitral valve is a concern for every patient undergoing mitral valve repair. SAM is the obstruction of the left ventricular outflow tract caused by the paradoxical movement of a large anterior leaflet into the outflow tract during systole. A thickened ventricular septum can also be a contributing factor. This was seen much more frequently with rigid encircling rings or when the height of the posterior leaflet was inadequately reduced. In most instances, the chordal structure of the mitral valve is able to prevent SAM. Nevertheless, the incidence of SAM is still between 2% and 16% in reported series and a problem that all surgeons are likely to encounter at some point. Cardiac anesthesiologist and/or cardiologists who do intraoperative echocardiography are quite expert at predicting which patients are at risk for SAM even before an incision is made. Echocardiographic parameters predictive of SAM that the surgeon should be aware of are (1) prerepair anterior to posterior leaflet ratio <1.4 and (2) the so-called C-sept distance <2.6 cm ("C-sept" refers to the distance from the line of mitral leaflet coaptation to the interventricular septum). In addition, excessive leaflet slack postrepair should alert the surgeon to a high likelihood of SAM. This occurs when there is an enormously elongated and enlarged anterior leaflet with a profound dilation of the annulus. When attempting to size the annuloplasty ring to the anterior leaflet, the annuloplasty ring necessary to properly fan out the leaflet will be far larger than the largest commercially available rings, as the maximum size available in North America is only 38 to 40 mm.

A technique that we have found very effective in this scenario, where a large myxomatous mitral valve makes SAM a major concern, is the edge-to-edge technique (Fig. 42.11). The edge-to-edge repair in this situation is very helpful when all other attempts to reduce SAM have failed. In our practice, this entails a figure-of-eight stitch with a multifilament suture (for strength), as opposed to a monofilament suture. We have also used this technique in patients with severely reduced left ventricular function and prolapsed valves with ruptured chords of the anterior leaflet. This is a short operation and a real-time saving maneuver. We do this after the ring is in place and all of the reparative techniques are done so as to align the anterior leaflet in the right position to be approximated. We also use a caliper to make sure that the two side orifices are large enough to allow a relatively normal pressure gradient between the left atrium and left ventricle. Ideally, the two orifices should have a 2 cm × 2 cm area of clearance. This technique works very well for patient with a prolapsed myxomatous valve but we never use this technique in patients with ischemic mitral valve disease or any other condition where the valve is relatively small due to concerns for mitral stenosis.

Mid- and late-term studies from our institution with the edge-to-edge repair when used solely for the prevention of SAM in large prolapsed valves have shown excellent repair durability, SAM prevention to be maintained, and no significant stenosis

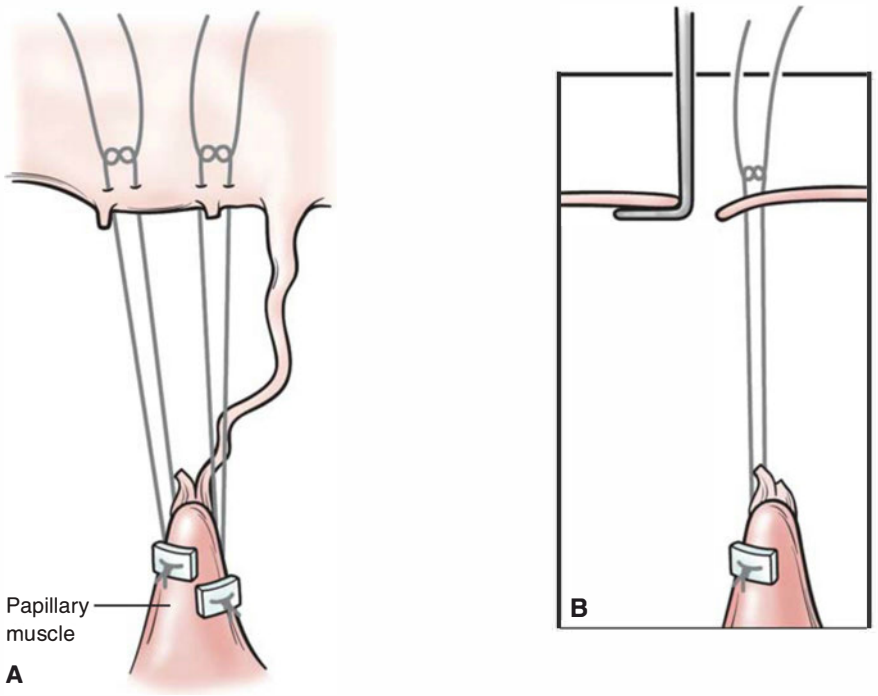


Fig. 42.8. Classic technique for neochord insertion. (**A**) The artificial chords are first attached to the papillary muscle. (**B**) Maneuvers to find the line of apposition with the adjacent leaflet are employed. Once the line of apposition is determined, the artificial chords are then tied down at the free edge of the leaflet at the appropriate height.

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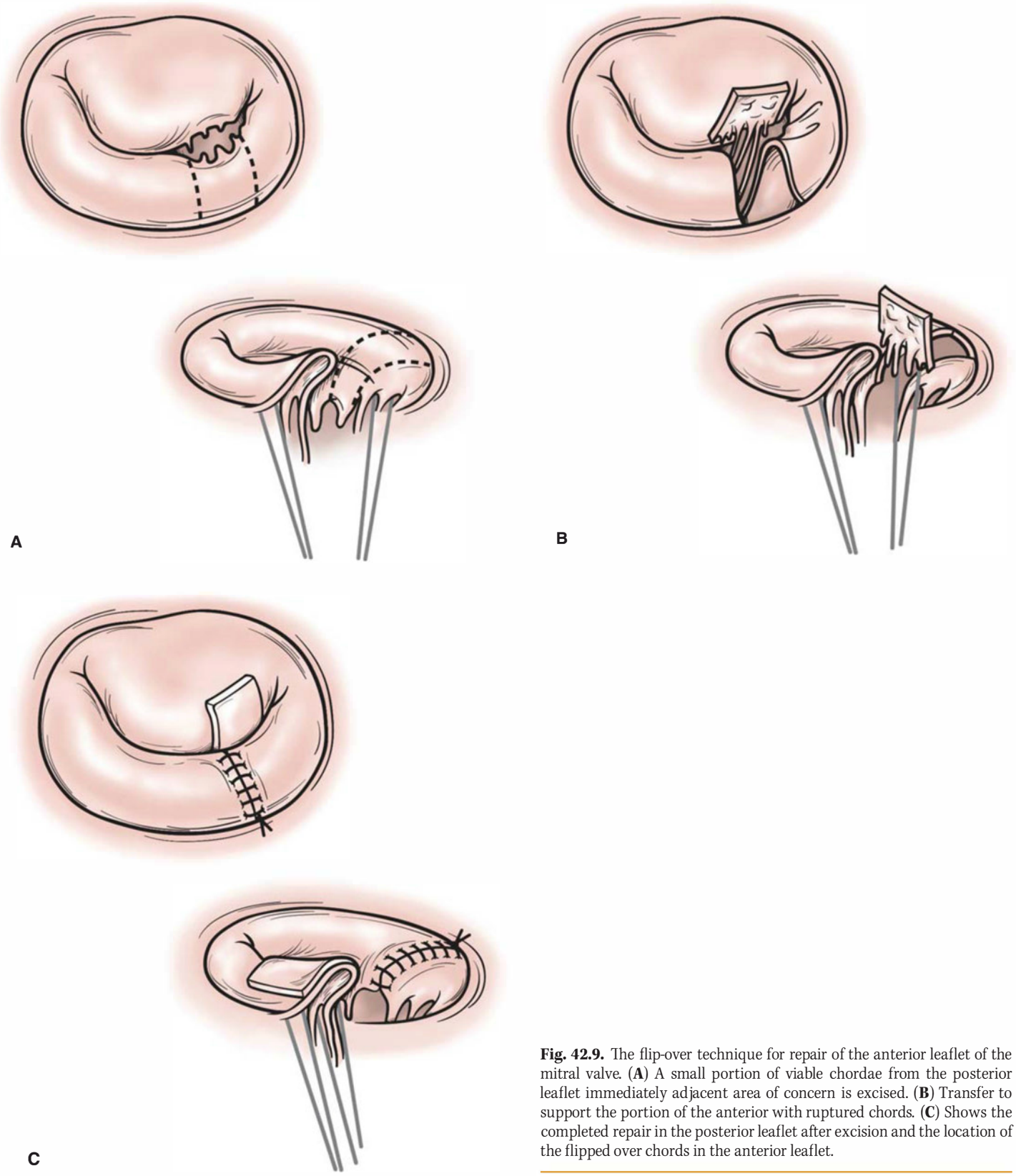


Fig. 42.9. The flip-over technique for repair of the anterior leaflet of the mitral valve. (**A**) A small portion of viable chordae from the posterior leaflet immediately adjacent area of concern is excised. (**B**) Transfer to support the portion of the anterior with ruptured chords. (**C**) Shows the completed repair in the posterior leaflet after excision and the location of the flipped over chords in the anterior leaflet.

to have occurred. If, however, the edge-to-edge repair is used as a "bail-out" in conjunction with other surgical techniques for complex forms of mitral repair, where the initial techniques did not achieve an adequate repair necessitating the edge-to-edge at the end of the procedure, the results are not nearly as good and the potential for recurrent regurgitation or stenosis is fairly significant. Triangular resection is another choice that may be used for SAM prevention though we feel the edge-to-edge repair to be a better option.

Finally, the ultimate judgment regarding the success of one's mitral valve repair is

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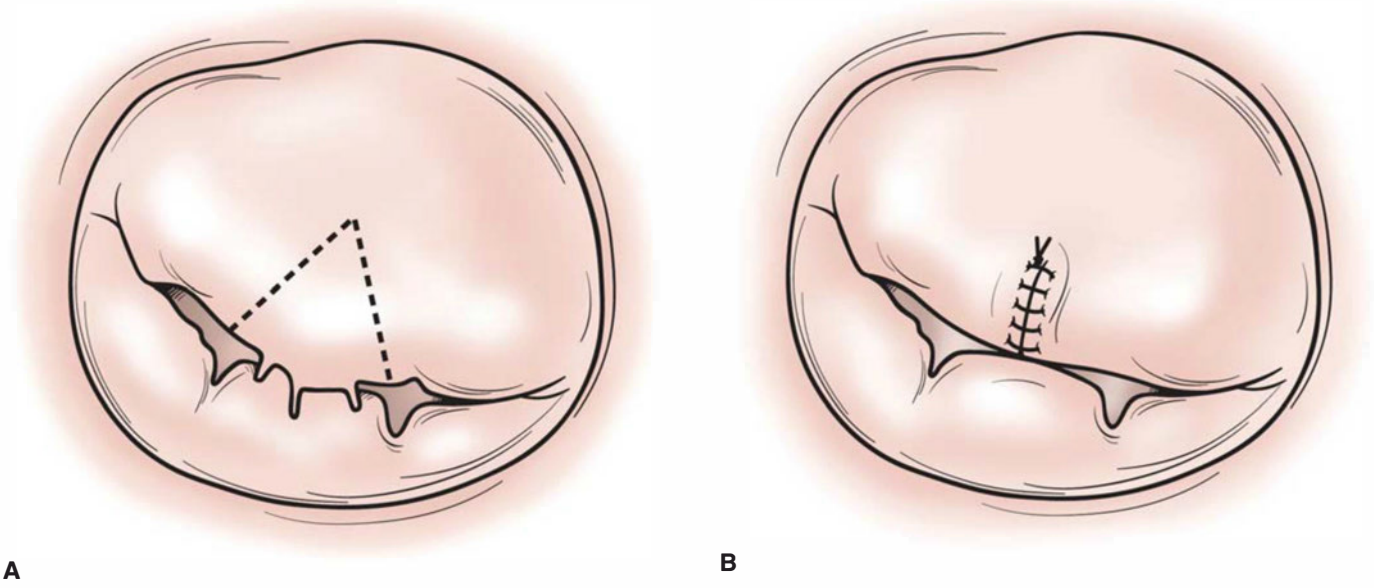


Fig. 42.10. (**A**) Shows a triangular resection of the anterior mitral leaflet and (**B**) shows the completed suturing of a small segment of the anterior leaflet. Triangular resection is an additional reparative option for isolated segmental prolapse both the anterior and posterior leaflets.

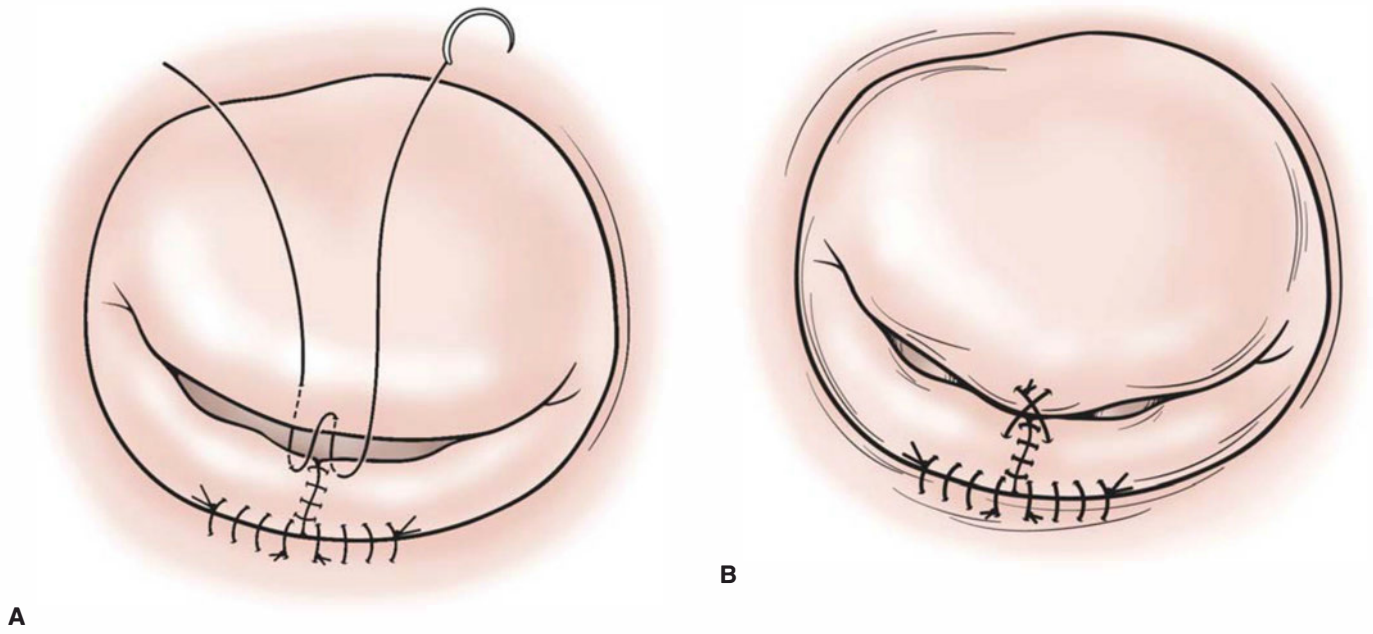


Fig. 42.11. The edge to edge technique. This is an adjunctive technique utilizing patients with a high propensity for systolic anterior motion based on pre-bypass echocardiographic findings or post-repair intra-operative systolic anterior motion. (**A**) Replacement of a figure of 8 suture at the mid-portion of the anterior and posterior leaflet and (**B**) shows the figure of 8 multi-filament suture after tying and knot cutting to create a double orifice valve.

determined using the postcardiopulmonary bypass transesophageal echocardiogram. Transesophageal echocardiography is absolutely necessary in every patient undergoing mitral valve surgery. In patients with contraindications to transesophageal probe insertion, we employ epicardial echocardiography with close collaboration from anesthesia. Should there be moderate residual mitral regurgitation this has to be addressed. After a suitable period of reperfusion, one should go back on cardiopulmonary bypass and look at the repaired valve to determine where one can improve the coaptation line and reduce the residual mitral regurgitation. There are now 20- and 30-year outcome data available for reparative procedures on the mitral valve from select centers including our own, the Cleveland Clinic and Dr. Carpentier's personal series. The probability of a re-repair at 10 years should be $<10\%$ and at 20 years $<15\%$. Mastery of the simplistic techniques that have been discussed thus far will provide the surgeon with an armamentarium necessary to repair the vast majority of myxomatous degenerated valves that present in clinical practice. These simple techniques provide excellent long-term results. Though patients will present with the same underlying degenerative process the exact mechanism for their regurgitation will vary. The ability to incorporate the many options discussed above allows the surgeon flexibility

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so as to treat every patient individually and adjust the corrective repair to fit each individual patient's needs.

MITRAL VALVE RE-REPAIR OF THE MYXOMATOUS MITRAL VALVE

Re-repair of the myxomatous mitral valve after a previous repair can be accomplished. A review of the re-repairable cases of the mitral valve from our series of 2000 mitral valve repairs has indicated five important causes of repair failure: (1) progression of disease with rupture of additional chords within the mitral valve apparatus unrelated to the previous repair, (2) failure to lower the height of the posterior leaflet, (3) failure to lower the height of the anterior leaflet so bulging of the anterior leaflet persists, (4) failure to use an annuloplasty ring, and (5) insertion of too small an annuloplasty ring largely due to inappropriate sizing with intertrigonal distance utilized instead of using the size of the anterior leaflet. This latter issue is probably the most common cause of mitral valve re-repair in our experience. When this is the case, where at the original valve repair the valve was large and an undersized ring was used related to the intertrigonal distance, simply inserting a new larger ring often resolves the problem. If, however, there is an accumulation of calcium on the valve, either because it was left untreated from the initial operation or it has accrued with time, valve replacement is required.

CONCLUDING REMARKS

The clinical outcomes of the lead author's 40-year experience with mitral valve repair for all etiologies of mitral disease have been summarized in a recent presentation to the American Association for Thoracic Surgery. In comparison to other etiologic conditions for mitral regurgitation, surgical outcomes with myxomatous mitral valve repair are unmatched. Postrepair survival endures the longest, and the incident of recurrent regurgitation is the lowest (Fig. 42.12). Though these differences are in many respects attributable to the different patient populations and their varied comorbidities, with regard to the pathology, the results are a shining testament to the effectiveness of reparative surgery for myxomatous disease.

To close, though we have expounded on the techniques to repair the myxomatous valve once it has been exposed, it is worth mentioning the varied surgical options available to gain exposure. Most important, a full median sternotomy for maximal exposure is always an appropriate option. A technically skilled repair is paramount and takes precedence above anything else. However, it is noteworthy to mention that many of these mitral valve repair techniques are amenable to less invasive surgical methods. Our personal preference is a lower hemisternotomy. Late clinical and echocardiographic data from our institution with this approach have validated its safety and efficacy. Still, other groups have preferentially used a right minithoracotomy, thoracoscopic techniques, or robotic assistance, with equally impressive results. Regardless of which approach one chooses, adhering to the simple techniques discussed in this chapter will go far to ensure excellent outcomes in clinical practice.

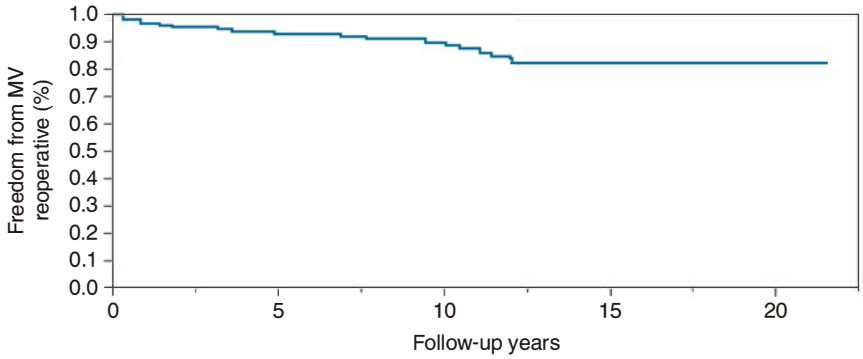


Fig. 42.12. Durability of the repaired myxomatous mitral valve. Actuarial freedom from mitral valve reoperation after repair for myxomatous mitral valve disease to 22 years postoperatively. Freedom from reoperation at 10 and 20 years is 90% and 82%, respectively. (From DiBardino DJ, ElBardissi AW, McClure RS, et al. Four decades of experience with mitral valve repair: analysis of differential indications, technical evolution and long-term outcome. J Thorac Cardiovasc Surg 2010;139:76-83, with permission from Elsevier.)

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EDITOR'S COMMENTS

Dr. Cohn and Dr. McClure's chapter on mitral valve repair is about as comprehensive approach as one can imagine. Dr. Cohn's 40-year experience with mitral valve disease certainly gives him expertise to have dealt with every type of mitral valve pathology. All the basic principles are discussed and the authors have alluded that many techniques truly work. The most important concept the authors have emphasized is that mitral valves must be repaired. I have heard Dr. Cohn state multiple times that sliding plasty may have been the worst thing to happen to mitral valve repair. The techniques obviously are excellent but the perceived complexity pushes inexperienced surgeons to end up replacing the valve.

I agree with most of the techniques elaborated on by the authors. I think the limited resection of the posterior leaflet mitral valve is an outstanding technique and allows for confident mitral valve repair in the majority of cases. I very much admire the approach to cords then there is rupture involving the same sections of anterior and posterior leaflets. Dr. Cohn's technique of simply closing this adjoining sections of leaflet with a suture adding a ring does work and makes a complex situation fairly simple. In our practice, we have not been as concerned with the height of the posterior leaflet. This is a classic Carpentier concept. I think the solution for this problem is just to use bigger rings. Since we have begun using larger rings in our practice, we have almost never seen SAM that was a major issue in the early days of mitral valve repair. Finally, I agree with the concept of either limited resection or chordal replacement for annular leaflet pathology. The bottom line is that all degenerative disease is repairable and can be repaired. The most certain way to harm a patient in the long term is to replace a repairable valve.

ILK

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43 Mitral Valve Repair—Robotic Minimally Invasive

Alan P. Kypson, L. Wiley Nifong, and W. Randolph Chitwood

INTRODUCTION

Bailey (1951), Harken (1953), Davila (1954), and Glover (1956) made early attempts to repair mitral valves. These methods are usually related to epicardial reduction of the mitral annulus size or reducing a regurgitant orifice by "plugging" with a cone-shaped prosthesis. Later, Kay (1961), Woller (1962), and Reed (1965) developed intracardiac suture annuloplasties to reduce the posterior mitral annulus length, thereby improving leaflet coaptation. McGoon (1960) performed the first repairs for ruptured chordae tendineae, and Austin (1965) first replaced a mitral valve for a papillary muscle rupture. The advent of mechanical valves impeded the progress of mitral valve repair surgery until the early 1970s, when Carpentier and Duran pioneered novel reconstructive techniques. Despite mitral valve repair successes in Europe, skepticism pervaded U.S. surgeons until Carpentier showed in 1983 excellent long-term repair results. These outcomes were based on his elucidation of the functional anatomy in insufficient mitral valves and the development of reproducible repair methods. By the late 1980s, numerous published reports from other centers validated the long-term durability of repairs, freedom from anticoagulation, and decreased mortality compared with mitral replacements.

Mitral valve surgery has been performed traditionally through a median sternotomy, which provides generous operative exposure and global cardiac access. However, during the past 15 years, improvements in instruments and endoscopes, as well as patient demand, have resulted in a substantial increase in minimally invasive general and subspecialty surgical procedures. Endovascular port-access technology first enabled less invasive surgical approaches to mitral valve surgery that avoided a median sternotomy. Advances in closed-chest cardiopulmonary bypass and myocardial protection, as well as intracardiac visualization, instrumentation, and robotic telemanipulation all have hastened a shift toward efficient and safe minimally endoscopic cardiac surgery. Today, mitral valve surgery, done through small incisions using robotic assistance, has become standard practice for an increasing number of cardiac surgeons and centers.

ROBOTIC TECHNOLOGY

Computer-assisted robotic cardiac surgery platforms have been developed to facilitate surgeon hand motions in limited operating spaces. These devices offer advantages of improved access, magnified vision, and stabilized instrument implementation. Six degrees of freedom are required to allow any free orientation of instruments in any space. Endoscopic instrumentation with four degrees of freedom reduces the dexterity significantly that is needed for delicate cardiac surgical procedures. Furthermore, the loss of depth perception with two-dimensional video monitors further increases operative difficulty. Computer-enhanced (robotic) instrumentation systems overcome these and other limitations. In fact, their impact has been so great that a paradigm shift for both the patient and the surgeon has changed dramatically.

Robotic systems consist of telemanipulators with end-effectors, or microinstruments, being controlled remotely from a console. The da Vinci™S system (Intuitive Surgical, Mountain View, CA) is composed of a surgeon console, an instrument cart with four integrated arms, and a visioning platform (Fig. 43.1A and 43.1B).The operative console is removed physically from the patient and allows the surgeon to sit comfortably, immersed into the operative field using high-definition three-dimensional imaging. Digital images are translated to analog natural depth perception with high-power magnification (10×). Finger and wrist movements are registered through sensors and translated into motion-scaled tremor-free movements that avoid the fulcrum effect and instrument shaft shear forces, which are common to endoscopic instrumentation. Wrist-like articulations at the ends of microinstruments bring the pivoting action to the plane of the operative field, improving dexterity in tight spaces and allowing truly ambidextrous suture placement. A clutching mechanism enables constant readjustment of surgeon hand positions that are necessary to maintain an optimal ergonomic attitude with respect to the visual field.

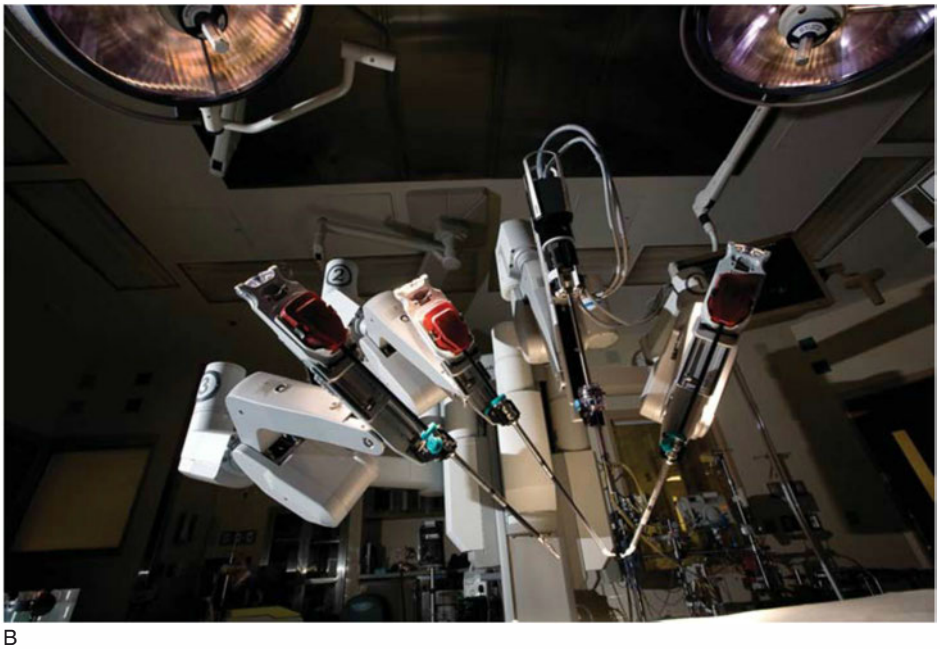
EVOLUTION OF MINIMALLY INVASIVE MITRAL VALVE SURGERY

Initially, minimally invasive mitral valve surgery was based on modifications of previously used incisions and performed under direct vision. Minimal access incisions provided adequate direct vision mitral valve exposure, and surgeons showed that mitral valve operations could be done as safely and precisely as done through a larger incision. Large series by Cohn and Cosgrove showed initially that surgical mortality (1% to 3%) and morbidity were comparable to those of conventional mitral surgery. By employing familiar approaches and relying on direct vision, the initial steps in minimally invasive mitral surgery became less daunting.

With acquired experience, a shift from direct vision to video-assistance occurred and operations began to be performed using secondary vision. In 1996, Carpentier performed the first video-assisted mitral valve repair through a right minithoracotomy using ventricular fibrillation. In 1997, Chitwood described the first mitral valve replacement using videoscopic vision. The operation was done using a 6-cm right anterior mini-thoracotomy, peripheral cardiopulmonary bypass, and a percutaneous transthoracic aortic cross-clamp, and retrograde cardioplegia. In 1998, Mohr reported the Leipzig Heart Center experience of 51 minimally invasive mitral operations done using port-access technology,



A



B

Fig. 43.1. (**A**) da Vinci™ S Robotic Tele-manipulation System: The operative console where the surgeon is seated. (**B**) da Vinci™ S Robotic Tele-manipulation System: The instrument cart with four arms.

a 4-cm incision, and three-dimensional videoscopy. In these patients, video technology was helpful for replacements and simple repair operations; however, complex reconstructions were still approached under direct vision. Concurrently, our group at East Carolina University reported 31 patients operated using video assistance with a two-dimensional 5-mm camera. Complex repairs, including quadrangular resections, sliding valvuloplasties, and chordal replacements, were done with no major complications and the operative mortality was <1%.

In the late 1990s, innovations in computer-assisted telemanipulation occurred rapidly. In 1998, Carpentier performed the first truly endoscopic mitral valve repair using an early prototype of the da Vinci™ surgical system. Within a week, Mohr performed five mitral repairs with the same device. In May 2000, our group performed the first complete mitral valve repair in North America using the da Vinci™ system. In that operation, a large P_2 trapezoidal resection was done with the defect closed using multiple interrupted sutures, followed by implantation of an annuloplasty band. Robotic technology has progressed to a point where at our center totally endoscopic mitral procedures have become routine for patients with isolated mitral valve pathology.

PATIENT SELECTION

All patients with isolated degenerative mitral valve disease are considered for a robotic mitral repair. Complex mitral disease is always approached with the techniques described herein. Contraindications include a prior right thoracotomy and circumferential annular calcification (Table 43.1). Moderate annular calcification alone does not preclude patients from undergoing a minimally invasive approach. However, extensive "bar" calcification remains for us a definite contraindication.

Patients with poor lung function undergo pulmonary testing to ascertain whether they will tolerate single-lung ventilation. Should patients not be able to tolerate isolated lung ventilation, cardiopulmonary bypass is instituted earlier for intrathoracic access. The transesophageal echocardiogram (TEE) remains the gold standard for perioperative planning. It is important to correlate the dynamic echocardiographic anatomy with both the Carpentier functional class and the intraoperative pathology. In patients over 40 years old, and/or with a strong family history and/or symptoms of coronary disease angiography is done preoperatively.

Table 43.1 Robotic Mitral Surgery Exclusion Criteria

- Previous right thoracotomy
- Renal failure
- Liver dysfunction
- Bleeding disorders
- Pulmonary hypertension (PAS >60 Torr)
- Significant aortic or tricuspid valve disease
- Coronary artery disease requiring surgery
- Recent myocardial ischemia (<30 d)
- Recent stroke (<30 d)
- Severely calcified mitral valve annulus

ROBOTIC MITRAL VALVE SURGERY

Anesthesia and Monitoring

To provide single-lung ventilation, either a dual lumen endotracheal tube or bronchial blocker is used. Hemodynamic monitoring is done with a radial arterial line and a pulmonary artery catheter. For superior vena caval (SVC) drainage the right jugular vein is cannulated percutaneously with either a 15F or 17F thin-walled Biomedicus™ (Medtronic, Inc., Minneapolis, MN) arterial cannula using the Seldinger guide-wire technique. This is performed before surgical draping by our anesthesiology colleagues. Three-dimensional TEE is used to evaluate cannula placement, annular size and quality, the leaflet coaptation plane, leaflet length, and the subvalvular mitral apparatus. The intensity and direction of regurgitant jets help us to determine the significance of leaflet pathology. The transgastric view is very helpful for defining precisely which mitral segments need correction. Currently, we select the annuloplasty band size based on TEE measurements of the anterior leaflet length, annular diameter, and septal thickness. Patient-side surgeon comfort and direct vision are most important when gaining ideal intracardiac access for the placement of the robotic instrument arms and the camera. Table-side surgeon long focal-length loupes (3.5×) are most helpful for early preparation as well as passing sutures and specimens through the 3-cm incision during a da Vinci™ repair.

Operative Techniques and Technology

Patients are prepared with the right thorax elevated 30 degrees on a towel roll and the right arm secured by the side. A 3-cm mini-thoracotomy incision is made in the sub-mammary fold in the anterior axillary line. After right lung deflation, the pectorals muscle is spared, and the fourth intercostal space is entered. Pericardial exposure is obtained without rib resection or division, and a soft tissue retractor (Applied Medical Resources Corp., Rancho Santa Margarita, CA) is inserted to displace the skin, fat, and muscle layers, leaving an oval space between the ribs. If the right hemi-diaphragm obstructs the operative field, a suture is placed in the central tendon and pulled through the chest wall with a "crochet hook" instrument, retracted, and secured. The pericardium is opened 2 cm anterior to the phrenic nerve and extended toward the inferior vena caval (IVC) and aortic reflections. Transthoracic pericardial retraction sutures should be placed near the SVC reflection and the atrio-IVC juncture to distract the posterior (dorsal) pericardial edge laterally. To minimize intracardiac air retention, a 14 G catheter is placed through the chest wall for continuous carbon dioxide insufflation (2 to 3 L/min). Instrument arm trocars are then placed through the third and fifth intercostal spaces and sighted to be in direct line with the mitral annulus (Fig. 43.2). It is important to maintain at least 8 to 10 cm between these arms to avoid intracardiac instrument collisions during the repair.

For cardiopulmonary cannulation, an oblique 2 cm right groin incision is made and superficial purse-string sutures are placed in both the femoral artery and vein. Then, coaxial dilators are introduced over a guide wire. Under TEE guidance, a 22F femoral vein venous cannula (Estech, San Ramon, CA) is positioned in the central right atrium. This cannula should not be suture-anchored, as intraoperative manipulation may be needed to provide optimal venous return. If SVC drainage is not possible because of patient small body habitus, a single 23/25F venous cannula (Estech) should be positioned in the SVC. By the same method, the femoral artery is cannulated with either a 17F or 19F Biomedicus™ arterial cannula. Using (kinetic-assisted) suction venous drainage, patients are cooled to 28°C systemically. Thereafter, an ascending aortic antegrade cardioplegia/vent catheter is placed just distal to the right coronary ostium. This position allows deployment of the transthoracic clamp without crowding the cardioplegia/vent catheter. Retrograde coronary sinus catheters also can be placed either across the right atrial wall or from the jugular vein and positioned using TEE.

The interatrial (Waterston's, Sondergaard's) groove is dissected minimally and the oblique sinus is opened behind the IVC. To occlude the aorta under either endoscopic or direct visual direction, the transthoracic aortic clamp (Scanlan, Inc., Minneapolis, MN) is placed through the second interspace in the mid-axillary line with the posterior fixed-tine directed through the transverse sinus, passing posterior (dorsal) to the ascending aorta. Care must be taken to avoid injury to the right pulmonary artery, the left atrial appendage, or the left main coronary artery. After the clamp is applied, the heart is arrested either with antegrade and/or retrograde cold blood cardioplegia.

A short left atriotomy is made inferior to the right superior pulmonary vein with extension toward and inferiorly behind the IVC. The arm of a percutaneous left atrial retractor is positioned just lateral to the sternal edge and medial to the incision, avoiding internal mammary vessels (Fig. 43.2). The dynamic retractor blade is inserted into the left atriotomy. Ventral retraction should elevate the interatrial septum to provide optimal mitral valve exposure. A left superior pulmonary vein sump sucker scavenges residual left atrial blood. Instrument arms are then passed through trocars and positioned in the left atrium, and the three-dimensional endoscope is positioned through the superior part of the incision. Valve function is evaluated using cold saline injections. The table-side surgeon is responsible for exchanging the various microtipped instruments. Standard reconstructive methods have been used in all of our da Vinci™ mitral valve repairs (Table 43.2). We are comfortable in performing leaflet resections, sliding-plasties, chordal transfers, chordal replacements, as well as annuloplasties. We consider patients with Barlow's bileaflet disease as optimal for robotic repairs.

MITRAL VALVE DISEASE AND REPAIR TECHNIQUES

Degenerative Mitral Disease

At our center, robotic instruments and techniques are used to repair degenerative (myxomatous) mitral valves. Classic features of this disease include leaflet prolapse and annular dilatation. Carpentier's functional classification of mitral insufficiency is based on leaflet motion characteristics. Insufficiency with normal leaflet motion is Type 1, with exaggerated (prolapse) leaflet motion is Type 2, and with restricted leaflet motion Type 3. Most common robotic mitral repairs are amenable to Type 1 and 2 pathologies. Chordae tendineae may be thinned, elongated, and/or ruptured. Posterior (P_2) chordal rupture and/or elongation with a dilated annulus are the most common problems that we repair. In the past, only symptomatic patients with 3+ or 4+ mitral regurgitation and ruptured/elongated posterior leaflet chords have been referred for surgery. With the advent of safe, effective minimally invasive and robotic mitral surgery, more asymptomatic

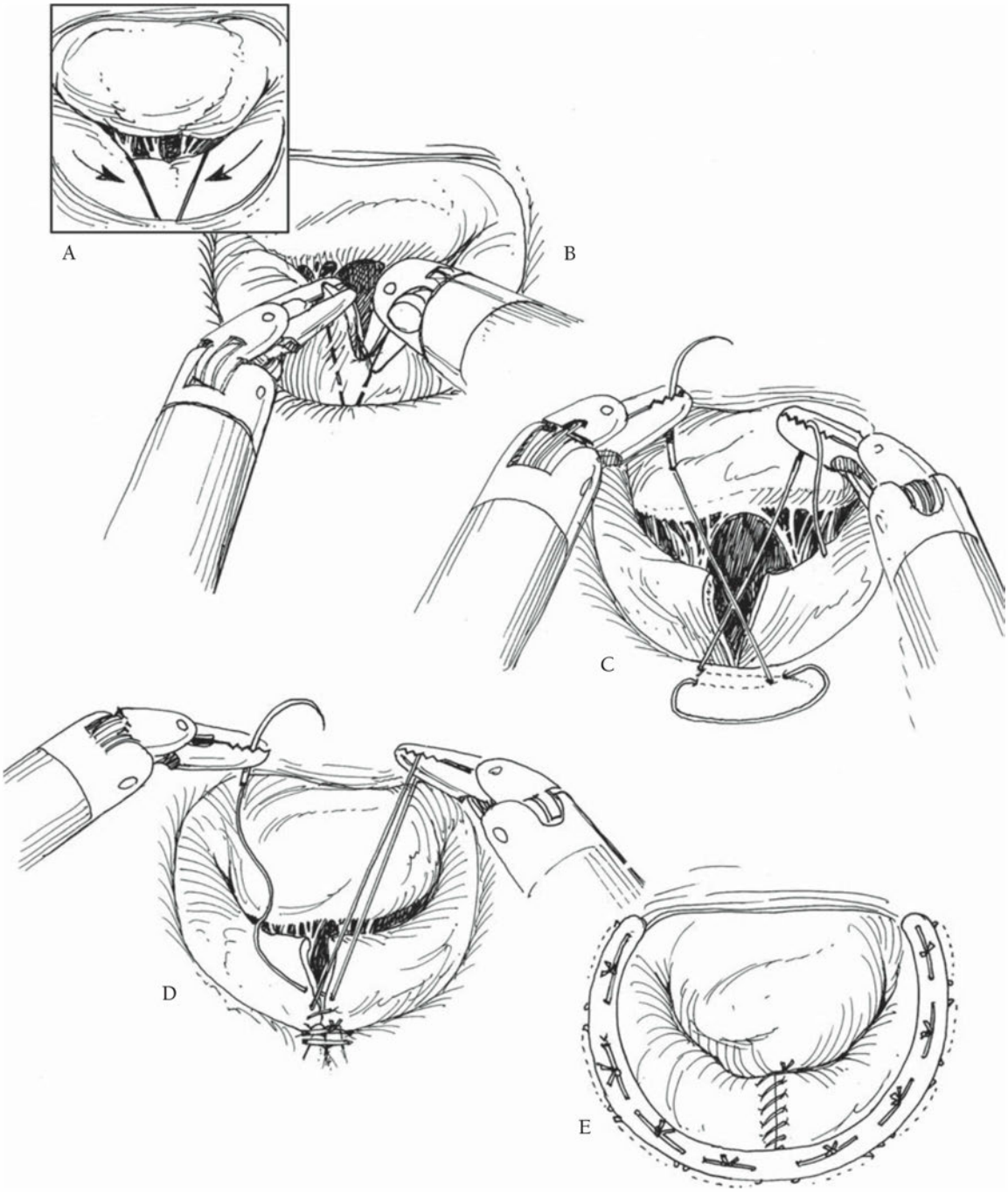


Fig. 43.2. Robotic posterior leaflet resection. A trapezoidal incision is created in P_2 (**A** and **B**) and annular compression sutures are placed (**C**). The leading edges of the remaining leaflet are then brought together using interrupted 5-0 Cardionyl (**D**).The repair receives a band annuloplasty (**E**).

patients with moderate-to-severe regurgitation are being referred earlier. This is especially true for patients who have developed any left ventricular functional impairment or dilatation. Moreover, the presence of early atrial fibrillation indicates significant detrimental changes in progression of valvular disease. To improve long-term survival, there is clear evidence that patients with ruptured chords and a flail leaflet segment should be repaired. In asymptomatic patients, newer indications are predicated on preventing ventricular dysfunction, left atrial dilatation, and the development of atrial fibrillation.

Posterior Leaflet Prolapse

As mentioned, posterior leaflet prolapse/flail occurs most commonly. Robotic micro scissors and atraumatic tissue forceps are used for leaflet resections. Robotic needle holders should never be used to grasp delicate leaflet and chordal tissues. Posterior leaflet prolapse is treated either by triangular, trapezoidal, or quadrangular resection of the diseased chordal leaflet segment (Fig. 43.3). We limit the length of the annular gap, especially in patients who do not require a sliding leaflet plasty to reduce the posterior leaflet height. Any annular defect is then reduced robotically using 2-0 Ticron

Table 43.2 Robotic Mitral Procedures ($N = 114$)

Valve replacements	2 (1.8%)
Valve repairs	112 (98.2%)
Ring annuloplasty	21 (18.4%)
Posterior leaflet resection	47 (41.2%)
Quadrangular/trapezoidal resection + sliding annuloplasty	32 (28.1%)
Posterior/anterior cleft repair	4 (3.5%)
Chordal replacements (GORE)	11 (9.6%)
Chordal transfer	23 (20.2%)
Alfieri	8 (7.0%)

figure-of-eight sutures. Residual leaflet edges are approximated with 4-0 or 5-0 Cardionyl (Peters, Inc., Paris, France) interrupted sutures. This monofilament suture has less memory and more strength than polypropylene material. In robotic operations, we have most frequently used the Cosgrove–Edwards™ annuloplasty band (Edwards Lifesciences, Irvine, CA), which is placed between each fibrous trigone using either tied 2-0 braided sutures, nitinol U-clips™ (Medtronic, Inc., Minneapolis, MN), or the Cor-Knot™ device (LSI Solutions, Inc., Victor, NY) (Fig. 43.4). Recently, the nitinol U-clips™ have been taken off of the market.

When we are concerned about systolic anterior motion (SAM) of the anterior leaflet, a sliding leaflet repair is indicated and can be performed with the da Vinci™ system. Postrepair SAM can develop in patients with excess posterior leaflet tissue (posterior height >1.5 cm), a long anterior leaflet (>34 mm), left ventricular outflow-track septal hypertrophy, and in the presence of an acute angle (<120 degrees) between the aortic and mitral annulus valve planes. Moreover, SAM can be caused by an undersized annuloplasty ring or band. It is not true that SAM can never develop when using a flexible ring or inter-trigonal annuloplasty band. The SAM

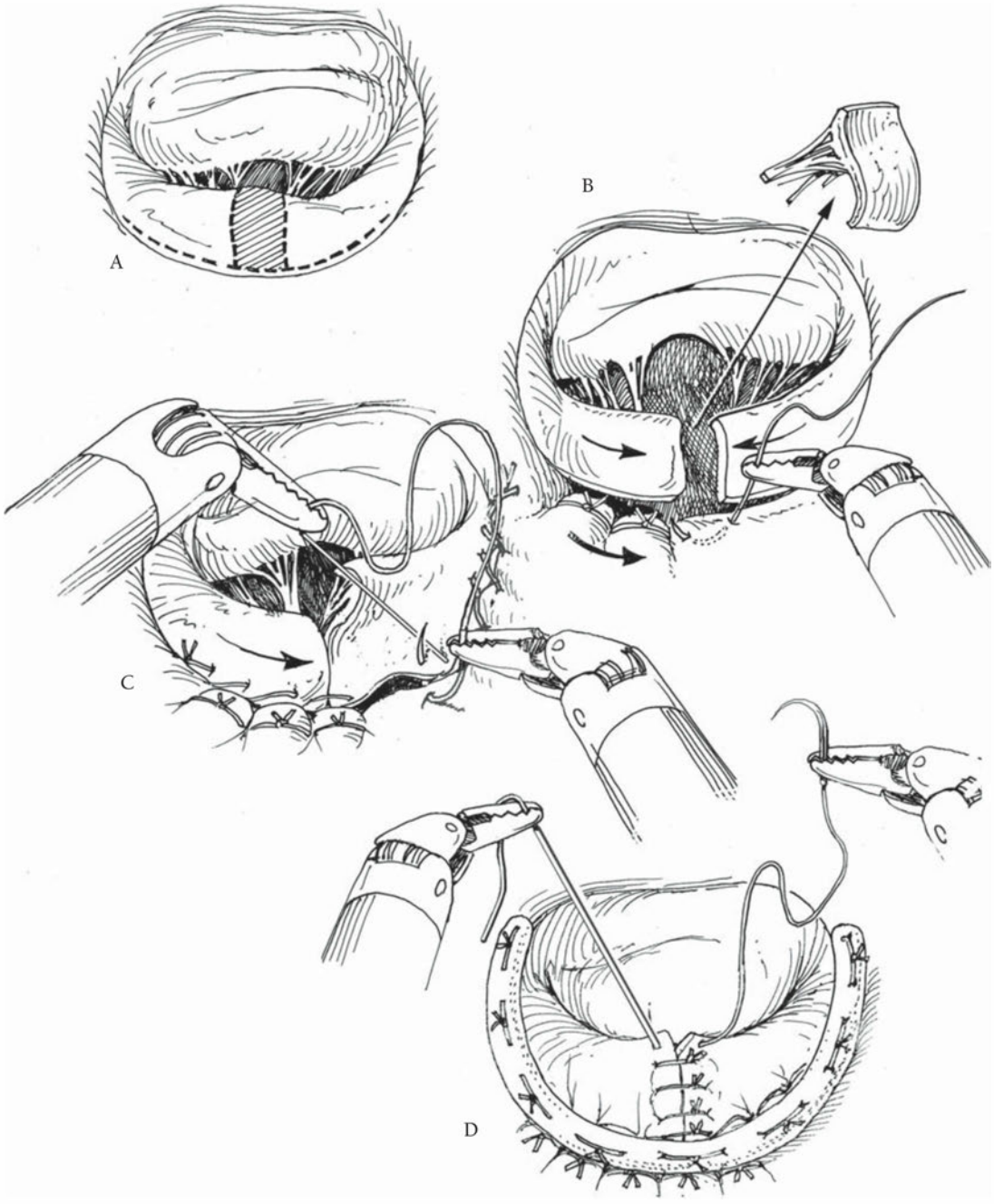


Fig. 43.3. Robotic sliding leaflet repair. After the P_2 resection (**A**), and after annular compression sutures are placed (**B**), a running suture sliding plasty is performed from both the *right* and *left* sides toward the middle of the annulus (**C**). These sutures are then tied together and the valve undergoes final repair (**D**).

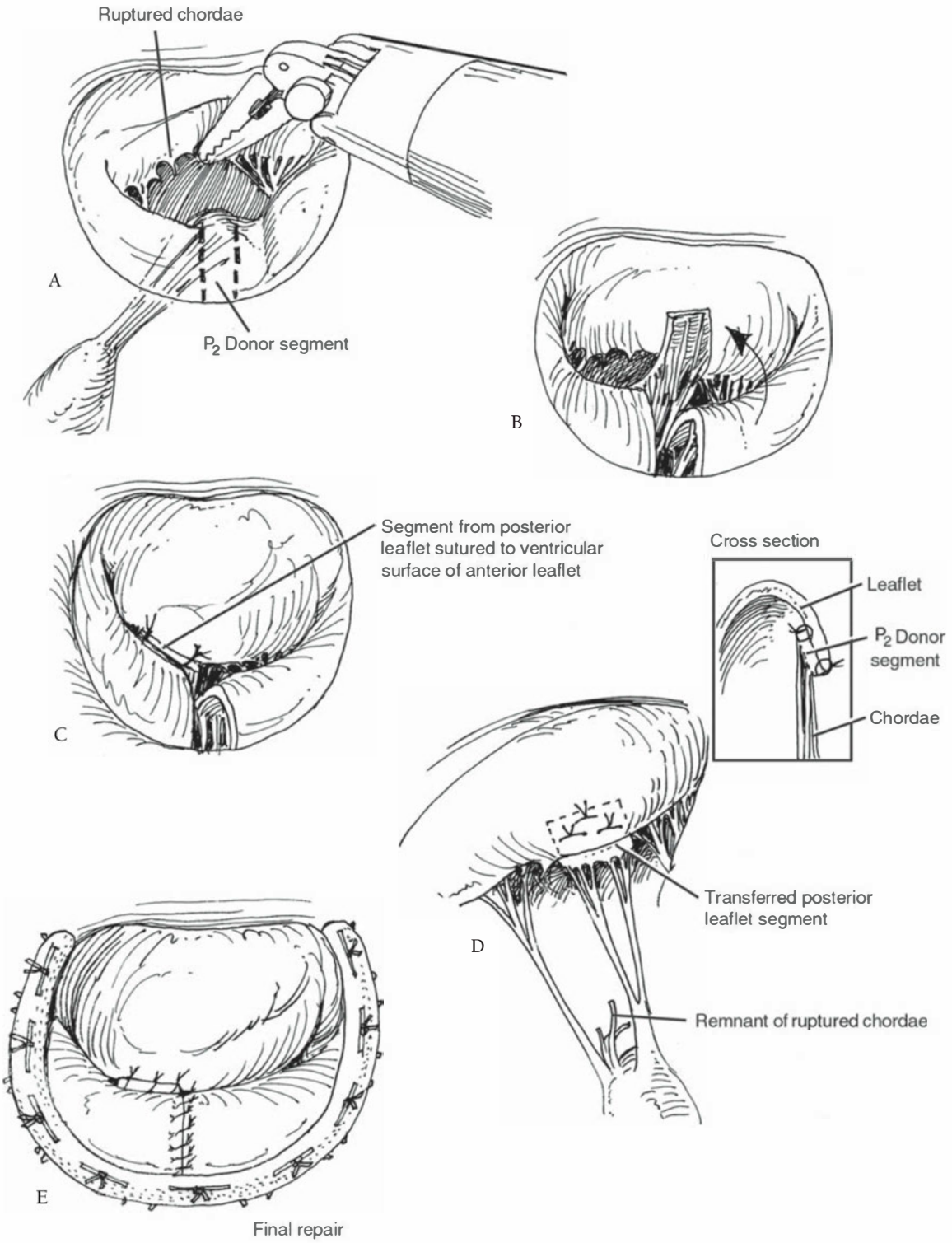


Fig. 43.4. Robotic posterior chordal transfer for anterior leaflet prolapse. A portion of P_2 is resected (**A** and **B**) and sewn to the underside of A2 with interrupted 5-0 Cardionyl suture (**C** and **D**). Final repair seen in **E**.

potential is created when the mitral reconstruction forces the middle or basal anterior leaflet to coapt with the tip of posterior leaflet. During systole, the anterior leaflet tip moves toward the septum and the outflow-jet "lift" causes unphysiologic leaflet displacement toward the outflow tract septum. This displacement can open the leaflet coaptation point, causing significant residual mitral regurgitation. Insufficiency can occur even with a competent valve "saline test" in the arrested heart. It makes sense that ventricular hypertrophy and inotropic support can decrease the leaflet–septum distance thus increasing SAM and the outflow tract pressure gradient.

The purpose of the *sliding leaflet repair* (Fig. 43.5) is to reduce the height of the posterior leaflet, which moves the anterior leaflet coaptation point more posterior

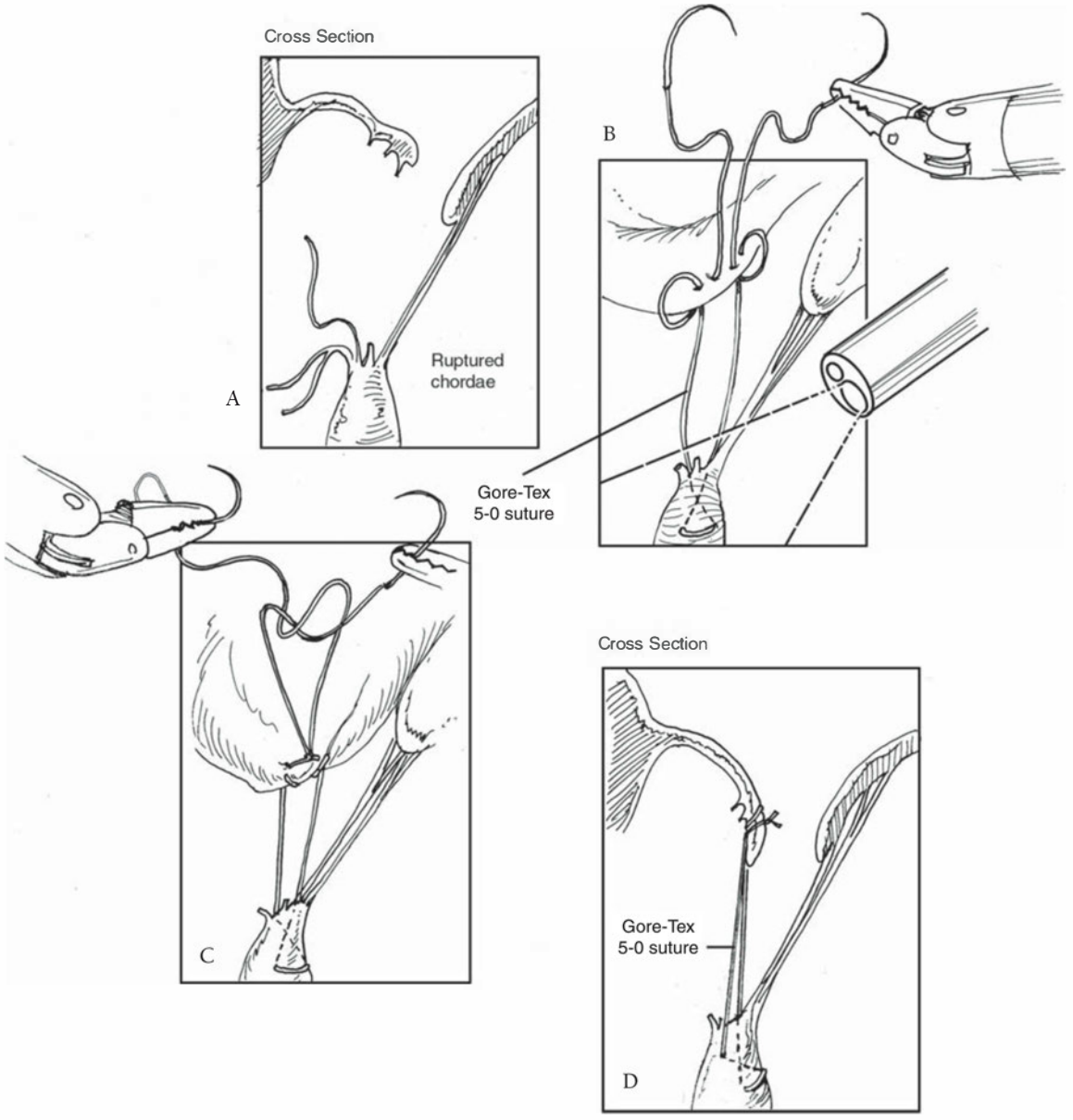


Fig. 43.5. Gore-Tex™ neochords for anterior leaflet prolapse (**A**). Here, a flail anterior chorda is being replaced. A 4-0 Gore-Tex™ is passed in "criss-cross" manner through a viable papillary muscle head. Sutures are passed through the edge of the leaflet and looped around the edge to repenetrate the leaflet (**B**). The suture is then tied while we perform the saline test and remeasure chordal lengths to ensure perfect leaflet coaptation (**C** and **D**).

and away from the ventricular septum. To perform a sliding plasty following a liberal quadrangular leaflet resection, the posterior leaflet is detached radially from the annulus on either side of the resection. Thereafter, multiple horizontal annular compression sutures are placed to reduce the gap symmetrically. Each leaflet remnant is then advanced along the annulus using a running suture. Both leaflet segments must be advanced centrally to reduce the gap. Leaflet edges are then re-approximated as described above, and the annuloplasty is performed to complete the repair.

Anterior Leaflet Prolapse

A variety of techniques have been used to repair mitral anterior leaflet prolapse. Currently, the most common methods include *chordal transfers and prosthetic chord replacements*. Using da Vinci™, we prefer to reduce the anterior leaflet to the coapting plane by either transferring marginal or large basal chords. The three-dimensional high magnification camera and microscissors allow us to detach anterior leaflet basal chords and transfer them directly to the flail edge of the leaflet. Moreover, posterior leaflet primary chordae can be transposed to the unsupported region of the anterior leaflet. Many prolapsing or flail posterior leaflet segments still have some good, thick chords, and these can be transferred anteriorly with a patch of leaflet for firm anchorage. Generally, by swinging either basal (secondary) anterior leaflet or residual posterior leaflet primary chords to the anterior leaflet edge, the radial movement geometrically reduces the anterior leaflet prolapse. The defect in the posterior leaflet is repaired as described for a standard quadrangular resection (Fig. 43.6). For significant anterior leaflet prolapsed, multiple chords

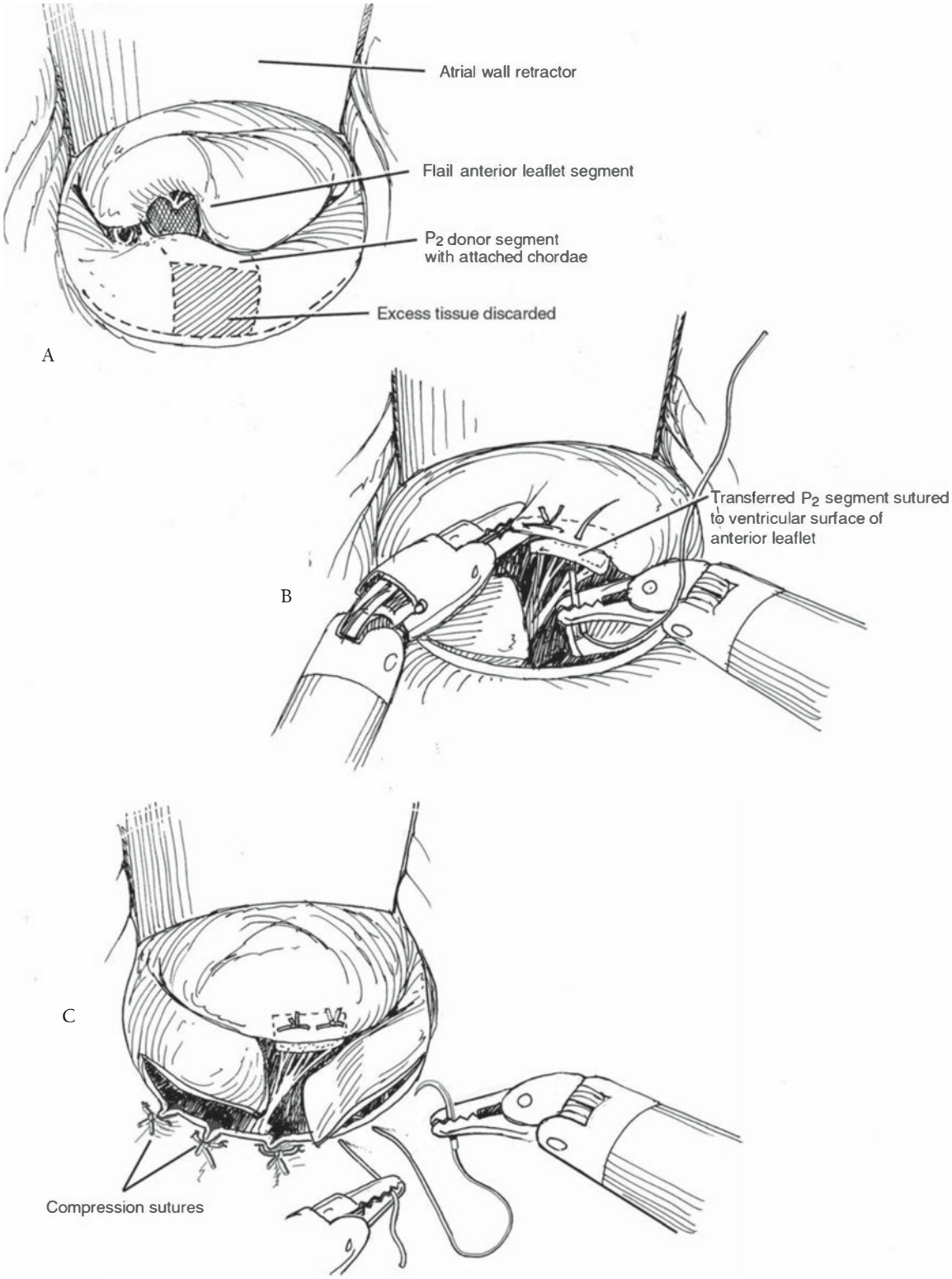


Fig. 43.6. Robotic Barlow's repair. A segment of P_2 is resected and transferred to the underside of A_2 (**A** and **B**). Compression sutures are placed in the annulus and tied immediately (**C**). A running sliding annuloplasty is often performed (**D**). Final repair (**E**).

must be transferred with the saline test used to confirm appropriate edge reduction. Because of the lack of tremor, precise visioning system, and microinstruments, with improved robotic access it is much easier to transfer chords using da Vinci™ system than operating through a sternotomy using conventional instruments and direct vision.

Our second choice for reduction of anterior leaflet prolapse is by using chordal replacements with 5-0 or 4-0 PTFE suture (Gore-tex™, Gore, Inc., Phoenix, AZ). Using the da Vinci™ system, papillary muscle access is magnificent. A short (16 cm) double armed 4-0 Gore-Tex™ suture is passed in a figure-of-eight manner through the papillary muscle fibrous head.

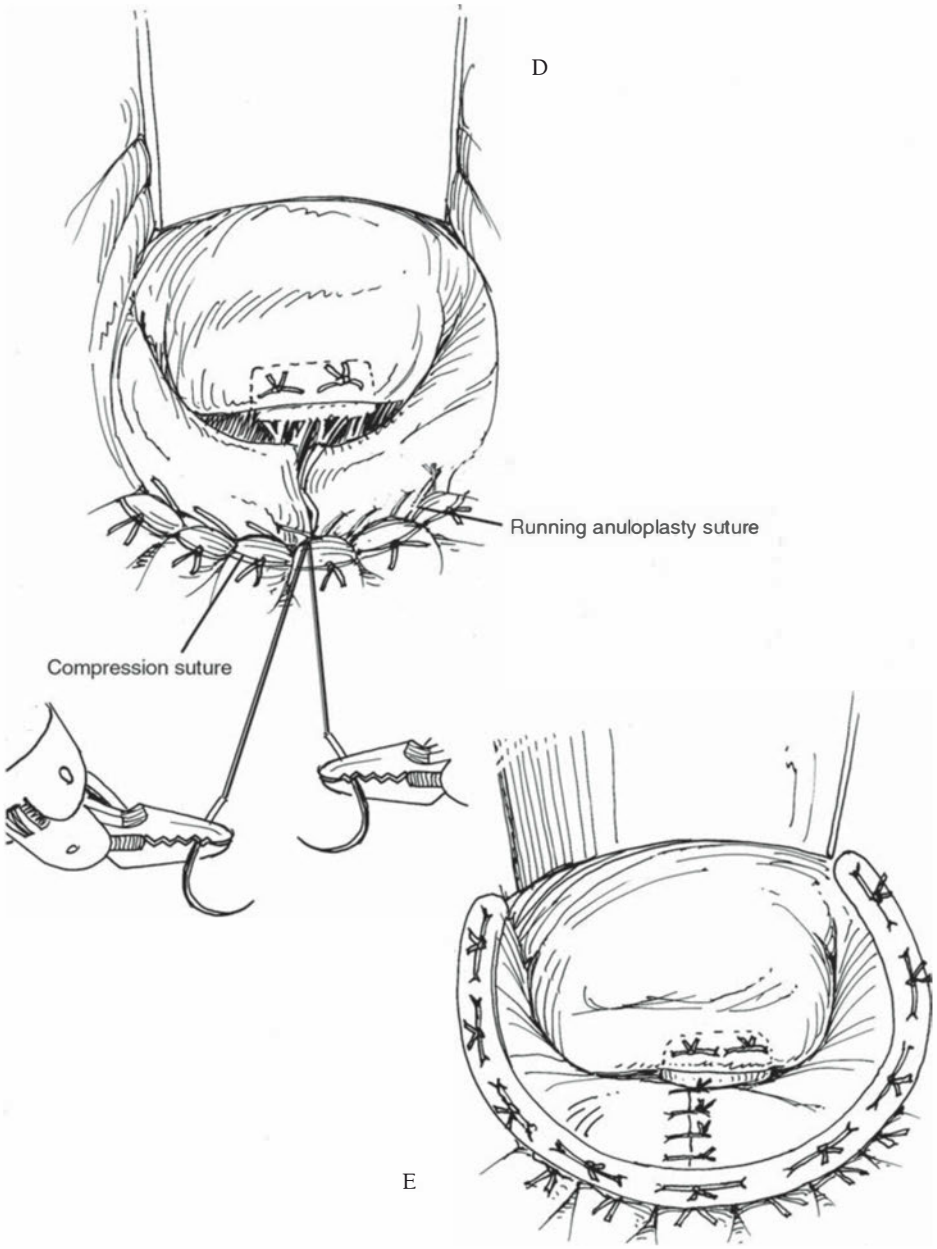


Fig. 43.6. (Continued)

Thereafter, the needles are passed through the leaflet edge at the rupture/prolapse site. Then, the same suture is looped around the coapting edge and back through the leaflet, creating a "locking loop" (Fig. 43.7). The second chord is placed in a similar manner with a 3-mm gap between the sutures. After chordal length adjustment and before the two chords are tied with robotic instruments, saline testing is done to assure optimal neochordal length and optimal leaflet coaptation.

In all of our mitral valve repairs, we have abandoned Carpentier's method of chordal shortening by tucking an elongated chord into a papillary trench. Simpler methods have been very satisfactory and reports of late chordal ruptures with the trench method have shifted our focus from correction at the papillary end to the leaflet end. Robotic methods lend themselves better to the methods described herein, and to date we have had no late ruptures in chordal transfers or replacements. In severe Barlow's bileaflet disease, we generally transfer a long strip of chord-bearing posterior leaflet (usually P_2) to prolapsing segments of A_1–A_3. Then the resected part of the posterior leaflet is closed using the reducing sliding plasty and commissural prolapse is corrected with the edge-to-edge method. Of course, a properly sized annuloplasty band or ring is selected to complete the robotic repair.

Annuloplasty

Annular dilatation is present in most patients with degenerative mitral disease. An annuloplasty is performed in conjunction with all repairs to restore the native geometry, reduce the annular size, prevent further dilatation, and reinforce the repair. Reducing the anterior–posterior annular diameter increases the leaflet coaptation surface. We select an annuloplasty support prosthesis that is either flexible or rigid and surrounds the mitral annulus either completely or incompletely. The latter notion suggests that the mitral-aortic curtain or central fibrous body of the heart, which extends between the fibrous trigones,

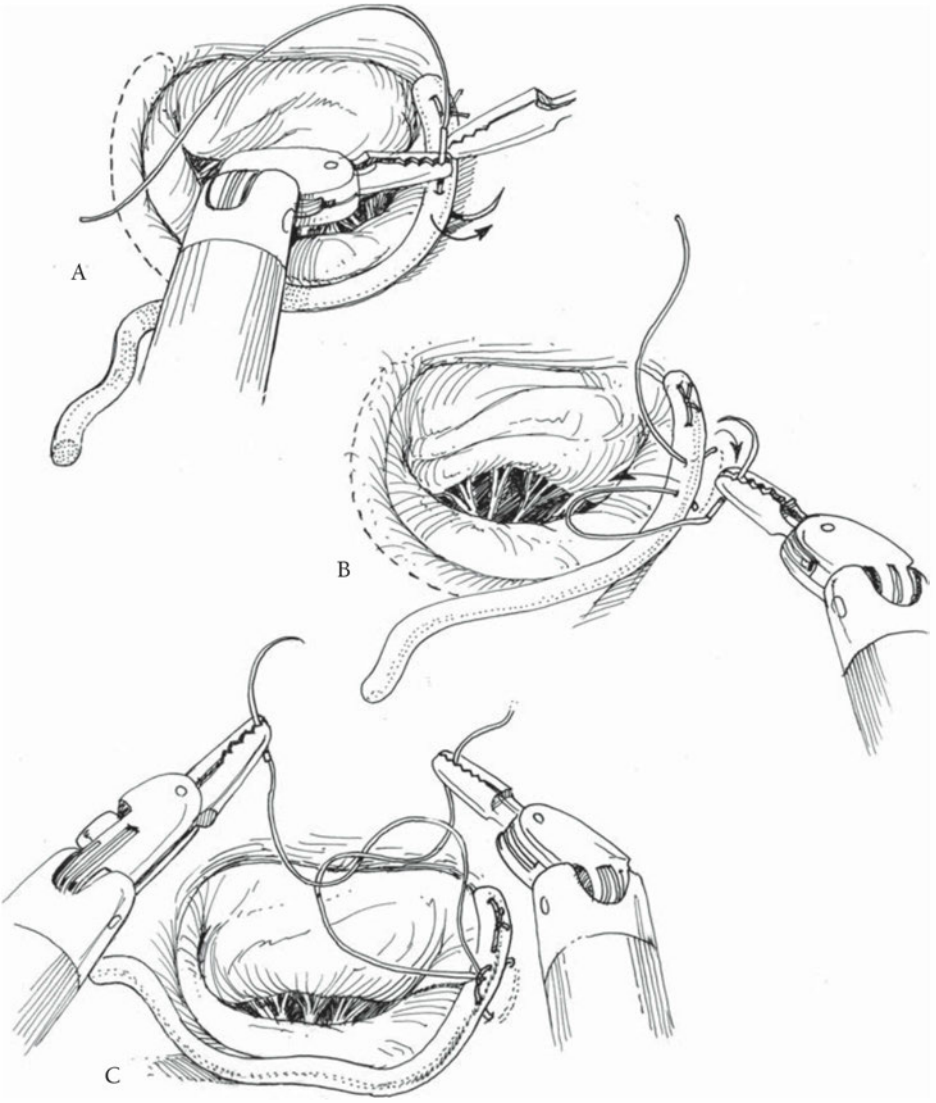


Fig. 43.7. Robotic suture placement for mitral annuloplasty (**A** and **B**). Twelve-centimeter sutures are used and tied serially **(C)**.

is nonmuscular and does not dilate in patients with degenerative mitral disease. The Cosgrove–Edwards™ (Edwards Lifesciences, Irvine, CA) annuloplasty band is anchored at each trigone and for the remaining muscular annulus both reduces and remodels the valve geometry. An ideal prosthesis should be flexible, soft, and easy to handle with robotic instruments. To date, no complete ring satisfies these requirements, and the Cosgrove–Edwards™ band has served well for our repairs as the major etiology has been from degenerative causes and not ischemic deformation.

Short (14 cm) double armed 2-0 sutures currently are unavailable and therefore, the band is attached using the single-arm method shown. Initially, sutures are placed into each fibrous trigone to aid in exposure. Also, we detach the band from the template in order to manipulate it in the confined interatrial space. Without careful implantation, this could be suboptimal as nonlinear placement of sutures or variations in spacing can cause kinking of the band and annular asymmetry. To obviate kinking and uniform implantation, the band is first anchored to the right fibrous trigone with measured suture placement proceeding clockwise. The last two sutures at the anterior commissure and left fibrous trigone are placed before tying either. Care must be taken in this region to avoid injury to the aortic root and the circumflex coronary artery. Good anchoring bites can be obtained here but the needle must remain perpendicular to the annulus and not tangential toward the aorta or circumflex region. The da Vinci™ system high-definition camera enables precise placement of deep precise bites along the annulus. From our experimental studies we have seen adequate annular compression, ease of placement, and band security to be optimal using either suture-tying or automatic securing clips. Using the Core-knot™ clip device, recently we have found that our band implantation times have decreased significantly compared with the suture technique described.

CLINICAL OUTCOMES

Currently, we have performed over 700 robotic mitral valve repairs with the daVinci™ system. Our training program has trained over 400 surgeons in mitral repair surgery using da Vinci™ system, and many of these groups are already successfully doing repairs.

da Vinci™ Robotic Mitral Valve Repairs

In the first Food and Drug Administration (FDA) safety and efficacy trial of robotic mitral valve repair, quadrangular leaflet resections, leaflet sliding plasties, chord transfers, polytetrafluoroethylene chord replacements, reduction annuloplasties, and annuloplasty band insertions were performed in 20 patients at East Carolina University. The first overall cardiac arrest times were long at 150 minutes but only were 52 minutes for leaflet repairs. Moreover, it took an additional 42 minutes to anchor the Cosgrove–Edwards™ annuloplasty bands (mean 7.5 sutures). The operative times seemed long but the results were good, and there were no device-related and few procedure-related complications. The average postoperative stay was 4 days (3 to 7 days). At 3-month follow-up, echocardiographic studies revealed nothing more than trace mitral regurgitation in these patients. Moreover, all patients returned to normal activity 1 month following surgery.

A subsequent multicenter, phase II FDA trial studied 112 da Vinci™ patients operated in 10 U.S. centers. Again, repairs included quadrangular resections, slidingplasties, edge-to-edge approximations, and both chordal transfers and replacements. Leaflet repair and annuloplasty times had fallen to 37 and 39 minutes, respectively, compared with phase I trial outcomes. Aortic cross-clamp and cardiopulmonary bypass times averaged 2.1 and 2.8 hours, respectively. There were little differences in operative times between centers. One-month follow-up by transthoracic echocardiography revealed that nine patients (8.0%) had ≥grade 2 mitral regurgitation and six (5.4%) of these had reoperations. Although the reoperative rate was disturbing, failed repairs were distributed evenly across the overall cohort and some centers had performed <10 robotic operations. There were no deaths, strokes, or device-related complications. This study demonstrated that multiple surgical teams could perform robotic mitral valve surgery safely early in the development of these techniques, albeit with a learning curve as far as operative times and repair results. Based on these two studies, the daVinci™ robotic surgical device was FDA approved in November of 2002 for intracardiac use in the United States.

Learning curve data from our first 38 da Vinci™ mitral repairs were encouraging. Patients were evaluated in two cohorts: *Early (N=19)* and *Late (N=19) follow-up*. Operative times were chronicled starting after the robot was positioned to begin the repair. Total robotic operating times decreased from 1.9 to 1.5 hours ($P = 0.002$), respectively, between the two groups. Concomitantly, leaflet repair times fell from 1.0 to 0.6 hours ($P = 0.004$), respectively. Both cross-clamp and bypass times decreased significantly with experience as well. More recently, a learning curve analysis of over 450 robotic mitral valve repairs performed at our institution demonstrated a logarithmic learning curve, with a learning percentage of 95%. Multiple regression analysis defined significant variables that affected total robot operative times and included: type of repair, band size, use of clips alone for band implantation, and the presence of a postgraduate fellow at bedside.

To date, we have completed over 700 robotic mitral valve repairs with the da Vinci™ system and have published results on the first 300. There was a 0.7% 30-day mortality and a 2.0% late mortality. No patients underwent valve replacement or conversion to a sternotomy. Complications included two (0.7%) strokes, two transient ischemic attacks, three (1.0%) myocardial infarctions, and seven (2.3%) reoperations for bleeding. The mean hospital stay was 5.2 ± 4.2 days. Sixteen (5.3%) patients required a mid-term or late reoperation. Mean postoperative echocardiographic follow-up studies at 2½ years demonstrated the following degrees of mitral regurgitation: none/trivial, 192 (68.8%); mild, 66 (23.6%); moderate, 15 (5.4%); and severe, 6 (2.2%). From these data, we concluded that robotic mitral valve repair was safe and associated with good mid-term durability.

LIMITATIONS/FUTURE DIRECTIONS

Despite rapid procedural advances, our early clinical experience with computer-enhanced mitral surgery has defined many of the technologic limitations. Force or haptic feedback is absent in these devices; however, new strain sensors may soon allow for more control of force applied to robotic end-effectors. Thus far, we have been able to overcome the limitations of "touchless" surgery by relying on "ocular tactility." That is with a high degree of visual acuity and magnification we can sense when knots are tight, bands are approximated closely to tissues, and calcifications are present. Nevertheless, the addition of haptic feedback assuredly will provide more latitude with these devices.

We have examined both experimental and clinical learning curves that are associated with robotic mitral valve repairs. A retrospective review of the first 80 mitral valve repairs using the da Vinci™ system suggested that learning curves can be flattened for learners (unpublished data). To study these curves, the group of 80 patients was divided into eight equal groups of 10 chronologic cohorts. Valve resection, repair, suture placement, knot tying, total robotic, cross clamp, and bypass times were analyzed in the first five 10-patient cohorts. All showed significant reductions in times when we compared the first group with the most recent operations. As expected, we proved that intraoperative robotic mitral repair surgery times decreased as more procedures were performed. Interestingly, the learning curve was most evident during the first 20 operations but thereafter was incremental improved. Depending upon further refinements and development of adjunctive technologies, computer-enhanced cardiac surgery should evolve and promises to be beneficial for many more patients. Moreover, instrument arm sizes must be reduced and a greater variety of end-effector tips developed before, we will see the robotic era in mitral surgery roll out to become standard of care. Future surgical imaging and positioning systems may allow us to do these repairs using 1 cm ports and perhaps even inside the beating heart using echocardiographic guidance.

CONCLUSIONS

A renaissance in cardiac surgery has begun, and robotic technology is providing benefits to cardiac surgeons and their patients. To be sure, improved optics and instrumentation, progressively smaller incisions, and simulated three-dimensional vision, and enhanced surgeon hand-eye coordination all will facilitate greater adoption of these surgical techniques. The placement of wrist-like articulation instrument ends moves the pivoting instrument tip actions to the plane of the mitral annulus. This improves dexterity in tight spaces and allows ambidextrous suture placement. Furthermore, as an educational tool, the robotic system may have an expander potential for use by residents and experienced surgeons alike. In the near future, surgical vision and training systems may be able to model most surgical procedures through immersive technology. Thus, a surgeon may be able to simulate, practice, and perform the operation based strictly on preoperative imaging data.

This new science is a trek and not a destination. In this era of outcome-based medicine, surgical scientists must continue to

evaluate intracardiac robotics and related technology critically. Despite initial enthusiasm, caution cannot be overemphasized. Surgeons must proceed carefully as indices of operative safety, speed of recovery, level of discomfort, procedural cost, and long-term operative quality remain poorly defined. Traditional valve operations still enjoy long-term success with ever-decreasing morbidity and mortality, which remain as our measure for comparison. Nevertheless, robot-assisted mitral valve surgery has developed to become a safe and successful modality.

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EDITOR'S COMMENTS

Dr. Kypson and colleagues have given a very balanced approach to the use of the robot in mitral valve repair. The senior author Dr. Chitwood has pioneered this technology and has provided outstanding results. I think Dr. Chitwood has always been fair about what this technology does. As he has stated frequently, the robot is just a pair of scissors and ability to suture with better visualization. There is no doubt that the techniques have continued to improve and certainly have become more wide spread. One must know the basic techniques for mitral valve repair prior to developing a robotic learning curve.

There are certainly some issues that need to be better solved. Authors have talked about methods in improving venous drainage. Presently, the cross-clamp times have been a little longer in must robotic mitral valve series. Having said, the technique is now useful for essentially all cases of degenerate disease and I believe the technology to continue to evolve.

ILK

44 Mitral Valve Repair for Ischemic Mitral Regurgitation

Vincent Chan, Javier G. Castillo, and David H. Adams

INTRODUCTION

Ischemic mitral regurgitation (MR) is a mechanical complication of coronary artery disease. Most commonly, ischemic MR is due to chronic left ventricle remodeling following myocardial infarction. The resulting papillary muscle displacement leads to tethering of the mitral valve leaflets and varying degrees of annular dilatation (Fig. 44.1). This entity should be distinguished from coronary artery disease and concomitant MR due to other causes such as degenerative mitral valve disease. Naturally, the repair techniques and long-term outcomes following repair are very different depending upon the underlying etiology. Acute ischemic MR is rare and is secondary to papillary muscle rupture or chordal elongation.

Ischemic MR is common and associated with significant morbidity and mortality. MR is observed in approximately 10% of patients undergoing cardiac catheterization and may be associated with 40% mortality within the first year of diagnosis. The increasing prevalence of coronary artery disease has resulted in a rise in the number of patients with ischemic MR requiring surgery. Despite advances in medical and surgical management, longitudinal data have shown poor survival for patients with ischemic MR compared with patients who present with MR due to other causes. Nevertheless, new technologies and recent studies have provided new insights into this challenging surgical problem. In this chapter, we review ischemic MR with regard to its pathophysiology, clinical presentations, diagnosis, and management.

PATHOPHYSIOLOGIC TRIAD OF MITRAL REGURGITATION

MR is defined as the backward flow of blood in systole from the left ventricle into the left atrium via the mitral valve. Minimal lesions might cause MR by reducing mitral leaflet coaptation. Therefore, a careful interrogation (identification, localization, and magnitude) for mitral lesions is essential to determine the chances of successful valve repair and to proceed with a tailored therapeutic plan for each patient. Three decades ago, Carpentier described a systematic analytic approach to patients with MR known as the "pathophysiologic triad of mitral valve regurgitation." The triad emphasizes the importance of distinguishing between the medical conditions causing MR (etiology), the resulting lesions, and finally how these lesions affect leaflet motion (dysfunction).

Patients with type I dysfunctions have normal leaflet motion, and MR is due to annular dilation or leaflet perforation (Fig. 44.2A). There is increased leaflet motion in patients with type II dysfunction, with the free edge of at least one of the leaflets overriding the plane of the annulus during systole (Fig. 44.2B). Lesions responsible for type II dysfunction include chordal elongation, chordal rupture, and papillary muscle rupture. Type III dysfunction refers to restricted leaflet motion, with the free margin of one or both leaflets pulled below the plane of the annulus into the left ventricle, thereby reducing leaflet mobility and coaptation during systole and leading to MR. Patients with type IIIa dysfunction have restricted leaflet motion during both diastole and systole, as in the case with rheumatic valve disease. Type IIIb dysfunction is caused by restricted leaflet motion during systole, which occurs in the setting of left ventricular dysfunction and dilation; apical and posterolateral papillary muscle displacement causes this type of valve dysfunction (Fig. 44.2C).

Ischemic MR can result from type I, II, or IIIb dysfunction (Table 44.1). Isolated type I dysfunction with annular dilation is uncommon but may occur in basal myocardial infarction. An associated type I lesion in the setting of a primary type IIIb lesion is a common finding in ischemic MR, but the amount of annular dilatation is often less than that seen in degenerative disease. Type II dysfunction after myocardial infarction results from papillary muscle rupture, which usually involves the posteromedial papillary muscle or occurs when a fibrotic papillary muscle elongates and causes leaflet prolapse, particularly in the commissural area (this a rare lesion). In rare instances, an isolated chordal rupture can cause type II dysfunction. Carpentier's type IIIb dysfunction is the *most common and significant* form of ischemic MR.

NATURAL HISTORY OF MITRAL REGURGITATION

Ischemic MR following myocardial infarction is associated with significant morbidity and mortality. Notably, more adverse events are observed with increasing MR severity. Data from the Controlled Abciximab and Device Investigation to Lower Late Angioplasty Complications study showed that in patients following ST elevation MI, the absence of MR was associated with an 1.4% 30-day mortality rate compared with 3.7% in patients with mild MR and 8.6% in patients with moderate-to-severe MR. Late survival is also worse with a reported 5-year total mortality and cardiac mortality of 62% and 50%, respectively, for patients with ischemic MR compared with 39% and 30%, respectively, for patients without ischemic MR. Beyond survival, ischemic MR is associated with more frequent hospitalization for congestive heart failure, with 30% of patients requiring hospital admission within 3 years of diagnosis.

TYPE IIIB ISCHEMIC MITRAL REGURGITATION

Pathophysiology

Normal mitral valve function involves a complex three-dimensional interaction

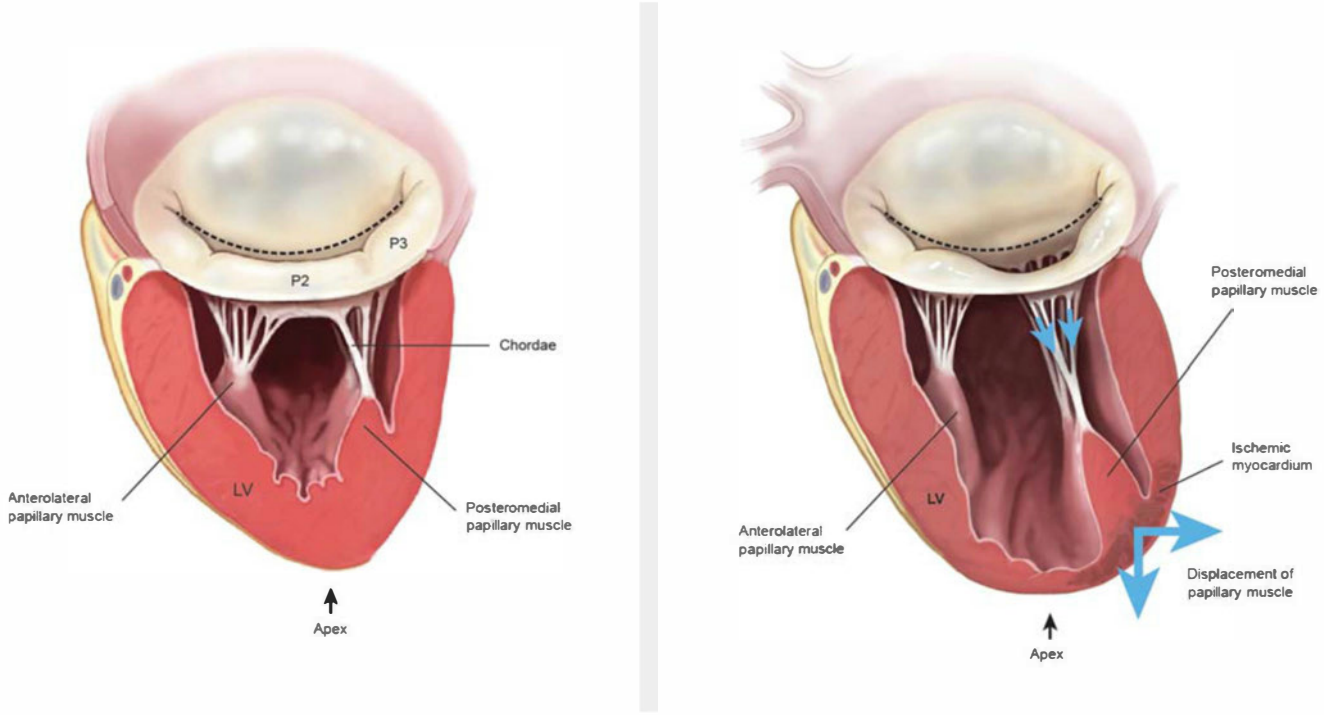


Fig. 44.1. Mechanism of functional mitral regurgitation. Normal mitral valve and subvalvular apparatus (*left*). Ischemic mitral valve with pronounced posterior restriction in P3 after an episode of ventricular ischemia (*right*). LV, left ventricle.

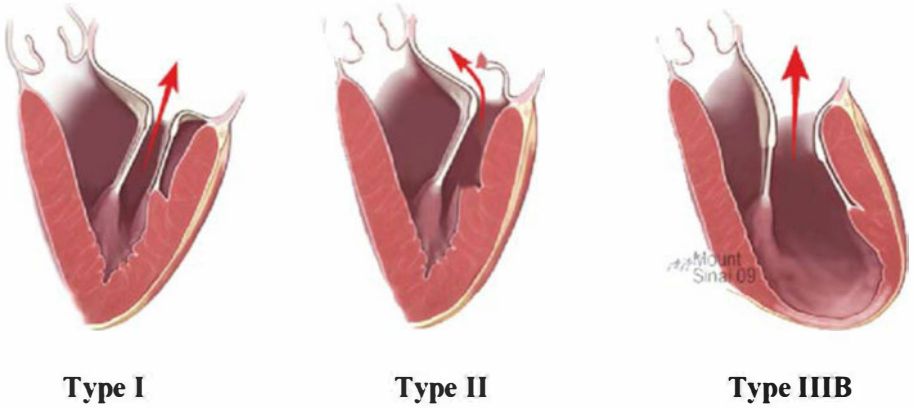


Fig. 44.2. Carpentier's functional classification i n ischemic mitral regurgitation. Type I has normal leaflet motion, and mitral regurgitation is on the basis of annular dilation, which may occur in the setting of basal infarction. Type II dysfunction implies excess leaflet motion with the free edge of the leaflet(s) traveling above the annular plane during systole, which may occur after myocardial infarction due to papillary muscle rupture (acute) or papillary elongation (chronic). Type IIIb is the most common form of ischemic mitral regurgitation and results from restricted leaflet motion during systole secondary to apical and lateral papillary muscle displacement.

among the leaflets, annulus, subvalvular apparatus, and left ventricular wall. Several anatomic and pathophysiologic changes are associated with the pathogenesis of ischemic MR. They include ventricular changes (wall-motion abnormalities and/or dilation with increased sphericity), subvalvular changes (papillary muscle infarction, displacement, or tethering), and annular changes (distortion, dilation). The initial insult in chronic ischemic MR is due to remodeling of the left ventricle after myocardial ischemia or infarction. This remodeling converts the shape of the left ventricle from ellipsoidal to spherical, which leads to regional annular and subvalvular distortion and ultimately to poor leaflet coaptation. It appears that left ventricular sphericity is more important than the actual ventricular volume or ejection fraction in the progression of ischemic MR. The primary ventricular alteration leading to ischemic MR is papillary muscle displacement. The pattern of displacement is complex and cannot simply be described as apical tethering. The papillary muscle tips are displaced away from the midseptal (anterior) annulus, that is, posterolaterally, apically, and away from each other. The tethering distance has been shown to correlate with the severity of ischemic MR. Papillary muscle tethering leads to *apical tenting* of the leaflets (restriction of the motion of the free margins of the leaflets), which prevents them from rising to the plane of the annulus to coapt with one another (Fig. 44.3). Recent animal studies also suggest that the remodeling process accelerates as the degree of MR increases. Clinically, this tethering of the secondary chordae can be seen echocardiographically in the form of the "sea-gull" deformation of the body of the leaflet.

Annular dilation is a common *associated finding* in chronic ischemic type IIIb MR. However, the degree of dilation can vary and does not necessarily correlate with the degree of MR. Annular flattening of the saddle also occurs in ischemic MR. Laboratory data support the notion that annular dilation is not a fundamental component in the pathogenesis of ischemic MR. It has been demonstrated that mild degrees of annular dilation observed during acute occlusion of the left anterior

Table 44.1 Pathophysiologic Triad in Ischemic Mitral Regurgitation

Dysfunction	Lesions	Natural history
Type I	Annular dilation	Chronic
Type II	Chordal rupture	Acute
	Papillary muscle rupture	
	Papillary muscle elongation	
Type IIIb	Papillary muscle displacement	Acute or chronic
	Leaflet tethering	

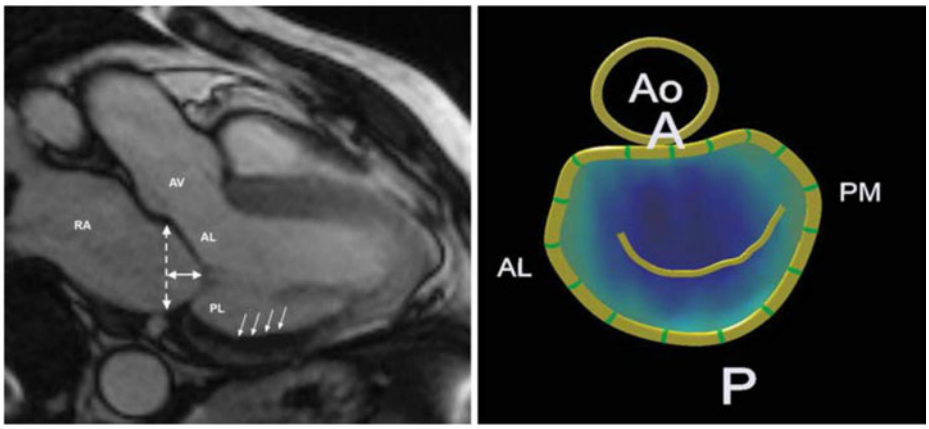


Fig. 44.3. Characterization of ischemic mitral regurgitation. Cardiac magnetic resonance imaging (vertical long axis, two-chamber view, maximum systolic pressure) reveals severe leaflet tethering and the area of ischemic insult involving the posteromedial papillary muscle (multiple arrows) (*left*). Three-dimensional echocardiography reveals severe leaflet tethering (*blue*) (*right*). AL, anterior leaflet; AV, aortic valve; continuous line, tenting height; discontinuous line, physiologic plane of coaptation); LA, left atrium; PL, posterior leaflet.

descending or distal left circumflex in an animal model did not result in ischemic MR. The aforementioned changes in left ventricular geometry, papillary muscle position, and annular dimensions interact to result in poor leaflet coaptation during systole, which is the final common pathway for type IIIb ischemic MR.

Clinical Presentation

Type IIIb ischemic MR can occur in an acute or chronic setting. *Acute postinfarction ischemic MR*, without papillary muscle rupture, can be documented in many patients by physical examination, ventriculography, or echocardiography. Moderate-to-severe MR is present in up to 13% of these patients. Acute severe type IIIb ischemic MR often presents with the sudden onset of shortness of breath and/or angina. In most patients, this incident is preceded by an acute myocardial infarction, which may be silent particularly in diabetic patients. A minority of patients will present with symptoms of severe heart failure and/or low cardiac output. Although MR resolves over time in some patients, it will persist in others and lead to *chronic postinfarction ischemic MR*. In these patients, chronic ischemic MR will appear up to 6 weeks (median 7 days) after myocardial infarction as the left ventricle remodels. Risk factors for postinfarction ischemic MR include advanced age, female gender, prior acute myocardial infarction, large infarct size, recurrent ischemia, multivessel coronary artery disease, and congestive heart failure.

In patients with chronic ischemic MR, two clinical scenarios are commonly encountered. Patients may present with moderate-to-severe MR, symptoms of congestive heart failure, or worsening left ventricular function, and are referred primarily for mitral valve intervention. The preoperative coronary angiogram in these patients shows significant multivessel coronary artery disease, which may or may not have lead to symptoms. These patients often have clear evidence of prior myocardial infarction and at least moderate left ventricular dysfunction. Other patients present with symptomatic multivessel coronary artery disease and are referred for coronary artery bypass grafting (CABG) with a varying degree of MR on preoperative ventriculography or echocardiography. An acute coronary syndrome or chronic stable angina is often the dominant presenting symptom in these patients. In addition, they may present with symptoms of shortness of breath and/or congestive heart failure.

Diagnosis

Patients often have changes on their electrocardiogram (ECG), and a prior myocardial infarction may be noted in >80% of patients with chronic ischemic MR. When ECG changes are noted, evidence of a prior inferior wall infarct is more common than an anterior or a lateral infarct. Most patients are in sinus rhythm; however, in patients with left atrial enlargement, p-wave abnormalities and atrial fibrillation may occur. Conduction abnormalities are uncommon in these patients. In the acute setting, the chest radiograph may show pulmonary edema. With disease progression, enlargement of the cardiac silhouette (left atrial enlargement, ventricular dilation) is a common finding. Cardiac catheterization will typically show multivessel disease and, likely, segmental wall motion abnormalities on ventriculography.

Two-dimensional echocardiography/Doppler is essential in determining the MR mechanism and severity, along with the function of the left ventricle. The severity of MR can be determined by semiquantitative measurements using jet geometry and area in multiple views. With restricted leaflet motion (type IIIb), the direction of the jet is often toward the restricted leaflet, or central if associated annular dilation is present. In this context, imaging has revealed two major patterns of leaflet tethering in the setting of ischemic MR: symmetric and asymmetric. Asymmetric tethering is defined by the displacement of the posteromedial papillary muscle, causing posterior leaflet restriction and leading to an eccentric jet toward the posterior wall of the left atrium. As opposed to asymmetric tethering, symmetric tethering results from the displacement of both papillary muscles, and a central jet is typically observed. This symmetric pattern is the one observed in the setting of functional MR secondary to overlapping dilated cardiomyopathy (Fig. 44.4). The severity of MR is quantified via calculation of regurgitant volume (the difference between the

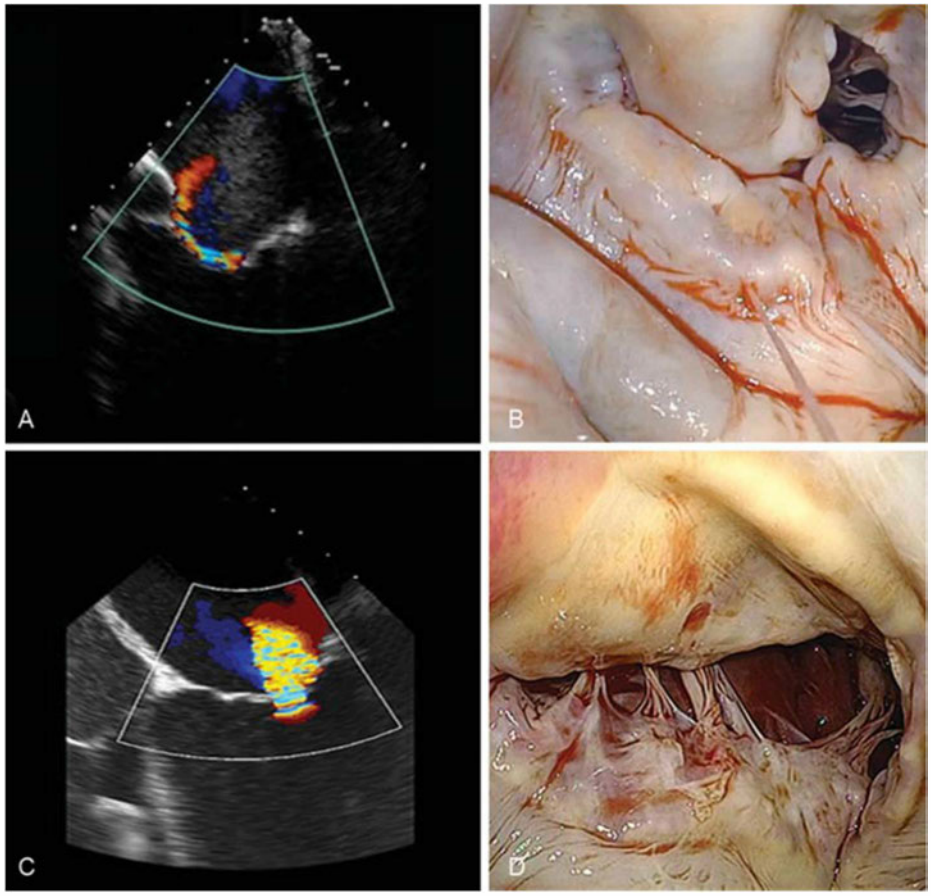


Fig. 44.4. Asymmetrical (**A** and **B**) and symmetrical (**C** and **D**) pattern of ischemic mitral regurgitation.

mitral and aortic stroke volumes) and effective regurgitant orifice (ratio of regurgitant volume-to-regurgitant time velocity integral). Intraoperative transesophageal echocardiography (TEE) can be used to further delineate the mechanism of MR; however, afterload reduction associated with general anesthesia is known to ameliorate the intensity of MR.

SURGICAL INDICATIONS

Severe Ischemic Mitral Regurgitation

Current practice guidelines do not separate recommendations for surgery according to the etiology of MR, but rather the severity of MR and the degree of symptoms. Most patients with severe ischemic MR, however, have symptoms of congestive heart failure. Therefore, per current guidelines, patients with severe, symptomatic MR have a class I indication for mitral valve surgery if their left ventricular ejection fraction (LVEF) is >30% and/or their end-systolic left ventricle dimension is ≤55 mm. Patients with severe symptomatic MR, LVEF <30% and/or an end-systolic left ventricle dimension >55 mm have a class IIa indication for mitral valve surgery, provided that chordal preservation is likely.

Mild-to-Moderate Ischemic Mitral Regurgitation

In this clinical scenario, patients are referred for myocardial revascularization secondary to symptomatic/severe coronary artery disease. Mild-to-moderate type IIIb ischemic MR is often an incidental finding. Current evidence demonstrates that CABG alone does not completely correct ischemic MR. One of the early seminal studies describing ischemic MR found that 40% of patients with moderate MR who underwent CABG alone were left with moderate or severe (3 to 4+) residual ischemic MR, and an additional 50% of patients had persistent mild (2+) ischemic MR.

More recently, data from the Surgical Treatment for Ischemic Heart Failure Trial, which enrolled patients with a LVEF ≤35% and coronary artery disease amenable to CABG, confirmed that mitral valve surgery with CABG is associated with better survival compared with CABG alone (hazard ratio 0.41, 95% confidence interval 0.22 to 0.77). These data have been criticized as biased secondary to confounding by indication. As yet, no prospective randomized data are available, although several such studies are ongoing.

SURGICAL MANAGEMENT

Perioperative Considerations

Standard monitoring is used in patients undergoing combined mitral valve repair and CABG. Insertion of a Swan–Ganz catheter allows assessments of pulmonary pressures and cardiac output, which is particularly helpful in patients with left or right ventricular dysfunction, or pulmonary hypertension. TEE also allows further assessments of the mechanism of MR and LV function. Epiaortic scanning of the ascending aorta is also recommended to rule out the presence of atherosclerotic lesions before aortic cannulation.

Surgical Approaches and Cardiopulmonary Bypass

Median sternotomy is the surgical approach of choice in patients undergoing combined mitral valve repair and CABG, as well as in the reoperative setting. A right *anterolateral thoracotomy* may be used in select patients referred for isolated ischemic MR with prior CABG and patent grafts. These patients are placed in a 30 degree left lateral decubitus position, and a 12 to 15 cm right anterolateral thoracotomy is performed through the fourth intercostal space. Because the right thoracotomy approach does not require extensive mediastinal dissection, it is also an alternative if dense mediastinal adhesions are suspected (e.g., previous mediastinitis). In general, a right thoracotomy is contraindicated in patients with severe chronic obstructive pulmonary disease, moderate-to-severe aortic insufficiency, or a history of previous right-sided chest surgery.

Myocardial Protection

Mitral valve surgery is performed on cardiopulmonary bypass with intermittent antegrade or combined antegrade and retrograde blood cardioplegic arrest. Further myocardial protection can be obtained with moderate systemic hypothermia and local hypothermia with topical ice. In reoperative mitral valve surgery, specifically through a right anterolateral thoracotomy, it may be difficult to cross-clamp the ascending aorta. Therefore, other types of myocardial

management such as beating-heart or moderate to deep hypothermia and fibrillatory arrest should be considered, provided that the degree of aortic insufficiency is not more than mild.

Exposure of the Mitral Valve

After completion of coronary bypass grafting, careful exposure of the mitral valve is essential before valve analysis. Our preferred approach is the interatrial approach through *Sondergaard's groove*. The interatrial groove is incised, and the two atria are dissected and divided up to the fossa ovalis. This dissection exposes the roof of the left atrium, which is incised close to the septum and, thus, mitral valve. A transseptal approach may be useful in selected patients with a small left atrium or prior aortic valve replacement.

MITRAL VALVE REPAIR

Mitral valve repair is the procedure of choice for correction of type IIIb ischemic MR. The main goal of valve repair is to restore a large surface of coaptation by reducing the septolateral dimension of the valve with a downsized remodeling (complete, rigid, or semirigid ring) annuloplasty.

Segmental Valve Analysis

The valvular apparatus is examined systematically with a nerve hook to assess tissue pliability and identify leaflet restriction. The anterior paracommissural scallop of the posterior leaflet (P1) constitutes the reference point. Applying traction to the free edge of other valvular segments and comparing them with P1 determines the extent of leaflet restriction. Using two hooks, it is then possible to confirm leaflet restriction (type IIIb dysfunction) due to posterior papillary muscle displacement, most commonly affecting the P2 and P3 segments. The mitral annulus is examined to assess the severity of *associated annular dilation*, which is common and may be asymmetric in type IIIb ischemic MR.

Undersized Remodeling Ring Annuloplasty for Type IIIb Ischemic Mitral Regurgitation

Remodeling annuloplasty using an undersized ring is the technique of choice in type IIIb dysfunction (Fig. 44.5). Most commonly, 2-0 braided sutures are used to implant an annuloplasty ring. In general, sutures in the septal and lateral portions of the annulus should be placed using a backhand orientation, and sutures in the anterior and medial portions of the annulus are placed with a forehand orientation. The full curve of the needle should be used to encourage deep, wide placement of individual sutures along the annulus. Because of the potential increased tension in the setting of type IIIb dysfunction with associated annular dilation, it is preferable to place the sutures very close together along the annulus, and suture crossover may be warranted. The anterior commissure is usually the most difficult area to expose and is usually approached last after placement of sutures along the septal, medial, and lateral portions of the annulus to place tension on prior sutures to expose this area of the annulus. Once sutures are placed around the annulus, standard ring sizers are used to select an appropriate ring size. Placing gentle traction on marginal chords in the A-2 portion of the anterior leaflet with a hook allows the height and surface

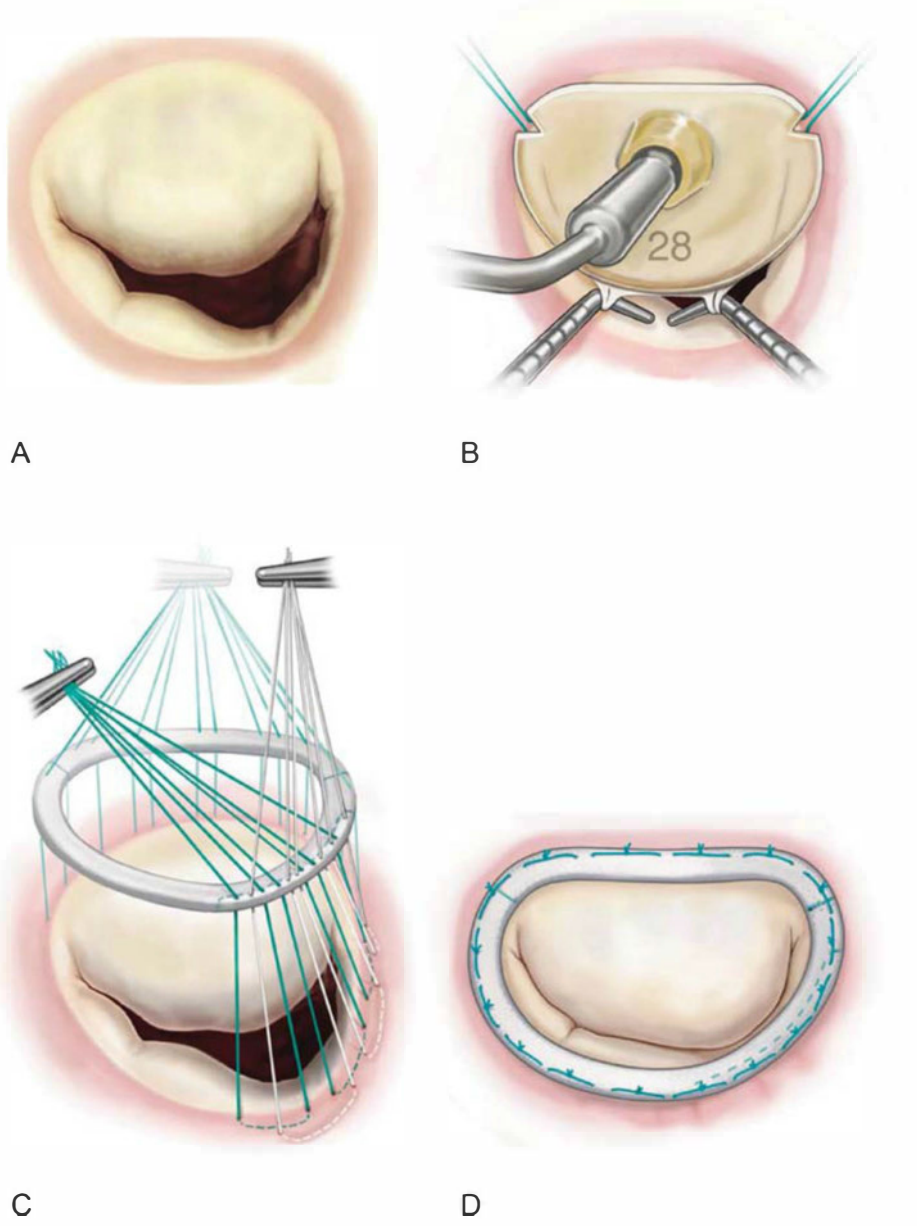


Fig. 44.5. Surgical approach to ischemic mitral regurgitation. Typical findings with leaflet restriction predominantly in the P2–P3 region (**A**). Sizing the annulus with a Carpentier–Edwards sizer is based primarily on the surface area and height of the anterior leaflet (**B**). One places 2-0 braided annular sutures into the mitral annulus, taking advantage of the full curve of the needle, with the angle directed toward the ventricle to ensure passage through the annulus. The sutures in the annulus at the position of the anterior commissure and trigone are placed last, taking advantage of previously placed sutures to expose this area (**C**). After placement of a full remodeling Carpentier–McCarthy–Adams IMR Etlogix ring, the surface of coaptation is restored (below the plane of the annulus) (**D**).

area of the anterior leaflet to be measured. An additional measurement to consider is the intercommissural distance. Because leaflet restriction in ischemic MR results in less leaflet tissue available for coaptation, it is necessary to downsize a *complete remodeling ring* such as the Carpentier–Edwards Physioring by one or two sizes or to use a true-sized Carpentier–McCarthy–Adams IMR Etlogix ring to ensure an adequate surface of coaptation following annuloplasty. The Etlogix ring combines the principles of undersizing with the specific asymmetric deformation observed in type IIIb ischemic MR. Compared with a symmetric Carpentier–Edwards Physioring, the Etlogix leads to increased leaflet coaptation due to the reduced anteroposterior (AP) dimension in patients with ischemic MR. This ring downsizes the D3 dimension by two sizes and the D2 dimension by one size compared with the Physioring. Systolic anterior motion will not occur despite aggressive downsizing, because the restricted posterior leaflet cannot displace the anterior leaflet into the outflow tract. After a ring is selected (typical size 24 to 28 mm), the interrupted annular sutures are passed through it, respecting the associated geometry of the annulus. The individual sutures are then tied, securing the ring to the annulus. Once the downsized remodeling annuloplasty is completed, a saline test is used to confirm the line of coaptation along the margin of the leaflets. Nearly the entire orifice is occupied by the anterior leaflet, allowing the entire restricted posterior leaflet to contribute to coaptation (Fig. 44.6).

De-airing Process

Careful de-airing at the end of the procedure is essential. Carbon dioxide insufflation is used in all patients to reduce intracardiac air. A small vent should be left across the mitral valve to facilitate de-airing after closure of the left atrium. When most of the air is expelled from the heart, the aortic cross-clamp is removed. Additional de-airing is performed during the reperfusion period. The aortic vent is maintained on suction until the patient is totally weaned from cardiopulmonary bypass and complete air removal is confirmed by TEE.

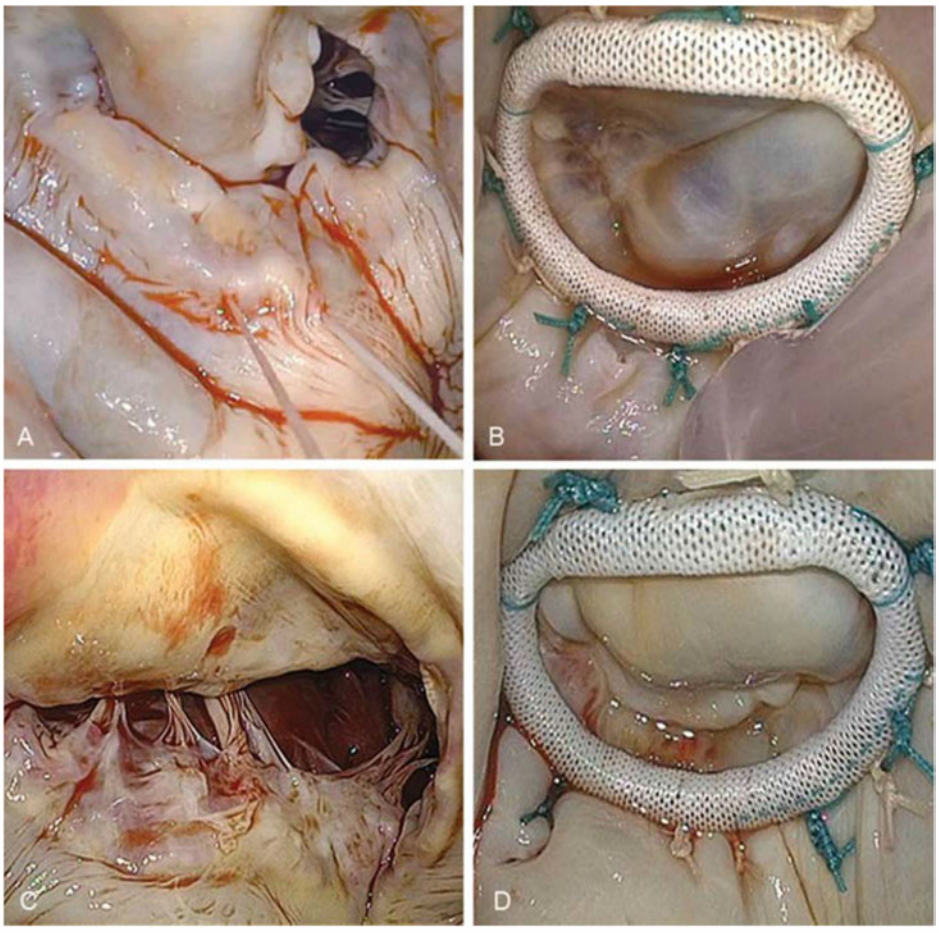


Fig. 44.6. Intraoperative views of two mitral valves with type IIIb dysfunction but different patterns of leaflet tethering (asymmetrical pattern—**A** and **B** versus symmetrical pattern—**C** and **D**). Note the angulation of the line of coaptation in patients with asymmetrical pattern (**B**).

RECURRENT MITRAL REGURGITATION FOLLOWING REPAIR

Recurrent regurgitation following repair of ischemic MR is complex and may be related to the viability of the coronary territory grafted, type of annuloplasty used at surgery, as well as anatomic factors related to the degree of ventricular remodeling. Advancements in imaging technologies have provided insights into the failure of mitral valve repair. One study using single photon emission computed tomography has shown that moderate ischemic MR typically improves when CABG is performed in patients with at least 5/16 viable segments. In fact, global LVEF alone does not completely predict recurrent MR. The detection of regional wall motion abnormalities is critical; as such quantitative measures such as Doppler strain are now used to predict mitral valve tenting.

Remodeling annuloplasty (complete, rigid, or semirigid ring) is also strongly recommended in patients with ischemic MR since flexible annuloplasty systems or suture annuloplasty techniques have been associated with recurrent MR. Midterm results on 100 patients undergoing combined CABG/mitral annuloplasty using a Duran flexible ring in patients with ischemic MR included a 29% rate of recurrent MR >2+ at a mean follow-up of 36 months. The Cleveland Clinic also reported a 30% incidence of recurrent 3+ or 4+ MR 18 months following mitral valve repair with the flexible Cosgrove–Edwards annuloplasty band. These observed failures in mitral valve repair may be related to the asymmetric tethering of the mitral valve, which is more severe toward P2–P3 in patients with ischemic MR. This observation led to the development of the Carpentier–McCarthy–Adams IMR Etlogix ring developed by Edwards Lifesciences (Irvine, CA). Clinical studies have confirmed that the Etlogix reduces the mitral tethering area and tenting height with survival free from recurrent MR ≥2+ being 95% at 15 months and 89% at 25 months. Echocardiographic factors associated with repair failure include posterior leaflet angle more than 45 degrees, distal anterior leaflet angle more than 25 degrees, tenting height more than 10 mm, and tenting area more than 2.5 cm^2. The size of the left ventricle may also have prognostic importance, with an end-diastolic dimension >65 mm associated with poor reverse remodeling following CABG even in the presence of viable myocardium (Table 44.2).

Table 44.2 Echocardiographic Predictors of Failed Mitral Valve Repair for Ischemic MR

Length of coaptation	≥1 cm
Tenting area	>2.5 cm^2
Tenting height	>10 mm
Distal anterior leaflet angle	>25 degrees
Posterolateral angle	>45 degrees
Distance between papillary muscles	>20 mm
Ventricular akinesia	
Left ventricular end diastolic diameter	>65 mm
Left ventricular end systolic diameter	>51 mm
Sphericity index	>0.7

FUNCTIONAL MITRAL STENOSIS

In a recent study, 24 patients with ischemic MR who underwent restrictive ring annuloplasty were subjected to dobutamine stress echocardiography and a 6-minute walk test. None of these patients had MR, but compared with control patients, those who underwent mitral repair had higher resting and stress peak mitral valve gradients. Six-minute walk distances were also shorter in patients who underwent restrictive ring annuloplasty. The long-term significance of functional mitral stenosis, however, remains unclear. Other authors have suggested that persistent pulmonary hypertension following mitral valve repair of ischemic MR is due to left ventricle dysfunction rather than persistent mitral valve gradients.

ADJUNCTIVE TECHNIQUES

It should be remembered that revascularization of all viable myocardial segments is recognized to reduce ischemic MR and should be considered an important "adjunct procedure." Cutting of secondary chords has been shown to decrease recurrent ischemic MR, although long-term data supporting this approach are lacking. Others have described patch augmentation of the posterior leaflet in order to increase the coaptation surface. Ischemic MR has been treated with papillary muscle repositioning using a pledgeted suture attached to the annulus, or interventricular papillary transposition. Resection of secondary chordae, ventricular plication, and septolateral "cinching" have also been shown to be of benefit in preclinical models. In our own practice, in addition to downsized rigid ring annuloplasty, we tend to cut the posterior strut chord to the anterior leaflet if we see a "hockey stick" deformity and tethering of the body of the anterior leaflet, in addition to closure of all clefts and indentations in the posterior leaflet if severe leaflet tethering is present (Fig. 44.7). If a residual leak is still present, we will cut restricted marginal chords, and either attach the leaflet margin to an adjacent chord, or add a neochordae.

New technologies as adjuncts to traditional surgery have also been reported. The Randomized Evaluation of a Surgical Treatment for Off-Pump Repair of the Mitral Valve study suggests that the Coapsys device (Myocor, Inc., Maple Grove, MN) decreases left ventricle volumes and MR at 2 years while also improving survival. Five-year follow-up with the Acorn CorCap device (Acorn Cardiovascular, St. Paul, MN) suggests that addition of this device to mitral valve surgery results in a greater reduction in left ventricle volume compared with mitral valve surgery alone. Neither of these devices are currently approved for use in the United States.

MITRAL VALVE REPAIR VERSUS REPLACEMENT

The higher rates of recurrent MR following repair in patients with ischemic MR compared with other causes of MR have led some to consider valve replacement instead of repair. Although some have found no difference in late survival between treatment approaches, others have found a benefit with repair. A recent meta-analysis of nine studies conducted between 2004 and 2009 suggests that mitral valve repair is associated with better early and late survival compared with replacement in patients with ischemic MR.

RESULTS

Operative Mortality

Early outcomes following CABG and mitral valve surgery in patients with ischemic MR have improved over the last 20 years. Some series have reported no difference in early mortality between patients receiving valve repair compared with valve replacement,

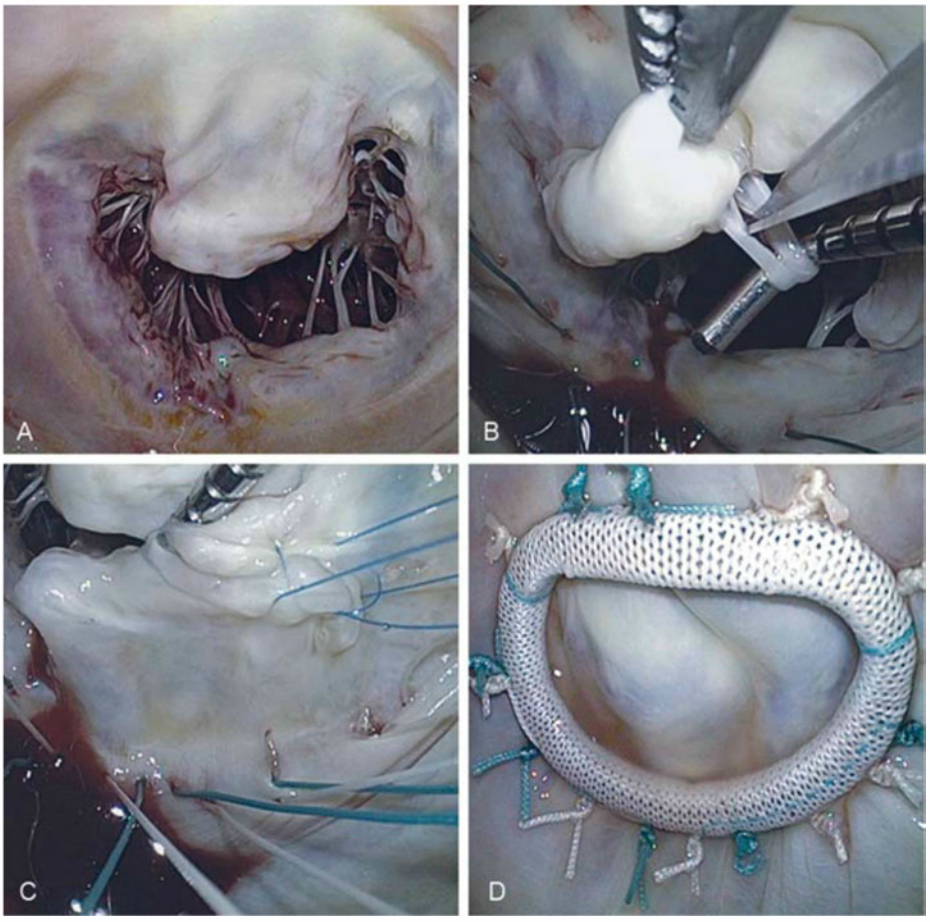


Fig. 44.7. In the setting of ischemic mitral valve disease (**A**), Adjunctive techniques in addition to downsized rigid ring annuloplasty (**D**) include cutting the posterior strut chord to the anterior leaflet if we see a "hockey stick" deformity and tethering of the body of the anterior leaflet (**B**) and closure of all clefts and indentations (**C**).

whereas others have shown a difference of more than 10%. Overall, operative mortality of patients undergoing cardiac surgery with ischemic MR ranges between 2% and 18%. Recent data from the Society of Thoracic Surgeons demonstrate an early mortality of 7.4% for patients undergoing mitral valve repair and CABG compared with 11.6% for patients undergoing mitral valve replacement and CABG.

Late Outcomes

Late survival for patients with ischemic MR is worse compared with patients with MR due to other causes. Medium-term (3 to 5 years) survival ranges from 60% to 85% depending on the risk profile of the population studied. In patients with less than severe MR, mitral valve surgery concomitant to CABG is associated with better long-term survival.

TYPE II ISCHEMIC MITRAL REGURGITATION

A minority of patients may present with type II ischemic MR. Leaflet prolapse results from partial or complete rupture of one papillary muscle, papillary muscle elongation, or, more rarely, chordal rupture.

Pathophysiology

Papillary muscle rupture is an increasingly rare mechanical complication of myocardial infarction. Papillary muscle rupture can occur during the acute phase of myocardial infarction; however, most patients present within 2 to 7 days after the initial ischemic insult. The rupture typically involves the posteromedial papillary muscle (75% vs. 25% for anterolateral papillary muscle) due to its singular vascular blood supply (either the right coronary artery or the circumflex artery in right- or left-dominant systems, respectively). In contrast, the anterolateral papillary muscle is supplied by the left anterior descending and circumflex arteries. Complete papillary muscle rupture will result in bileaflet prolapse with severe MR. Partial rupture of one papillary muscle involving one or more heads remains the most common lesion in patients with type II ischemic MR. Myocardial infarction often involves a limited area of the myocardium, which may explain the relatively well-preserved left ventricular function in most patients. Chronic *papillary muscle elongation* with leaflet prolapse due to remote myocardial infarction is also rare, and results from fibrotic transformation of a papillary muscle after myocardial infarction.

Clinical Presentation and Diagnosis

Patients with papillary muscle rupture often present with sudden development of congestive heart failure and cardiogenic shock. It is important to note that the sudden appearance of a systolic murmur and hemodynamic compromise may result from acute severe ischemic MR secondary to papillary muscle rupture as well as from ventricular septal rupture.

Clinically, it may be difficult to distinguish between these two entities despite differences in the characteristics of murmurs (the systolic murmur associated with ventricular septal rupture is loud, prominent at the left sternal border, and associated with a thrill, whereas the systolic murmur of ischemic MR due to papillary muscle rupture is softer, intense at the apex, and without thrill). Extensive myocardial infarction with cardiogenic shock associated with varying degrees of ischemic MR without papillary muscle rupture (acute type IIIb) is also part of the differential diagnosis. Preoperative examinations including two-dimensional transthoracic echocardiography, TEE, and cardiac catheterization are crucial to making the accurate diagnosis of ischemic MR due to papillary muscle.

Medical and Surgical Treatment

The principal goal of preoperative medical management is hemodynamic stabilization. This is best achieved with the use of inotropic agents and prompts insertion of an intra-aortic balloon pump. However, this stabilization phase should not delay surgical intervention in the form of mitral valve repair/replacement and coronary revascularization.

Chordal-sparring mitral valve replacement is the procedure of choice in most patients. This technique preserves postoperative left ventricular function, contributing to improved long-term survival of these patients.

Mitral valve repair using classic Carpentier's techniques can be applied to patients with papillary muscle elongation. Papillary muscle shortening is best performed by resecting the appropriate length of papillary muscle and suturing the fibrous cuff supporting the chords to the papillary muscle. Four or five circumferential simple sutures without pledgets should be used for this fixation. If the elongation involves only one papillary muscle head resulting in the prolapse of one valve segment, more conventional techniques of repair such as leaflet resection combined with sliding plasty or annular plication can be performed safely. Papillary tip transposition, which involves sewing the fibrous tip of the elongated muscle to a papillary tip at the normal height, can also be considered.

Results for Type II Ischemic Mitral Regurgitation

Published series have reported an operative mortality as high as 19%, with 5-year survival approximately 50%. Given the rarity of type II ischemic MR, most studies have involved small numbers of patients and included both mitral repair and replacement. More contemporary studies suggest that a perioperative mortality of close to 0% may be possible.

SUMMARY

Ischemic MR is a progressive and dynamic disease with important clinical implications for patients who present either acutely or in the chronic setting after myocardial infarction. As our understanding of ischemic MR evolves, the technologies used to manage these patients will continue to be refined. Currently, mitral valve repair can be performed with favorable early and late results. Despite the wealth of retrospective data, no prospective randomized data are available describing outcomes following the surgical correction of ischemic MR. Undoubtedly, our understanding of this challenging disease will evolve as these data are available.

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EDITOR'S COMMENTS

Dr. Adams and his colleagues have truly been experts in the surgical treatment of ischemic mitral regurgitation. They have done an outstanding job discussing the pathophysiology as well as appropriate surgical types of repair. The point that I would like to emphasize include proper definition of mechanisms as well as the annular downsizing approach. I totally agree with the authors that a semirigid ring is preferably used in this particular situation.

The difficult area is deciding between repair and replacement for these types of patients. The positive for repair include that there seems to be some survival disadvantage. The down side includes a higher recurrence rate than replacement. These issues I believe will be settled based on randomized trials including through the cardiothoracic surgical network. The data will be analyzed and hopefully we will be able to learn the best practice approach for this issue. My own bias is that those patients with severe tethering probably have the highest recurrence rate. If one is not going to do a procedure to deal with the tethering in addition to the treatment of annular dilatation, then these patients should be considered for chordal-sparing mitral valve replacement.

ILK

45 Percutaneous Mitral Valve Repair

D. Scott Lim and Gorav Ailawadi

INTRODUCTION

As the preceding decades have witnessed the maturity of surgical therapies for valve and coronary artery disease, the next decade will witness a tremendous change in our approach to percutaneous therapies for heart valve disease. Recently, we have seen a remarkably explosive growth of percutaneously implanted heart valves in Europe, and a myriad of novel devices being developed for repair or replacement of diseased heart valves from a transcatheter approach. This chapter will focus on novel percutaneous options for repair of mitral valve regurgitation.

PERSPECTIVE ON MITRAL REGURGITATION

Mitral regurgitation (MR), from nonrheumatic etiology, may be due to either degenerative valve disease, such as prolapse, chordal rupture, or myxomatous degeneration, or functional etiologies such as ischemic cardiomyopathy and secondary MR. Additionally, as transcatheter therapies focus on functional etiologies of MR, our knowledge base of how the disease of the ventricle affects the mitral apparatus. Significant MR (3 or 4+) occurs in 0.5% of the population, with approximately 250,000 new cases annually in the United States. For patients with significant MR, medical therapy has been shown to be ineffective in treating the underlying pathophysiology and is unable to retard disease progression. Surgical repair or replacement is the standard of care, as it is efficacious and low risk. However, it has been estimated that only ~20% of patients with significant MR undergo surgery. Therefore, there is a significant unmet therapeutic need for patients with mitral valve regurgitation, and particularly for patients at high surgical risk, a significant opportunity for novel catheter-based therapies.

HISTORICAL PERSPECTIVE ON PERCUTANEOUS THERAPIES FOR MITRAL VALVE DISEASE

The very first percutaneous therapy for mitral valve disease was the balloon mitral commissurotomy for rheumatic mitral stenosis. Since then, the technique has matured with the use of specific shaped balloons, and a better understanding of the types of rheumatic valve disease amenable to this therapy, as well as the long-term outcomes from this percutaneous approach. However, it is also important to note that the skills and techniques involved with this procedure are complex and involve a transseptal puncture, which is not commonly performed by most physicians trained today (Fig. 45.1).

Adapted from a Surgical Approach to Mitral Regurgitation

Currently, surgery for MR has become a mature therapy, with a wide range of different techniques involving leaflet, chordal, and annular approaches. Concepts behind each of these approaches have been utilized by novel transcatheter approaches to repair of MR. Additionally, work continues to create a transcatheter replacement option for mitral valve disease.

Indirect Annular Approaches to Mitral Regurgitation Therapy

The concept of a percutaneous annular approach mimics the surgical concepts of annular diameter reduction and bringing the posterior part of the annulus anteriorly, thereby pushing the posterior leaflet forward to create a better coaptation with the anterior leaflet.

While later percutaneous devices have focused on direct approaches to the annulus, early designs have attempted to follow the course of the coronary sinus. However, they have been limited by the anatomic fact that the coronary sinus does not run in the same plane as the mitral annulus, as well as the incidence of circumflex artery crossing the coronary sinus.

The coronary sinus devices were introduced from a percutaneous, transjugular approach. One of the first such devices tested was the Edwards Lifesciences MONARC system—it was implanted in the coronary sinus with distal and proximal self-expanding stent anchors, and had a variable length connecting bridge. The connecting bridge had dissolvable polymers that would foreshorten over a period of weeks, changing the radius of curvature of the coronary sinus and bringing the posterior portion more anterior. It was studied in 72 patients—mainly with moderate degrees of MR, but with limited efficacy (able to be implanted in 82%, and a sustained reduction of mitral regurgitaiton by 1 grade in ~40%) and an incidence of complications of 28% at 2 years. Likely due to the inability to study the affects and risks of this device acutely, further study on this device was stopped.

Other coronary sinus devices such as Cardiac Dimensions' Carillon device has had more success. It too is implanted with proximal and distal coronary sinus anchors, but with acutely observable affects on MR and is retrievable if needed due to inadequate efficacy or complication. It was studied in the European AMADEUS and TITAN trials in small numbers of patients also with mainly moderate degrees of functional MR. It was able to be implanted in ~66% of intended patients, and an ultimately low incidence of complications (~13% in the early AMADEUS trial and decreasing to ~2% in the later TITAN trial). Its efficacy was limited (~1 grade improvement in indices of MR) but promising in the appropriately selected patients. It has recently been awarded regulatory approval in Europe.

Similarly, Viacor's PTMA coronary sinus device was studied in the PTOLEMY 1&2 trials, demonstrating in a small number of

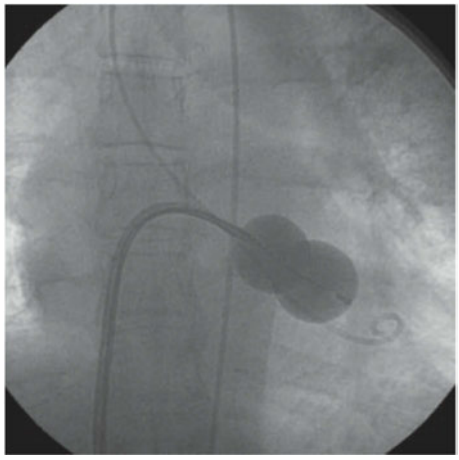


Fig. 45.1. Percutaneous transvenous transseptal approach is used for mitral balloon commissurotomy in a patient with rheumatic mitral stenosis. The Inoue balloon catheter (Toray Medical, Houston, TX) is utilized for this successful palliation.

patients low adverse events and an average of 1 grade MR reduction. It remains at present in early clinical study.

Direct Annular approaches to Mitral Regurgitation Therapy

Other percutaneous therapies have utilized sutures to plicate or wires to cinch the annulus directly, with catheters coming at the annulus from both the left atrium and left ventricle. However, these therapies remain in early research stages. A potentially promising approach uses radiofrequency energy to cause scarring and contraction of the collagen in the annulus. While this technology has remained in relatively early preclinical trials, it holds promise in that no hardware is implanted and the therapy may be applied to the annulus repeatedly.

Leaflet Approaches

One approach, which ultimately matured into a catheter-based therapy, is the edge-to-edge approach as described by Alfieri's group. In this surgical leaflet therapy, the central edges of the anterior and posterior leaflets are anastomosed creating a double-orifice mitral inflow. The concepts of this edge-to-edge approach were used by both the Edwards Lifescience's Mobius device and Abbott Vascular's MitraClip to create some of the first novel catheter-based therapies for MR. The Mobius device utilized a 17F delivery system from a percutaneous, transvenous, and transseptal approach. Its therapy catheter contained a vacuum port which was used to grab the leaflets, through which it would then throw a stitch, secured with a nitinol clip, mimicing the Alfieri edge-to-edge approach. This device was evaluated in the Milano II trial, which demonstrated limited efficacy both acutely and at 30 days, ultimately leading to its abandonment.

The MitraClip's early design iterations attempted to duplicate this same suture-based approach to leaflet apposition, and involved using hollow needles to pierce the central portions of the anterior and posterior leaflets to deliver pledgeted sutures (Fig. 45.2). This technique worked on the open, nonbeating heart but it was soon realized that an independent mechanism to grasp the leaflets while they were in motion was needed to be incorporated into the design concept.

MITRACLIP SYSTEM

This novel percutaneous approach was the first achieving success in prehuman and clinical trials for treatment of patients with nonrheumatic MR. The concept of leaflet grasping by a clip mechanism on the regurgitant portions of the mitral valve became the hallmark of the final design freeze of the MitraClip (Fig. 45.3; Abbott Vascular, Santa Clara, CA). The MitraClip is introduced from a percutaneous, transvenous, and transseptal approach to the mitral valve (Fig. 45.4). The traditional imaging modality in the catheterization laboratory of fluoroscopy is of limited utility in this procedure, as it cannot visualize the mitral leaflets. Therefore, the procedure is guided by simultaneous transesophageal imaging, ideally using both two- and three-dimensional echocardiography (see Figs. 45.5 and 45.6). Combined with the reality that the MitraClip delivery system is manipulated by a mechanical-based device, it is more important that the valve interventionalist be skilled in transesophageal imaging than the standard catheter and wire skills.

By placing the MitraClip on the central portions of the anterior and posterior leaflets, it acts to anchor prolapsing or flail segments, as well as coapt tethered leaflets so that it reduces the time and force required to close the valve. By decreasing MR, the left ventricular volumes are in turn reduced, leading to beneficial left ventricular remodeling. Anatomically, the MitraClip creates a tissue bridge between the two leaflets, which limits dilation of the mitral annulus in the septal-lateral dimension, which supports the durability of this repair.

EVEREST I AND II TRIALS

This MitraClip approach to percutaneous mitral valve repair was evaluated in the EVEREST trials. EVEREST I was the safety and feasibility registry, which enrolled 55 patients in a nonrandomized clinical trial. EVEREST II was the pivotal clinical trial in which 279 patients with 3 or 4+ MR were randomized to either the MitraClip therapy or standard surgical therapy (either mitral valve repair or replacement). It is important to note that this involved low- and moderate-risk patients, in comparison to the EVEREST—High Risk Registry, which enrolled 79 patients who were nonoperative candidates to the MitraClip therapy. In the early nonrandomized experience with the MitraClip, procedural success (reduction of MR to 2+ or less) was achieved in 74%, and 66% were free from death, mitral valve surgery, or MR >2+ at 12 months. These outcomes were similar for those with either degenerative (79%) or functional (21%) etiology of MR. It is also important to note that this represents the initial experience with this novel technology, which has a steep learning curve.

The randomized EVEREST II trial included operative-candidate patients with either moderately severe or severe MR. Patients were either symptomatic, with a left ventricular ejection fraction (LVEF) >25% and left ventricular end-systolic dimension (LVESD) of ≤55 mm, or asymptomatic with ventricular dysfunction (defined as LVEF 25% to 60% or LVESD ≥40 mm), or pulmonary hypertension. Patients were excluded if they had need for other cardiac surgery, recent acute myocardial infarction (within 12 weeks), severe renal insufficiency (creatinine >2.5 mg/dl), endocarditis, or rheumatic etiology of valvar dysfunction. Additionally, patients were excluded for certain anatomical issues of the mitral valve—stenosis with mitral valve area <4.0 cm^2, a severely broad flail width (≥15 mm) and flail gap (≥10 mm), or deficient coaptation length (<2 mm).

Compared with patients from either the Society of Thoracic Surgery database or those undergoing first time elective mitral valve surgery, those enrolling in the EVEREST II trial were significantly older, with more comorbidities. This is likely due to the preference for the initial experience with novel percutaneous therapies to be reserved for patients who are less ideal surgical candidates. This randomized trial was designed to have a primary safety endpoint powered for a superiority hypothesis, for major adverse events at 30 days. The primary effectiveness

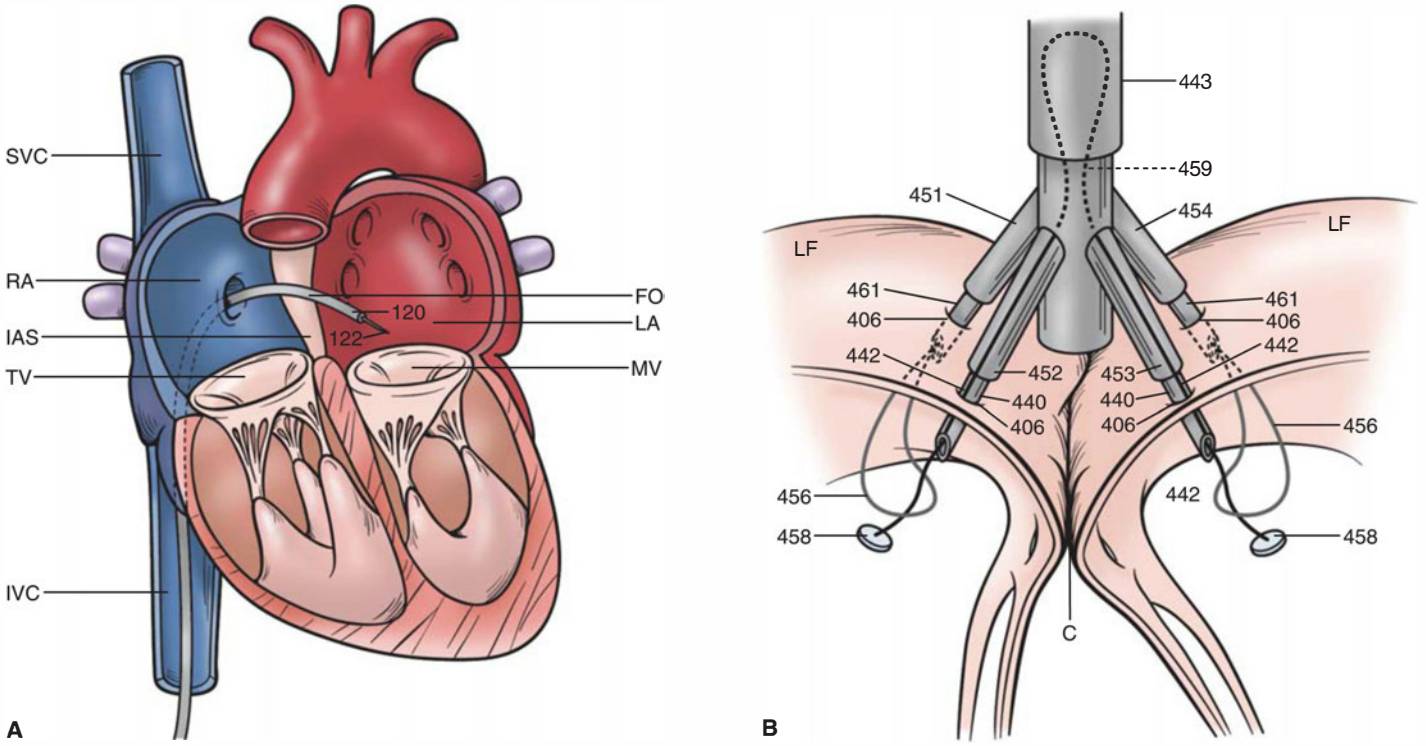


Fig. 45.2. Initial patent drawings of an early design iteration are shown in this device to create a percutaneous mitral valve repair. The idea behind this device is to use hollow needles to perforate the anterior and posterior mitral valve leaflets, and then throw pledgeted sutures, mimicking the Alfieri-type repair. Unfortunately, this early design did not account for mobility of the leaflets in the real patient, and a separate grasping mechanism was required. SVC: superior vena cava; RA: right atrium; IAS: interatrial septum; TV: tricuspid valve; IVC: inferior vena cava; FO: foramen ovale; LA: left atrium; MV: mitral valve; LF: valve leaflet.

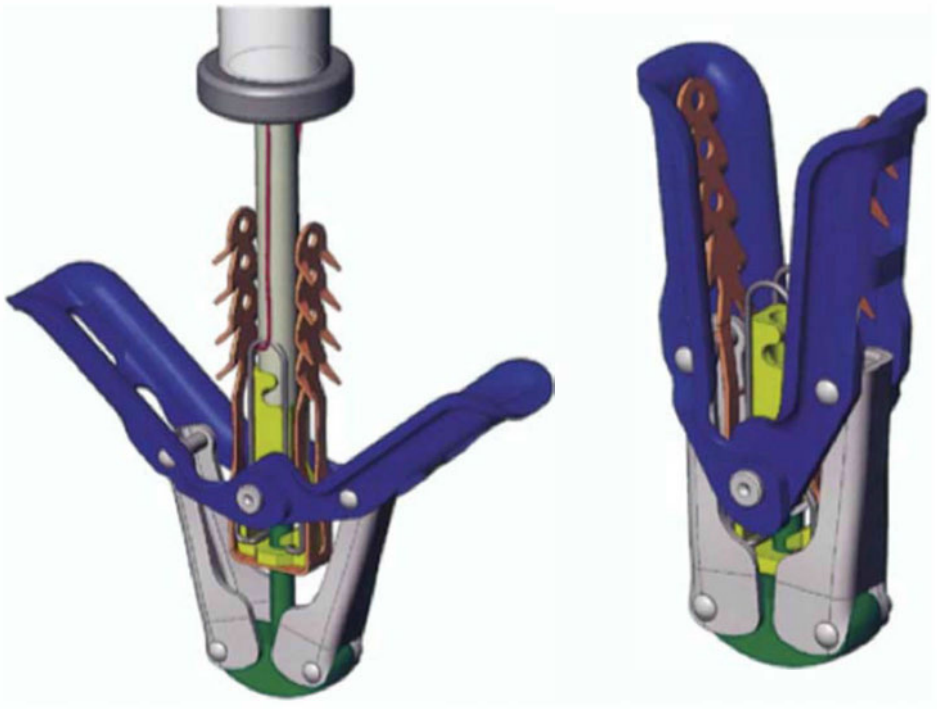


Fig. 45.3. A schematic view of the MitraClip is shown, which is used for percutaneous repair of mitral regurgitation. The *blue* arms are mechanically hinged and allow the operator to change the angle between them. These arms are designed to be pulled up from the ventricular side of the mitral leaflets to bring them together. The *reddish* spiked frictional elements may be lowered onto the atrial side of the leaflets, thereby sandwiching each leaflet between the *blue* arm and the *red* frictional element. Image provided courtesy of Abbott Vascular Structural Heart.

endpoint was powered for a noninferiority hypothesis, for the 12-month composite of freedom from death, mitral valve surgery or reoperation, or MR >2+. Additional endpoints included echocardiographic-assessed MR severity, left ventricular function, New York Heart Association functional class, and quality-of-life indices.

From the primary safety endpoint, by an intention-to-treat statistical analysis, those

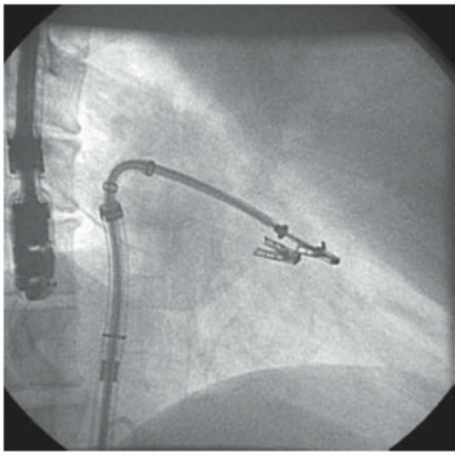


Fig. 45.4. Fluoroscopic image demonstrates the transvenous, transseptal approach to introduce the MitraClip to the mitral valve.

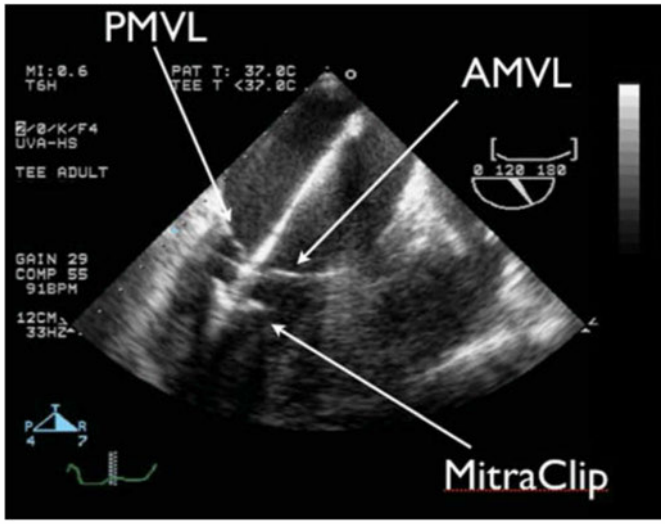


Fig. 45.5. Two-dimensional transesophageal echocardiographic image of the MitraClip and its orientation to the anterior and posterior mitral valve leaflets is shown. This three-chamber view of the mitral valve is used to orient the MitraClip in an anterior–posterior direction on the valve.

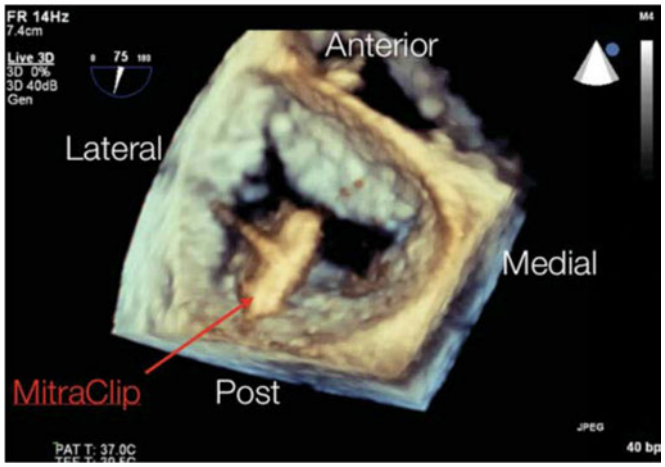


Fig. 45.6. Three-dimensional transesophageal echocardiographic image of the MitraClip and its orientation to the mitral valve is shown. The arms of the MitraClip are oriented perpendicular to the line of coaptation of the valve, and over the point of pathology. To maintain operator orientation, the atrial septum is on the *right* side of the screen, with the aortic valve at approximately 12 o'clock position.

randomized to the MitraClip arm had a major adverse event rate of 15% at 30 days, versus those in the surgical arm had an event rate of 48% ($P_{sup} < 0.0001$). Further detailed analyses of the major adverse events such as death, stroke, re-operation, or emergent surgery demonstrated no such events occurring in patients receiving the MitraClip alone, but occurred in those crossing over to surgical therapy following unsuccessful MitraClip therapy. Therefore, in a per protocol analysis, which evaluates just the performance of the device alone rather than a strategy approach, the major adverse event rate was under 10% at 30 days.

From the primary effectiveness endpoint, in terms of the clinical composite of freedom from death, mitral valve surgery/reoperation, or MR >2+ at 12 months by an intention-to-treat analysis, the MitraClip arm had a success rate of 55% versus the surgical arm had a success rate of 73% ($P_{NI} < 0.0012$). Further analysis of the events driving the composite endpoint demonstrates that there was no significant difference between the MitraClip arm and the surgical control arm with respect to death or freedom from severe MR, but that the difference in composite endpoints was due to a difference in subsequent surgery for mitral valve dysfunction ($P < 0.001$). MR reduction to the goal and maintenance at 12 months of 2+ or less was achieved in 81% of the MitraClip patients, versus 97% of the surgical patients, although this was achieved by mitral valve replacement in 12% of surgical patients. Despite this difference in echocardiographic-measured MR reduction, there were similar reductions in left ventricular dimensions and volumes achieved at 12 months. Additionally and paradoxically, despite the differences in apparent MR reduction, there were a greater percentage of patients from the MitraClip arm who were asymptomatic or minimally symptomatic at 12 months (98%) versus those in the surgical arm (88%). Some of these apparent inconsistencies may be due to the difficulties in quantifying the degree of residual MR after a MitraClip has been placed in the center of the valve.

Quality-of-life indices, as measured by an SF-36 survey, demonstrated improved scores on both physical and mental evaluations in the MitraClip arm at 30 days, versus impaired physical and indeterminate mental scores at 30 days in the surgical arm. At 12 months, there were improvements in all study arms.

EVEREST HIGH-RISK REGISTRY

A separate U.S. registry was created for nonoperative patients with severe MR, from either degenerative (prolapse or flail) or functional etiologies (MR secondary to ischemic or nonischemic cardiomyopathy)—the EVEREST High Risk Registry. This registry enrolled 79 patients rapidly, as there was significant support for it from surgical colleagues. The majority of patients in the High Risk Registry had functional etiology of their MR, as would be expected from finding more patients with ischemic etiologies in a higher risk subgroup. Additionally, patients were older (average age 76 years with 68% over age 75) and more symptomatic (New York Heart Association functional class III or IV in 89%) than the randomized EVEREST population.

Data presented showed that while at 30 days the predicted mortality for the group was 18%, the actual mortality was under 8%, with 76% surviving at 1 year and 79% of the survivors in New York Heart Association symptom class I or II. In a nonrandomized comparison to similar high-risk patients treated medically, there was a survival advantage at a year, with 76% alive in the MitraClip group versus 55% in the medically treated group ($P = 0.037$). These data give support to the concept that particularly for these nonoperative patients, novel percutaneous options are attractive.

In Europe, along with the MitraClip having become a commercially available therapy, there is an ongoing

nonrandomized registry collecting clinical and health economic data on treatment options for MR. Based on physician and patient choice, patients either receive MitraClip therapy, medical management, or standard surgical therapy. The patients undergoing MitraClip therapy continue to be older (41% of an age >75 years), with more comorbidities (49% cardiomyopathy, 45% with renal disease, and 19% with lung disease), and at high risk (average logistic EuroSCORE predicting mortality in 20%). Interestingly, unlike the U.S. experience, 87% had functional etiology (either underlying ischemic or nonischemic cardiomyopathy causes of MR) and 41% had LVEFs between 10% and 30%. It is evident that the sweet spot for patient selection for this therapy is those patients that are not ideal operative candidates.

Not all patients who have undergone percutaneous mitral valve repair with the MitraClip will have adequate reduction of MR nor perfect durability of that repair. Clearly, the time and energy spent up front, with appropriate patient selection, will pay dividends in terms of outcomes. However, for those patients who have inadequate or nonsustained MR reduction after MitraClip implantation, subsequent surgical repair has been performed. Rate of surgical repair of MR following MitraClip implantation was 89% ($n = 20$), which compares favorably to the repair rate in those randomized to the surgical control arm of EVEREST II (86%, $n = 69$), or as found in the 2007 Society of Thoracic Surgery database of 59%. Anterior or bileaflet disease was an independent predictor of mitral valve replacement following MitraClip therapy, but age, etiology, or surgeon experience was not. Additionally, successful mitral valve surgery has been performed up to 5 years after MitraClip implant.

COLLABORATIVE EFFORTS

Many of the aspects of percutaneous mitral valve therapies rely on skills and technology not usually available to the standard cardiac interventionalist. Therefore, collaboration between interventionalists, echocardiologists, and cardiac surgeons is imperative for centers wishing to become involved in these exciting, but early stage technologies. It was no accident that the first U.S. trial of the transcatheter aortic valve was called PARTNER—emphasizing the very close working relationship required across disciplines.

SUMMARY

Just as the previous decade comprised a marked explosion in the utility and maturity of transcatheter interventions for coronary artery disease, the next decade will bear a similar focus on transcatheter approaches to heart valve disease. The MitraClip system represents a first-generation step in this direction and is remarkable for its success as such. As is true with many things, the pendulum of excitement for these novel technologies will initially swing a bit farther than where the real utility of these approaches will truly lie, and careful patient selection will be imperative.

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EDITOR'S COMMENTS

Dr. Scott Lim is a structural cardiologist and a very important colleague of mine. He happens to be a skilled technician and had outstanding results with percutaneous mitral valve repair. This type of technology sounds like it should clearly be a failure. However, I must say that it has been extremely useful in some very complex patients and it definitely has a role in the treatment of mitral valve regurgitation.

The advantages to the percutaneous approach are very little mortality or morbidity. The down side is that it is not a perfect repair. Most patients had some residual mitral regurgitation and certainly the technology does not deal with all the mechanisms of mitral regurgitation.

Having said all that, there are patients that clearly are at high risk for open mitral surgery. They are patients who could have been easily treated with this technology. Many of them do not need a perfect repair and clearly patients improve with this technology. The real difficulty is not to find patients that are inoperable but rather which patients who are operative but high risk but would be better suited to a percutaneous approach.

ILK

46 Tricuspid Valve

Gorav Ailawadi

INTRODUCTION

The tricuspid valve is arguably the least understood valve in the heart. Most often, the tricuspid valve is affected in the setting of left heart disease, specifically with mitral valvular disease. In this scenario, the pathophysiology of tricuspid valve insufficiency is a result of elevated left heart filling pressures with subsequent development of pulmonary hypertension leading to right ventricular (RV) hypertrophy or dysfunction, and ultimately tricuspid annular enlargement. This is often termed "functional tricuspid regurgitation" as it is due to a failure of coaptation of structurally normal leaflets. Although tricuspid repair, in general, is a simple technique, according to the Society for Thoracic Surgeons (STS) National database, the operative mortality in patients undergoing tricuspid valve surgery with or without concomitant operation is roughly 10%. Moreover, data from individual centers report an operative mortality for reoperations for recurrent TR in excess of 30%.

As such, many surgeons have trepidation in treating patients with tricuspid valve disease. In fact, the need for surgical repair of tricuspid regurgitation (TR) is unclear and often debated by the surgical and cardiology community. First, the usually low-pressure, right-sided circulation is tolerant to imperfect function of the tricuspid valve. Second, tricuspid valve regurgitation most often occurs in the setting of left-sided pathology, resulting in a longer and more complex procedure with additive risk. The argument against addressing the tricuspid valve also stems from the improvement in tricuspid valve function often seen after correction of left-sided pathology. However, accurate predictors of tricuspid valve improvement after sole correction of left-sided pathology have not been convincingly or prospectively studied.

Isolated tricuspid disease is much less common, especially in adults. Patients typically present with right-sided failure symptoms. Patients with fixed pulmonary hypertension, severe RV dysfunction, and severe TR with no left-sided pathology are often the most challenging and have the greatest risk. The prognosis in this especially high-risk group of patients can be poor due to RV dysfunction, and operative intervention is often associated with dismal outcomes. Primary leaflet abnormalities of the tricuspid valve are less common and are caused most commonly by infectious endocarditis and carcinoid heart disease. Finally, Ebstein's anomaly, although most commonly treated in childhood, may also be first diagnosed in adulthood in its less severe forms.

The ACC/AHA has provided guidelines for operative intervention for tricuspid disease. Class I indications for surgical intervention include (1) patients with severe TR undergoing left-sided surgery, (2) primary severe TR without severe RV dysfunction, and (3) severe primary or secondary tricuspid stenosis. Class IIa indications include patients with moderate TR in the setting of left-sided surgery with or without tricuspid annular dilation (>40 mm by echocardiography). It should be noted that all guidelines are based on level C evidence indicating that this field has been largely guided based on retrospective series.

EVALUATION AND OPERATIVE INDICATIONS

Functional Tricuspid Regurgitation

Tricuspid annular dilation of the tricuspid valve due to left-sided pathology leads to functional TR. Generally, the annulus adjacent to the septal leaflet does not dilate, whereas the anterior and especially the posterior annulus dilates causing functional TR. Echocardiography is the most helpful modality in evaluating tricuspid valve regurgitation and distinguishes between annular problems and primary leaflet problems. Evidence of severe tricuspid insufficiency includes dilation of the right atrium, right ventricle, and inferior vena cava, along with a large color flow jet across the tricuspid valve. Hepatic vein flow reversal is likewise diagnostic. It is important to note that the degree of TR can be quite variable based on loading conditions and fluid status. Preoperative coronary artery catheterization is usually done given the typical patient age. Importantly, the assessment of right-sided hemodynamics should be done to evaluate RV function and pulmonary hypertension as these are significant predictors of outcomes in patients with TR. It should be noted that the degree of tricuspid regurgitation is poorly assessed by cardiac catheterization, while tricuspid stenosis can be diagnosed with this invasive test.

We often perform tricuspid annuloplasty in patients with more than moderate (>2+) TR in the setting of mitral valve disease. Selected patients with concomitant 2+ TR are also treated if there is any (1) evidence of RV dysfunction/significant pulmonary hypertension, (2) significant tricuspid annular dilation (annulus >40 mm), (3) elevated central venous pressure (>18 mmHg), or (4) in patients undergoing concomitant maze procedure as we believe even lesser degrees of TR may be contributing to AF.

In patients with isolated tricuspid insufficiency, surgical correction is warranted when symptoms are refractory to maximal medical therapy in selected patients. Often these patients will have difficult-to-treat right-sided congestive heart failure with significant peripheral edema and clinically significant passive congestion of the liver. Intensified diuretic administration often results in worsening renal function rather than improvement in the patient's clinical status. Moreover, patients may

develop signs of liver dysfunction from passive congestion. Tricuspid operation should be cautioned in patients with severe liver dysfunction (MELD > 15) due to high operative mortality.

Endocarditis

Infectious endocarditis involving the tricuspid valve is the most frequent valve involved in cases of endocarditis secondary to intravenous (IV) drug abuse. Endocarditis in these patients involves the tricuspid valve in three-fourths of these patients, whereas involvement of the mitral and aortic valve occurs in one-fourth of cases. Pulmonic valve involvement is much less common, accounting for <1% of cases. The other common groups of patients who develop tricuspid endocarditis are those who develop infections of indwelling pacemakers and leads. Transthoracic echocardiography is typically sufficient in screening and evaluating for tricuspid endocarditis. Transesophageal echocardiography can be performed in cases where the tricuspid valve is not seen well by surface echocardiography.

Surgical intervention for IV drug abusers with isolated right-sided endocarditis should be performed with caution. First, imperfect tricuspid valve function is overall well tolerated in these patients as the right side is a low-pressure system. Second, the consequences of embolization from the tricuspid valve to the lungs are less catastrophic than those from embolization from the left-sided valves. Most importantly, the risk of ongoing IV drug abuse with ongoing risks including reinfection and overdose also may necessitate a conservative approach to surgery in these patients. Two important indications for surgery for tricuspid valve endocarditis are (1) endocarditis caused by microorganisms that are difficult to eradicate such as fungal organisms or bacteria resistant to antibiotic therapy and (2) patients with tricuspid valve vegetations >2 cm, a dilated right ventricle, and recurrent pulmonary emboli or right-sided heart failure. When surgery is necessary, debridement of infected tissue and preservation of the native valve is preferred when at all possible. Patients usually have significant degrees of valve regurgitation, and restoration of complete competence is neither possible nor necessary. Complete excision of the tricuspid valve will negate the risk of reinfection of an implanted prosthetic valve by continued IV drug use. This approach is not advocated by most surgeons since patients undergoing excision often have a challenging postoperative course and some eventually require prosthetic valve insertion secondary to poor cardiac output. Patients with pulmonary hypertension, sometimes related to multiple septic pulmonary emboli, will tolerate the absence of the tricuspid valve poorly.

In patients with infected pacemaker leads, the vegetations commonly involve the tricuspid valve. Diagnosis can be challenging in these patients as most patients with indwelling pacemaker leads have sterile vegetations on their leads without infection. Persistent positive blood cultures without other clear source or evidence of septic pulmonary emboli should prompt consideration for lead removal. While lead extraction alone may be sufficient therapy, in patients with large tricuspid valve vegetations (>2 cm) or significant TR, surgical removal and tricuspid repair should be considered. An alternative approach is the use of a hybrid approach with lead extraction by electrophysiologists and surgical tricuspid valve debridement and repair. Patients who require biventricular pacing may benefit from epicardial lead placement after tricuspid valve has been addressed and atrium closed. Great care must be taken to avoid cross-contamination of these new epicardial leads.

Carcinoid Heart Disease

Right-sided cardiac involvement is common in patients with carcinoid syndrome, although clinically significant lesions are much less frequent. Endocardial plaquing, the typical lesion seen in carcinoid heart disease, may involve either or both the pulmonic and tricuspid valves as well as the RV endocardial surface. Both tricuspid stenosis and regurgitation can occur in carcinoid disease. Evidence of tricuspid (and likely pulmonic) valve dysfunction and progressive right heart failure is the usual clinical presentation of patients considered for surgery. With current treatment of malignant carcinoid tumors, reasonable survival rates (>50% at 5 years) can be expected. Thus, patients with valvular dysfunction and progressive heart failure should be considered for valve replacement if their tumor is not imminently life-threatening.

Ebstein's Anomaly

Ebstein's anomaly causes varying degrees of TR and may be clinically silent or present at any time in children or adults. The lesion is characterized by downward displacement of the septal and often posterior leaflets of the valve into the right ventricle (Fig. 46.1). This gives the characteristic "atrialized" appearance of the right ventricle. The annular circumference is often quite large, and the right atrium is quite enlarged. The quality of the anterior leaflet is critical to the success of any attempted valve repair for Ebstein's anomaly. The anterior leaflet is generally large, sail-like, and relatively thin. A thickened, muscularized anterior leaflet makes a poor substrate for repair. Ebstein's anomaly valve may present along a spectrum of severity and may become clinically significant at any time during life and require surgical intervention.

Adults with less severe forms of Ebstein's may become symptomatic and require surgical intervention. In adults presenting with Ebstein's anomaly, symptoms may include arrhythmias, fatigability, dyspnea, and cyanosis. In addition, patients with Ebstein's anomaly may have an associated atrial septal defect. Echocardiography is useful for characterizing the degree of valvular insufficiency and the quality of the leaflets, specifically the quality of the anterior leaflet when contemplating valve repair. Repair of the valve is obviously favored when possible. Replacement is necessary in some cases, however. In the neonatal period, transplantation or single-ventricle palliation (Starne's procedure) may be required.

Anatomy of the Tricuspid Valve

The septal leaflet lies closest to the surgeon, the anterior leaflet away from the surgeon, and the posterior leaflet closest to the inferior vena caval cannula (Fig. 46.2). The anterior leaflet is quadrangular and is the largest of the three leaflets. Its chordae are derived from the anterior and posterior papillary muscles. The posterior leaflet generally is the smallest of the three leaflets and is triangular. Its chordae are also derived from the anterior and posterior papillary muscles. The septal leaflet is semicircular, with chordal insertion directly into the ventricular septum at its inlet and membranous portions. The membranous portion of the ventricular septum generally lies beneath the portion of the septal leaflet nearest to the anteroseptal commissure when looking in from the right atrium.

The atrioventricular node lies in the apex of the triangle of Koch, which is bounded by the septal annulus of the tricuspid valve, the coronary sinus, and the tendon of Todaro (Fig. 46.2). The bundle of His continues on from the atrioventricular node to pierce the central fibrous body that runs along the posteroinferior margin of the membranous septum. This close proximity of the atrioventricular node to the septal leaflet is of prime

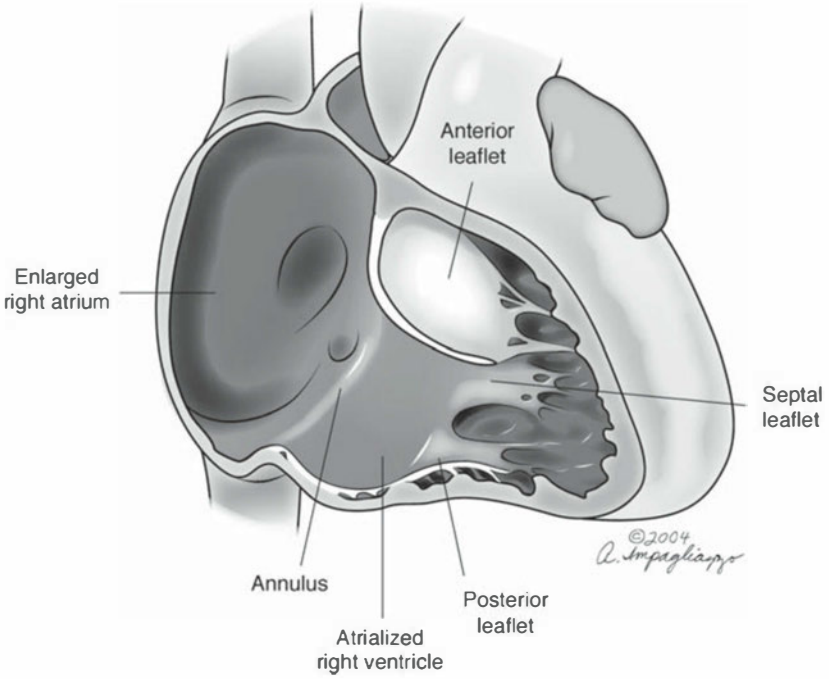


Fig. 46.1. Ebstein's anomaly of the tricuspid valve, showing downward displacement of the septal and posterior tricuspid leaflets as well as "atrialization of the right ventricle."

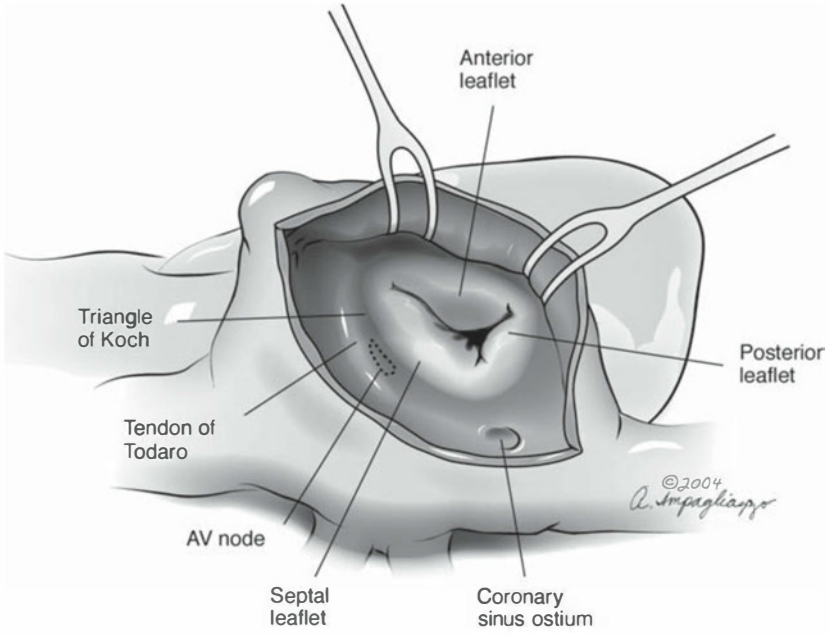


Fig. 46.2. Tricuspid valve anatomy. Surgeon's view of the tricuspid valve as seen via *right* atriotomy (AV, atrioventricular).

importance during tricuspid valve surgery. During tricuspid valve replacement, it is safest to leave a small margin of septal leaflet tissue in order to place the valve sutures so as to avoid injuring the conduction system. Most rings used for tricuspid valve annuloplasty are designed to specifically avoid placement of sutures in this critical area by leaving a gap in the ring in this area.

Operative Setup

The setup for cardiopulmonary bypass for operations involving the tricuspid valve depends on whether the operation is performed for isolated tricuspid valve disease or concomitant mitral and tricuspid valve surgery and also depends on the approach—via sternotomy or right thoracotomy. In most patients undergoing tricuspid surgery, the right atrium is enlarged, enhancing exposure of the tricuspid valve.

During conventional sternotomy, there are greater options for the placement of the venous cannulas and their exact placement will depend on the type of right atriotomy planned (Fig. 46.3A and 46.3B). During concomitant mitral and tricuspid surgery, both a transeptal approach to the mitral valve as well as separate left and right atriotomies provide excellent exposure to the mitral valve and should be dictated by surgeon preference. In exceptionally deep, barrel-chested patients, we prefer a transeptal approach for better visualization of the mitral valve. The superior vena cava cannula can be placed via the right atrial appendage (Fig. 46.3A) or directly into the superior vena cava preparing for a standard right atriotomy. If, planning a superior transseptal approach to the left atrium, the superior vena cava cannula can be placed more laterally on the right atrial wall as is done during heart transplantation (Fig. 46.3B). Various combinations of antegrade and retrograde cardioplegia can be used according to the surgeon's preference. If left-sided valvular surgery is to be performed, it is performed under moderate hypothermia, the left atriotomy is closed, and the tricuspid valve procedure is performed as the patient is rewarmed. Some surgeons prefer to remove the aortic cross-clamp before the tricuspid portion of the operation but does add to a more challenging operative view of the tricuspid valve due to blood return from the coronary sinus and thebesian veins.

For isolated tricuspid operation, mild hypothermia is used. We often perform these without cross-clamp as many of them have significant RV dysfunction. In this setting, a standard right atriotomy is made parallel to the atrioventricular groove and far enough away from it to allow safe closure of the atriotomy without endangering the right coronary artery. After the atriotomy is made, exposure can be facilitated by suspending the edges of the atriotomy with silk stay sutures or by using a retractor system. The valve leaflets and subvalvular apparatus are inspected carefully, and the mechanism of tricuspid dysfunction is confirmed and dealt with accordingly.

In a right thoracotomy approach to the tricuspid valve, right internal jugular and femoral venous cannulation is performed. Arterial cannulation can be performed via a femoral approach or through the ascending aorta directly. A standard atriotomy is performed. The use of video-assisted thoracoscopy can improve visualization and lighting to the tricuspid valve.

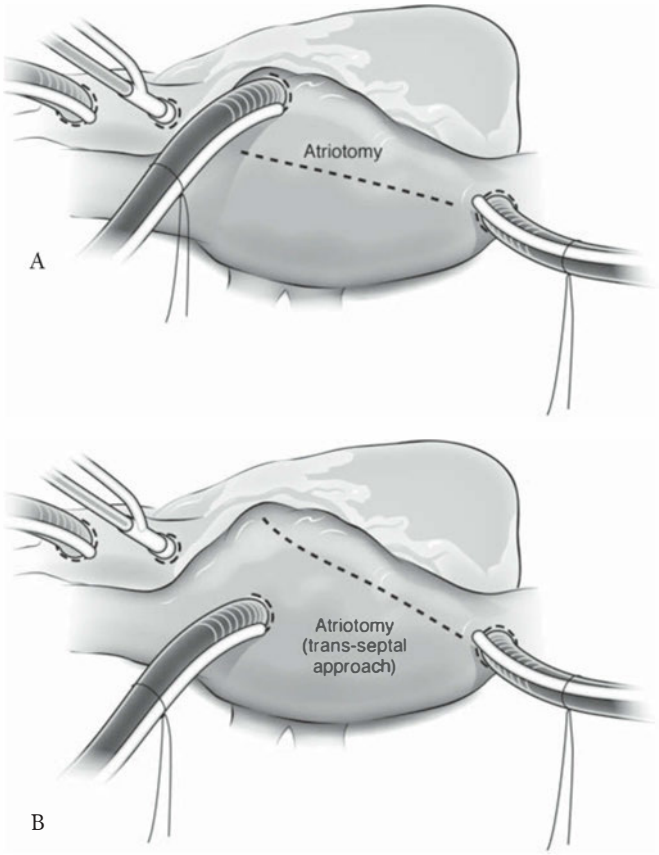


Fig. 46.3. (**A**) Operative setup for standard *right* atriotomy parallel to the atrioventricular groove. (**B**) Operative setup for trans-septal approach to the mitral valve for operations involving both atrioventricular valves.

TRICUSPID VALVE ANNULOPLASTY

Rigid Annuloplasty

Our standard approach for the functional TR is annuloplasty with a rigid or semi-rigid incomplete ring. The gap is in the area of the conduction system adjacent to the septal leaflet. There are little data on how to adequately size the tricuspid annulus. The septal leaflet is often used for reference as this part of the annulus usually does not dilate. Moreover, according to a report by Mohr and colleagues, the dehiscence rate is as high as 10% even in experienced centers.

We typically use 8-10 nonpledgeted 2-0 Ticron overlapping sutures. Larger travels are undertaken along the anterior and posterior annulus to create greater reduction annuloplasty in these areas. We often reinforce the first and last suture with an additional pledgeted 3-0 or 4-0 prolene suture as these are the areas at greatest risk for dehiscence (Fig. 46.4).

In those patients with extremely poor quality annular tissue, we use horizontal mattress sutures consisting of 2-0 Ticron with pledgets. Typically, there will be 3 to 4 mm space between each suture in the annulus and 1 mm of space between each suture as it is placed through the annuloplasty ring. This will ensure even distribution of the annular plication and tension around the circumference of the annuloplasty band. Each stitch enters the atrial tissue just outside the annulus and exits within the annulus (Fig. 46.5B and 46.5C). The suture is then placed through the inside of the ring to get the ring to seat in a directly annular position. The ring is lowered down to its annular position (Fig. 46.5C). I prefer to tie down the sutures starting at the junction of the anterior and posterior leaflets and work counterclockwise to the anterior leaflet. The septal annular sutures are tied last as this often has the greatest amount of tension.

DeVega

Long-term data with the DeVega technique have not been consistent. As such, we use the DeVega annuloplasty technique only rarely such as endocarditis where avoidance of any prosthetic material is preferred. A 3-0 Prolene double-arm pledgeted suture is used. The inner suture line is begun approximately 1 mm off of the tricuspid annulus and starts at the anteroseptal commissure. The bites are approximately 3 mm in depth and 5 mm in length, skipping 5 mm between bites. This inner suture line proceeds in a clockwise manner around the annulus to the posteroseptal commissure. The outer suture line is 3 mm outside the first and is placed in a similar manner, in a clockwise direction to the posteroseptal commissure. A second pledget is passed onto the free ends of the suture. The suture is tightened down over a 28-mm tricuspid ring sizer or a 60 cm^3 syringe.

Suture Bicuspidization

An additional technique popularization by Cohn and colleagues involves suture

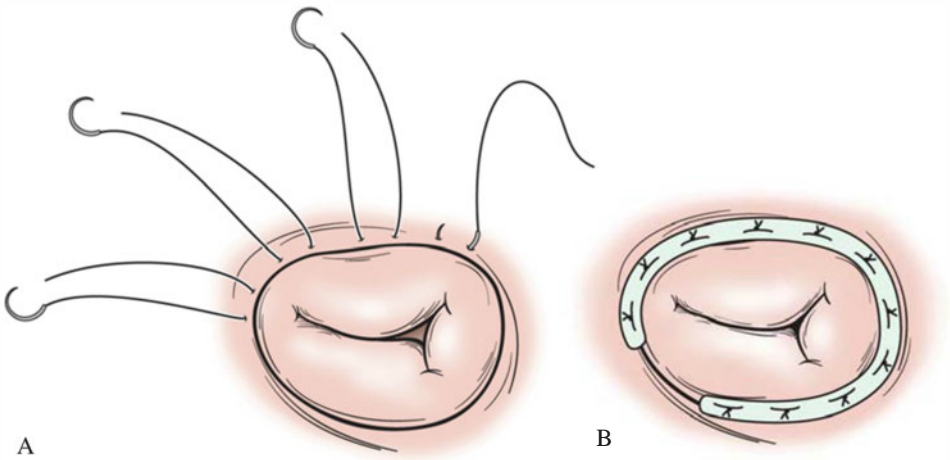


Fig. 46.4. (**A**) Tricuspid valve annuloplasty. Placement of annuloplasty sutures in horizontal mattress manner with needles passed through the annulus. (**B**) Completed tricuspid valve annuloplasty using Carpentier–Edwards ring.

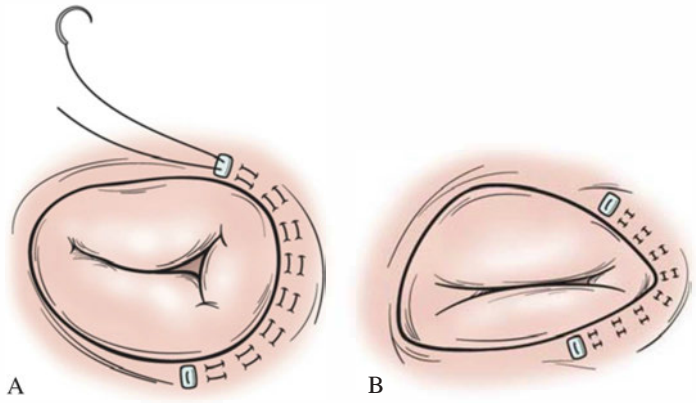


Fig. 46.5. Bicuspidization. 2-0 pledgeted suture placed from 2 o'clock to 5 o'clock making the annulus smaller shortening the annulus around the posterior leaflet.

bicuspidization of the tricuspid valve. With this technique, a pledgeted 2-0 Ethibond or Ticron suture on a large needle is run from the anteroposterior to the posteroseptal commissure along the posterior annulus. The second suture is passed in a similar manner and an additional free pledget is placed (Fig. 46.5). The suture is tied down with a dilator (similar to the DeVega technique). The net result is exclusion of the posterior leaflet and a functional bicuspid tricuspid valve.

Other Techniques

Other methods that are established and proven for the mitral valve including leaflet resection, Alfieri plication, and artificial cord placement can also be applied in rare cases of prolapse of the tricuspid valve or in cases of endocarditis.

TRICUSPID VALVE REPLACEMENT

Tricuspid valve replacement may be indicated for infectious endocarditis, failed previous tricuspid repair, failing previously placed valve prosthesis, or the more rare instances of carcinoid heart disease or rheumatic disease. Prosthetic replacement of the tricuspid valve with either a bioprosthetic or mechanical valve has been generally shown to be equally safe, although most would place a bioprosthetic valve in patients with concern for compliance with anticoagulation.

The setup for tricuspid valve replacement is similar to that for a tricuspid valve repair. We tend to debride only the diseased portions of the tricuspid valve and preserve as much choral apparatus as feasible similar to chordal-sparing mitral valve replacement (Fig. 46.7A). If the septal leaflet requires excision, leaving a 2-mm margin of septal leaflet adjacent to the annulus for suture placement may help to avoid injury to the conduction system. Whether a bioprosthetic or mechanical valve is chosen, 2-0 Ticron-pledgeted horizontal mattress sutures are

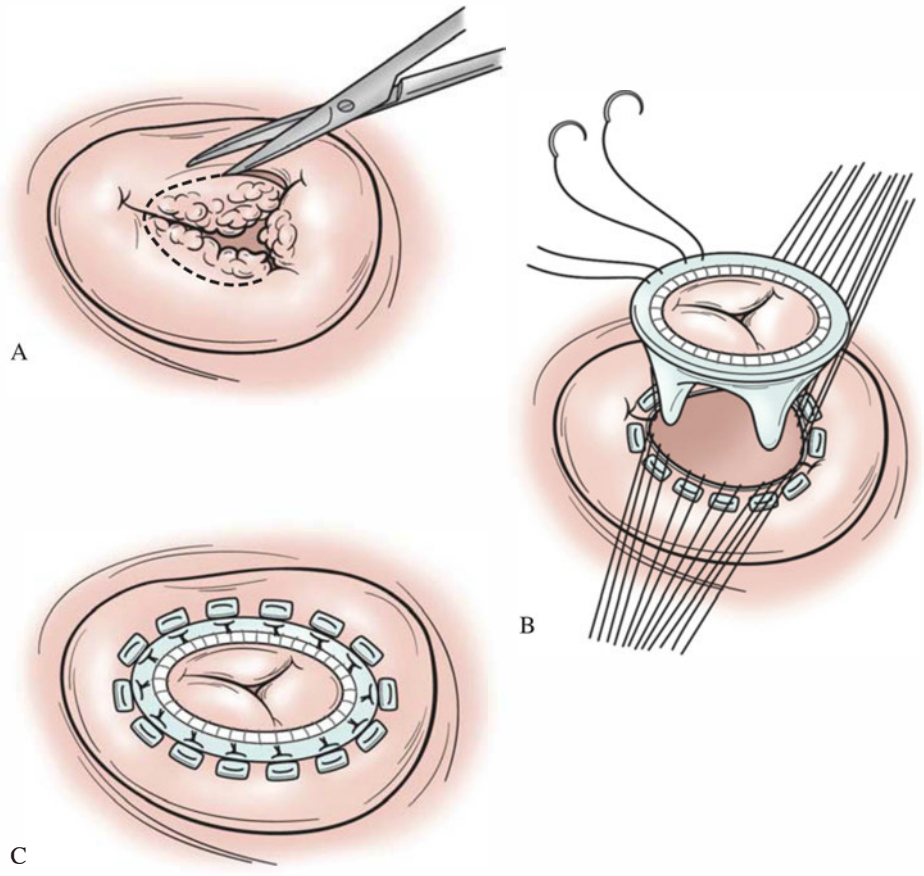


Fig. 46.6. (**A**) Tricuspid valve excision leaving a 2-mm margin of septal leaflet tissue adjacent to the annulus. (**B**) Suture placement for tricuspid valve replacement. (**C**) Completed tricuspid valve replacement.

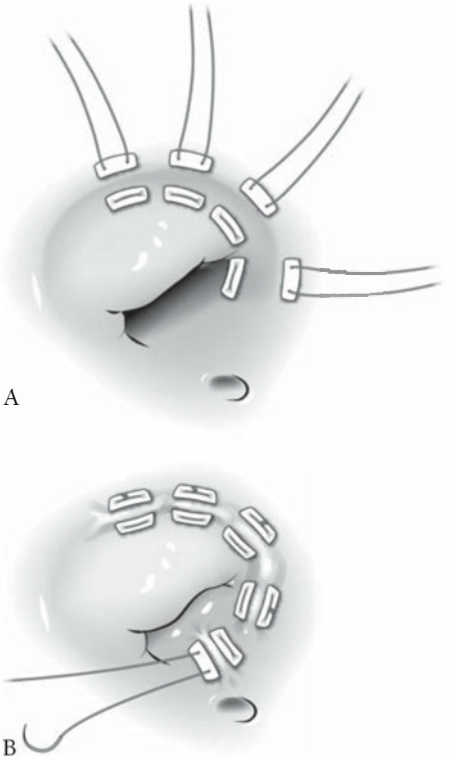


Fig. 46.7. Danielson repair. (**A**) Pledgeted horizontal mattress sutures draw the tricuspid leaflets up to the tricuspid annulus. (**B**) Posterior annuloplasty suture lessens the annular circumference, creating a functional monocuspid valve based on the anterior leaflet.

placed with the needles entering just outside the tricuspid valve annulus and exiting from within the tricuspid annulus from atrium to ventricle and through the leaflet (Fig. 46.7B). In the area of the septal leaflet, the sutures are passed through the remnant of the leaflet to avoid injuring the conduction system. The valve sutures are then passed through the sewing ring, the valve is lowered into position, and the sutures are tied and cut (Fig. 46.7C). In the case of placement of a bioprosthetic valve, the orientation of the valve is not critical because RV outflow tract obstruction by one of the struts is unlikely.

TECHNIQUE OF VALVE REPAIR FOR EBSTEIN'S ANOMALY

For repair of Ebstein's anomaly, the usual preparation is made for cardiopulmonary bypass. Bicavalvenous cannulation is used. Usually, the right atrium is enlarged and cannulation of the superior vena cava via the right atrial appendage is used. Mild hypothermia is used. The aorta is cross-clamped and antegrade cardioplegia is delivered. Once adequate myocardial preservation has been attained, a generous right atriotomy is made keeping in mind that in cases of a markedly enlarged right atrium an ellipse of right atrial wall may be removed. The tricuspid valve and atrial septum are evaluated, and if an atrial defect is present, it is closed after the tricuspid has been repaired. A cardiotomy sucker or vent catheter can be placed through the atrial septal defect during the tricuspid repair to facilitate visualization. The leaflets of the tricuspid valve are evaluated with particular attention to the morphology and quality of the anterior leaflet. Less satisfactory results can be predicted by the presence of a thickened muscularized anterior leaflet with fused or indistinct chordae. The Danielson and Carpentier methods of repair have both been used successfully; however, we have found the Carpentier repair to provide excellent results, given the appropriate candidate valve.

The Danielson repair (Fig. 46.8) in the adult involves the vertical plication of the atrialized portion of the right ventricle and creation of a functional monocuspid valve based on the anterior leaflet. A portion of the right atrium is also removed to normalize its size. Pledgeted horizontal mattress 3-0 Prolene sutures are placed from the downwardly displaced tricuspid valve to the tricuspid annulus. These sutures are tied down, drawing the tricuspid valve up to the tricuspid annulus and thus excluding the atrialized portion of the right ventricle. A 3-0 Prolene mattress posterior annuloplasty suture is also placed to reduce the annular circumference. Additional sutures may be placed to obliterate the remainder of the posterior tricuspid annulus. The atrial septal defect, if present, is closed.

The Carpentier repair uses longitudinal plication of the right ventricle and tricuspid annulus with resultant reduction in annular circumference. The anterior leaflet is detached along most of its annular circumference, and a sliding plasty is performed following annular plication using 4-0 Prolene running suture. This redistributes the anterior leaflet

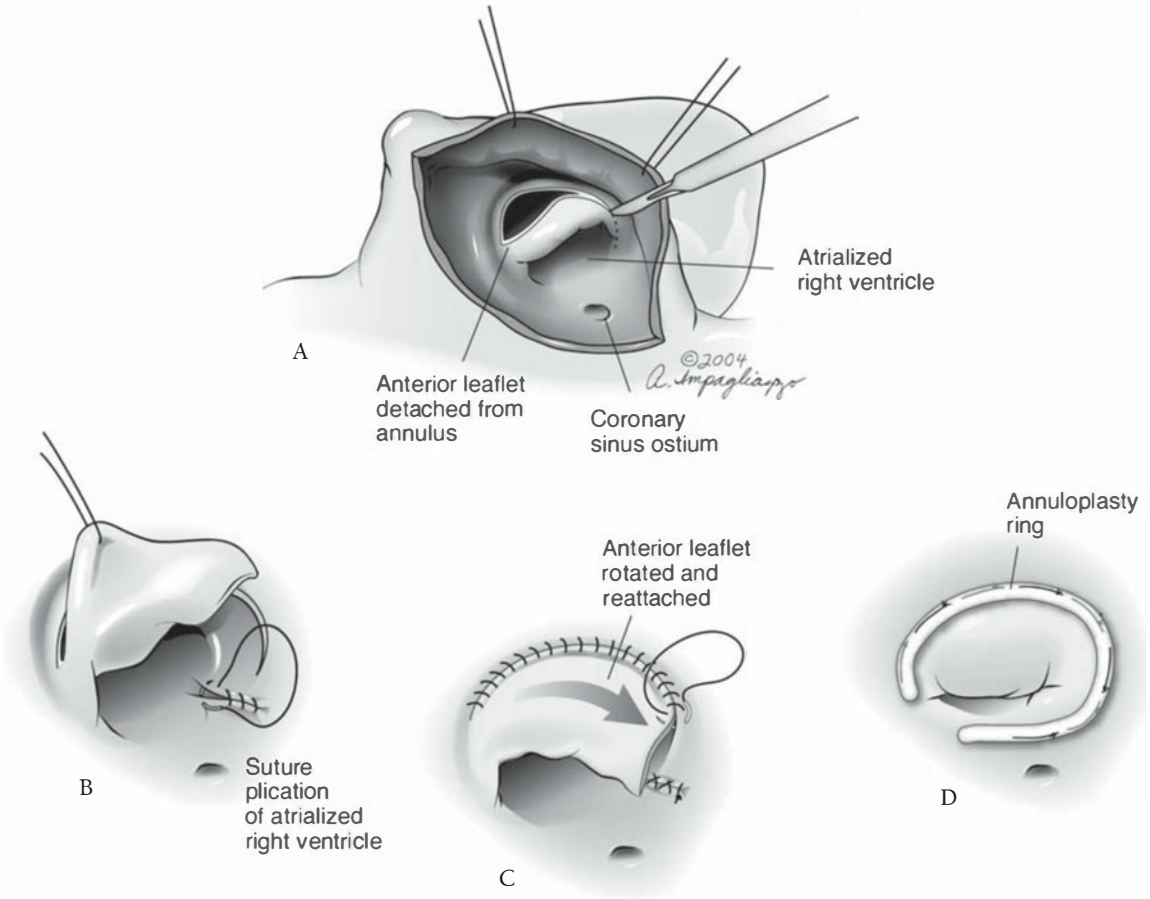


Fig. 46.8. Carpentier repair. (**A**) Anterior leaflet is detached from the tricuspid annulus. (**B**) Vertical plication of tricuspid annulus. (**C**) Redistribution and reattachment of the anterior leaflet to the remodeled tricuspid annulus. (**D**) Finished repair is supported by an annuloplasty ring.

around much of the refashioned annulus. The result is effectively a monocuspid valve that spans the tricuspid orifice. The repair is supported by the placement of a Carpentier annuloplasty ring using the method described previously in this chapter. Given an adequate annular reduction and an anterior leaflet of reasonable quality, a dramatic reduction in TR can be expected.

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EDITOR'S COMMENTS

The surgical treatment of the tricuspid valve may be one of the more controversial areas in cardiac surgery. There are multiple decent techniques for repair but unfortunately all have their issues. I believe presently there are enough good studies that if one truly has to do a repair for an isolated tricuspid valve disease, a ring repair is probably the best approach. Other suture approaches may be appropriate in patients who require the tricuspid valve fixed at the time of mitral valve surgery.

Surprisingly, though this is an operation on the right side of the heart, tricuspid valve surgery carries a significant mortality. Most frequently, this occurs due to comorbidities particularly congestion of the liver, which does not get better immediately. I think it is clear now that tricuspid repair should be done in patients who have right-sided congestion when done as an isolated procedure. Repair should be done in conjunction with left-sided valve procedures when there is moderate or severe tricuspid regurgitation or in some case when the annulus is markedly dilated.

ILK

47 Pulmonary Valve Replacement

Damien J. LaPar, Carlos M. Mery, and James J. Gangemi

PULMONARY VALVE REPLACEMENT AND TETRALOGY OF FALLOT

Dysfunction of the right ventricular outflow tract (RVOT), whether is be due to stenosis or regurgitation, is a common and challenging condition in both children and adults with congenital heart defects. Some of these defects include pulmonary atresia, d-transposition of the great arteries with pulmonary stenosis, truncus arteriosus, double outlet right ventricle, and most commonly tetralogy of Fallot (TOF). Pulmonary valve replacement (PVR) remains the gold-standard of care for the definitive surgical treatment of progressive right ventricular (RV) failure from these defects due to long-standing pulmonary regurgitation (PR) and pulmonary stenosis. Often occurring two to three decades after surgical repair of TOF, PR has become an increasingly more common clinical situation confronting congenital and adult cardiac surgeons as outcomes and survival have improved for TOF patients over the past several decades. TOF still remains the most common type of cyanotic congenital heart defect. Surgical palliation of TOF was first popularized with the introduction of the Blalock–Taussig systemic-to-pulmonary artery shunt in 1945, and surgical management of this condition for a majority of patients has evolved over the past 50 years to include primary repair of each associated defect during infancy. With advances in surgical technique, diagnostic imaging and technology, postoperative and critical care management, and long-term follow-up capabilities, outcomes following primary TOF repair have dramatically improved with current mortality rates of 2% to 3% and 20-year survival approaching 90%. Despite these successes, significant anatomic and functional abnormalities remain following surgical treatment of TOF that confer important long-term cardiopulmonary implications.

The recognition of certain adverse functional sequelae following TOF repair has been increasingly reported and emphasized in recent years. These abnormalities include RV failure from progressive volume overload and PR, persistence of atrial and/or septal defects, RV outflow obstruction, pulmonary artery stenosis, and tricuspid regurgitation. While these lesions are often benign during childhood and adolescence, they have the potential to progress and significantly compromise patient functional status, exercise tolerance, and survival by the second or third postoperative decade. Moreover, severe, progressive chronic PR is considered a treatable cause of RV failure and has been the focus of increased study and investigation in recent years.

This chapter reviews the general principles that apply to the pathophysiology of pulmonary valve dysfunction, indications and timing for treatment of severe and clinically significant pulmonary valve lesions, selection and availability of pulmonary valve prostheses, surgical technique of PVR, and recently reported outcomes for PVR.

PATHOPHYSIOLOGY OF PULMONARY VALVE DYSFUNCTION

The pathophysiology of pulmonary valve dysfunction following TOF repair is multifactorial, and the severity of pulmonary valve dysfunction has been shown to increase over time. While for many years, the impact of PR after surgical correction of TOF was considered relatively benign, evidence has demonstrated that progressive PV dysfunction results in right heart failure. Four primary factors influence the degree of PR following TOF repair: (1) regurgitant orifice size, (2) length of diastole, (3) diastolic PA to RV pressure gradient, and (4) pulmonary artery resistance and capacitance. Early after TOF repair, the effect of pulmonary valve regurgitation is minimal due to an increased heart rate and shortened diastole, decreased RV compliance due to RV hypertrophy, and decreased pulmonary artery capacitance. However, over time, physiologic changes occur, including an increase in RV compliance and stroke volume, increased PA compliance, and slowing of heart rate with a lengthening of diastole. These changes, therefore, predispose to the eventual development of worsening PR. Moreover, the progressive development of PR results in a cyclical pattern of worsening PV and cardiac dysfunction. Increased PR results in RV dilation and increased RV compliance that serves to increase RV stroke volume, ultimately resulting in elevated PA pulse pressures, increased PA dilation, and worsening of PR.

The development of improved cardiac imaging modalities has demonstrated important changes to RV mechanics following primary TOF repair that play an important role in the progressive decline in pulmonary valve function. In fact, a close relationship between RV end-diastolic volume and percent PR exists. While the effects of RV systolic function are preserved for several years despite significant PR, eventually compensatory mechanisms fail. Failure of RV compensation results in decreased RV mass-to-volume ratios, increased end-systolic volume, and ultimately decreased RV ejection fraction. In addition, PV function is further degraded due to the effects of age of repair; RV dyskinesia due to ischemia, fibrosis, RV aneurysm, or RVOT patch; impaired RV diastolic function; and increased LV size and dysfunction.

INDICATIONS AND TIMING OF PULMONARY VALVE REPLACEMENT

PVR has documented beneficial effects for PV dysfunction including improved ventricular function, reduced atrial and ventricular arrhythmias, and improved cardiac functional status. These benefits notwithstanding the clinical decision as to the timing of PVR and the identification of

clinical indicators for surgical valve replacement often remain a dilemma.

Several clinical and physiologic indications exist for PVR following TOF repair. Clear clinical indication for PVR include symptomatic physical deterioration, decreased exercise tolerance, onset of clinical arrhythmia, clinical signs and symptoms of right heart failure as a result of severe PR, need for cardiac medications, and onset of syncope. However, indications for PVR in the asymptomatic patient are much less defined. In an era of advancing technology, several different cardiac functional metrics have been identified to aid in the decision to proceed with PVR. In the presence of moderate-to-severe PR, PVR is indicated in the presence of the following homodynamic factors as reported by Geva:

- RV end-diastolic volume index $\geq$ 160 ml/m^2.
- RV end-systolic volume index $\geq$ 70 ml/m^2.
- LV end-diastolic volume index $\leq$ 65 ml/m^2.
- RV ejection fraction $\leq$ 45%.
- Presence of RVOT aneurysm.

Other important factors include the presence of other hemodynamically significant lesions including moderate-to-severe tricuspid valve regurgitation or the presence of atrial and/or ventricular septal defects, TOF, or other congenital pulmonary outflow tract reconstruction after the age of 3 years. Moreover, an MRI-based study by Therrien and colleagues demonstrated that the optimal timing for PVR in adults following TOF repair occurred when RV end-diastolic dimension was $<$170 ml/m. When dealing with pulmonary stenosis, surgical indications include a RV pressure two-third of systemic regardless of symptoms.

SELECTION OF AVAILABLE PULMONARY VALVE PROSTHESES

Reconstruction of the RVOT remains a common surgical procedure in congenital heart surgery. There are several congenital heart defects that require this operation to prevent or reverse RV dysfunction that can result from long-standing pulmonary insufficiency or stenosis. Despite the high frequency of this operation, controversy still exists regarding the best valve replacement in the pulmonary position.

Pericardial Valve

There have been encouraging results with the use of the bovine pericardial valve. It has been associated with low morbidity and no perioperative mortality. Shinkawa et al. showed a freedom from reoperation of 97.7% at 5 years. At midterm follow-up, the mean pulmonary valve gradient was 19 mmHg, and 9% of the patients had freedom from more than moderate pulmonary insufficiency. Other studies supported these encouraging results. Allen et al. reported excellent early outcomes using bovine pericardial valves with 100% freedom reoperation at 5 years. Fiore et al. also showed promising early outcomes with 92% freedom from reoperation at 5 years. In that study, late valve dysfunction was lower in the bovine pericardial valve when compared with porcine valves and homografts (5.5% vs. 19% vs. 54%, respectively).

There are several theoretical advantages of pericardial pulmonary valves. The valve may deteriorate by both stenosis and regurgitation rather than regurgitation alone. Slow progressing stenosis may be less damaging to the right ventricle as compared with insufficiency alone, which can result in recurring RV dilation. Clearly, more studies specifically evaluating long-term performance and structural changes of these valves in the pulmonary position need to be completed to support this theory.

Mechanical Pulmonary Valves

The overwhelming experience of mechanical prosthesis has been in the aortic position. The benefits of these valves include excellent long-term durability and potential freedom from reoperation. Obviously, the requirement of anticoagulation is a concern and carries its own set of risks. For patients, particularly adults, with congenital heart defects involving the pulmonary valve, this seems like a viable option in certain circumstances. Unfortunately, there exists only a small body of literature evaluating the use of mechanical valves in the pulmonary position. There have been conflicting reports with the use of mechanical valves in the pulmonary position particularly with respect to the use of a bileaflet versus monodisc valve (bileaflet valves had a higher rate of thrombotic complications). The range of thrombotic complications for mechanical valve in the pulmonary position, being a lower pressure system, is from 25% to 80%. However, the majority of patients studied in the past were only anticoagulated with aspirin. Stulak et al. showed excellent durability of mechanical pulmonary valves with freedom from reoperation at 5 years of 100% (vs. 90% for bioprosthetic) and at 10 years of 100% (vs. 52% for bioprosthetic). Furthermore, the average gradient was 13 mmHg across the valve in follow-up. There were no thromboembolic events when patients were adequately anticoagulated with warfarin. In addition, there were no reoperations for pannus formation, paravalvular leak, endocarditis, valve thrombosis, or prosthetic dysfunction with long-term follow-up. Although bleeding with anticoagulation remains a concern and is often a deterrent to use of mechanical valve prostheses, particularly in children, there are several advances that may decrease the future risk of anticoagulant-related bleeding. Contrary to historic data, achieving a slightly lower target INR may be beneficial. Further, there are new antiplatelet agents and direct thrombin inhibitors that have a wider therapeutic window and fixed dose. The side-effect profile is much less as compared with warfarin. In addition, INR self-testing has demonstrated decreased risk in bleeding complications from 10.9% to 4.5% and decreased risk of thromboembolism.

Given the excellent durability and hemodynamic profile combined with the low rate of thrombotic events (when adequately anticoagulated), mechanical PVR is a potentially viable alternative in highly selected circumstances (multiple prior operations, those requiring anticoagulation for something else, or patients who have demonstrated early or accelerated deterioration of previously placed tissue valves).

Pulmonary Homografts

Pulmonary homografts have been a commonly used conduit to reestablish RV to pulmonary artery continuity in a variety of congenital heart defects. Initial use of these conduits, with early preservation techniques employing antibiotics or irradiation sterilization, was associated with early failure. Over the years, the development of cryopreservation techniques have improved both the availability and durability of allografts, which has resulted in increased use of RVOT reconstruction. The freedom from reoperation rates reported for series of cryopreserved in the pulmonary position in the young have been 55% to 80% at 4 to 6 years. Some of the early failure rates of the pulmonary homograft may be related to age of the patient. Hawkins et al. described an increased risk of valvular degeneration in infants when compared with older children. In their study, children older than 1 year of age experienced a freedom from valve explantation or death of

94% at 3 years after surgery compared with 50% in children younger than 1 year of age. This has subsequently been supported by Brown et al. showing an almost double incidence of dysfunction and failure at 5 years in infants younger than 1 year of age.

Standard allografts are widely used at many centers for several reasons. They have excellent handling properties, are easy to insert, and have an acceptable hemodynamic profile after implantation. Pulmonary homografts have been preferred over aortic homografts in the pulmonary position because they have less elastic tissue and lower rates of calcification. One main issue with the homograft conduit is its variable durability especially in young children, which frequently require repeat operations. It is theorized that homograft failure is partially a result of host immune response to antigens present in the homografts. Specific cellular and humoral responses by the recipient contribute to early graft deterioration and tissue calcification with subsequent clinical failure. Infants and children tend to have the greatest immunologic reaction to cryopreserved homograft material with a shorter period of freedom from reintervention after implantation than seen in adults.

Decellularized cryopreserved valved homografts were developed to ameliorate the recipient immune response to implanted conduits. These homografts were also designed to provide an acellular matrix that could be infiltrated by autologous cells. In theory, a decellularized cryopreserved homograft repopulated with recipient cells would be capable of repair and remodeling similar to native tissue giving it increased durability. Despite evidence that these homografts do not elicit a significant immune response, and even though autologous cells have been shown to infiltrate decellularized autografts, there has been no definitive data demonstrating a clinical advantage over standard homografts. Unfortunately, most studies have been limited by small patient sizes as well as short duration of follow-up.

Burch et al. studied long-term follow-up of 47 patients undergoing RV to pulmonary artery conduit replacement using decellularized cryopreserved homografts and compared them with 47 age-matched and diagnosis-matched controls receiving standard homografts. There was a nonsignificant trend toward lower peak valve gradient and reintervention in the decellularized cryopreserved homografts. Several other studies have evaluated the performance of decellularized homografts in the RVOT position as compared with standard homografts. There have been no statistical differences between the two types of homografts with regard to reintervention in any study of short- or long-term function.

Furthermore, decellularized cryopreserved homografts cost an average of $3,000 more than standard homografts. In addition, the shelf-life of these decellularized grafts is approximately one-tenth that of traditional cryopreserved homografts. There is, however, a particular patient population that might benefit from these decellularized grafts. Patients who progress to transplant may be a better choice for this homograft as they result in minimal activation of recipient immune system, thus preventing sensitization that would limit donor compatibility. It should be kept in mind that patients with congenital heart disease requiring RVOT reconstruction currently represent 15% of patients who ultimately progress to transplant.

Percutaneous Pulmonary Valve Replacement

The number of patients requiring RVOT intervention continues to grow. For patients who receive their first PVR at an early age, the number of operations becomes substantial. Percutaneous PVR, as first described by Bonhoeffer in 2000, has made remarkable progress over the years. The Melody transcatheter valve was FDA approved in 2010 for use in pediatric and adult patients who have failed RVOT conduits (stenotic or regurgitant).

Transcatheter PVRs are less invasive, avoid the risks associated with cardiopulmonary bypass, avoid risks of bleeding and infections, and potentially reduce the costs by avoiding postoperative intensive care. The valve that is a bovine internal jugular vein valve mounted in an expandable platinum iridium stent can be delivered percutaneously via the femoral or internal jugular veins. In addition, the valve can be delivered with a hybrid approach directly through the right ventricle without having to go on cardiopulmonary bypass. Percutaneous pulmonary valve implantation is currently approved to be performed in a >16 mm conduit with either a mean gradient of 35 mmHg or at least moderate pulmonary valve insufficiency and is expandable to 22 mm.

Several studies have shown a high rate of procedural success and encouraging short-term function of the percutaneous Melody valve. This could reduce the number of operations and the cumulative hemodynamic burden on the right ventricle over the total lifetime of children and young adults. This may in turn potentially improve the life expectancy of patients with congenital heart disease involving the RVOT. While initial trials have shown promising early results in certain subset of patients, long-term outcomes are currently lacking and complication rates in addition to reintervention rates are being closely evaluated.



SURGICAL TECHNIQUE

Reconstruction of the RVOT is a common surgical procedure in congenital heart disease. Preoperatively, a cardiac catheterization or magnetic resonance imaging scan is performed. Duplex studies may also be considered to evaluate the groin vessels for possible groin cannulation. All patients have an intraoperative transesophageal echocardiography performed to assess function and particularly any residual atrial or ventricular communications. The overwhelming majority of these surgeries are performed in the redo sternotomy setting. Once the chest has been opened and the mediastinal structures are dissected out, we typically place a single aortic cannula in the distal ascending aorta and a two-staged venous cannula in the right atrium. The patient is placed on cardiopulmonary bypass. The conduit replacement can be performed on pump, with a beating heart and no cardioplegic arrest assuming there is no atrial or ventricular communication. If there is, then bicaval venous cannulas should be placed and the heart should be arrested to repair the residual communications. Once completed, the aortic cross-clamp can be removed and the conduit replaced with a beating heart. Once on pump, the main pulmonary artery is transected at the distal confluence. The distal confluence and proximal branch pulmonary arteries are completely dissected out and freely mobilized for an easy distal anastomosis. The transannular patch is completely resected back to healthy RV tissue. Similarly, a previously placed conduit is also completely excised. The distal anastomosis is typically performed first between the distal conduit and the confluence of the pulmonary artery. Once completed, the proximal conduit can be trimmed accordingly and sewn to healthy RV muscle wall. A homograft can be inserted in a similar manner although some would prefer to perform the proximal anastomosis first and then trim the distal conduit before sewing the anastomosis to the pulmonary arteries. The entire operation can be performed no lower than moderate hypothermia (34°C).

POSTOPERATIVE OUTCOMES FOLLOWING PULMONARY VALVE REPLACEMENT

Outcomes following performance of PVR are encouraging. Operative mortality remains low with most series reporting rates of <1%. Perhaps more importantly, long-term survival remains high with 5- and 10-year survival rates of approximately 90% to 92% and 75% to 85%, respectively. Freedom from reoperation and valve failure rates approach 80% at 5 years, 60% at 10 years, and 40% at 15 years. Young age at the time of PVR remains a well-established risk factor for reoperation and valve failure.

CONCLUSIONS

PVR following TOF repair remains the gold-standard treatment for RV dysfunction. With improved surgical outcomes and advances in modern postoperative care, the number of patients requiring PVR is likely to increase in coming years. Several different choices in valve prostheses are available to the surgeon, and advances in the percutaneous, transcatheter delivery of pulmonary valves may offer an attractive alternative for select patient populations. While outcomes following surgical PVR remain excellent, future comparative effectiveness study will help to determine the future place of percutaneous technology in the treatment of clinically significant PR.

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EDITOR'S COMMENTS

The surgical treatment of the pulmonary valve has been one of the least understood chapters in adult cardiac surgery. Many of these patients have had previous repair of tetralogy of Fallot. Previously, surgeons tended to cut out all pulmonary valve tissue to prevent obstruction. We now know that this results in late right ventricular dysfunction due to pulmonary insufficiency.

The conduits for pulmonary valve replacement have changed over time as well. I use to favor homografts but with time even pulmonary homografts tend to have earlier dysfunction than expected. The use of valve conduits with tissue valves seems to be the best approach at this point with low mortality and certainly the ability to best preserve a damaged right ventricle. There is another huge advantage to the use of pulmonary valve conduits. It becomes a resting place for percutaneous pulmonary valves that are truly excellent technology. We use these at our institution and they work extremely well inside a degenerated conduit.

ILK

48 Aortic Valve Replacement

David A. Fullerton

THE SURGICAL ANATOMY OF THE AORTIC VALVE

The normal aortic valve is composed of three thin, pliable leaflets or cusps attached to the heart at the junction of the aorta and the left ventricle. The leaflets are attached within the three sinuses of Valsalva to the proximal aorta and joined together in three commissures that create the shape of a coronet. Because the coronary arteries arise from two of the three sinuses of Valsalva, the aortic leaflets are named after their respective sinuses as the left coronary leaflet, the right coronary leaflet, and the noncoronary leaflet. However, because of the oblique position of the aortic root, the sinuses themselves are rarely in a strict left or right position. The attachment of the leaflets to the left ventricular outflow tract is termed an annulus; however, in the strictest terminology this is not a true annulus because it is not truly circular: the points of attachment of the leaflets do not all lie in the same plane. There are two important surgical landmarks. First, the commissure between the left and noncoronary leaflets is positioned along the area of aortic–mitral valve continuity. Beneath this commissure is the fibrous subaortic curtain. The commissure between the noncoronary and the right coronary leaflets is positioned over the left Bundle of His. Injury to this conduction bundle during aortic valve surgery may create heart block.

INDICATIONS

Aortic Stenosis

The diagnosis and severity of aortic stenosis are determined by echocardiography. The normal aortic valve area (AVA) is approximately 3 to 4 cm^2, and it has very little gradient across the valve until the AVA has been reduced by approximately one-half. Therefore, the flow velocity across the normal aortic valve (determined by Doppler echocardiography) is $\leq$1.0 m/s. With mild aortic stenosis, the AVA is decreased to $>$1.5 cm^2, and the flow velocity across the valve is increased to 2.5 to 2.9 m/s. Aortic stenosis is considered moderate when AVA is reduced to 1.0 to 1.5 cm^2, and the flow velocity across the valve increases to 3.0 to 4.0 m/s. Severe aortic stenosis is diagnosed by an AVA $<$1.0 cm^2 and a velocity across the valve of $>$4.0 m/s. When normalized for patient body surface area (BSA), severe aortic stenosis is an AVA $\leq$0.60 cm^2/m^2.

Over a period of years, the valve progressively narrows. During this "latent" period, patients are typically asymptomatic. However, the progressive narrowing of the valve is not linear; it occurs in an unpredictable, stepwise manner. Patient survival is not significantly diminished until patients develop symptoms. Thereafter, survival is quite limited. The three principal symptoms of aortic stenosis are angina, syncope, and heart failure. With the onset of angina, the mean survival of a patient with aortic stenosis is 4.7 years. Once a patient develops syncope, the mean survival is typically $<$3 years. Patients with dyspnea and heart failure have a mean survival between 1 and 2 years. Heart failure is the presenting symptom in at least one-third of patients with aortic stenosis. Once a patient crosses the threshold from asymptomatic to symptomatic, approximately 3% to 5% of patients will die within weeks to months. Hence, it is extremely important to accurately identify the presence of symptoms. The presence of symptoms in a patient with aortic stenosis is an indication for an aortic valve replacement (AVR). Management of the asymptomatic patient is discussed below.

Aortic Regurgitation

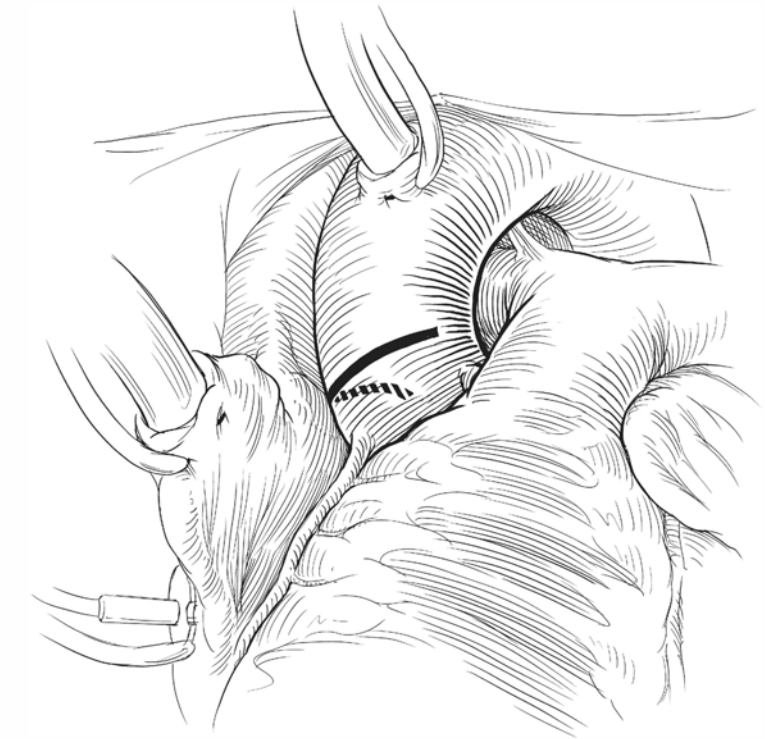
The diagnosis and severity of aortic regurgitation are also determined by echocardiography. Patients with mild-to-moderate aortic regurgitation are typically asymptomatic and have an excellent prognosis without surgery. Severe aortic regurgitation may produce symptoms of heart failure, and AVR is indicated in the symptomatic patient. The asymptomatic patient is discussed below.



SURGICAL TECHNIQUE

The standard surgical approach for AVR is via a median sternotomy (Fig. 48.1). Cardiopulmonary bypass is established by aortic and right atrial cannulation. After initiation of cardiopulmonary bypass, the aortic root is vented and a left ventricular vent is inserted via the right superior pulmonary vein. If the aortic valve is competent, cardiac arrest may be achieved by antegrade cardioplegia with subsequent administration of cardioplegia in retrograde manner. Otherwise, all cardioplegia may be administered retrograde. The myocardial temperature is monitored in the interventricular septum and kept below 10°C by administration of cold blood cardioplegia every 20 minutes throughout the period of aortic occlusion. I routinely cool the patient to a bladder temperature of 28°C.

There are several important technical caveats. First, it is important to prevent the introduction of air into the left atrium with the insertion of left ventricular vent via the right superior pulmonary vein. This can be done by temporarily pinching the venous line, thereby filling the left atrium with blood. I typically vent the aortic root before placing the left ventricular vent to evacuate any air that might be introduced. Second, the ascending aorta in patients undergoing AVR may be very thin because of poststenotic dilatation, advanced age, annuloaortic ectasia, etc. Hence, aortic cannulation stitches must be placed very carefully to avoid tearing the aorta. I often use felt pledgets to reinforce each bite of the aortic cannulation stitches to minimize this problem. Third, when operating for aortic regurgitation, the heart is prone to ventricular fibrillation once cardiopulmonary bypass has been initiated. If the heart fibrillates, the left ventricle will



A



B

Fig. 48.1. Operative technique: aortic valve replacement. (**A**) The initial transverse aortotomy is made 3 to 4 cm distal to the right coronary artery. The valve is visualized, and the aortotomy is extended at least 1 cm distal to the zenith of the commissure between the right and noncoronary leaflets. (**B**) The aortotomy is extended to the half-way point over the noncoronary leaflet. It is then opened down toward the annulus, stopping at least 1 cm from the annulus itself. (**C**) The leaflets are removed. (**D**) The calcified annulus is debrided with a Rongeur. It is helpful to place a small gauze sponge in the left ventricle to help catch any flecks of calcium during the debridement. (**E**) Interrupted horizontal pledgetted sutures are placed with pledgets in the subannular position. (**F**) After the sutures are brought the valve sewing ring, the valve is seated and the sutures tied.

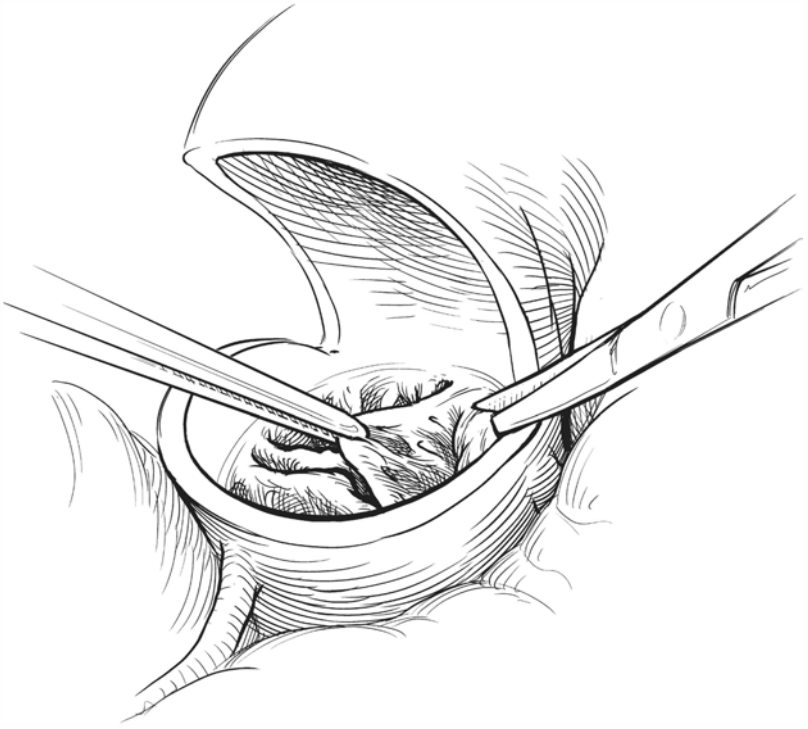
immediately distend, which may sometimes be lethal. To avoid this, I avoid systemic cooling until the left ventricular vent has been placed. The heart typically fibrillates soon after the initiation of systemic cooling, at which point I immediately cross-clamp the aorta and administer retrograde cardioplegia.

Once cardiopulmonary bypass has been initiated, the plane between the aorta and pulmonary artery is dissected. This is important to optimize visualization of the aortic valve and to facilitate aortic closure. One of the most important technical nuances of this operation is to identify the surface anatomy of the right coronary artery as it originates from the right sinus of Valsalva. This may be done by gentle dissection of the fat pad overlying the right sinus of Valsalva. One must be sure that the aortotomy is not too close to the right coronary ostium, for the ostium may be damaged or distorted with aortic closure or by the valve itself. Once the origin of the right coronary artery is identified, I mark the aorta with the electrocautery at the point of the anticipated aortotomy, approximately 2 to 2.5 cm distal to the origin of the right coronary artery.

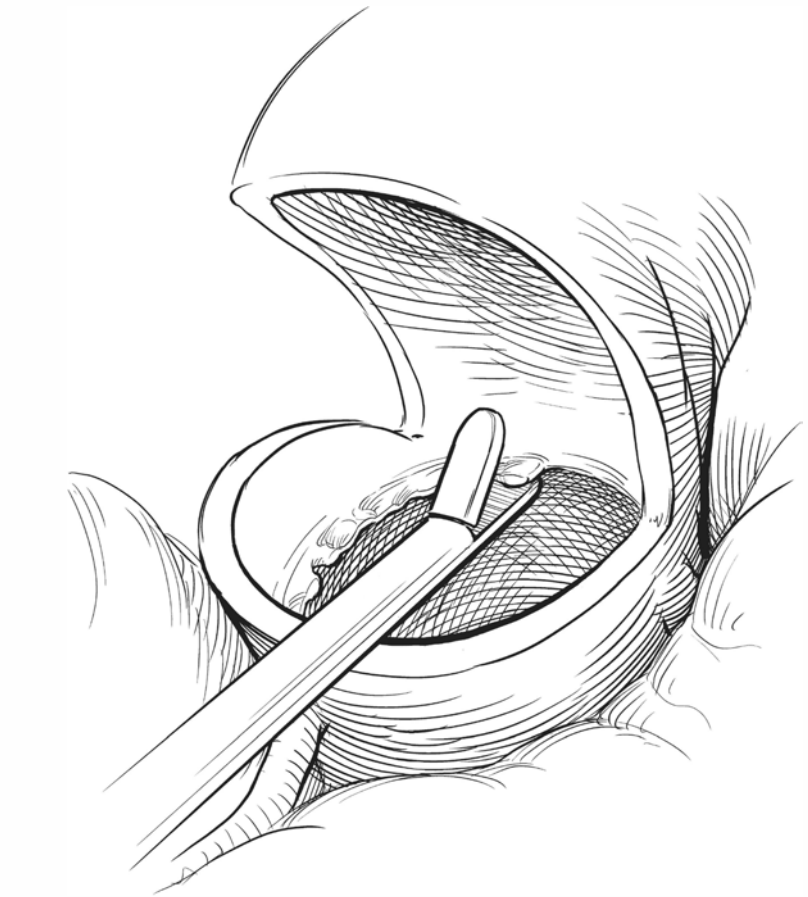
As the patient is systemically cooled, the heart will fibrillate. The aortic cross-clamp is then applied, and cardioplegia is administered.

A small, transverse aortotomy is made at the point of the previously placed mark. Through this initial aortotomy, one may visualize the aortic valve. The aortotomy is then extended transversely across the anterior surface of the aorta. It is important to stay approximately 1 cm distal to the zenith of the commissures of the aortic valve leaflets. Having extended the aortotomy to the patient's right, once the incision is exactly over the half-way point of the noncoronary leaflet, the aortotomy incision is directed to the axis of the aorta down toward the aortic annulus. This portion of the aortotomy incision should stop at least 1 cm distal to the aortic annulus.

The aortic valve is best visualized with the operating table in a bit of reverse Trendelenberg position and rotated a bit to the patient's left. Traction sutures are then placed through the top of each commissure and snapped to the surgical drapes. This brings the aortic valve up toward the surgeon. The aortic valve leaflets are then excised with scissors. After the leaflets have been removed, a moist gauze sponge is placed in the lumen of the left ventricle to help catch any small pieces of calcium. A Rongeur instrument is then used to gently debride the annulus of calcium.



C



D

Fig. 48.1. (Continued)

During this process, it is helpful for the assistant to follow along with an open-tipped suction catheter to help sweep up any small pieces of calcium. Once the annulus has been sufficiently debrided of calcium, the sponge is removed from within the ventricle, and the lumen of the left ventricle is liberally irrigated with cold saline to flush out any calcium debris. The annulus is then sized.

It is very important not to attempt to place an oversized valve. Regardless of the choice of prosthesis, I routinely implant a valve one size smaller than the patient's annulus might accept as judged by valve sizers. Horizontal pledgetted mattress sutures are placed in the aortic annulus with the pledgets in the subannular position. Note that the annular size will be somewhat smaller once all the valve sutures have been placed. Therefore, I finalize the choice of valve size only after all the valve sutures have been placed. The aortic valve prosthesis is then brought to the field and the sutures passed through the valve sewing ring. To facilitate symmetrical suture placement, it is helpful to mark the sewing ring in thirds.

Once the sutures are passed through the sewing ring, the valve is seated into the aortic annulus, and the sutures tied. To minimize difficulty in seating the valve, the sutures at each of the three commissures should be tied first. Next, a suture midway between each commissure should be tied. In this manner, the surgeon may be assured that the valve will seat appropriately.

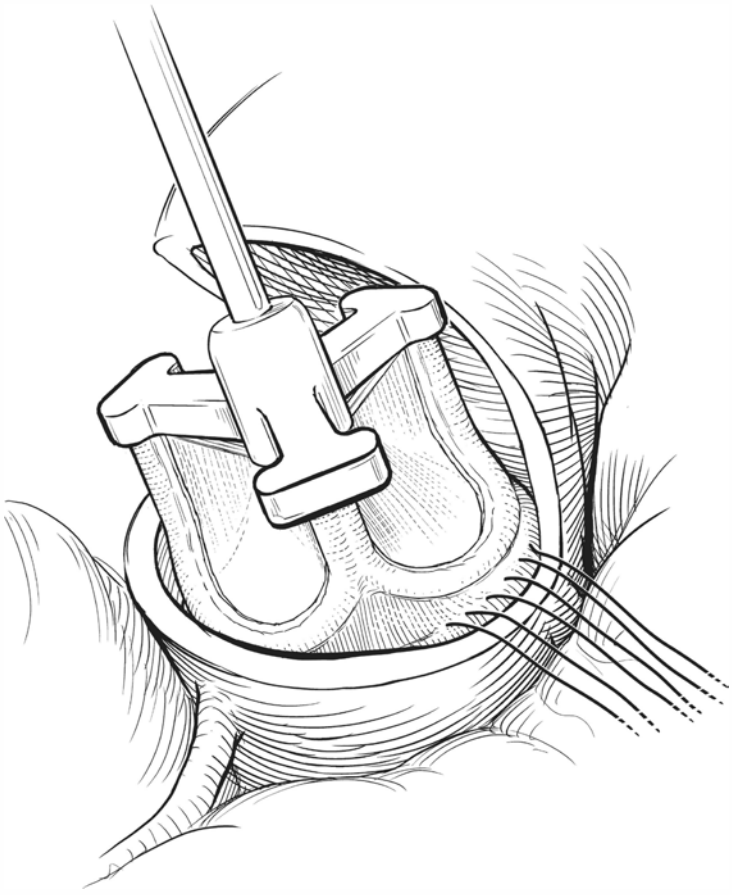
Once the valve is sewn in place, the aortotomy is closed with 5-0 polypropylene sutures in two layers. The first layer is a running horizontal mattress stitch, and the second is an over-and-over running stitch. Once the aortotomy is closed, I routinely administer antegrade cardioplegia in order to test the suture line. It is easier and safer to repair any leaks before removal of the cross-clamp.

In anticipation of removing the aortic cross-clamp, I typically infuse warm blood in retrograde manner. The purpose of this is to flush air out of the coronary arteries and to begin increasing the myocardial metabolic rate before reanimating the heart.

During this infusion, it is helpful to begin de-airing the left ventricle by partial occlusion of the venous line and the resumption of ventilation while the left ventricular and aortic vents are on suction. After the retrograde administration of 500 cm^3 of warm blood, I administer warm blood in an antegrade manner until the heart reanimates. With the heart beating, the aortic cross-clamp is removed. With the heart beating, one should assess the adequacy of the de-airing maneuvers by transesophageal echo. The usual maneuvers include filling the heart while on cardiopulmonary bypass, rotating the operating table from right-to-left and Valsalva maneuvers to express air out of the pulmonary veins. Once satisfied that the left heart is completely de-aired, the



E



F

Fig. 48.1. (Continued)

left ventricular vent is removed and the patient is weaned from cardiopulmonary bypass.

SURGICAL OUTCOMES

According to the Society of Thoracic Surgeons (STS) National Cardiac Surgery Database in 2010, approximately 35,000 valve operations are performed in the United States annually; just over one half are cases of isolated AVR. The operative mortality rate in the STS Database for isolated AVR is approximately 2.6%, and the incidence of stroke is 1.3%. The operative mortality rate for combined AVR/CABG is approximately 6.8%.

Certain patient-specific risk factors markedly increase the odds ratio (OR) of death following AVR. Operative mortality is significantly increased with surgery performed under salvage status conditions (OR 7.12), dialysis-dependent renal failure (OR 4.32), emergency status (OR 3.46), nondialysis-dependent renal failure, and first reoperation (OR 1.70).

CHOICE OF PROSTHETIC AORTIC VALVE

The operative risks of cardiac valve replacement are not associated with the choice of prosthesis. Further, the hemodynamic performances of contemporary valves are similar. Traditionally, the choice of aortic valve prosthesis has focused on whether a patient would be committed to the risks of life-long anticoagulation (mechanical valve) or the presumed need for reoperation for structural valve deterioration (bioprosthetic valve). However, this approach is an over simplification and the choice of prosthetic valve must be patient-specific. In addition to appropriate consideration of a given patient's comorbidities, one of the most important considerations is the age of the patient at valve implantation. In this light, it is important to recognize that approximately 80% of all valve replacements in the United States are performed in patients above the age of 60 years. Currently, the use of bioprosthetic valves has risen to 80% of all implanted valves.

Following AVR, the median survival for patients <80 years is approximately 11 years; for octogenarians, it is approximately 6 years. The 10-year survival for patients following AVR ranges from 40% to 70%, with an average in the literature of 50%. The type of prosthesis does not impact survival, but other patient-specific factors such as age at operation and presence or absence of coronary artery disease do impact survival following valve replacement.

Regardless of the type of prosthetic valve implanted, approximately one-third of patients die of valve-related causes. Given that valve-related complications occur at a frequency of about 3% to 6% per year, it is important to ask if the risks in a specific patient may be minimized by the choice of a mechanical versus a bioprosthetic valve. The choice of valve prosthesis must be individualized.

Mortality among heart valve recipients is valve-related in approximately 30% of patients. The principal causes of valve-related death following valve implantation include thromboembolism (12%), reoperation (10%), bleeding (4%), and prosthetic valve endocarditis (PVE) (3%). The risk of PVE is not different between mechanical or tissue valves. It is approximately 4% spread over the patient's lifetime. However, if PVE does occur, it is associated with up to a 50% mortality rate.

The leading cause of valve-related death is thromboembolism. Largely because mechanical valves are thrombogenic, the risk of thromboembolism is greater with mechanical valves. At 10 years following AVR, the risk of thromboembolism is 20% for mechanical valves and 9% for bioprosthetic valves.

Because a mechanical valve obligates the patient to chronic anticoagulation therapy (warfarin sodium), the choice of a prosthetic valve must consider the risks of chronic anticoagulation. The risk of bleeding complications from chronic anticoagulation is between 1% and 2% per year. In fact, 4% of valve-related deaths result from bleeding. Mechanical valves should be avoided in patients with contraindications to anticoagulation because of occupation or because of coexistent medical conditions. Similarly, patients who are medically noncompliant or whose level of anticoagulation may not be closely monitored should not receive mechanical valves.

Ten percent of valve-related deaths result from reoperation. It was traditionally assumed that following implantation of a tissue valve, patients would require reoperation for structural valve deterioration. Mechanical valves were, therefore, recommended for patients with a life expectancy longer than 10 years. This reasoning requires refinement. First, placement of a mechanical valve does not eliminate the potential for subsequent valve reoperation. While mechanical valves will not structurally fail, approximately 10% of mechanical aortic valves require reoperation within 5 to 10 years, primarily for paravalvular leak, endocarditis, or nonstructural valve dysfunction such as scar tissue or pannus in-growth. Second, the structural durability of newer bioprosthetic valves is superior to prior generations of valves. Third, it is now appreciated that on average, patient death (all-cause mortality) occurs prior to reoperation for structural valve deterioration. In fact, the incidence of reoperation for structural valve deterioration of a bioprosthetic valve is $<$15% for patients older than 60 years.

THE ASYMPTOMATIC PATIENT

Asymptomatic Aortic Stenosis

In general terms, asymptomatic patients do not require AVR. However, some do carry a low but real risk of sudden death. The challenge is the identification of the subset of asymptomatic patients in whom the risk of death is greater if AVR is not performed, or in whom the likelihood of AVR in the near-term is probable.

The Risks without Operation

The risk of sudden death in asymptomatic patients is considered to be quite low. However, in a rigorous study of nonoperated patients with asymptomatic aortic stenosis, the incidence of sudden death was approximately 6%. Further, one must also consider that the mortality is 3% to 4% soon after the onset of symptoms.

Unmasking the Symptoms

Given the patient with significant aortic stenosis by echocardiogram, it is therefore important to determine that the patient is truly asymptomatic. This may be objectively confirmed by exercise stress testing (treadmill). While unnecessary and risky in the symptomatic patient with aortic stenosis, exercise testing has been found safe in asymptomatic patients. A modified Bruce protocol should be employed under careful observation. An exercise test is considered positive if symptoms occur, systolic blood pressure falls by more than 10 mmHg, dysrhythmias occur, or ST segment changes are noted. Given a positive exercise test, the patient should be considered symptomatic and offered surgery. As many as 66% of asymptomatic patients may have a positive stress test.

Outcomes of Those with a Negative Stress Test

Following a negative stress, patients must be closely followed for hemodynamic progression of aortic stenosis. On average, the aortic flow velocity increases by 0.3 m/s per year and the AVA decreases by 0.1 cm^2 per year. However, the flow velocity is largely dependent upon ventricular contractility and should ventricular function decline, velocity may not change despite a smaller AVA. Further, the rate of progression of aortic stenosis is quite variable, making it difficult to predict the clinical course of a given patient.

Several characteristics have been identified, which help stratify asymptomatic patients. Otto and colleagues identified aortic flow velocity as the most important patient-specific variable. The highest risk group were those asymptomatic patients with an aortic flow velocity of $>$4 m/s, as only 21% were alive without valve replacement at 2 years.

Patients with "very severe" aortic stenosis are defined as those with a peak aortic jet velocity $>$5 m/s, a mean gradient $>$60 mmHg, and a valve area $<$0.6 cm^2. Asymptomatic patients with very severe aortic stenosis have a very poor prognosis; with event-free survivals was 64% at 1 year and 36% at 2 years. In the only published direct comparison of surgery and conventional therapy in asymptomatic patients with very severe aortic stenosis, Kang and colleagues examined 197 consecutive patients. Early surgery was performed in 102 patients, and a conventional treatment strategy was followed in 95 patients. In the conventional treatment arm, surgery was deferred until the onset of symptoms. At 6 years, the cardiac mortality rate in the early surgery group was 0%, and 24% in the conventional treatment group. Provided that AVR may be performed with low morbidity and mortality, asymptomatic patients with very severe aortic stenosis should undergo AVR.

In addition to the absolute baseline aortic flow velocity, longitudinal follow-up may reveal patients with a rapidly increasing flow velocity on serial echocardiograms; these patients are at risk for cardiac events. Rosenhek and colleagues noted that among patients who remained asymptomatic, the average rate of progression of aortic flow velocity was 0.14 m/s per year. But among those patients who experienced cardiac events, it was 0.45 m/s per year.

Patient age and aortic valve calcification may also be important. In a study of asymptomatic patients, all of whom had aortic flow velocity $>$4 m/s, those above age 50 years were much more likely to experience cardiac events in follow-up. Interestingly, in this same group, moderate or severe aortic valve calcification was strongly associated with subsequent cardiac events.

From a practical standpoint, I have found the following helpful. For the patient who appears to be asymptomatic following carefully obtained history, an exercise stress is obtained. Patients with a positive stress test are offered surgery. If the stress

test is negative, I have found the recommendations of the European Society of Cardiology helpful and recommend AVR if the aortic flow velocity is >4 m/s, the aortic valve has moderate or severe calcification and the rate of progression of aortic flow velocity is >0.3 m/s per year. I also recommend AVR if the left ventricular ejection fraction (LVEF) is <50% as this implies ventricular dysfunction secondary to aortic stenosis.

Asymptomatic Aortic Regurgitation

AVR is clearly indicated in the patient with symptomatic aortic regurgitation. Conversely, the natural history of asymptomatic severe aortic regurgitation is not well defined. Ten years after the diagnosis of severe aortic regurgitation, as many as 75% of patients will have died or undergone AVR. It is now clear that upon diagnosis, certain subsets of asymptomatic patients with severe aortic regurgitation who should be offered AVR. The recommendation for AVR should be based on left ventricular size and function.

There is consensus that the asymptomatic patient with severe aortic regurgitation with subnormal LVEF at rest should undergo AVR. This consensus is based on a 25% progression per year to heart failure or death in these patients. Evidence of left ventricular dilation is strongly associated with progression to heart failure or death; a left ventricular end-systolic dimension >55 mm (or >25 mm/m^2) or a left ventricular end-diastolic dimension >80 mm are an indication for AVR. Similarly, if longitudinal echocardiographic follow-up reveals either "rapid" diminution in LVEF or a "rapid" increase in left ventricular dimensions, AVR is indicated. While normal at rest, myocardial performance may be found abnormal with exercise. Hence, exercise stress testing may be used to unmask myocardial contractile dysfunction; a fall in LVEF with exercise >5% is a strong relative indication for surgery.

LOW-GRADIENT AORTIC STENOSIS

Low-Gradient "Severe" Aortic Stenosis with Preserved Left Ventricular Function

The management of these patients is controversial. Unfortunately, up to 30% of patients evaluated for aortic stenosis are found to have echocardiographic discrepancies among the parameters aortic stenosis. Such patients are found to have an AVA that would be considered severe aortic stenosis, but not severe as judged by mean pressure gradient in the setting of a normal LVEF. The explanation for such discrepancies is unclear. Using retrospective data, some authors have recommended that these patients should undergo AVR, suggesting that such patients have advanced aortic valve disease and left ventricular impairment with a reduced stroke volume. However, this may not be the case.

Jander and colleagues prospectively followed 1,525 asymptomatic patients with low-gradient "severe" aortic stenosis (AVA <1.0 cm^2 and mean gradient <40 mmHg) and 184 patients with moderate aortic stenosis (valve area 1.0 to 1.5 cm^2 and mean gradient 25 to 40 mmHg). At 46 months of follow-up, there were no differences in valve-related events, major cardiovascular events or cardiovascular deaths. These data suggest that this subset of patients have a prognosis akin to moderate aortic stenosis, and that surgery may be deferred.

Low-Gradient Aortic Stenosis with Left Ventricular Dysfunction

Among the most challenging patients with aortic stenosis are those with a low transvalvular gradient and severe left ventricular dysfunction. Low-gradient aortic stenosis is defined as less than 30 mmHg. Treated medically, the median survival of these patients is usually <2 years; the 1- and 4-year survivals are 41% and 15%, respectively. This subset represents less than 5% of patients with aortic stenosis. The clinical outcomes of AVR in these patients have not been well characterized, but the operative mortality rate for AVR in this group of patients has historically been quite high, leading some authors to recommend against surgery.

When the patient with severe left ventricular dysfunction associated with low-gradient aortic stenosis is first seen, it may be difficult to know if the left ventricular dysfunction is primary or secondary. The pathogenesis of left ventricular dysfunction secondary to aortic stenosis is derived from afterload mismatch. With significant aortic stenosis, the left ventricle compensates for pressure overload by hypertrophy, thereby normalizing wall stress. In this manner, LVEF and cardiac output are initially maintained. When the ventricle is no longer able to compensate for the increased wall stress, left ventricular systolic function declines secondary to afterload mismatch. The transvalvular pressure gradient may be low despite the presence of severe aortic stenosis. Provided the left ventricle still has the ability to contract (contractile reserve), clinical outcomes with AVR are quite good; operative mortality is low, and left ventricular function improves after AVR.

On the other hand, the left ventricular dysfunction may be primary rather than secondary to the aortic stenosis. The dysfunctional left ventricle may be unable to open a mildly calcified aortic valve fully if the stroke volume is significantly decreased. This condition is termed "relative aortic stenosis." These patients have a cardiomyopathy (ischemic or otherwise) with only mild aortic valve narrowing.

For the surgeon and the patient, the principal questions relate to the risks of an operation and whether the patient will be improved afterward. The answers must rely upon a paucity of data regarding the clinical outcomes of patients with low-gradient aortic stenosis and severe left ventricular dysfunction. Taken together, the available data suggest that patients with low-gradient aortic stenosis and severe left ventricular dysfunction may be stratified according to the severity of aortic stenosis and the contractile reserve of the left ventricle. This can be accomplished with a pharmacological challenge with dobutamine. During an infusion of dobutamine up to 40 μg/kg/min, the hemodynamic response may be monitored by echocardiography or cardiac catheterization. In response to dobutamine infusion, stroke volume and thereby cardiac output should increase, raising the transvalvular gradient. An increase in stroke volume of at least 20% is considered evidence of contractile reserve.

In my own experience, I have come to rely heavily upon the echocardiographic appearance of the aortic valve. If the valve is densely calcified with severely restricted leaflet motion, I feel confident the patient has severe aortic stenosis. Conversely, if the leaflets are not badly calcified, or the leaflet motion subjectively seems better than the patient's clinical condition suggests, I examine the hemodynamics during dobutamine infusion. A dobutamine stress echocardiogram is useful in two ways. First, if the stroke volume (and thereby cardiac output) increases by at least 20%, the patient has contractile reserve. If the increase in stroke volume is associated with an increase in transvalvular gradient, I feel confident the patient will survive AVR with a postoperative improvement in LVEF and functional status. But if no increase in stroke volume and transvalvular gradient are noted with dobutamine, the patient has no contractile reserve or may not have severe aortic stenosis. AVR is not recommended.

"PROPHYLACTIC" AORTIC VALVE REPLACEMENT

The finding of moderate aortic stenosis in patients undergoing concomitant cardiac operations is not uncommon. Whether or not to replace the aortic valve may be a difficult and controversial decision, particularly in patients undergoing coronary artery bypass surgery. In 1994, Collins and colleagues reported a high operative mortality rate (18.2%) in patients undergoing AVR who had had prior coronary artery bypass surgery. Shortly thereafter, several other groups reported similar results. Following these reports, an international trend toward "prophylactic" AVR began for patients with mild-to-moderate aortic stenosis undergoing coronary artery bypass surgery. The decision to perform a "prophylactic" AVR has always been controversial, and significant variation in the management of these patients continues.

Proponents of "prophylactic" AVR at the time of CABG argue the aortic stenosis will inevitably progress, committing the patient to a second operation associated with a high morbidity and mortality. Those favoring an expectant approach argue that "prophylactic" AVR unnecessarily commits an unacceptable number of patients to the risks of valve-related morbidity and mortality. While progression from asymptomatic to symptomatic aortic stenosis in a relatively short time is clinically recognized, it is very difficult to identify those patients likely to do so. At the heart of the argument is the need for an understanding of the natural history of mild-to-moderate aortic stenosis and the contemporary risks of AVR after previous CABG.

The natural history of moderate aortic stenosis (AVA 1.0 to 1.5 cm^2) is difficult to define but is clearly worse than that of mild aortic stenosis. Rosenchek and colleagues reported an ominous natural history for patients with moderate aortic stenosis (aortic flow velocity 2.5 to 3.9 m/s). Of 176 patients followed for an average of 48 months, 33 underwent AVR and 34 patients died. The 5-year event-free survival was 60%. The strongest predictors of patient events were aortic valve calcification, a peak aortic flow velocity at study entry >3 m/s, age >50 years, and coronary artery disease. The data from Rosenchek are consistent with those of Otto and clearly indicate that patients with moderate aortic stenosis may experience hemodynamic progression during relatively short follow-up.

The operative mortality rate reported for AVR subsequent to CABG has significantly diminished in recent years. Certainly advances in myocardial protection, surgical and anesthetic techniques as well as greater collective experience with cardiac reoperations have contributed to improved outcomes. In recent series, the mean interval between CABG and subsequent AVR is typically 7 to 9 years. In these reports, the operative mortality rate of the AVR at reoperation has been approximately 7% and not significantly different from the operative mortality associated with AVR/CAB done in the same institutions, suggesting that an expectant approach is warranted.

Smith and colleagues utilized the STS National Cardiac Surgery Database and employed Markov decision analysis to analyze quality-adjusted outcomes of patients with mild-to-moderate aortic stenosis undergoing CABG alone versus CABG/AVR. Assuming a constant rate of progression of aortic stenosis of 5 mmHg/year, the authors concluded that "prophylactic" AVR may be indicated in all patients once the valve gradient exceeds 28 mmHg. However, in patients above the age of 70 years, the threshold to "prophylactically" replace the valve steadily rises; the morbidity of AVR plus CABG is higher than with CABG alone. Dagenais and colleagues reported that in patients >70 years with moderate aortic stenosis, the operative mortality of AVR/CABG was not different than CABG alone. But the incidence of perioperative neurological injury, mediastinitis, and renal failure were all significantly higher. Further, there was no difference in at 5-year survival.

Patients with mild AS at the time of CABG (AVA >1.5 cm^2; aortic flow velocity <2.5 m/s) have an excellent prognosis from their aortic valve and should not have AVR. On the other hand, patients above age 65 years who undergo CABG with moderate aortic stenosis and an AVA <1.2 cm^2 and/or an aortic flow velocity >3m/s associated with significant aortic valve calcification are likely to have symptomatic aortic stenosis within the next several years. In this subset of patients, it is reasonable to perform AVR with CABG at the initial operation.

AORTIC VALVE REPLACEMENT WITH A PATENT INTERNAL MAMMARY

AVR in patients who have had previous coronary artery grafting, particularly if the internal mammary artery (IMA) graft is patent, can be technically challenging. It is associated with mortality rates of 6% to 17%, considerably higher than first-time AVR procedures. Contributing to this mortality rate is the fact that the IMA may be damaged during the procedure, or its presence may compromise myocardial protection during the procedure.

To minimize the risk of injury to the IMA, some advocate moderate-to-deep systemic hypothermia (20°C) while on cardiopulmonary bypass, aortic occlusion, administration of cardioplegia, but avoidance of dissection of the IMA pedicle. Others also leave the IMA open during the period of aortic occlusion but administer continuous retrograde cardioplegia. With either technique, flow through the IMA may washout cardioplegia and warm the heart during the procedure. A third option is to utilize deep hypothermia and circulatory arrest.

While these techniques may be helpful in situations in which the IMA is densely adherent to the posterior sternum and cannot be safely dissected, it is preferable to dissect out the IMA and occlude it during aortic cross-clamping. The IMA may be isolated even before re-sternotomy using a supraclavicular incision. With interruption of IMA flow, the surgeon may be confident that cardioplegia will not be washed out and the heart will be made uniformly cold.

It may also be difficult to expose the aortic valve through the standard aortotomy because of the presence of saphenous vein grafts anastomosed to the proximal aorta. In my own practice, I have found it helpful to cannulate the axillary artery in these cases. In so doing, the aorta may be cross-clamped just proximal to the innominate artery, which provides several more centimeters of ascending aorta unencumbered by the aortic cannula. This extra length of ascending aorta then permits an aortotomy incision (which avoids proximal anastomoses) made in the axis of the aorta, directed posteriorly along the patient's right. Excellent exposure of the aortic valve is achieved without injury to previously placed proximal anastomoses.

THE SMALL AORTIC ANNULUS

The use of all mechanical and stented bioprosthetic valves results in some degree of residual obstruction to flow because of the sewing rings and stents. Nonetheless, the goal of AVR is to leave the patient with minimal gradient across the valve. In so doing, the increased left ventricular mass index (developed in compensation for aortic stenosis) typically resolves following aortic valve replacement. Traditional concern has surrounded the potential for a high residual transvalvular gradient should too-small a prosthesis be implanted, creating

the phenomenon of "prosthesis–patient mismatch" (PPM). The term "prosthesis–patient mismatch" was coined in 1978 by Rahimtoola and used to describe his observation that a patient's aortic stenosis may not be completely relieved by a prosthesis. This concern has led to a persistent controversy over the management of the small aortic annulus.

While some authors have demonstrated that implantation of small aortic valve sizes has no effect on long-term survival, others have suggested otherwise. One factor confounding the interpretation of these data is the fact that the sizing of prosthetic valve is inconsistent from one manufacturer to the next. Depending on the manufacturer, a size 19 valve may vary in effective orifice area (EOA) from 1.0 to 1.3 cm^2. Another confounding factor is the fact that population of patients (with particular regard to patient age, gender, and size) has varied from one report to the next.

The patient's size (BSA) and age must be considered when choosing the appropriate aortic valve prosthesis. Acknowledging some of the uncertainties provided by the literature in this area, one should avoid implantation of an aortic prosthesis with a calculated EOA/BSA <0.8 cm^2/m^2; it is generally accepted that the likelihood of PPM is minimized if the prosthesis provides the patient with an EOA/BSAi >0.85 cm^2/m^2. An EOA/BSAi of 0.65 to 0.85 cm^2/m^2 is considered moderate PPM, and <0.65 cm^2/m^2 is considered severe PPM. This recommendation is derived from the fact that an AVA of 0.6 cm^2/m^2 is considered severe aortic stenosis. The use of this guideline is helpful as it places the consideration of valve size in the context of the patient's hemodynamic needs, rather than in absolute terms.

The implication of this strategy is the anticipation that the patient with a larger BSA will require a higher flow rate across the valve (cardiac output) than will the patient with a smaller BSA. Knowledge of the patient's BSA and the EOA of given sizes of a particular type of prosthesis permits an estimation of the valve size needed to leave the patient with an appropriate EOA/BSA ratio. Since PPM may not affect a difference in survival for 5 to 7 years other patient-specific factors must be considered such as age and activity level; the 60-year-old distance runner will require a larger EOA that the 80-year-old sedentary person. Nonetheless, this strategy serves as a useful guideline in choosing valve size.

The problem of the large valve relative to the small aortic annulus has plagued cardiac surgeons since the outset of aortic valve surgery. Within a few years of the first AVR by Harken in 1960, surgical techniques to surmount the problem by enlargement of the annulus emerged. By the end of the 1970s, Nicks, Konno, and Manouguian had each described techniques to enlarge the annulus in order that a larger valve may be implanted. Nicks (1970) and Manouguian (1979) each described techniques of patch enlargement of the posterior aspect of the aortic annulus. Konno described a more extensive procedure of aortoventriculoplasty of the anterior aspect of the aortic annulus in 1975. More recently, Otaki described a method of patch enlargement of both the posterior and anterior aortic annulus (1997).

Implantation of stentless bioprosthetic aortic valves was reported by 1990. Without the valve struts of their stented counterparts, the design of stentless valves offers an inherently larger EOA. In most centers, stentless valves are implanted by one of the two techniques: subcoronary or as a total aortic root replacement. While some controversy exists in the literature, it is generally accepted that the hemodynamic performance of stentless bioprosthetic valves is better than that of stented valves. In particular, the use of the root replacement technique is felt to minimize the risk of PPM.

Patch enlargement, stentless valve, and the Konno procedure may all be used effectively to avoid PPM. In my opinion, posterior patch enlargement of the annulus is the easiest and most straightforward technique. According to the STS National Cardiac Surgical Database, approximately 2.4% of AVR procedures performed in 2010 included an annular enlarging procedure. Early concerns were expressed that operative mortality was higher with the use of the technique, and it was therefore avoided by many. Contemporary experience, however, demonstrates that surgical outcomes are probably not different from those of AVR alone. The annular dimensions should be known from the preoperative echocardiogram, allowing the surgeon to plan to enlarge the annulus from the outset of the procedure. As a rule of thumb, a Nicks procedure (incision of the annulus in the mid-point of the noncoronary leaflet portion of the annulus) will generally permit one-size larger valve. On the other hand, a Manouguian procedure (incision of the annulus through the commissure of the non- and left valve leaflets) will generally permit the placement of a valve two sizes larger. In my experience, a Nicks procedure is typically sufficient and therefore my preferred technique (Fig. 48.2):

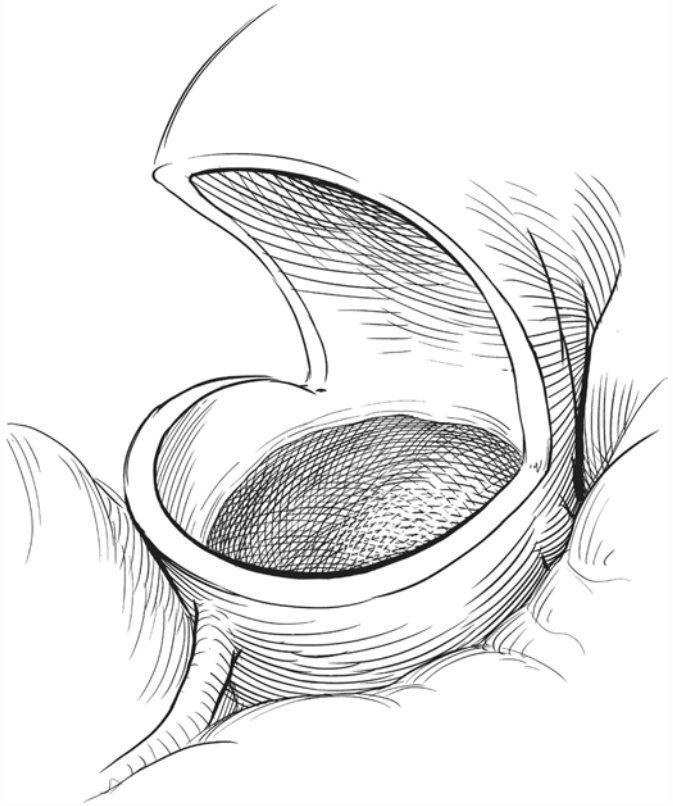
1. Enlarge the annulus at the midway point of the noncoronary leaflet. This is accomplished by extension of the aortotomy incision.
2. Extend this incision into the fibrous apron between the mitral and aortic annuli, but it is unnecessary to incise the mitral annulus or the anterior leaflet of the mitral valve.
3. Beginning at the apex of the incision, a fashioned piece of woven Dacron is sewn in place. The patch material is typically 2 cm wide, and beveled at the point of insertion into the annulus. A pledgetted 5-0 polypropylene suture is brought through the patch material, next the roof of the left atrium, and then the apex of the aortic incision, and tied. The patch material is sewn to the aorta for a distance of about 2 inches. I always use a second suture line of 5-0 polypropylene suture to aid with hemostatis.
4. The valve stitches are placed. In the area over the patch material, four pledgetted sutures of 4-0 polypropylene suture are brought from outside-in through the patch material.

Technical Caveat: At each edge of the patch, a horizontal mattress stitch is brought outside-in with one needle through the aorta and the other through the patch material. Between these two stitches, two additional valve sutures are typically placed. I prefer to use 4-0 polypropylene for these four valve sutures because the needle hole through the patch material is small, and bleeding less. The remainder of the valve sutures are pledgetted braided 2-0 sutures.

5. The patch material is then brought anteriorly to complete aortic closure.

Technical Caveat: If the patch material is too wide over the anterior aorta, the proximal aorta may be displaced anteriorly. In so doing, the orifice of the right coronary may be displaced and kinked. This may be avoided by cutting the patch material no wider than about 1.5 to 2 cm in this area. Should the right coronary artery ostium be distorted despite this, the patch material may be imbricated anteriorly in order to restore the normal position of the right coronary artery ostium.

6. Once the aorta is closed, test for hemostasis by administration of antegrade cardioplegia. A small amount of oozing through the patch material is to be expected until heparin reversal. But any bleeding sites, particularly near the apex of the patch closure, should be repaired before release of the aortic cross-clamp.

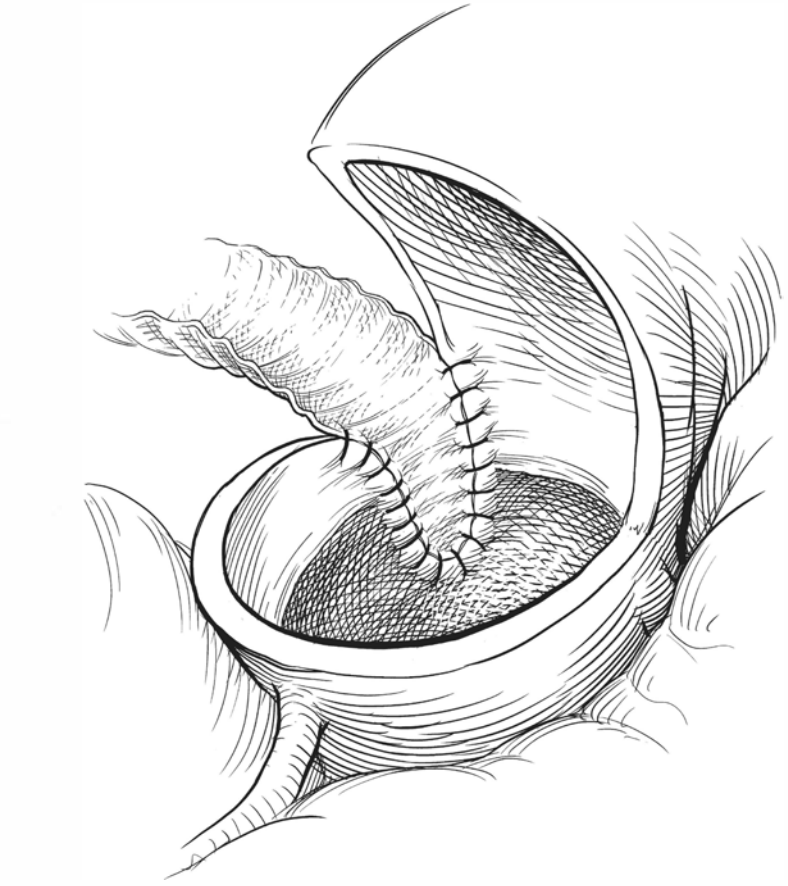


A

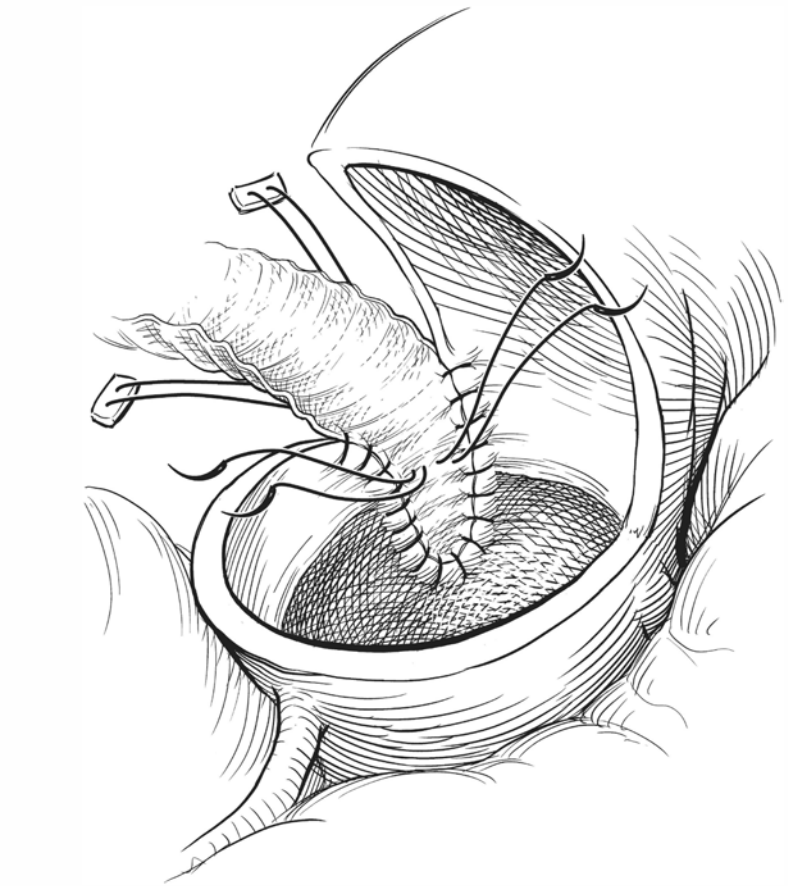


B

Fig. 48.2. Operative technique: aortic annulus enlargement. (**A**) The aortotomy incision is extended across the aortic annulus at the midpoint of the noncoronary leaflet. (**B**) Vascular patch material is partially sewn in place. (**C**) The valve is implanted. Across the patch material, the pledgetted valve sutures are placed from "outside-in." (**D–F**) The patch material is brought anteriorly to close the aorta.



C



D

Fig. 48.2. (Continued)

THE PORCELAIN AORTA

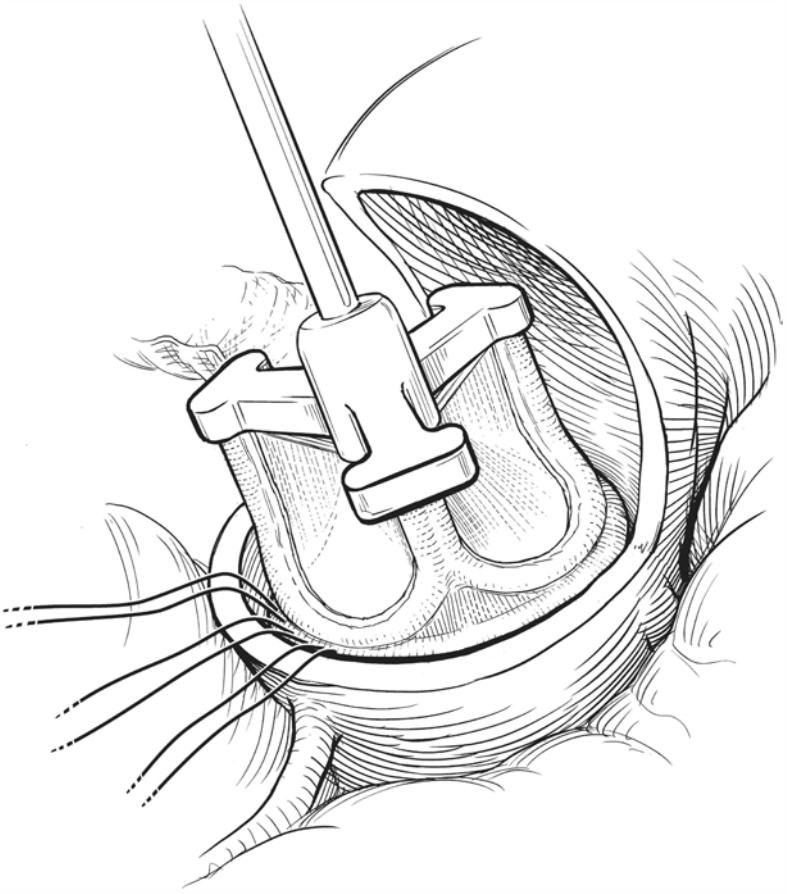
The presence of extensive, dense calcification of the aortic root and ascending aorta complicates a small but important percentage of patients undergoing AVR. In the setting of a porcelain aorta, AVR using the standard technique and aortic cross-clamp results in an unacceptably high risk of stroke and death. It may, in fact, be a contraindication for surgery in some patients. Hence, alternative surgical strategies must be utilized.

Hypothermic Circulatory Arrest

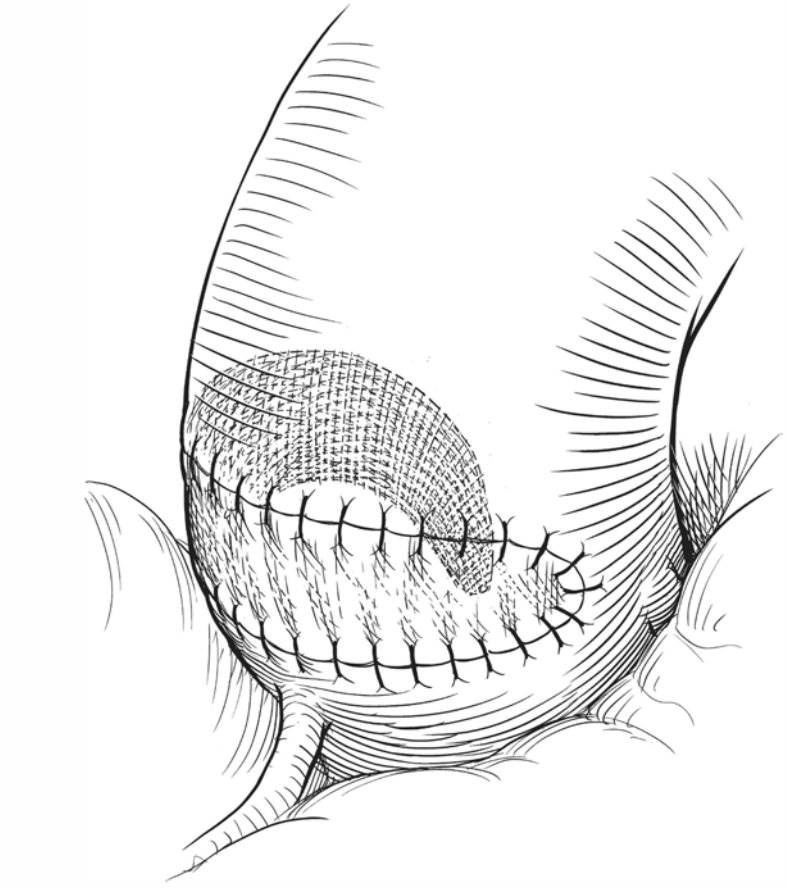
Peripheral arterial cannulation via the right axillary artery and the use of deep hypothermia and circulatory arrest without aortic cross-clamping can be an effective strategy for AVR with possible aortic replacement. Cosseli first described AVR using circulatory arrest in 1986, and Byrne likewise reported its use in 1998. Both reported excellent outcomes in a total of five patients. It may, in fact, be necessary to completely excise a badly calcified aortic root, and Wareing and colleagues first described aortic replacement under circulatory arrest in 10 patients in 1992. The operative mortality was 10%, and no patients sustained a stroke. More recently, Girardi and colleagues reported their experience of aortic valve and aortic replacement in 25 patients under circulatory arrest. The operative mortality was 8% and the incidence of stroke was 4%. In the opinion of this author, this is the preferred strategy. Right axillary artery cannulation provides the ability to employ selective cerebral perfusion during the period of circulatory arrest. Replacement of the calcified aorta obviates the need to perform aortic endarterectomy in order to secure aortic closure. Both operative mortality and stroke are acceptably low with the use of the technique.

Aortic Valve Bypass

Aortic valve bypass with the use of an apical–aortic conduit was first used clinically in 1962. It was reintroduced by J.W. Brown with a report of 23 patients in 1984. The technique is not commonly utilized, as fewer than 200 patients have been reported in the literature. Significant aortic regurgitation is considered a contraindication to the procedure. The procedure may be performed with or without the use of cardiopulmonary bypass, but in the experience of this author, the procedure is greatly facilitated by its use. As described by Gammie



E



F

Fig. 48.2. (Continued)

and colleagues, the procedure is performed through a left anterior–lateral thoracotomy incision. A valve-conduit is placed from the left ventricular apex to the descending aorta. The valve-conduit consists of three parts: a rigid apical connector, a stentless porcine aortic root, and a prosthetic graft. The three parts are sewn into continuity on the back table of the operating room. The use of a prosthetic graft with an 8 mm side branch permits arterial cannulation for cardiopulmonary bypass through the side branch. Success of the procedure requires that the apical connector be securely placed within the left ventricular apex, as the apex may be friable. This is best accomplished by full thickness bites through the ventricular muscle. Failure to securely place the apical connector may result in significant bleeding or pseudoaneurysm formation. As the prosthetic graft is anastomosed to the descending aorta, it is very important to be certain that the valve conduit will lay in a manner that will not be kinked and that its length is appropriate.

Using this technique in 47 high-risk patients, Gammie and colleagues recently reported an 11% operative mortality. The incidence of stroke was 6%. Blood flow through the valve-conduit was found to be a mean of 60% of total cardiac output. Survival at 1 year was approximately 70% and approximately 50% at 2 years. However, given the small number of patients that have undergone the procedure, the durability of the procedure is not well characterized.

SUGGESTED READINGS

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EDITOR'S COMMENTS

The chapter on Aortic Valve Replacement by Dr. Fullerton is truly a complete and learned discussion of this very common part of cardiac surgery. I agree with virtually everything with some minor acceptations. I agree that the technical aspects of this valve replacement are critically important. One should truly avoid impingement of the coronary ostia. There are some surgeons who enlarge the aortic root frequently and I believe our philosophy is closer to Dr. Fullerton's to enlarge less frequently. It makes no sense to put a large valve in a very small elderly patient.

The only minor disagreement we have is handling the patent internal mammary artery bypass in a reoperative situation where valve replacement is necessary. We have published that attempting to find the internal mammary in order to control it occasionally leads to injury of this important blood vessel. We, therefore, prefer now to use moderate hypothermia and intermediate cardiologic just as we do in every case and leave the internal mammary artery flowing. We have incurred extremely low mortality in this situation and little evidence that we create any LV dysfunction. In fact, mortality for redo aortic valve operations with patent coronaries is in the 2% range.

This is truly an excellent chapter. I think that Dr. Fullerton gives a great amount of detail about techniques to use in some really complex situations.

ILK

49 Ross Procedure

John D. Oswalt

The Ross procedure, first performed by Mr. Donald Ross in 1967, is the replacement of the aortic valve with the autologous pulmonary valve. It is considered the valve of choice for aortic valve replacement in children. It is also ideal for replacement in younger patients, athletes, women of child-bearing age, and patients with endocarditis. The operation since 1967 has evolved with better standardization of techniques and in multiple long-term studies has produced similarly excellent results.

INDICATIONS

Patients with a life expectancy of more than 20 years are candidates for the Ross procedure. Likewise, patients who cannot safely take or have a lifestyle not desirous of permanent anticoagulation are candidates for the procedure. Athletes whose sport requires extended periods of highly elevated heart rates will also see better performance with an autograft than a mechanical valve. Over the last 20 years, the Ross procedure has proven highly effective for aortic endocarditis. The autograft, with the flexibility of tissue and used as a root, allows for increased cure of the infection and has better longevity than other valve choices. Initially, discrepancy in aortic annulus to pulmonary annulus size was a contraindication. With improved techniques of dealing with the enlarged aortic annulus, reduction and fixation in a size suitable to the recipient, the Ross procedure is now performed in these patients. In the pediatric age group, the Ross procedure can be combined with the Konno procedure (myotomy and myectomy), particularly useful in combination with left ventricular outflow tract (LVOT) obstruction with aortic stenosis. An extension of right ventricular outflow tract (RVOT) muscle can be harvested with the autograft to fill in the myotomy for the enlargement of the LVOT. The viability of this graft then allows for continued growth with the child.

CONTRAINDICATIONS

Patients with Marfan's or other genetic disorders known to affect fibrillin or elastin of the aortic valve are not candidates for the Ross procedure because the pulmonary valve is likely to be affected by the same disease process. This includes patients with bicuspid aortic valve associated with root enlargement. Patients afflicted with significant immune-complex disease (juvenile rheumatoid arthritis, lupus erythematosus, and active rheumatic heart disease) as the etiology of their aortic valve disease have had early failure or degeneration of the pulmonary autograft valve. These patients are not considered candidates for a Ross procedure. In general, patients who fall into these categories have had early failure of the autograft through dilatation of the root into an aneurysm and/or development of aortic insufficiency.

SURGICAL TECHNIQUE

A standard median sternotomy is recommended, as adequate exposure of the great vessels and roots is mandatory for safe harvesting of the autograft and implantation of the coronary ostia. The operation then proceeds as for an aortic valve replacement. Cannulation high on the aorta is done in case remodeling or resection of some of the ascending aorta is necessary. Either bicaval venous cannulation or two-stage single venous cannulation is acceptable; however, care must be taken to avoid air lock in the venous return when the RVOT is opened. Antegrade cardioplegic and aortic sump line is placed as well as retrograde coronary sinus cannula for intermittent or continuous retrograde blood cardioplegia. Manipulation of this cannula can be facilitated by the echo-anesthesiologist without having to open the atria. A left ventricular sump is placed via the right superior pulmonary vein (Fig. 49.1). As cardiopulmonary bypass is begun, dissection between the great arteries is performed until the take-off of the right pulmonary artery is seen. Systemic cooling is accomplished to 32°C to 34°C. With aortic clamping antegrade followed by retrograde blood cardioplegia is delivered for complete arrest. This is facilitated with saline slush in the pericardial well. Initially, the autograft is harvested by making a transverse arteriotomy proximal to the right pulmonary artery take-off (Fig. 49.2). Inspection of the valve leaflets is done to rule-out a deformity (bicuspid or fused) in the pulmonary valve (Fig. 49.3). A bicuspid pulmonary valve should not be used. Finding this, one would abort the Ross and use the backup valve planned with the patient preoperatively. The cryopreserved pulmonary homograft can then be thawed, so it will be ready for implantation later. The harvesting of the autograft is begun posteriorly using the electrocautery. One must dissect close to the pulmonary artery. Caution is taken as the left main coronary artery lies posterior and superior at the base of the pulmonary artery. Dissection is carried down to the level of ventricular muscle (Fig. 49.4). A right-angled clamp is then placed through the opening of the pulmonary valve visualizing the RVOT muscle beneath the annulus. The clamp can then be pushed through the muscle about 3 to 4 mm from the annulus (Fig. 49.5). From this opening, sharp dissection is accomplished with scissors and direct visualization of the annulus. Three millimeters of subannular muscle is taken with the graft. Initial dissection is to the patient's left to the point where the posterior wall of the RVOT begins. Similarly, the dissection is taken to the patient's right, again to the posterior wall. Scalloping the underlying muscle is useful always maintaining the same 3 to 4 mm of muscle (Fig. 49.6). The posterior wall is incised with a scalpel and dissection is carried to a depth in the muscle similar to the thickness of the RVOT muscle cut through anteriorly. Embryologically, the RVOT is separate

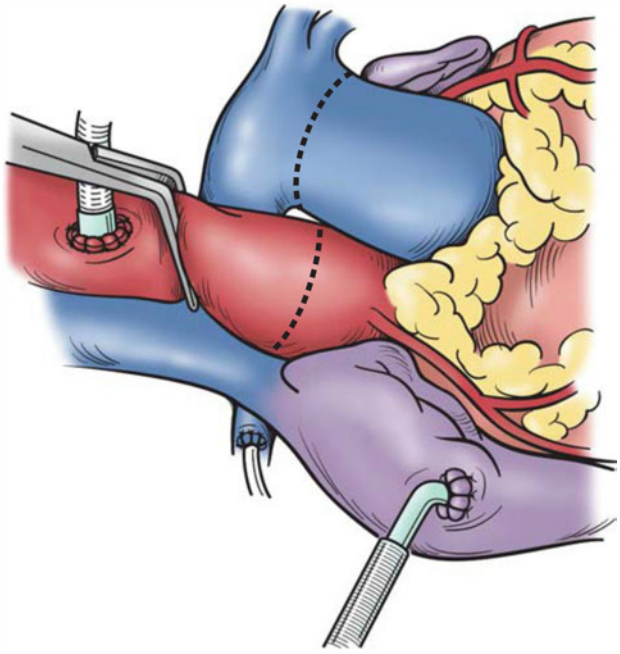


Fig. 49.1. Cannulation: Distal aorta, bicaval cannulation with the superior vena caval cannula placed through a purse string in the vena cava, the left ventricular vent through the right superior pulmonary vein, and the retrograde cardioplegia cannula through the right atrium. All illustrations are oriented as seen by a surgeon standing on the right side of the patient.

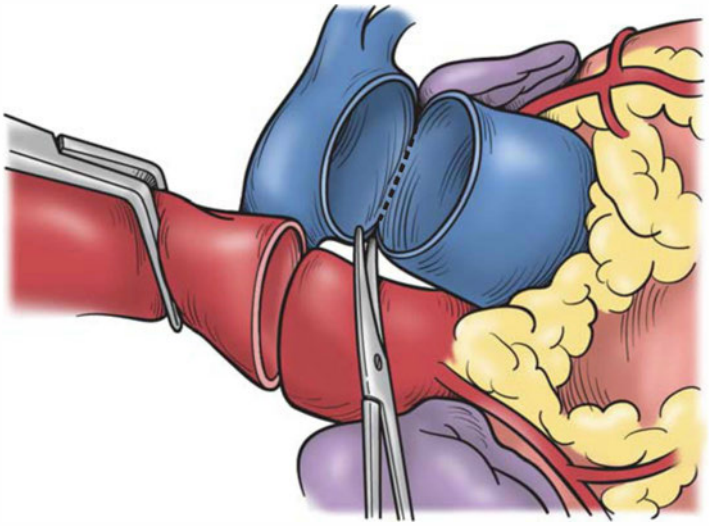


Fig. 49.2. The distal pulmonary artery is incised at the origin of the right pulmonary artery. A transverse arteriotomy is made that is adequate for careful inspection of the pulmonary artery.

from the septal muscle and this plane can be seen as one dissects directly posterior. When this plane is reached, care must be taken to avoid the first septal perforator which runs in this layer. The most common course of the artery is from the anterior descending coronary to the middle papillary muscle of the tricuspid valve, which is easily visualized. The angle of dissection is changed to be more horizontal and is carried craniad until the level of the previous posterior dissection is reached. This is the enucleation of the valve. Caution around the sinuses is important as too close dissection may injure the sinus wall. The valve can then be inspected for injury and then placed in the pericardial well for safe keeping. This harvested valve can later be used as a subcoronary (scalloped) valve or as a root.

SUBCORONARY IMPLANTATION

Most surgeons now use one of the two techniques for the implantation of the autograft. The subcoronary technique, Mr. Ross' original technique, uses a hockey-stick aortotomy. The aortotomy is started 2 to 3 cm above the right coronary ostium and angled down into the noncoronary sinus. The aortic valve is excised and the annulus sized. The autograft is then trimmed of excess epicardial fat and tissue, and the subannular muscle is trimmed to 2 to 3 mm. The sinuses of what will be the left and right sinus of the autograft are excised leaving 2 to 3 mm of arterial wall above the annulus. The distal artery is trimmed to just above the commissures. This leaves the noncoronary sinus intact. Implantation is accomplished by interrupted 4-0 polypropylene sutures or in running single or triangulated-running suture. The triangulation is done by placing three separate sutures through the aortic annulus and in the subannular autograft at the nadir of the cusp. Interrupted sutures are then placed at 2 mm separation between the triangulated sutures. Some surgeons will reinforce this proximal suture line with a strip of autologous pericardium. If a running triangulated suture technique is used, each suture can then be run to the commissures. Similarly, a single running suture can be done from the nadir of the left cusp (beneath the left coronary ostium) and run first counterclockwise to the right noncommissure followed then by the other end of the suture run clockwise to meet the suture at the right noncommissure. Some surgeons facilitate the exposure by inverting the autograft valve into the LVOT. As the muscle of the autograft was somewhat scalloped with the harvesting, the line of the anastomosis will follow the natural annulus. With the proximal anastomosis completed, the distal implant is accomplished with a running 4-0 polypropylene suture. The right–left commissural post is tacked to the sinus wall 3 to 5 mm above the existing native commissure. It is important to have the post in this suspended position as laxity will result in prolapsing of the leaflets. The noncoronary sinus maintains the relation of its connected commissural posts at 120 degrees. Tacking of the other posts is facilitated by leaving the noncoronary sinus intact. The suture line proceeds in scallop manner beneath the coronary ostia attaching the autograft to the sinus walls in a hemostatic manner. The noncoronary sinus is tacked to the remaining aortic wall to remove any free space behind it. Closure of the aorta then incorporates aorta to autograft in a manner as to not buckle

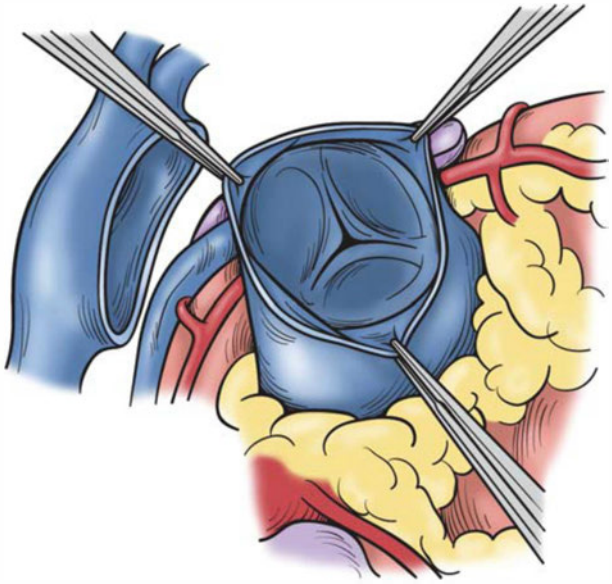


Fig. 49.3. The normal trileaflet pulmonary valve with three equal sinuses and no significant fenestrations or other abnormalities of the leaflets.

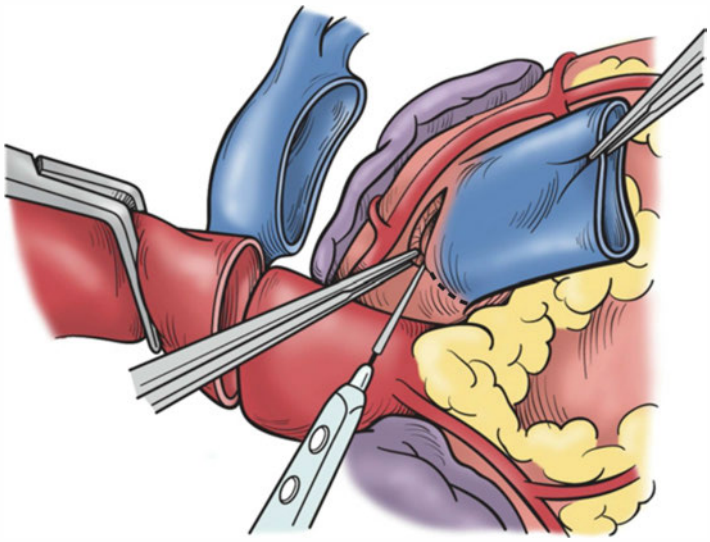


Fig. 49.4. Dissection of the pulmonary autograft is initiated on the posterior aspect of the proximal pulmonary artery. Dissection is continued in this plane, adjacent to the pulmonary artery, until septal myocardium is encountered. The left main coronary artery and left anterior descending coronary artery are protected.

the noncoronary sinus of the autograft. Advantages of this implantation technique include not having to reimplant the coronary ostia and avoidance of late sinus dilation as may be seen with the root technique. Disadvantages include inability to perform if there are dilated sinuses and inability to do if calcifications exist in the sinus walls prohibiting the suturing in the wall of the sinus. Groups with long-term results using this technique have achieved similar results with those surgeons using the root implantation technique.

ROOT IMPLANTATION

The root implantation technique has become the most popular for insertion of the pulmonary autograft. It is easier to insure a competent aortic valve as no manipulation of the commissural posts is required. Since beginning the use of the Ross procedure, we have exclusively used this technique. Patients with dilated sinuses, dilated sinotubular junction, true bicuspid valve with 180-degree coronaries are all candidates for a root replacement. Cannulation is the same for the root replacement as with the subcoronary technique. Bypass and autograft harvesting are likewise the same. Preparation of the autograft is different as the adventitia is left intact but the muscle below the annulus is trimmed to 2 to 3 mm. The aorta is transected at the sinotubular junction with dissection of tissue behind the aorta to enter the transverse sinus. The aortic valve is excised and the annulus measured. Each coronary ostium is excised with a portion of aorta creating "buttons" that will be reimplanted later into the autograft. Mobilization of each ostium is important to allow for proper reimplantation. The noncoronary sinus is left intact and not excised. This will be used as reinforcement to the root in patients who have a dilated annulus, dilated sinuses, or sinotubular junction. If the patient has aortic insufficiency or has an annular size larger than the normal aortic size for the patient's body surface area, an annular reduction and fixation is accomplished using a double row of 2-0 polypropylene placed in a intra-annular plane and brought to the outside of the noncoronary cusp for tying through a pledget. This double row of suture run in purse-string manner can then be tightened over a sizer to achieve the desired annular size for the specific patient (Fig. 49.7). Aortic stenosis patients without annular enlargement have not required the fixation and do not require the use of the native noncoronary sinus. The proximal anastomosis is then performed as described with the subcoronary implantation technique. If annular fixation is used, then the suture line must incorporate the fixating sutures. The autograft is oriented so that the posterior sinus of the pulmonary valve will be the left coronary sinus in the root. This positions the autograft in the most anatomic position. We have used the single running technique almost exclusively. It is paramount to visualize each suture placement to avoid placing a suture into the annulus or the leaflet. The sutures should be placed in a tangential manner from endocardial muscle angled to a higher level exit on the epicardium, in line with the annulus. This telescopes the autograft annulus into the remaining native aortic annulus. The suture line is reinforced with a thin strip of pericardium, which acts as a hemostatic gasket. Inspection of the leaflets following the anastomosis insures no injury to the leaflets. Should a suture be placed in a leaflet, it must be removed and replaced as eventual perforation of the leaflet will occur. An incision is made in the left sinus and enlarged with an aortic punch. The left coronary button is then sewn to the opening, without rotation, using 5-0

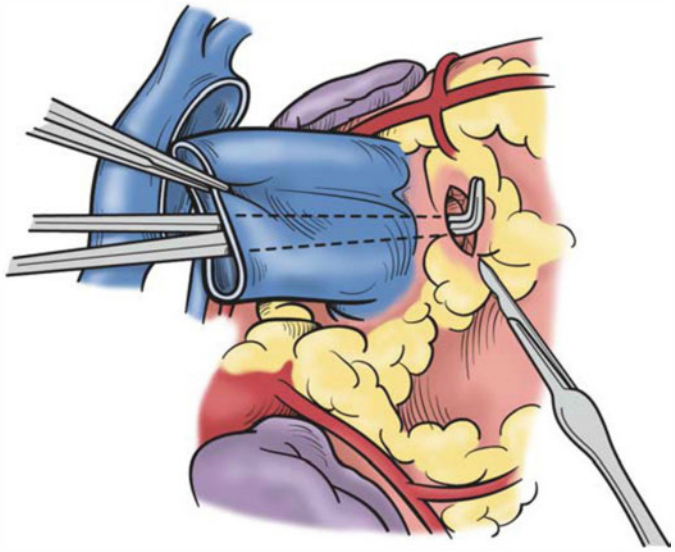


Fig. 49.5. Identification of the anterior right ventriculotomy is facilitated by placing a right-angled clamp through the pulmonary valve and indenting the myocardium 3 to 4 mm below the pulmonary valve annulus.

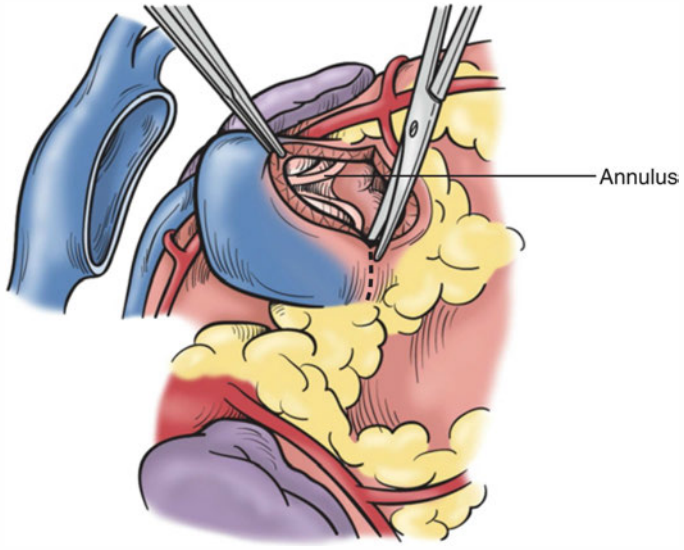


Fig. 49.6. Completion of the posterior enucleation of the pulmonary autograft from the outflow tract of the right ventricle. Note the usual location of the first large septal perforating coronary artery. It arises adjacent to the first diagonal coronary artery of the left anterior descending coronary artery and traverses the septal musculature toward the conal papillary muscle of the tricuspid valve.

polypropylene running suture (Fig. 49.8). Excellent hemostatic suturing is helpful as this is difficult to see after distention of the aorta. Inspection is made from the interior also. The autograft is then trimmed so only 2 to 3 mm of artery remains above the commissures. Marking of the commissures on the outside of the autograft will facilitate the placement of the right coronary after the distal closure of the autograft to the aorta. The distal anastomosis is then performed between the autograft and ascending aorta. It is this suture line that the remaining noncoronary sinus of the native aorta can be attached in patients with insufficiency. The reinforcement covers the most vulnerable portion of the root for dilation. The left sinus is protected by the left atrium and a portion of the right by the pulmonary homograft. In addition, the left and right sinuses will have the coronary ostia implanted to help reinforce them. It is our feeling that aortic stenosis patients are not at the same risk of late dilation. If annular downsizing has been used, it is important to make the sinotubular (distal) anastomosis the same size as the annulus. Example: 23 mm annulus = 23 mm sinotubular junction. This can be accomplished by ascending aortoplasty (by posterior wedge resection) or interposition of Dacron graft. This insures against late dilation at the sinotubular junction with ensuing aortic insufficiency. With completion of the distal anastomosis, the aortic clamp, with low flow in cardiopulmonary bypass, can be temporarily released to allow for distention of the root. The mobilized right button can then be measured to a high position on the right sinus and a hole made where the implantation will take place. The previous commissural markings on the exterior will help to not injure the leaflets, which will be coapted with the aortic pressure. The hole is enlarged with an aortic punch. Rotation of the right coronary button is more of a risk than with the left and must be avoided. Placement in a higher position in the sinus also avoids kinking of the artery with distention of the right ventricle.

RIGHT VENTRICULAR OUTFLOW RECONSTRUCTION

Replacement of the "switched" autograft has been with a pulmonary homograft in our experience. There are reports of heterograft and aortic homografts being used, but most surgeons performing the Ross procedure choose a pulmonary homograft. There is evidence of better performance than with the other choices. More recently, decellularized pulmonary homografts have been used with good success. The repopulation with native endothelium into the decellurized matrix remains a question. We have had success with both grafts. We universally choose a homograft that is 2 to 3 sizes larger than the size of a normal aortic valve size for the patient. Example: 23 mm aortic valve = 27 to 28 mm pulmonary homograft. We believe that this is responsible for our very low incidence of homograft explant secondary to late pulmonary stenosis. Some surgeons will attempt the reconstruction with the cross-clamp off. We have found that with the safety of modern cardioplegic techniques that a dry motionless field actually speeds the operation. The thawed homograft is trimmed to mimic the excised autograft. Generally, the main pulmonary artery is transected proximal to the bifurcation and the excess RVOT muscle is trimmed to 4 mm below the annulus. The distal anastomosis is performed first using 4-0 polypropylene. Care is taken not to narrow the anastomosis. A finger can then be passed into both the right and left pulmonary arteries through

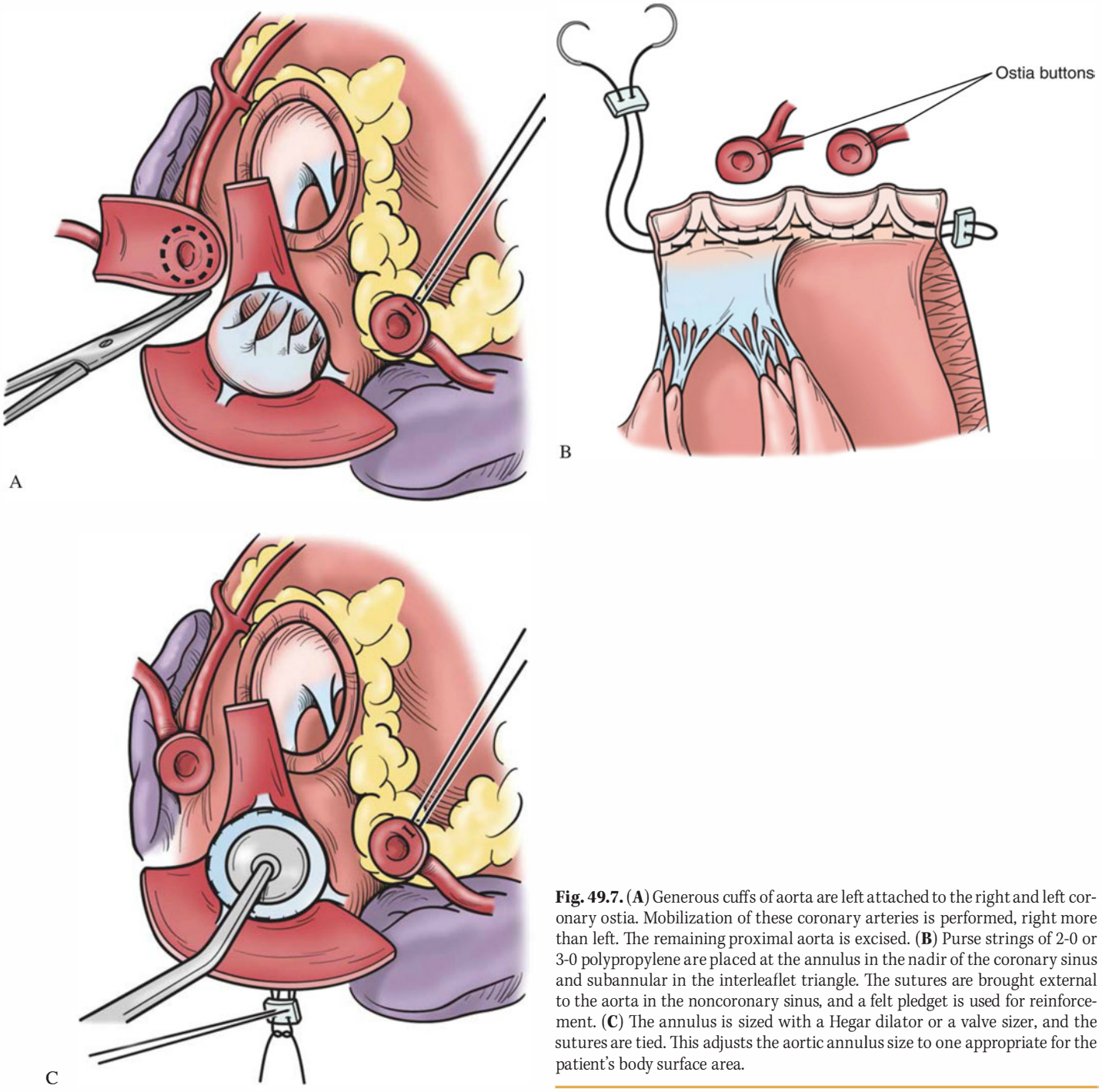


Fig. 49.7. (**A**) Generous cuffs of aorta are left attached to the right and left coronary ostia. Mobilization of these coronary arteries is performed, right more than left. The remaining proximal aorta is excised. (**B**) Purse strings of 2-0 or 3-0 polypropylene are placed at the annulus in the nadir of the coronary sinus and subannular in the interleaflet triangle. The sutures are brought external to the aorta in the noncoronary sinus, and a felt pledget is used for reinforcement. (**C**) The annulus is sized with a Hegar dilator or a valve sizer, and the sutures are tied. This adjusts the aortic annulus size to one appropriate for the patient's body surface area.

the valve. Lastly, the proximal anastomosis is performed with 4-0 polypropylene. Hemostasis in the bed of the enucleated autograft is paramount prior to the proximal closure. Electrocautery and possibly sutures can insure this. One must insure not to rotate the valve and suturing is best started at the patient's left side of the RVOT. There are trabeculations in this region that require accurate closure. The suture line then proceeds on the posterior wall with tangential sutures so as to not injure the first septal perforator. The anterior running closure then follows (Fig. 49.9). Care must be taken on the left side to not take too much tissue in the suture that could damage the left anterior descending artery. Following rewarming, which is done while the homograft is placed, the cross-clamp is released and de-airing accomplished. Usual weaning from cardiopulmonary bypass is then accomplished. Transesophageal echocardiography is then performed to evaluate the neoaortic valve. It is typical to see a small central insufficiency jet. Finding significant insufficiency warrants investigation into source, whether prolapse of leaflet or rotation of graft.

POSTOPERATIVE MANAGEMENT

Postoperative management of the patient actually begins in the operating room. Management of the aortic blood pressure is paramount. We keep the pressure <110 systolic and lower if tolerated by the patient. This continues throughout the stay in the ICU with IV drips. When the patient tolerates oral intake, oral antihypertensives are added to the regimen. As these are generally younger patients, the lower blood pressure is nicely tolerated. We try to manage the

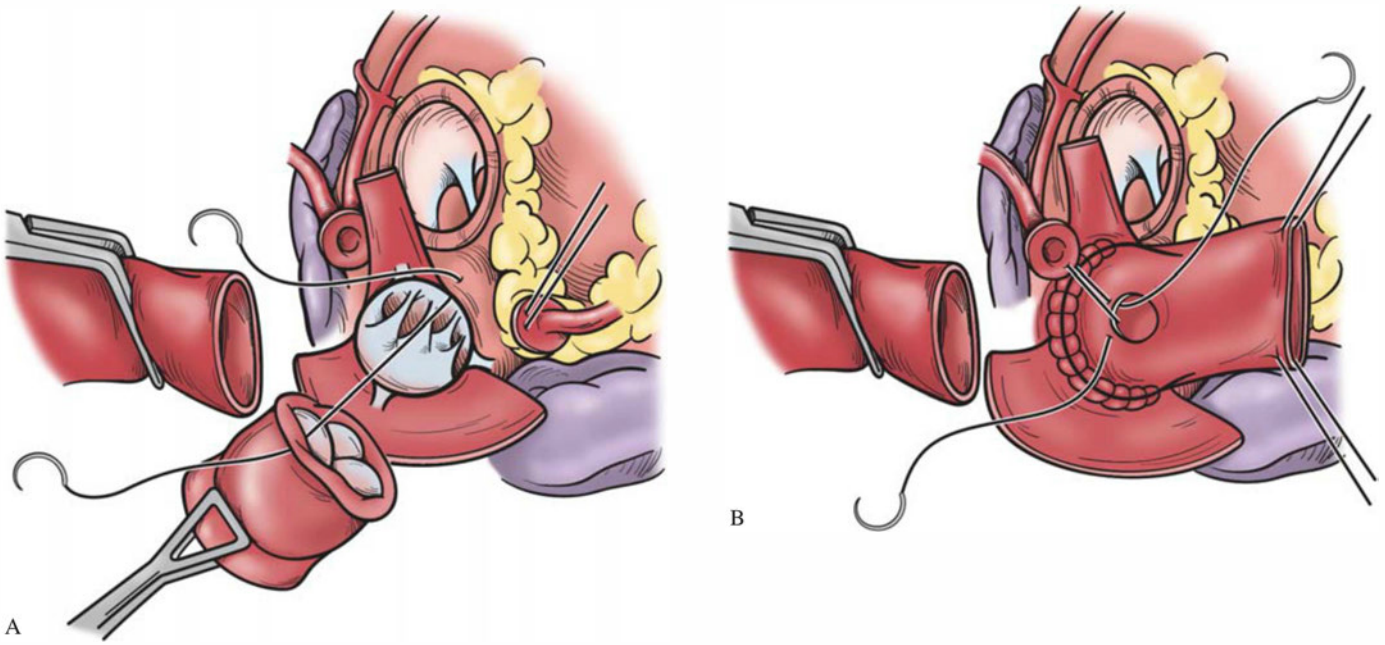


Fig. 49.8. (**A**) The pulmonary autograft is in an anatomic position with the posterior sinus of the autograft becoming the new left coronary sinus. (The stay suture in this sinus is not shown for clarity.) The remaining sutures for orientation are placed to position the new right coronary sinus and to trifurcate the aortic annulus. (**B**) The left coronary artery is implanted with a continuous suture of polypropylene.

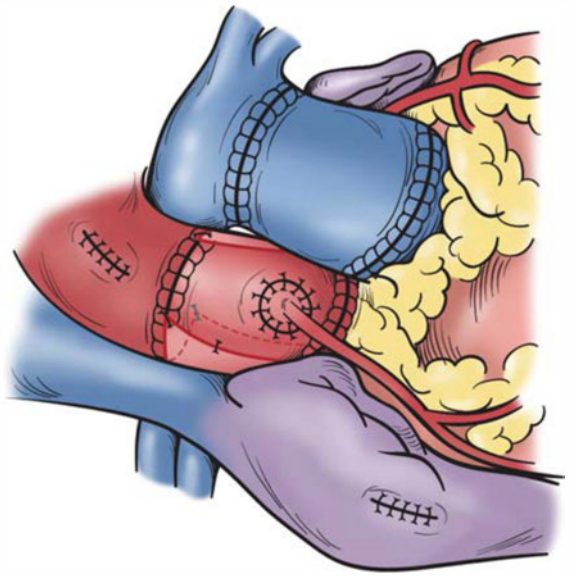


Fig. 49.9. Completion of the pulmonary autograft root implantation with selection of the site of implantation of the right coronary artery with the autograft distended. The pulmonary homograft reconstruction of the outflow tract of the right ventricle is done with two continuous suture lines.

blood pressure for a minimum of 3 months. Should the patient have hypertension, a lifetime management is very important. We strongly believe that this has led to a lowered level of late dilation and valve incompetence. Another postoperative management is the addition of anti-inflammatory medication, usually in the form of nonsteroidals. The pulmonary homograft can cause an intense postpericardiotomy syndrome, and the addition of these drugs may help prevent some immune responses, which could lead to homograft constriction. This phenomenon is also helped by the oversizing of the homograft as was described in the technique section of homograft implantation. The nonsteroidals can be discontinued in 3 to 4 weeks.

Since 1967, the Ross procedure has undergone multiple variations in the technique of implantation and indications for use. We have also learned some contraindications. Each modification has been added or deleted in an effort to improve the long-term outcomes, or to facilitate the ease of performing the operation. Following Mr. Ross' publication of his 20-year results, there was a sharp increase in the number of cases performed. Not all were performed in the same technique with many variations of autograft implantation used. Some of the variations were useful while others were not so well-performed. However, the results of the multiple changes were all reported as Ross procedures. This has allowed for variations in outcomes as one would expect. Many of the changes, when followed over the years, have added to better results. These many variations early in the history have lead to poorer acceptance of the procedure. Those who have persisted with sound autograft techniques with long-term follow-up have had very good results, which are duplicated by the other programs following similar techniques. Through accurate clinical follow-up, progress has definitely been made in delineation of appropriate and inappropriate candidates. Technical features such as reduction and fixation of the aortic annulus with concomitant fixation of the sinotubular junction have

been extremely beneficial. Postoperative management of blood pressure and use of anti-inflammatory drugs have boosted long-term results in the series that have incorporated this protocol. The use of the Ross procedure in endocarditis has produced outstanding results in cure rate and re-infection rates and provides the patient with a valve that has significantly better longevity than other valves. The viability of the autograft, with cellular levels of preoperative antibiotics, insures increased cure rates when placed in an adequately debrided LVOT. Placement as a root completely exteriorizes to the pericardium space all infection leaving none exposed to the endothelium and blood stream. The flexibility of the graft allows for modeling of the valve to the irregularities of the debrided annulus, thus providing a more secure hemostatic anastomosis. Flexibility rather than rigidity, as with a stented bioprosthesis or mechanical valve, markedly decreases the incidence of late paravalvular leak or pseudoaneurysm. When compared in a prospective randomized study, the autograft has outperformed the aortic homograft in both earlier valve failure and mortality rates. With improved cardioplegic techniques, the use of the Ross procedure for endocarditis, while requiring possibly longer ischemic times, has not lead to any unfavorable outcomes.

Our series of Ross patients began in 1989. Since then, 386 patients aged 1 day to 71 years, mean age 39.6, follow-up range 0 to 19 years have undergone a root replacement with the autograft. The operative mortality was 4.1%. Kaplan–Meier survival at 15 years is 86%. Freedom from explant of the autograft is 71% and freedom from pulmonary homograft explant is 93%. In our cohort of patients with a mean age of 55 years, freedom of mortality was 85%. Programs that have performed the Ross procedure for more than 15 years all show very similar survivals and reoperation rates, both in the United States and Europe (Table 49.1). Comparison to other valve series, with similar or longer length of follow-up, in mechanical valves, bioprosthetic valves, and homografts are shown (Tables 49.2 to 49.6). Each of these series have a mean age of 50 years or greater, similar to the Ross series. Although cross-comparisons may be difficult, a definite difference in survival rates is seen. The final table groups the valve series in survival percentages for comparison (Table 49.7). Besides the follow-up data presented, there remains the subjective advantages to the Ross procedure. There is a profound lack of thromboembolism throughout the life of the patient as well as the freedom of frequent and inconvenient management of anticoagulation in a patient with a mechanical prosthesis. The option of child-bearing in women of this age group is extremely important. We have many patients who have born children post-Ross procedure. There is no noise related to the autograft, and the physiologic function and ventricular remodeling have been well documented.

The Ross procedure remains an excellent procedure for aortic valve replacement. Proper selection of candidates is mandatory and performance of an exact operation allows for excellent outcomes. More than other procedures, attention to postoperative management needs to be in the surgeon's control. It is the valve of choice for the pediatric age group and a good choice for patients of child-bearing age, athletes, patients with endocarditis, and patients who must avoid anticoagulation.

Table 49.1 Ross Series

Length of Follow Up		Operative mortality	Survival	Freedom from replace autograft
Elkins	16 y	4.6%	89%	82%
Yacoub	10 y		96%	96%
Stelzer	20 y (424 patients)	2%		Late 15 patients
Seivers	10 y	0.4%	95%	95%

Table 49.2 Homograft Series

Length of Follow Up		Age	Survival
Yacoub	10 y	43	88%
O'Brien	18 y		63%

Table 49.3 Mechanical Valve Series

Length of Follow Up		Age	OR mortality	Survival	Freedom from replace autograft	
Emery (ST.J)	25 y	64	4%	25%	20 y, 30%	
Palatianos (ON-X)	7 y	60	2.2%	82%		
Williams (ON-X)	5 y	33		74%	40%, poorly or not A/C	
Garcia-Rinaldi (ST.J), 122 patients	7 y	64		17 Deaths	Plavix 300 ASA 325	Plavix 75 ASA 325/81

Table 49.4 Pericardial Valve Series

Length of Follow Up		Age	Operative mortality	Survival	Freedom from replace autograft
Biglioli	15 y	64	4%	56%	
Aupart	18 y	>70	2.8%	22%	Elderly cohort
Frater	14 y	65	n/a	40%	
Banbury	15 y	65		26%	10 y, 52%

Table 49.5 Porcine Stented Series

Length of Follow Up		Age	Operative mortality	Survival
Mosaic valve				
Botzenhardt	8 y	73 y old	3%	60%
Fradet	7 y	69 y old		84%
Jamieson	5 y	70 y old		73 %
Biocor valve				
Eichinger	10 y	72 y old		45%

Table 49.6 Porcine Stentless

Length of Follow Up			Operative mortality		Survival
Root					
Kon	Freestyle	8 y	3.9%		60%
Multicenter group (8)	Freestyle	12 y		≤60, ≥60	65%, 33%
Mixed					
Bach	Freestyle	10 y		Subcor root	44%, 47%
Subcor					
Desai	Toronto SPV	10 y			68%

Table 49.7 Fifteen-Year Comparisons Survival

Ross	Homograft	Mechanical	Pericardial	Porcine stented	Porcine stentless
80–85%	60–65%	45–60%	40–50%	50–60%	40–60%

SUGGESTED READINGS

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EDITOR'S COMMENTS

This is a chapter by Chip Oswalt, who is one of the experts at the Ross procedure. He has published extensively on endocarditis. We too use the Ross procedure though our uses are more limited than they have been previously. As Dr. Oswalt has mentioned, patients with bicuspid valves and with aortic dilation often go on to develop ascending aneurysms late. We have seen this. In the vast majority of these situations, we have been able to preserve the autograft but this is a significant complication of the Ross procedure. We now reserve the Ross procedure for children or very young adults. Even in this situation, it has potential to be a lifetime valve with the ability to grow. This is unique and therefore it is worth the extra surgery required. In addition, even though the pulmonary valve is replaced with a homograft, the use of percutaneous pulmonary valves can deal with any late failures of the homografts.

I think the Ross procedure still has a place in cardiac surgery. There are other options that are available for older patients with less surgery required. However, there is a subset of patients that Dr. Oswalt has mentioned that include children, young adults, and perhaps certain cases of endocarditis.

ILK

50 Transcatheter Aortic Valve Replacement

Isaac George, G. Russell Reiss, and Mathew R. Williams

INTRODUCTION

Aortic stenosis (AS) is the most common form of acquired valvular heart disease in Westernized nations and its prevalence increases with age. The natural history of untreated AS portends a grave prognosis with 1- and 5-year survival rates of 60% and 32%, respectively. Currently, the only effective treatment for patients symptomatic from AS is surgical aortic valve replacement (AVR). In the ideal candidate, the estimated mortality from surgical AVR performed by a skilled operator should be <3%. Historically, the rate of mortality and postoperative complications for surgical AVR have climbed rapidly after the eighth decade of life and in those with multiple pre-existing conditions and comorbidities, such as depressed left ventricular (LV) function, associated coronary artery disease (CAD), prior cardiac surgery, renal insufficiency, peripheral vascular disease, and chronic lung disease. Because of this increased perioperative risk in the elderly and often frail patient, as many as 30% of patients diagnosed with critical AS never get referred to a surgeon.

First introduced nearly a decade ago, transcatheter AVR (TAVR) has been established as a safe and effective alternative to AVR in patients previously considered "high risk" or inoperable due to prohibitive predicted preoperative mortality. There are currently two transcatheter heart valve (THV) replacement devices approved for use in the United States, each using a unique platform for delivery and having individual nuances regarding patient workup, clinical management, and device deployment. The Edwards Lifesciences (Irvine, CA) SAPIEN™ THV system (transfemoral: Retroflex 3 [RF3] and newer generation XT; transapical: Ascendra) are balloon expandable devices consisting of bovine pericardial valve leaflets attached to stent platforms placed in the subcoronary position. The transfemoral SAPIEN RF3 is currently the only THV that is FDA approved for commercial use in the United States for inoperable patients. In contrast, the Medtronic (Minneapolis, MN) CoreValve™ Revalving system is a self-expanding, nitinol cage with porcine leaflets, which seats in the LV outflow tract and spans through the aortic valve to above the coronary ostia. Both THV systems will be discussed in this chapter. In addition, several next generation devices are in various stages of development and worthy of brief comment.

PATHOPHYSIOLOGY

AS progresses over time and is characterized by thickening and calcification of the valve leaflets. Increasing obstruction to flow occurs at valve areas <1.5 cm^2, at which point hemodynamic changes are manifest and concentric LV hypertrophy occurs in order to normalize wall stress. As LV pressure and wall stress increase beyond the limit of hypertrophy, the pathologic changes in AS are seen, including diastolic dysfunction, impaired coronary reserve with resulting ischemia, and systolic ventricular dysfunction. Decompensation may rapidly ensue in patients with myocardial ischemia/infarction, loss of atrial systole to aid filling, or heart failure symptoms. In addition, patients with severe LV hypertrophy are at risk for sudden death. Indeed, prognosis from the time of symptom onset is poor with medical therapy, with time to death estimated at 2 years for those in heart failure, 3 years for those with syncope, and 5 years for those with angina. Clinical symptoms upon presentation include classic signs: angina pectoris, syncope, and heart failure symptoms, as well as other less common symptoms of gastrointestinal bleeding (from angiodysplasia), atrial fibrillation, and pulmonary hypertension.

A detailed description of severe AS can be found in Chapter 50—Aortic Valve Disease.

RISK STRATIFICATION, PATIENT SELECTION, AND SCREENING

Despite the mismatch between incidence of critical aortic valve disease and surgical referral for AVR, TAVR is currently only available to the highest risk patient population with symptomatic, calcific severe AS (see Table 50.1 for accepted echocardiographic criterion for severe AS). who are currently without a surgical option or whose risk of mortality exceeds 15%. Hence, the typical TAVR patient is in the top 10% for operative mortality with a Society of Thoracic Surgery (STS) PROM score typically >10 and having a corresponding Logistic European System for Cardiac Operative Risk Evaluation score (EuroSCORE) of >30. Data from multiple well-designed prospective randomized trials have demonstrated that TAVR, although carrying a higher incidence of stroke, is a viable alternative to surgical AVR in patients with this extreme predicted operative risk. However, due to unproven long-term durability and efficacy, TAVR has not been offered in lower risk patients and further study is needed to determine if TAVR will be appropriate in these populations.

Risk assessment for TAVR is not solely for the purpose of evaluating periprocedural mortality, however. A more important function of risk assessment in this population is to better understand the natural history of patients treated with TAVR. It is well documented that surgical models of risk fail to capture the true risk of patients with severe comorbidity while overestimating risk in elderly patients, and offer little validation in the current TAVR population. Furthermore, models of surgical risk provide no information regarding important clinical variables such as cost-effectiveness, long-term morbidity, and noncardiac mortality, which are all vital in determining which patients should be treated with TAVR. For that reason, risk assessment must take into account both traditional methods of risk scoring, such as the STS and EuroSCORE systems, which

Table 50.1 Echocardiographic Criteria for Severe Aortic Stenosis

- AVA <0.8 cm^2 *or* indexed EOA <0.5 cm^2/m^2

And

- AVG_{mean} >40 mmHg *or* jet velocity >4.0 m/s

AVA, aortic valve area; AVG, aortic valve gradient.

Table 50.3 Relative Contraindications to Transcatheter Aortic Valve Replacement

Congenital unicuspid or bicuspid, or noncalcific
Mixed aortic valve disease
Hypertrophic cardiomyopathy with or without obstruction
Ejection fraction $<20\%$
Severe pulmonary hypertension or severe right ventricular dysfunction
Echocardiographic evidence of intracardiac mass, thrombus, or vegetation
Annulus sizing <18 or >29 mm
Significant aortic disease
Severe mitral regurgitation
Life expectancy <12 mo
Moderate-to-severe dementia
Extreme frailty
Futility of medical care due to comorbidities or severity of cardiac disease

are based on historical data of outcomes from coronary and valvular surgery, as well as newer, nontraditional methods, such as frailty scoring and adjusted incremental risk. A frailty index used at our institution has proven to be invaluable to our assessment and its components are described in Table 50.2. In addition, taking into account factors not integrated in the STS or EuroSCORE, such as pulmonary hypertension, cirrhosis, and dementia, can weigh heavily in final patient selection. In general, any patient with a life expectancy of less than 12 months due to a noncardiac condition or with such severe medical comorbidity (i.e., extreme frailty, moderate-to-severe dementia, etc.) that will prevent meaningful recovery should not be offered TAVR. A comprehensive list of relative exclusion criteria pooled from existing clinical trials is provided in Table 50.3.

Appropriate patient selection and screening is crucial to maximize the chance of procedural success. Once a patient has been deemed a nonoperative or high-risk surgical candidate, the primary determinants of TAVR eligibility initially focus on aortic annulus sizing and anatomy and route of device delivery. Table 50.3 lists the current annulus sizing criteria for patients eligible for both the SAPIEN and Core Valve THV. Patients with extremely small or large annuli (<18 or >25 mm) may face a higher risk of aortic/ventricular rupture or severe paravalvular leak (PVL), respectively, with current devices, and should be avoided for THV use until suitable devices are available. There are a number of anatomic considerations that may also preclude TAVR success: (1) noncalcified valves may not provide adequate annular support for an expanded valve stent and have not been studied, (2) patients with unicuspid or congenital bicuspid aortic valves may not allow proper THV stent expansion, (3) patients with leaflet thrombus or vegetation are not THV candidates, (4) placement of a THV in the presence of low-lying coronary ostia, bulky calcified leaflets, or an effaced sinus of Valsalva may result in coronary obstruction, and careful consideration for coronary vessel access must be made prior to THV deployment, and (5) patients with severe aortic insufficiency should be evaluated with caution. Inadequate characterization of this complex anatomy may lead to serious procedural morbidity; complications have included valve migration and embolization, annulus rupture, aortic dissection, and severe PVL. Many of these complications can be avoided with careful planning.

Table 50.2 Frailty Index

Must fulfill 3 out of 4 criteria

- Grip strength <18 kg
- 5-m walk <7 s
- Serum albumin <3.5 mg/dl
- Katz ADLs <4 of 6 criteria
- Visual appearance (subjective "eyeball test")

ADL, activity of daily living; AVR, aortic valve replacement.

The delivery of THV may be performed by either a transfemoral or transapical approach. The transfemoral approach consists of retrograde percutaneous delivery through femoral artery access, while a transapical approach requires a mini-left thoracotomy for LV apical exposure, followed by percutaneous antegrade delivery via the LV apex. Advantages to both routes exist but patient considerations usually dictate the approach. In patients with suitable femoral access, a transfemoral approach may be preferable to transapical, as pain and recovery from mechanical ventilation may be reduced. However, in patients with poor vascular access, the transapical approach offers a safe and direct means for device delivery. To date, both access routes have shown equivalent outcomes versus surgical AVR. Vascular size criterion for transfemoral access for both the Edwards SAPIEN and Medtronic CoreValve systems, as well as inclusion/exclusion factors, is described in Table 50.4.

Successful alternative methods of THV delivery have been reported as off-label use and/or outside the United States. These routes, such as direct-aortic (through a hemi-sternotomy or right third intercostal space parasternal exposure), subclavian or axillary artery cutdown, or via abdominal aorta conduit, present separate technical challenges in terms of surgical exposure and device deployment, and should only be performed when necessary and by highly qualified operators. To date, placing a THV into a bioprosthetic valve has been performed worldwide with some success in isolated cases, but full evaluation of these techniques have not been documented and are beyond the scope of this chapter.

PREOPERATIVE EVALUATION

Preoperative testing for patients being considered for TAVR is comprehensive, as

Table 50.4 Annulus Sizing Criteria

	Annulus size (mm)	Valve size
Edwards SAPIEN RF3	18–21	23
	21–22	23 or 26
	23–25	26
Edwards SAPIEN XT	18–21	23
	21–22	23 or 26
	23–25	26
	26–27	29
Medtronic CoreValve	20–23	26
	23–27	29
	26–29	31

both eligibility for valve placement from an anatomic standpoint must be determined, as well as testing for fitness to tolerate the procedure in this sick cohort of patients.

Evaluation of the aortic valve, aortic root, and peripheral vasculature provides the necessary clinical information to confirm eligibility for TAVR. The echocardiogram is the primary modality to document the severity of AS and size the aortic valve annulus (see Table 50.1 for echocardiographic description of severe AS). Valuable information regarding the calcium severity of the leaflets, valve type (uni-, bi-, or tricuspid), annulus to coronary ostia height, and leaflet length and bulkiness may also be obtained. Note that measurement in the parasternal long axis views and during systole most accurately size the annulus (Figs. 50.4A and B). Preoperative transthoracic echocardiography may be sufficient for most patients, but transesophageal echocardiography (TEE) should be utilized in the event of any discrepancy or concern. Low gradient in the presence of low-ejection fraction is frequently encountered in high-risk and elderly patients; in these cases, dobutamine stress echocardiography is mandatory to evaluate contractile reserve and candidacy for TAVR.

Multidetector computed tomography (MDCT) angiography (CTA) of the chest, abdomen, and pelvis with iliofemoral runoff is performed, detailing the anatomy of the aorta and aortic root, with special attention given to aortic root size, aortic tortuosity and angulation (especially at the arch), calcification, areas of stenosis, and adequacy of peripheral vessels for the transfemoral approach. Iliac angiography or computed tomography (CT) reconstruction of peripheral vessels is mandatory to confirm vessel adequacy for the transfemoral approach (see Table 50.5 for minimum vessel size for TAVR). Aortography may be used selectively as an adjunct to echocardiography and CT scanning. Iliac intravenous ultrasound can be substituted for CTA scan to avoid intravenous dye administration in patients at risk of renal failure.

A standard right and left heart cardiac catheterization should be performed to evaluate the presence of pulmonary hypertension and any concomitant CAD. Depending upon the severity, certain coronary lesions may need to be addressed prior to consideration for TAVR. The use of systemic anticoagulation or platelet inhibitors should not affect TAVR candidacy, although TAVR within 1 month of bare-metal stent placement and within 6 months of drug-eluting stent placement is not recommended. Finally, other comorbidities, such as advanced pulmonary disease, chronic renal insufficiency, and ongoing gastrointestinal disease should be addressed as part of a standard preoperative workup.

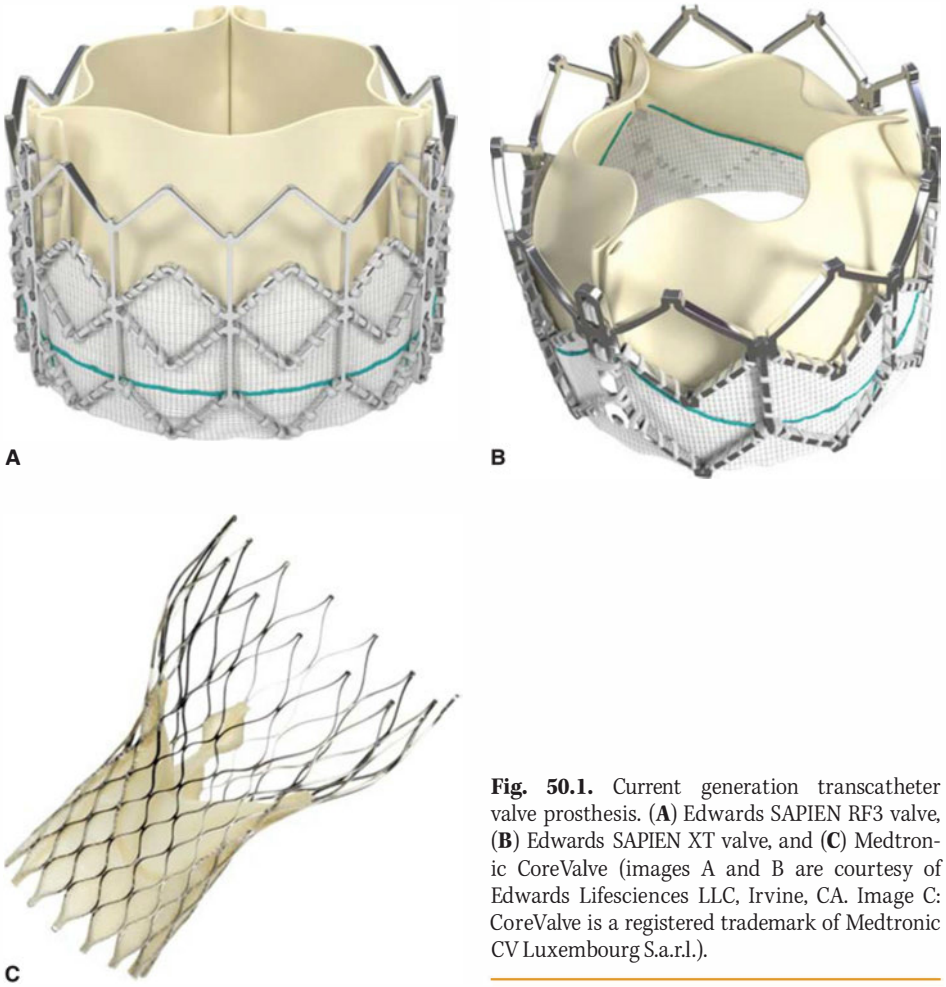


Fig. 50.1. Current generation transcatheter valve prosthesis. (**A**) Edwards SAPIEN RF3 valve, (**B**) Edwards SAPIEN XT valve, and (**C**) Medtronic CoreValve (images A and B are courtesy of Edwards Lifesciences LLC, Irvine, CA. Image C: CoreValve is a registered trademark of Medtronic CV Luxembourg S.a.r.l.).

CURRENT TRANSCATHETER HEART VALVE TECHNOLOGY

Edwards SAPIEN and SAPIEN XT

The Edwards SAPIEN THV is a balloon-expandable bioprosthesis that is available in two sizes, 23 and 26 mm (Fig. 50.1A). The valve leaflets are made from bovine pericardium that has been pretreated to

Table 50.5 Vascular Access Sizing

		Valve size	Device sheath (F)	Sheath OD (mm)	Minimum vessel diameter (mm)
Edwards SAPIEN	RF3	23	25	8.38	7
		26	28	9.14	8
	TA-RF3	23/26	26	8.6	
	XT	23	21	7.20	6
		26	22	7.50	6.5
Medtronic CoreValve		26	18	6	>6
		29	18	6	>6
		31	18	6	>6

reduce calcification and functional deterioration. The leaflets are suspended on a stainless steel cage, which is 14.3 or 16.1 mm high for the 23 and 26 mm (Fig. 50.1C), respectively. There is a fabric cuff in lieu of a sewing ring residing on the ventricular side that covers approximately one half of the valve. This fabric is designed to limit overexpansion and reduce PVL. The SAPIEN XT is a newer generation THV mounted on a cobalt–chromium alloy cage that allows for a lower profile and smaller device sheath size at the equivalent radial strength (Fig. 50.1B). The effective orifice area of both the SAPIEN and CoreValve valves are larger and provide lower hemodynamic profiles than their corresponding surgical valves.

For the transfemoral approach, SAPIEN and SAPIEN XT THVs are delivered across the aortic valve through the Retroflex 3 Guiding Catheter or Novaflex Delivery System, respectively. For transapical access, the Ascendra I system is the only delivery system available for SAPIEN THV. All SAPIEN THV are designed to seat at the annulus and to provide clearance of the coronary ostia. A unique feature of the Retroflex 3 and Novaflex catheters is the ability to manually "steer" the catheter tip, facilitating the safe passage of the catheter across a highly angulated aortic arch.

The SAPIEN valves are stored prepackaged in an expanded state and require crimping prior to delivery. The valve is crimped onto a 30-mm long delivery balloon using a specialized crimping tool. The diameter of the delivery balloon is either 23 or 26 mm depending on which bioprosthetic size valve is being deployed. The crimping tool also has an incorporated measuring ring for exact calibration of balloon expansion size and saline-contrast volume.

Medtronic CoreValve ReValving System

The Medtronic CoreValve THV is a self-expanding bioprosthetic valve available in three sizes: 26, 29, and 31 mm (Fig. 50.1C). Three porcine pericardial leaflets are attached at the level of smallest diameter on a 53 to 55 mm tall nitinol stent frame. A pericardial skirt at the inflow is aimed at securing the valve and minimizing PVL, similar to the Edwards SAPIEN valve. The valve is seated at the annulus at its lowest level, while the stent frames cross over the coronary ostia at the upper stent segment. Differential radial strength (high at lower inlet portion, low with limit at the middle portion at the level of coronaries, and flared at the upper portion) is designed to reduce the incidence of coronary obstruction; however, a possible downside to this feature is potential difficulty in future coronary interventions. All CoreValve THVs are delivered via transfemoral access at this time.

The CoreValve requires manual loading onto the delivery system by means of a disposable loading apparatus. Given the overall long length of the stent frame, manipulation of the device catheter across an angulated aortic arch may pose technical challenges.

SURGICAL TECHNIQUE

Heart Valve Team Approach

The members of the multidisciplinary group that is assembled for a successful TAVR include cardiac surgeons, interventional and imaging cardiologists, anesthesia, perfusion, and ancillary staff. Due to the complex nature of the workup, multiple imaging modalities, and the varied skill sets needed at the procedure table, a highly functional heart team is essential for success. This model has been established in all current U.S. TAVR centers, as outlined in the latest ACCF/AATS/SCAI/STS Consensus Document on Transcatheter Aortic Valve Replacement (2012).

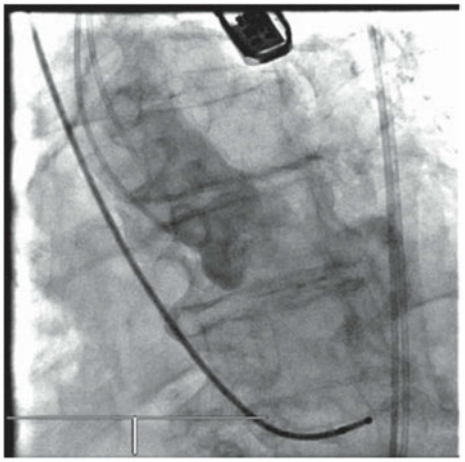
A hybrid room with full catheterization, hemodynamic, and cardiopulmonary bypass surgical capabilities has been utilized for TAVR to allow centralization of the team, manage various access routes in the same operating room, and deal with complications rapidly. Nursing, angiography support staff, and surgical staff may require cross-training in other specialties in order to be fully functional in the hybrid room and with all aspects of the procedure.

Anesthesia

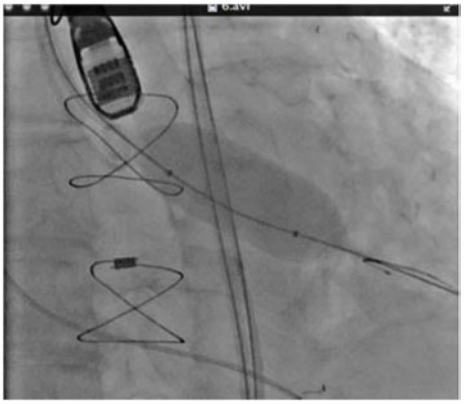
Although percutaneous THV is being performed under conscious sedation in many countries outside the United States, most operators require general anesthesia administered by a cardiovascular trained anesthesiologist. This approach also provides a controlled motionless patient for TEE, which is a critical modality for initial valve imaging, positioning during deployment, and measurement of PVL and complications postdeployment. Double-lung ventilation is not required for either transfemoral or transapical approach. Large-bore venous access for rapid volume/drug infusion arterial-line measurement and transcutaneous defibrillation pads are mandatory for every case; however, pulmonary artery catheterization is at the discretion of the operative team and found to be prudent in sicker patients prone to acute heart failure. Extubation at the end of the case in the operating room is the expectation for all patients, independent of comorbidity or route of access. Finally, careful maintenance of hemodynamics is an important determinant of success in TAVR, as AS patients with severe diastolic dysfunction or hypertrophied ventricles possess little hemodynamic reserve and may experience labile hemodynamics during key points (balloon valvuloplasty (BAV), rapid pacing, etc.). Optimal fluid management and vasopressor administration are generally required for temporary support perioperatively.

General Preparation

To reduce infection risk, prophylactic antibiotics are administered similar to any open surgical aortic valve replacement



A



B



C

Fig. 50.2. Balloon aortic valvuloplasty. (**A**) Optimal deployment view, (**B**) balloon inflated in aortic valve, and (**C**) hemodynamics and pacing strip during balloon inflation.

and should be continued through the perioperative period, usually 24 to 48 hours. Aspirin and clopidogrel are given preprocedure and are continued for at least 6 months. Patients are scrubbed and draped for sternotomy in the event of an emergency. In addition, selection of peripheral cannulas for cardiopulmonary bypass should be made prior to intervention should cardiopulmonary bypass need to be instituted from the groin.

Transfemoral Approach

Vascular Access

Vascular access for TAVR may be achieved for the transfemoral approach by either a fully percutaneous approach or surgical cutdown. Early THV using a surgical cutdown may be preferred until sufficient experience with the procedure is obtained.

If a percutaneous approach is chosen, meticulous technique for venous and arterial access is paramount, given the large sheath sizes, need for anticoagulation, duration of procedure, and potential for unrecognized retroperitoneal bleeding.

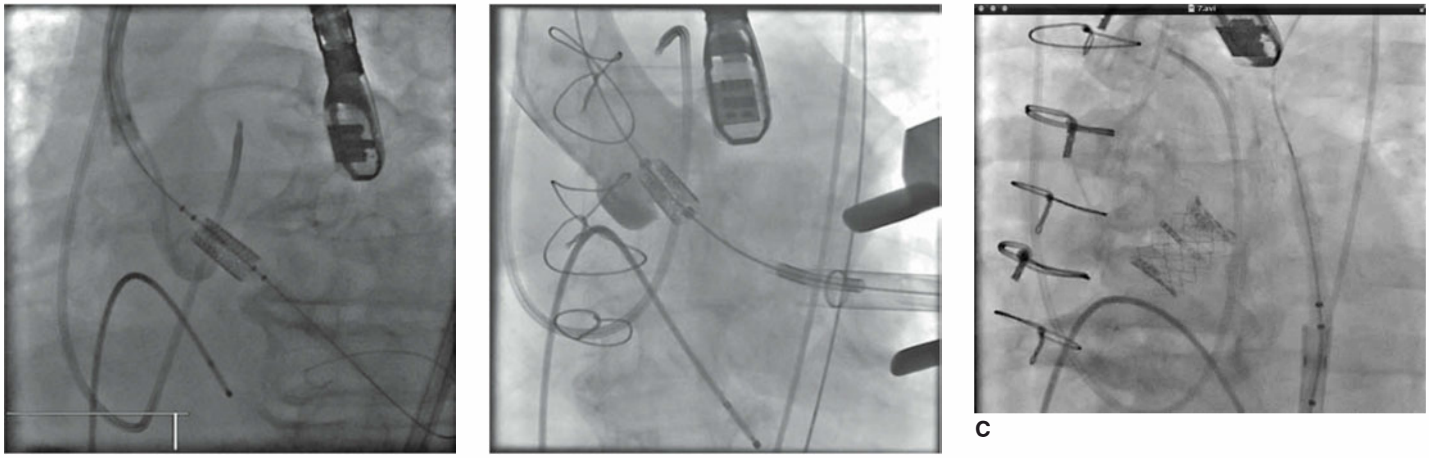


Fig. 50.3. Valve positioning and deployment. **(A)** Edwards SAPIEN valve positioned across aortic valve—transfemoral. **(B)** Edwards SAPIEN valve positioned across aortic valve—transapical. **(C)** Transfemoral Edwards SAPIEN RF3 valve postdeployment.

Percutaneous femoral access is first achieved on the contralateral side (side opposite to that intended for THV deployment sheath): a 5F arterial sheath is placed first, accommodating a 5F pigtail catheter, which is inserted into the aortic root. This is followed by a 6F venous sheath in the femoral vein for the passage of a temporary transvenous pacemaker (TVP). Once positioned in the posterior wall of the right ventricle, the TVP is tested for capture with a low diastolic threshold. Prolonged hypotension should be avoided at this point, as AS remains uncorrected at this stage and systemic recovery from profound hypotension may be compromised.

Shortly after vascular access is obtained, initial aortic angiography via the pigtail catheter is performed confirming the best image projection for deployment. An ideal projection plane is one that lines up all three aortic cusps in a single coplanar image, while making the valve cusps orthogonal to the image intensifier (Fig. 50.2A). The importance of correctly identifying this image cannot be understated; inaccurate deployment views may lead to severe PVL, coronary obstruction, valve migration, or embolization. Frequent views that we have used include left anterior oblique-caudal, straight anteroposterior, and slight left anterior oblique-cranial, but standardization is impossible due to differing patient anatomy. Third-party imaging software such as angiographic reconstruction programs (DynaCT, Siemens, Germany; Paieon, Inc., Rosh Ha'ayin, Israel) may aid in finding this optimal view.

Access on the delivery sheath side (ipsilateral) is accomplished either by surgical cutdown or percutaneous methods, as previously mentioned. If a surgical cutdown is chosen, a 3 cm transverse incision above the inguinal crease is made. Dissection down to the femoral sheath is performed to expose the anterior surface of the common femoral artery and vein, located below the inguinal ligament. Vascular control of the iliac and common femoral artery upon device sheath removal after TAVR completion may be obtained either by direct clamping of the vessel or by contralateral endovascular balloon control. If direct surgical control is chosen, surgical exposure should be made to allow clamping of the artery proximally and distally. Alternatively, a fully percutaneous method of device sheath delivery is now more common than surgical exposure. In experienced hands and with smaller delivery sheaths, a percutaneous success rate >95% can be achieved. In this approach, the 10F Prostar (Abbott Vascular, IL) two-suture percutaneous closure can be used to "pre-close" the THV access artery. To begin, ipsilateral arterial access is carefully gained with a 5F sheath, and proper location is confirmed angiographically. Entry should be located between the inferior epigastric artery and the femoral bifurcation over the femoral head. Prostar stitches are delivered through the arterial wall over a standard 0.035 J-wire, and untied stitches are secured for tying postprocedure. Concurrently, intravenous heparin is administered to maintain an ACT >200 throughout the case.

The ipsilateral access side is serially dilated with increasingly larger French sheaths over a extra stiff wire, typically beginning with 12F to 16F and ending with a dilator 1F to 2F smaller than the device sheath size. Continuous fluoroscopy during advancement of dilators is necessary, and issues of tortuosity, calcification, or wire kinking may need to be addressed at this time. After final dilatation, the large device sheath is carefully advanced and positioned in the descending thoracic aorta.

Balloon Aortic Valvuloplasty

Attention is turned to preparation for crossing the stenotic aortic valve. In the deployment view, the aortic valve is crossed using a straight stiff glide wire through a Amplatz left 1 or 2 catheter (Cook, Bloomington, IL) positioned in the aortic root. Once across the aortic valve, the catheter is advanced into the left ventricular outflow tract and the glide wire is exchanged for a 0.035 Amplatz extra stiff guidewire with a large preshaped loop to allow seating in the LV and minimize ventricular ectopy. Careful control of the wire in the ventricle must be maintained at all times throughout deployment, as aggressive manipulation can easily result in ventricular perforation. A baseline gradient should be measured just prior to this wire exchange to confirm the severity of AS and allow for hemodynamic comparison post-THV deployment. Next, a BAV is chosen based on annulus size (2 to 4 mm less than annulus size). At this point, prior to BAV, the THV should be crimped and ready on the back table in the event of severe aortic regurgitation (AR) or hemodynamic instability. The BAV is performed expeditiously under temporary rapid ventricular pacing when the mean blood pressure is <40 mmHg (Figs. 50.2B and C). There should be minimal distraction in the room at this time, and clear and deliberate commands to the person operating the TVP must be given. Rapid pacing is continued

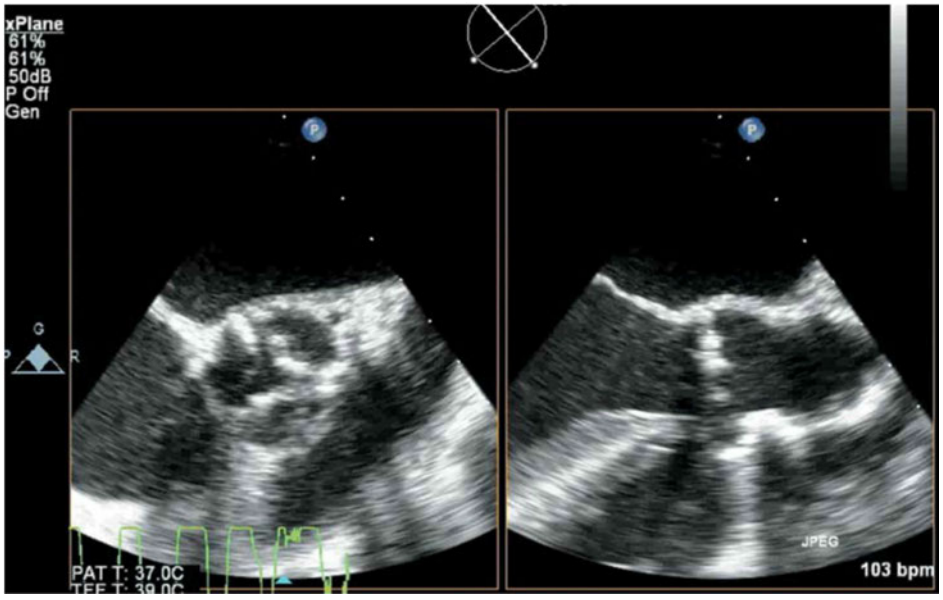
until full inflation. The balloon is then rapidly deflated, the pacer is turned off, and the balloon is removed. Improved excursion of the valve leaflets with increased regurgitation should be apparent by TEE with a suitable BAV (Fig. 50.4B).

Valve Deployment

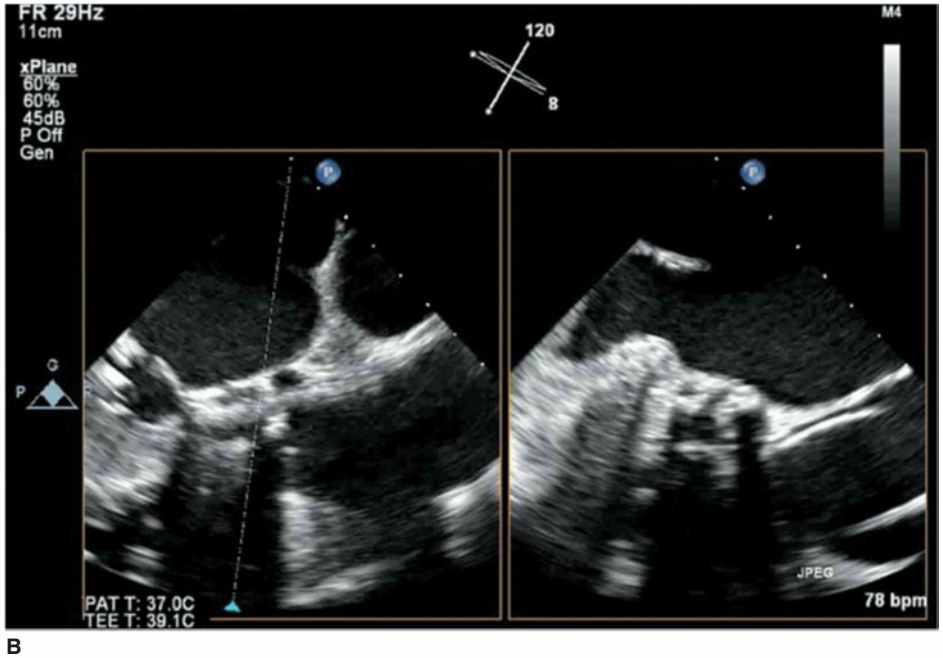
Next, the valve itself is checked for orientation on the valve delivery system prior to insertion into the sheath. The combination THV delivery system is inserted through the large sheath and slowly advanced retrograde through the aorta. At the level of the aortic notch, the delivery system is carefully guided across the aortic arch (the retroflex steering system is activated if an Edwards SAPIEN THV is used, creating a curved angle in the catheter). At the level of the stenotic aortic valve, slight backward tension is placed on the wire while advancing the valve across the aortic valve. Once across the aortic valve, the flexible portion of the delivery catheter is moved back on the wire away from the valve.

The position of the valve is checked both by TEE as well as fluoroscopy. For the Edwards SAPIEN valve, an ideal position seats the mid-portion of the stent directly across the presumed aortic annulus in approximately 50% aortic, 50% ventricular proportions (Figs. 50.3A, B and 50.4C). Once position is confirmed, rapid pacing is again instituted; the position is confirmed by TEE while pacing, capture is confirmed by visual and hemodynamic standstill, and the valve balloon is smoothly inflated, deploying the valve. The pacer is then turned off, and the rhythm and blood pressure are allowed to stabilize. For the Medtronic CoreValve, the valve is seated at the annulus and extends into the ascending aorta. Deployment occurs without ventricular pacing or balloon expansion, and some tolerance for repositioning of the valve exists, unlike the SAPIEN valve.

The device balloon is then rapidly deflated (Edwards valves only), and the balloon is drawn back into the ascending aorta. The stiff wire is left in place while immediate assessment of valve integrity and function is confirmed by TEE (Fig. 50.4D); immediate complications such as annular disruption, ventricular perforation, and valve migration may be readily apparent if present. At this point, hypotension is often present but tends to be transient with proper valve deployment. Communication with anesthesia should be continuous here as to not overcompensate with medication, as severe hypertension has been associated with stroke, periaortic hematomas, and root rupture. The TEE is the most useful tool for immediate assessment of valve function and the presence of AR and PVL. In the presence of greater than mild PVL, we frequently add an additional 0.5 to 1.0 cm^3 of saline to the deployment balloon and a repeat inflation is performed under rapid pacing. This can generally improve the PVL by one grade, although the long-term effect on valve durability has yet to be determined.



A



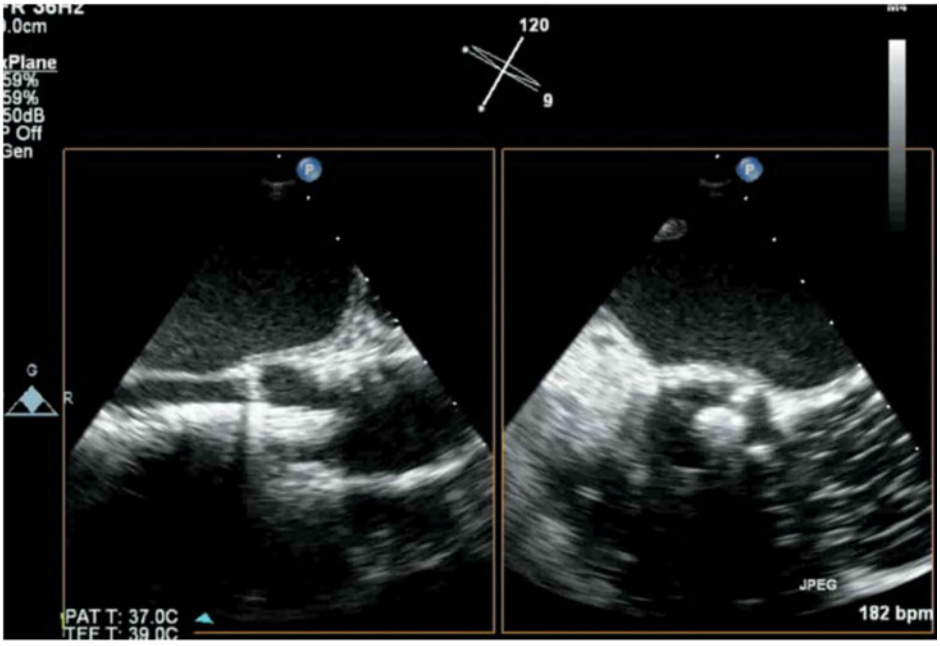
B

Fig. 50.4. Representative transesophageal images. (**A**) Optimal deployment view, (**B**) balloon aortic valvuloplasty, (**C**) device sheath located across aortic valve, and (**D**) valve deployed.

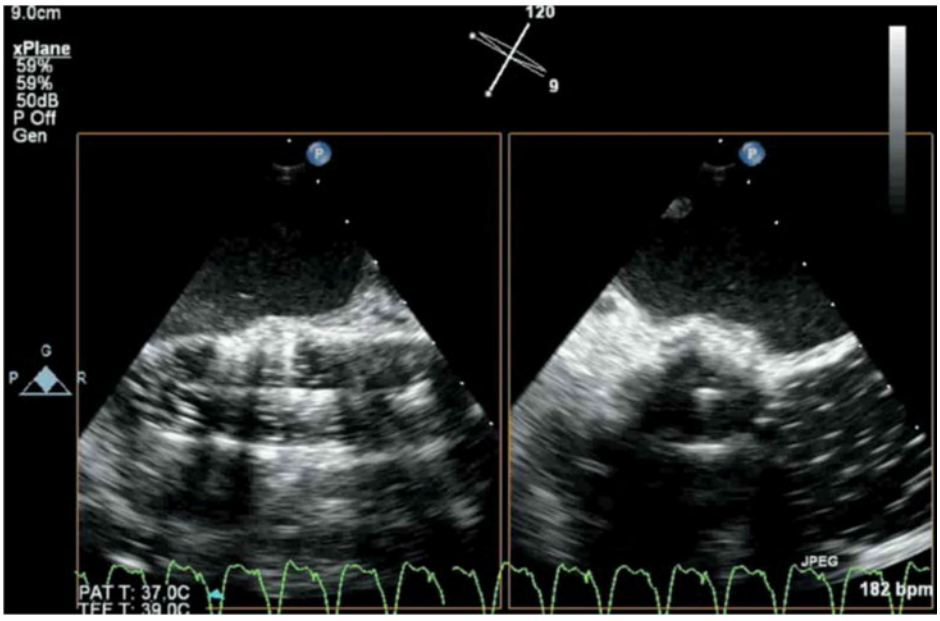
Vascular Access Site Closure

Once the valve position and function are confirmed to be satisfactory, the THV delivery system can be removed with deactivation of the retroflex function upon exiting the aortic arch to straighten the catheter and removed from the device sheath (Fig. 50.3C). If a surgical cutdown was performed for access, surgical closure is performed following device sheath removal under direct vision.

For fully percutaneous access, we routinely perform a cross-over balloon technique that utilizes a contralateral balloon for the control of the iliac. In brief, an Omni catheter (Angiodynamics,



C



D

Fig. 50.4. (Continued)

Latham, NY) over an angled glide wire is placed in the contralateral side and maneuvered into the device sheath distally until at the ipsilateral access site. The Omni catheter is replaced with a 7F Ansel catheter (Cook Medical, IN) over a stiff wire. A peripheral occlusion balloon (usually an 8 or 10 mm) is placed in the ipsilateral common iliac through the Ansel catheter, and the Prostar sutures are prepared. The device sheath is then moved back to just above the ipsilateral access site, the peripheral balloon is inflated to fully occlude flow distally in the ipsilateral iliac, and the Prostar sutures are tied as the device sheath is removed. Once the sutures are secure, the iliac balloon is deflated and a femoral angiogram is performed. Most post-THV extravasations less than severe can be adequately controlled with redeploying the occlusive balloon at the site of the arteriotomy, reversing the heparin, and applying manual pressure. In the event of a percutaneous closure failure, the presence of percutaneous balloon control of the proximal iliac allows for controlled open vascular repair with a minimal incision or peripheral covered stenting from the contralateral access.

If no significant changes in heart rhythm are encountered, the TVP may be safely removed at the conclusion of the case, and manual pressure applied to the venous catheter site.

Transapical Approach

The transapical approach mirrors the transfemoral approach conceptually: transapical access, device sheath placement, crossing the valve, BAV, and then valve deployment. Hemodynamic changes and AR after BAV are tolerated less with a large device sheath impeding flow in the ventricle; therefore, operators must move quickly between steps should hemodynamic compromise occur. Anesthetic and team considerations all apply to the transapical approach, and the hybrid room may need to be modified to accommodate the image intensifier, operators, and echocardiogram machine all near the head of the bed. Note that at this time, the transapical route of delivery is approved only for the Edwards SAPIEN THV, using the Ascendra delivery system.

Transapical and Vascular Access

The patient is positioned on the table with special attention given to the left chest area where a mini-anterolateral thoracotomy incision will be made. In women with large breasts, a plastic surgical drape may be used to pull the breast toward the head and away from the breast crease.

Arterial (5F) and venous (6F) access in the femoral vessels is gained on side best suited for cardiopulmonary bypass in an emergency, and a TVP is positioned as described previously. A clamp placed on the left chest is then imaged to identify the LV apex and proper rib space in which to enter—this location is used to make a 4 cm incision, usually in the fourth or fifth intercostal space. Dissection is carried down to the intercostal fascia. The pleura is carefully entered after ventilation is temporarily held, and LV apex location is palpated. If the apex is not accessible from the current incision, entry one rib space above or below is still an option. An area just superior to the apex is usually most suitable for entry, and this spot is verified by manually identifying the ventricular wall with a finger while observing the location on TEE. The true apex is avoided as the entry site, due to thinning of the apical wall and frequent scarring after myocardial infarction. Adhesions and pericardial fat surrounding the LV apex

are cleared. A longitudinal pericardiotomy is made large enough to expose an area for apical sutures; stay sutures are placed if the patient has not had prior cardiac surgery. If the patient has had prior cardiac surgery, we make a small pericardiotomy just large enough to accommodate the device sheath, but place our apical sutures directly through the pericardium—the adhesed pericardium acts to buttress the ventricular tissue when tying sutures. Apical sutures with soft felt pledgets are placed in a 3 × 3 cm area centered on the previously identified entry site. We typically place two U-shape stitches with felt pledgets using a 2-0 polypropylene suture on a large needle, although others have used a purse-string with success. Given the fragility of elderly ventricular tissue and labile blood pressure after valve deployment, all stitches must be precise, exact, and with perfect technique to prevent tearing of the ventricular tissue. Stitches placed too shallow may not hold, and apical site bleeding can quickly become fatal. Once the apical stitches are placed, the patient is heparinized for an ACT >230.

The optimal deployment view is determined at this stage. The deployment view may vary greatly in the transapical approach, and the positioning of the operators near the left head-side of the table often limits the full range of the C-arm. We find a slight left anterior oblique with slight caudal views allows good visualization of the surgical field as well as a reasonable deployment view.

Access to the apex is achieved by placing an introducer needle directly into the center of the sutures. Extreme care must be made to ensure that this needle stick is centered within the sutures—if outside, the sutures will tighten the tissue but will not close the bleeding apical entry site. A J-wire is passed retrograde across the aortic valve through the access needle, and positioned in the descending aorta. A Judkins right catheter is placed in the descending aorta over the J-wire. An Amplatz stiff wire is passed through the Judkins catheter into the descending aorta, and the catheter is removed. Minimal bleeding from the access site is seen at this point. The stiff wire is held perpendicular to the access site, as angulation of the exposed wire may saw into the fragile ventricular tissue and cause more bleeding.

The 26F Ascendra I device sheath is now advanced into the ventricle to the 4 cm mark under fluoroscopy over the stiff wire. Significant bleeding around the sheath occasionally occurs, and gentle tightening of the sutures may lessen bleeding. Note that this maneuver can also result in unwanted tearing of ventricular tissue leading to life-threatening disruption of the LV free wall. Hence, aggressive cinching of apical sutures should be avoided, with expeditious THV deployment followed by quick removal of the sheath being favored whenever perisheath bleeding is encountered. Control of the device sheath must be maintained at all times by an operator—motion of the device sheath may severely impact BAV, valve deployment, and bleeding.

Balloon Aortic Valvuloplasty

A BAV balloon is next brought across the aortic valve, and a BAV is performed under rapid pacing, as previously described for the transfemoral approach. Sizing for the BAV balloon follows the same guidelines as the transfemoral approach. The balloon is rapidly removed after deflation.

Valve Deployment

The THV delivery system is now loaded into the device sheath, and the valve is advanced to beyond the end of device sheath (still inside the ventricle). An important detail for every case is to check the orientation of the valve—for the transapical retrograde approach, the THV is loaded in opposite manner from the transfemoral approach. Once well beyond the device sheath yet still in the ventricle, the delivery sheath is pulled back from the THV. Preparation for deployment is made by all members of the team: operators, anesthesia, and ancillary staff. Tension is held on the stiff wire, while the THV crosses the native stenotic aortic valve. Positioning is confirmed by TEE as well as pacing aortogram if necessary; subtle movements of the valve can be controlled by moving the entire device sheath rather than the THV, to avoid stored tension in the wire. Slack on the stiff wire can also alter the position of the valve—removing the slack with tension will move the valve closer to the left coronary cusp. The valve is positioned 50% ventricular and 50% aortic based on patient anatomy.

Deployment occurs under rapid pacing with balloon inflation after position confirmation by TEE and fluoroscopy. After deployment, the balloon is quickly moved back into the device sheath, and PVL is evaluated by echocardiography. If no significant PVL is identified, the wire and balloon are removed from the device sheath. Ventricular apical sutures are prepared for tying, while ventricular pacing is used to reduce the systemic blood pressure below 100 mmHg. Once a target blood pressure is reached, the sheath is removed and the two sets of sutures are tied down. Rarely, central leaking from the suture line is seen and is easily controlled with a single suture through the pledgets. Bleeding with systole from outside the pledgeted suture area not only may be benign and resolve with pressure or hemostatic agents but may also signal a more serious ventricular tear beneath the visible sutures. Additional sutures may help, but significant bleeding from the apex should be addressed by instituting cardiopulmonary bypass peripherally, followed by appropriate repair of the apex with a decompressed ventricle.

Closure

Reversal of heparin with protamine is performed if any concern for bleeding exists. The femoral access site is closed with a Perclose device, while the TVP is removed and pressure applied to the venous access site. In patients with no prior cardiac surgery, one 3-0 absorbable suture stitch is placed to reapproximate the pericardium. The thoracotomy is closed in layers with absorbable suture; sutures to close the rib space are unnecessary. A generous intercostal nerve block with topical anesthesia is given and appears to have a noticeable reduction of postoperative pain. A 9F blake drain is placed in the pleural space.

Clinical Outcomes

The Edwards SAPIEN RF3 THV valve and delivery system were approved for use in symptomatic severe AS in inoperable patients based on data from the PARTNER trial, a prospective, randomized controlled trial conducted in the United States. Each potential patient was screened jointly by investigators and enrolled into cohort A, high-risk surgical candidates, or cohort B, inoperable patients, and met criteria for severe AS.

Patients in cohort A were randomized to either surgical AVR or TAVR (transfemoral or transapical based on vascular access), while patients in cohort B were strictly randomized to medical treatment versus TAVR by transfemoral access; transapical delivery was not an option in cohort B patients with inadequate vascular access. Patients were otherwise defined as inoperable if the risk of mortality or serious morbidity exceeded 50%, roughly corresponding to a logistic STS score >15. The primary endpoint in cohort A was freedom from all-cause death at 1 year, with secondary endpoints of rate of death from cardiovascular causes, NYHA functional class, the rate of repeat hospitalization due to valve-related or procedural

Table 50.6 PARTNER Trial Cohort A Clinical Outcomes

	30 Days			1 Year		
	TAVR ($n = 348$)	Surgical AVR ($n = 351$)	*P*-value	TAVR ($n = 348$)	Surgical AVR ($n = 351$)	*P*-value
All-cause death (%)	3.4	6.5	0.07	24.2	26.8	0.44
All-cause death or rehospitalization (%)	7.2	9.7	0.24	34.6	35.9	0.73
All stroke (%)	5.5	2.4	0.04	8.3	4.3	0.04
Major stroke (%)	3.8	2.1	0.20	5.1	2.4	0.07
All-cause death or major stroke (%)	6.9	8.2	0.52	26.5	28.0	0.68
Major vascular complications (%)	17.0	3.8	<0.01	18.0	4.8	<0.01
Major bleeding (%)	9.3	19.5	<0.01	14.7	25.7	<0.01
Atrial fibrillation (%)	8.6	16.0	<0.01	12.1	17.1	0.07
Pacemaker insertion (%)	3.8	3.6	0.89	5.7	5.0	0.68
AV area (EOA) (cm^2)	1.7 ± 0.5	1.5 ± 0.4	0.001	1.6 ± 0.5	1.4 ± -0.5	0.002
AVG_{mean} (mmHg)	9.9 ± 4.8	10.8 ± 5.0	0.16	10.2 ± 4.3	11.5 ± 5.4	0.008

AV, aortic valve; AVG, aortic valve gradient; AVR, aortic valve replacement; EOA, effective orifice area; TAVR, transcatheter aortic valve replacement.

clinical deterioration, the distance covered in a 6-minute walk test, valve performance, and the rates of myocardial infarction, stroke, acute kidney injury (AKI), vascular complications, and bleeding. For cohort B, the primary endpoint was freedom from death over the trial period (1 year), and a composite endpoint of death and recurrent hospital readmission.

TAVR met criterion for noninferiority when compared with surgical AVR in cohort A patients at 1 year (overall mortality: 24.2% vs. 26.8%; hazard ratio: 0.93; 95% confidence interval: 0.71 to 1.22; $P = 0.001$ for noninferiority; see Table 50.6). The study was not powered to compare transfemoral versus transapical outcomes, but it is notable that the outstanding surgical outcomes at 30 days (mortality 6.5%) were far lower than expected by risk calculations (STS score: 11.8%). Each treatment arm experienced adverse events at 1 year: in TAVR patients, a higher rate of major stroke (5.1% vs. 2.4%) and vascular complication (18.0% vs. 4.8%) was seen, while major bleeding (25.7% vs. 14.7%) was more common after surgical AVR.

TAVR demonstrated a dramatic survival benefit for patients in cohort B: all-cause mortality was decreased from 49.7% in medically treated patients to 30.7% in patients undergoing TAVR at 1 year ($P < 0.001$; see Table 50.7). Event-free major

Table 50.7 PARTNER Trial Cohort B Clinical Outcomes

	30 Days			1 Year		
	TAVR ($n = 179$)	Medical therapy ($n = 179$)	*P*-value	TAVR ($n = 179$)	Surgical AVR ($n = 179$)	*P*-value
All-cause death (%)	5.0	2.8	0.41	30.7	49.7	<0.001
All-cause death or rehospitalization (%)	11.2	12.3	0.74	43.6	70.4	<0.001
Event-free MACCE (%)	90.5	94.4	NR	65.4	47.1	0.003
All stroke (%)	7.3	1.7	0.02	11.2	4.5	0.03
Major stroke (%)	5.6	1.1	0.04	8.4	3.9	0.12
All-cause death or major stroke (%)	8.4	3.9	0.12	33.0	50.3	0.001
Major vascular complications (%)	16.8	1.1	<0.0001	17.3	2.2	<0.0001
Major bleeding (%)	20.6	3.9	<0.0001	28.4	14.4	<0.001
Pacemaker insertion (%)	3.4	5.0	0.60	4.5	7.8	0.27
AV area (EOA) (cm^2)	1.5 ± 0.4	0.8 ± 0.2	<0.0001	1.6 ± 0.5	0.7 ± –0.32	<0.0001
AVG_{mean} (mmHg)	11.1 ± 6.6	33.0 ± 12.5	<0.0001	12.5 ± 10.3	44.4 ± 15.7	<0.0001

AVG, aortic valve gradient; EOA, effective orifice area; MACCE, major adverse cardiac and cerebrovascular events; TAVR, transcatheter aortic valve replacement.

adverse cardiac and cerebrovascular events and almost all functional endpoints were significantly improved after TAVR. Yet, concern has been raised over the higher rate of stroke experienced in the TAVR arm (11.2% vs. 4.5%; $P = 0.03$) and vascular complications (17.3% vs. 2.2%; $P < 0.0001$), although it is expected that vascular complications will lessen with smaller delivery systems.

POSTOPERATIVE CONSIDERATIONS

Complications

TAVR is associated with a steep learning curve during which time the operative team will face numerous unforeseen challenges. Expert opinion indicates an experience between 30 and 100 cases being needed before a competency plateau is reached where an individual heart team can reliably manage the majority of complications faced during a TAVR procedure. As such, most periprocedural complications are related to lack of operator experience, interpreting complex three-dimensional anatomy through two-dimensional imaging, and understanding the nuances of clinical management unique to the fragile TAVR patient. Rarely, a particular THV prosthesis may malfunction; however, this has become increasingly less common, particularly with later generation THVs. Although not strictly considered a complication, many heart teams find themselves managing numerous complications in their early experience secondary to poor patient screening and selection.

Strokes and Transient Ischemic Attacks

The incidence of clinically significant "major" stroke, defined as a modified Rankin score >2, after TVAR has been reported between 2.9% and 8.4% at 30 days and 1 year, respectively, depending upon the access approach and device deployed. This incidence is significantly higher than the 2.1% and 2.4% rates for surgical AVR reported in the literature for similar matched cohorts. The etiology of most cerebral vascular events in the TAVR patient is related to embolization of atherothrombotic material during device delivery and deployment. The source of embolic material is thought to be from the aortic arch and heavily calcified valve leaflets. In addition, periprocedural hypotension and hemodynamic instability, in conjunction with pre-existing patient comorbidities such as advanced age, hypertension, atrial fibrillation, peripheral vascular disease, and diabetes all contribute to the overall stroke risk. Recent magnetic resonance imaging-diffusion weighted imaging (MRI-DWI) microembolism studies have reported the incidence of cerebral ischemic lesions post-TAVR to be as high as 68% to 84%. However, the clinical significance of these data is not apparent. Currently, novel cerebral protection devices are under development, as stroke risk will need significant mitigation prior to TAVR becoming an acceptable option in younger or lower risk candidates. The incidence of stroke may also lessen as patient selection becomes more refined, delivery systems improve in their profile, and protocol-driven antithrombotic regimens are routinely used during TAVR.

Conduction System Disturbances

Conduction abnormalities are frequently seen in patients undergoing TAVR with an incidence higher than that historically seen in surgical AVR. Pre-existing conduction defects, extent of aortic leaflet and annular calcification, interventricular septal thickness and final prosthesis position, which can cause mechanical impingement of the conduction system, are all associated factors. The incidence of complete heart block after TAVR ranges from 19% to 22% with pre-existing right bundle-branch block being a risk factor contributing to its development and the need for subsequent pacing. The incidence of complete heart block requiring permanent pacemaker implantation has been higher with the CoreValve than with the SAPIEN valve potentially due to its larger profile and lower extension into the LV outflow tract. The incidence of new left bundle-branch block varies widely ranging from 14% to 83%. The majority of conduction abnormalities occur in TAVR prior to THV deployment, with nearly 70% attributable to wire-crossing of the aortic valve, BAV, and prosthesis positioning. The remaining events likely occur during THV expansion. Although permanent pacemaker implantation has not been shown to negatively effect survival, continuous perioperative telemetry should be routine in all patients undergoing TAVR, with longer monitoring required for those patients with known conduction abnormalities or those receiving a CoreValve device.

Vascular Complications

Pre-existing vascular disease ubiquitous to the TAVR population combined with large caliber sheaths necessary for THV delivery makes vascular complications the most frequent adverse outcome associated with TAVR. The incidence of major vascular complication for transfemoral access ranges from 2% to 26% and it has reported to be between 5% and 7% for transapical TAVR. Although of lower incidence, transapical complications, when they arise, are more likely to be fatal. Vessel size, tortuosity, degree of aortoiliac occlusive disease, calcium burden, and sheath-to-artery ratio along with operator experience are all predictors of major vascular complications. As operator experience increases, vascular complications become easier to anticipate and mortality decreases. Aortic occlusive balloons and cross-over catheters should be accessible to minimize bleeding and stabilize the patient in the event of a major vascular complication. The Valve Academic Research Consortium (VARC) has classified the major vascular complications associated with TAVR to include aortic dissection, perforation, rupture, bleeding requiring significant blood transfusions, or additional percutaneous or surgical intervention. In an attempt to limit vascular complications, a growing experience with alternative approaches including transaortic and subclavian access is underway in several centers.

Aortic Regurgitation and Paravalvular Leak

AR is commonly noted immediately post-TAVR, occurring at an incidence of 85% with up to 75% of patients still experiencing mild or more AR at 1 year, and one-third having more than mild. AR post-TAVR can be categorized according to etiology, as either valvular or paravalvular. In addition, it should be further characterized as to its location, severity, and cause with an estimate of overall volumetric impact calculated at time of recognition. Most valvular AR is central and attributable to the guide wire remaining across the prosthesis after deployment and immediately resolves upon withdrawal of the wire. Residual central AR, however, results from improper sizing or deployment. Diligent pre-procedural planning utilizing both echocardiography and MDCT for annular analysis is important in order to appropriately size the valve compared with the annulus. Rarely is AR caused by THV malfunction but when this scenario occurs it may be related to prosthesis damage imparted during the crimping process. In the setting of CoreValve, low positioning of the frame can result in significant AR and can be resolved by snaring the frame loops and correctly repositioning

the prosthesis. Significant central AR persisting after withdrawal of the guide wire requires rapid consideration of a valve-in-valve deployment.

PVL is also not uncommon post deployment and is most often secondary to inadequate expansion of the prosthesis or to a high calcium burden that prevents the valve from properly seating and sealing within the annulus. Trace PVL requires no intervention and the temptation to achieve perfection with hastily re-ballooning a deployed prosthesis should be resisted. Mild-to-moderate PVL, however, may need to be addressed and is worthy of cautious attempts at repeat balloon expansion of the prosthetic valve to obtain a better seal. An additional 0.5 to 1.0 cm^3 of saline can be added to the balloon prior to this repeat expansion. This advanced technique when used in experienced hands generally achieves good outcomes. Hemodynamically severe AR post-TAVR is rare. Currently, there are not sufficient long-term data that impart the clinical significance of residual PVL post-TAVR. However, it is clear that significant AR, if left untreated, is associated with adverse outcomes with even mild to moderate-severe AR patients developing chronic volume overload and LV remodeling and dysfunction. Two-year data from the PARTNER trial further support this, and suggest that even mild PVL may lead to worsened short-term survival outcomes. At this time, hemolysis is not a reported complication post-TAVR.

Valve Malposition and Embolization

Improper THV positioning can lead to instability, migration, and embolization. If recognized in a timely manner, an unstable prosthesis can be remedied by rapid placement of a second overlapping valve-in-valve. If embolization into the ascending aorta occurs, immediate efforts should be made to maintain wire access through the THV keeping its orientation such as to allow persistent forward aortic blood flow. Subsequent maneuvers are then aimed at delivering the valve into the descending aorta to a final resting location where it can be permanently fixed by balloon re-expansion in the appropriate orientation. Once the original valve is stabilized, a second valve deployment can be attempted depending on the reason for the initial deployment failure. Ventricular embolization requires urgent surgery; however, this complication is near uniformly fatal.

Coronary Obstruction

Coronary obstruction post-TAVR is a rare concern occurring with an incidence of <1.0%. It occurs less frequently with the SAPIEN THV than with the CoreValve system and can usually be avoided with careful MDCT imaging and thorough preprocedural aortic root analysis. Longitudinal remodeling, degenerative changes associated with AS, and sinotubular junction effacement can all result in a foreshortened distance between the coronary ostia and the aortic annulus. These factors combined with heavy leaflet and annular calcium burden predispose a patient to coronary obstruction. Acute occlusion can be recognized by characteristic regional hypokinesis, best appreciated from the transgastric view of the TEE and by evaluating flow in the coronary arteries themselves. In patients with prior CABG, the left main coronary artery may be protected by patent bypass grafts in which case occlusion of the coronary ostia is of less consequence; however, preparedness for rapid PCI or surgical revascularization is paramount should acute ostial occlusion with associated hemodynamic instability arise.

Acute Kidney Injury

AKI after TAVR occurs in up to one-third of patients and results in renal replacement therapy in up to 2%. As with surgical AVR, AKI portends a much higher mortality than those without injury (25% vs. 8%). In comparison to the surgical AVR, TAVR still carries an overall lower incidence of AKI and would appear to be one of its advantages over the conventional approach. Contrast load, pre-existing renal insufficiency, hypertension, transapical approach, and blood transfusion have been all implicated as risk factors for post-TAVR renal failure. In terms of procedural optimization against AKI, recommendations pertain to limiting contrast volume, avoiding intraprocedural extremes of blood pressure, and meticulous vascular access and management. Efforts to avoid other complications that contribute directly to AKI are the mainstay of prevention, with supportive care and optimization of fluid status being the only treatment once AKI occurs.

FUTURE DIRECTIONS

Expanding Patient Eligibility

Currently, only patients without a surgical option for severe AS are candidates for TAVR. However, as the technology for THV matures and clinical outcomes improve, there will be increased demand for TAVR in lower risk populations. Proliferation of TAVR among low-risk patients has been seen in Europe, despite the lack of randomized clinical data to support its use, and this practice has yet not been adopted in the United States. Currently, data are being reviewed by the Food and Drug Administration from the high-risk cohort (A) from the PARTNER 1 trial, as defined by patients with an STS score >10 for device approval. The PARTNER 2 randomized control trial will also address the use of the XT device in high-risk patients who are still operative candidates, also defined by an STS score >10. These data will hopefully provide justification for the use of TAVR in high-risk patients, but the issue of long-term durability and stroke will need to be fully evaluated before TAVR can replace surgical AVR in low- or medium-risk patients. An exciting application of THV technology is for bioprosthetic valve structural degeneration or failure, otherwise known as "valve-in-valve" use. As noted earlier, off-label use of THV for valve-in-valve procedures has been reported with success in the United States for both aortic and mitral valve replacement; both transfemoral and transapical routes have been described, although transapical may offer anatomic advantages for mitral valve replacement (easier access for crossing mitral valve, obviate need for transseptal puncture, etc.). Patients with concomitant AR may also be effectively treated with TAVR, and refinement of deployment techniques may be necessary for success. Future study of all of these patient groups in randomized clinical trials is warranted, and should be forthcoming in the following years.

Next Generation Technologies

One of the immediate needs in the current valve technology is availability of smaller delivery sheath sizes and less traumatic deployment systems. In addition, a broader range of valve sizes is in demand, especially as patient population eligible for TAVR expands. A number of companies have valves in various stages of production and testing, each with slightly different design features: Direct Flow (Lake Forest, CA), Sadra (Los Gatos, CA), AortTx (Redwood City, CA), Jena Valve (Munchen, Germany), Heart Leaflet Technology (Milan, Italy), Advanced Bioprosthetic Surfaces PercValve (San Antonio, TX), EndoTech SPA (Como, Italy), Ventor Embracer (Medtronic),

Symetis (Switzerland). It remains to be seen which valves will actually progress to clinical practice.

Route of Access

Alternative routes of access may further expand the patient population who are not transfemoral or transapical candidates. The routes frequently require some form of surgical exposure with modification of existing THV delivery platforms. The subclavian artery, via a right or left subclavian artery cutdown and/or conduit, and direct aortic access, via a parasternal incision or hemisternotomy, are the two most common routes described, both of which require surgical exposure. Current device sizes do not permit a fully percutaneous access from an alternative arterial site, such as the brachial artery, though this may be possible in the future.

Adjunctive Technology

Accessory technology in the THV field may revolutionize the deployment of THV, imaging of the aortic valve, hemostasis of surgical sites, and safety profile of the procedure. New balloon technology may prevent valve slippage or minimize PVL, while reducing the delivery system profile. A number of percutaneous transapical systems may provide a safe and bloodless entry site to the left ventricle. Cerebral protection devices, such as SMT (Herzliya, Israel), Embrella (Edwards Lifesciences), and Claret Medical (Santa Rosa, CA) may reduce stroke rates for transfemoral and transapical approaches by the use of deflectors and filters. Finally, advanced imaging techniques may offer increased aortic annulus resolution and anatomy that improves sizing, while positioning software programs automate valve positioning based on individual anatomy. These telemanipulation ideas offer tremendous potential for application in the field of THV.

Conclusions

TAVR has become a safe, effective, and appropriate first-line therapy for the treatment of symptomatic severe AS in patients with prohibitive surgical risk. Both the transfemoral and transapical approaches are available, depending on patient anatomy and characteristics. The Edwards SAPIEN and Medtronic CoreValve THV systems are under study in the United States, but an explosion of new technology and innovation is on the forefront in the field of THV. Results from the PARTNER study have established THV as a superior treatment when compared with medical therapy for severe AS, and further study of lower risk patient populations is underway.

ABBREVIATIONS AND ACRONYMS

AS—aortic stenosis
AVR—aortic valve replacement
BAV—balloon aortic valvuloplasty
CAD—coronary artery disease
EVAR—endovascular aortic aneurysm repair
IVUS—intravascular ultrasound
LAD—left anterior descending
LV—left ventricle
MDCT—multidetector row computed tomography
PVL—paravalvular leak
OR—operating room
SAVR—surgical aortic valve replacement
TAVR—transcatheter aortic valve replacement
THV—transcatheter heart valve
TVP—transvenous pacemaker

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EDITOR'S COMMENTS

Dr. Williams and colleagues at Columbia University have the leading experience and expertise in the United States regarding transcatheter aortic valves. This chapter is outstanding and provides us a detailed approach to this complexed intervention. The chapter descriptions are clear and they give us a terrific approach to this important new technology.

The important concepts include that of a heart team (cardiologist and surgeons), hybrid lab, and appropriate selection of candidates. I cannot overstate the importance of the selection criteria. There is no doubt that when used properly, this technology has outstanding results in high-risk patients. However, there are many patients who are just too high risks for any procedure let alone a valve operation. Despite the fact that the approach can be completely percutaneous, as an aortic valve surgery this cannot be ignored. Though we had outstanding results in the PARTNER trial, our commercial results lagged in the beginning because of issues with selection. Often the patient comorbidities are more important than their aortic valve disease.

There are still some issues that need to be learned and dealt with. In the initial experiences, vascular complications were the major cause of morbidity and mortality. I think things have gotten better with appropriate imaging and use of transapical and transaortic routes for deployment. An ongoing issue is aortic insufficiency which occurs in some fashion in the majority of patients. This relates to evolving technology and limited valve sizes. However, we will need to understand better what this means to overall survival, both short and long term.

ILK

51 Coronary Artery Bypass Grafting using Cardiopulmonary Bypass

Ahmet Kilic, Chittor Sai-Sudakar, Juan Crestanello, and Robert S.D. Higgins

INTRODUCTION

Atherosclerotic heart disease continues to be the leading cause of death in the United States. While coronary artery bypass surgery has enjoyed a storied evolution since the 1960s to be among the most spectacular advances in medicine, the field of cardiothoracic surgery is undergoing a significant evolution with the advent of more effective medical therapies, alternative percutaneous and hybrid revascularization techniques, and percutaneous valve procedures. Ironically as these alternative strategies evolve, the need for effective surgical myocardial revascularization techniques using cardiopulmonary bypass is becoming even more critical in patients with advanced atherosclerotic heart disease and left ventricular dysfunction. Central to this expanding need is the refinement of indications for coronary artery bypass procedures in critically ill patients. Older patients with advanced disease, left ventricular dysfunction, and multiple comorbidities create even greater challenges for the practicing cardiac surgeon.

In this chapter, we will review the evolving assessments of coronary artery anatomy using innovation imaging techniques. A full understanding of assessments of myocardial viability and suitability for revascularization continues to be of critical importance in this discussion. We will also review the impact of left ventricular dysfunction on surgical revascularization outcomes in patients with low ejection fraction. The importance of surgical risk reduction in complicated coronary revascularization particularly in patients on platelet inhibitor therapies, that is, clopidogrel as well as evolving perioperative management techniques, has significantly reduced the morbidity and perioperative complications of coronary revascularization. The indications for hybrid and multivessel disease management as well as left main coronary revascularization will also be reviewed.

No discussion of the management of coronary artery disease (CAD) by conventional revascularization techniques can ignore the importance of the development of appropriateness criteria for the management of multivessel disease and left main coronary disease. Evidence highlighting the benefits of coronary artery bypass grafting (CABG) continues to demonstrate significant survival advantage in patients with multivessel disease compared with percutaneous coronary intervention (PCI). In these recent contributions to the literature, critical evidence from randomized clinical trials and case-matched patient studies, CABG was associated with lower risk of death than stenting underscoring the comparative effectiveness of CABG in these patients. As we appreciate the comparative benefits of CABG versus PCI, we also have to be mindful of the challenges in the current reimbursement environment to manage preventable complications, so as to reduce the financial impact of readmissions in the current healthcare economy. Only if the surgical profession can minimize the risk in appropriately selected patients, enhance patient outcomes, reduce perioperative complications, and readmission rates after CABG, will we be able to continue to demonstrate the comparative benefits of CABG.

ASSESSMENT OF CORONARY ANATOMY

Coronary Angiography

Coronary angiography remains the gold standard in defining coronary anatomy and graft patency. It is an invasive procedure and is associated with procedural risks including stroke, native vessel dissection, acute myocardial infarction (MI), ventricular arrhythmias, and puncture site morbidities such as hematomas and pseudoaneurysm formation. In addition, its deficiency in three-dimensional definition has led to the investigation of other noninvasive imaging techniques to define the overall coronary pathoanatomy. Initial attempts to utilize electron beam tomography and helical scan computed tomography (CT) were not encouraging due to motion artifacts, the need for prolonged breath holding, and image distortion due to metallic clips. Newer imaging techniques such as multidetector computed tomography (MDCT) have begun to revolutionize the diagnostic field.

Multidetector Computed Tomography

MDCT allows for fast data acquisition in a single breath hold of 25 to 30 seconds with varying slice thickness collimation, and Gantry rotation speeds following intravenous administration of 80 to 100 ml of nonionic contrast agent and a predetermined delay period to allow the contrast to reach the arterial circulation. Preprocedure beta-blockade allows for a heart rate of <60 beats per minute since faster hearts along with atrial fibrillation and

ectopic beats impair the quality of images obtained. Image reconstruction and manipulation is facilitated by electrocardiographic gated reconstruction algorithms.

Evaluation of Native Coronary Anatomy

Martuscelli and coworkers investigated the accuracy of MDCT in detection of significant (>50%) stenosis using a scanner equipped for 16 × 0.625 mm collimation and compared it with coronary angiography in 64 patients with suspected CAD. Eighty-four percent of the angiographic segments were evaluable. Severe calcification, cardiac motion artifacts, poor opacification, and blending of the segments with veins hindered the evaluation of the remaining segments. In the segments that were evaluable, namely, vessels >1.5 mm in diameter, MDCT had a sensitivity, specificity, positive predictive value, and negative predictive value of 89%, 98%, 90%, and 98%, respectively. MDCT had a 100% specificity and sensitivity in detecting total occlusion. Overall, MDCT detected 78% of stenoses detected by angiography. Cury et al. demonstrated an excellent correlation between 16-slice MDCT and angiography in quantifying the degree of stenosis. However, MDCT tended to overestimate the stenosis. The evaluation of the coronary anatomy and the results were significantly better than those obtained with the 4-slice MDCT. With the advent of 64-slice MDCT, Rubinshtein et al. analyzed its efficacy in the emergency room in evaluating patients with chest pain. They observed that emergency room MDCT had a high positive predictive value for diagnosis of acute coronary syndrome and a negative study predicted a low rate of major adverse cardiovascular event. In this study, only 4.6% of the coronary segments were of low image quality testifying to improvements in MDCT technology and also aided by the low calcium burden. These diagnostic tests have also been helpful in planning for minimally invasive revascularization procedures. During the procedures of minimally invasive direct coronary artery bypass (MIDCAB) or totally endoscopic coronary artery bypass (TECAB), the visualization of the target vessels is often limited and it is critical to identify the target vessel correctly and bypass it at the correct site. A common source of error is the bypassing of a large diagonal instead of the left anterior descending (LAD) artery during minimally invasive surgery. In addition, the target vessels could be buried under epicardial fat, covered by a myocardial bridge, run an intramural course, or heavily calcified. Newer generation MDCT scanners have been shown to identify these variations in coronary anatomy better than coronary angiography. These findings can dictate the type of surgery offered to the patients. Coronary calcification is obscured by the dye used in angiography. In an open procedure, such areas can be palpated and anastomosis can be performed in the noncalcified segments of the coronary vessels. However, tactile feedback is absent in the minimally invasive procedures. Regions of coronary vessels burdened by calcification as evidenced by MDCT can be avoided for anastomosis.

Evaluation of Graft Patency

Reoperative cardiac surgery is associated with increased morbidity and mortality and the potential to inadvertently injure the bypass grafts. MDCT allows for the accurate three-dimensional visualization of the course of the left internal mammary artery (LIMA) graft in relation to the midline, location of other conduits, and the degree of adherence of the right ventricle to the posterior table of the sternum. Unusual location of the grafts and their proximal anastomoses pose difficulties in evaluation by conventional angiography, especially in the absence of radio-opaque markers for the proximal anastomoses. Faster data acquisition allows a manageable breath hold for the patient allowing for visualization of the most proximal portions of the internal mammary artery grafts not seen in the early generation scanners. Martuscelli et al. analyzed 285 conduits in 96 patients with 16-slice MDCT and compared the results with those obtained by conventional angiography. The sensitivity of MDCT in diagnosing significant stenosis was 96%. Evaluation of graft patency and occlusion improved with the advent of 64-slice MDCT. The accuracy of 64-slice multislice CT for graft occlusion and stenosis is given in Table 51.1. A recent analysis suggests that the sensitivity and specificity for graft occlusion approach 100% across the studies, with only two cases of missed occlusions in bypass grafts, which were not visualized on MDCT. There are currently no reports of false-positive occlusion. When diagnosing bypass graft stenosis, sensitivity ranged from 75% to 100%. Missed stenoses at distal anastomoses due partly to calcified wall plaque and membranous-like stenoses accounted for false-negative results. The specificity for diagnosis of stenosis of bypass grafts ranges from 89.3% to 100%. Poor opacification combined with small vessel size, vessel kinking, and surgical clip artifact account for false-positive results. The results from these eight studies of 1,169 coronary artery bypass grafts are similar to those obtained by coronary angiography. Several studies have reported that grafts not visualized on coronary angiography were visualized on MDCT, thus, providing important additional clinical information.

However, assessment of native coronary arteries is impaired in the presence of atherosclerosis, calcification, and small diameter vessels. In addition, MDCT does not provide the flow characteristics of either the grafts or the distal run off. Arrhythmias, metallic clips on the side branches of the grafts, and higher heart rates continue to hinder adequate evaluation of the bypass grafts. Improved temporal and spatial resolution of the 64-slice CT scanner overcomes the artifacts caused by metal clips. Evaluation of in-stent stenosis by MDCT is difficult. Coronary stents are usually 2.5 to 4 mm in diameter. Blooming artifacts are produced by the metal in both the bare metal and drug-eluting stents. Cardiac motion compounds the problem. Partial volume effects lead to decreased visibility of the stent lumen, underestimates the luminal diameter, and overestimates

Table 51.1 Diagnostic Accuracy of 64-Slice MSCT in Coronary Artery Bypass Grafting Assessment (Assessable Grafts Only)

				Occlusion		Stenosis	
Author	**Grafts**	**Art**	**Ven**	**Sens (%)**	**Spec (%)**	**Sens (%)**	**Spec (%)**
Dikkers	69	52	17	100	100	100	98.7
Feuchtner	70	46	24	100	100	75	95
Jabara	147	47	100	93.3	100	100	100
Malagutti	109	45	64	—	—	100	98.3
Meyer	397	144	253	100	100	97[a]	97[a]
Onuma	146	72	74	100	100	100	98.1
Pache	93	22	71	—	—	97.8	89.3
Ropers	138	37	101	100	100	100	94

Art, arterial grafts; Sens, sensitivity; Spec, specificity; Ven, venous grafts.

[a]Combined results for stenosis and occlusion.

From: Jones CM, et al. Coronary artery bypass graft imaging with 64-slice multislice computed tomography: literature review. Semin Ultrasound CT MR 2008;29(3):204-213.

the outer diameter. Inside the lumen of the stent, there is an increase in the attenuation that can be minimized by utilizing submillimeter collimation and a sharp reconstruction kernel. With improving technological areas, the larger diameter stents in the left main coronary artery and the proximal coronary vasculature can be analyzed for evidence of decreased luminal diameter due to intimal hyperplasia. The native coronary vasculature in the immediate vicinity of the stent is difficult to evaluate for stenosis due to beaming artifacts. In addition, beam hardening produces dark areas adjacent to the clips stimulating stenosis especially at the anastomotic points.

MDCT can be effectively used as a one-stop test to evaluate the different causes of chest pain following CABG. Dual source and 256-slice scanning allow for fast data acquisition, may further simplify the test, improve diagnostic accuracy, and eliminate the current limitations imposed by rapid hearts rates. MR coronary angiography is being evaluated to analyze its potential as a noninvasive diagnostic tool. The summary statement released by the American Heart Association in collaboration with other councils has recommended that neither coronary artery CTA nor MRA should be used to screen for CAD in patients who have no signs or symptoms of CAD (Class III, level of evidence C). The potential benefit of noninvasive coronary angiography is likely to be greatest and is reasonable for symptomatic patients who are at intermediate risk for CAD after initial risk stratification including patients with equivocal stress-test results (Class IIa, level of evidence B). Concerns regarding radiation dose limit the use of coronary CTA in high-risk patients who have a very low pretest likelihood of coronary stenoses; they are likely to require intervention and invasive catheter angiography for definitive evaluation; thus, CTA is not recommended for those individuals (Class III, level of evidence C). Pronounced coronary calcification may negatively impact interpretability and accuracy of coronary CTA and thus, the usefulness of CTA is uncertain in these individuals (Class IIb, level of evidence B)."

ASSESSMENT OF MYOCARDIAL ISCHEMIA AND VIABILITY

Total or sub-total occlusion of any portion of the coronary vasculature leads to varying spectra of myocardial viability including infarction, myocardial stunning, and hibernating myocardium. Reestablishment of the coronary perfusion by fibrinolytic therapy or percutaneous interventions is the cornerstone of the ST elevation myocardial infarction (STEMI) protocols adopted by nearly all the hospitals across the country. In less emergent cases or patients presenting in outpatient settings with symptoms of angina or congestive heart failure and documented evidence of critical CAD and depressed systolic function, there is considerable debate about the utility of further assessment for myocardial viability prior to any intervention.

Nuclear Imaging

Nuclear imaging utilizes technetium-labeled radioisotopes or positron emission tomography (PET) tracers under rest and stress conditions to detect the extent and severity of myocardial ischemia (Table 51.2), and single-photon-emission computed tomography (SPECT) myocardial perfusion imaging is considered a useful tool for risk assessment for CAD. Following the injection of the thallium or technetium-labeled tracers (sestamibi or tetrofosmin), their uptake by the myocyte is dependent on the regional myocardial blood flow. The subsequent washout of these radiopharmaceuticals is faster from normal myocardium compared with the ischemic myocardium. The images obtained at this time point are usually considered the resting images. Subsequently, stress images are obtained using exercise stress protocols (Fig. 51.1). In patients who are unable to exercise or who have other pathologies such as an abnormal EKG, left bundle-branch block, paced rhythm, chronic obstructive pulmonary disease, or asthma pharmacological stress is induced utilizing several agents (vasodilator therapy, adenosine, dipyridamole, or dobutamine. The images obtained at rest and stress are compared and myocardial perfusion defects are graded as myocardial ischemia, reversible or fixed perfusion defects. Myocardial perfusion imaging utilizing SPECT has been validated in several studies as a useful screening tool for CAD in patients with risk factors and in addition for the detection of extent and degree of myocardial ischemia. Glucose metabolism substitutes free fatty acid metabolism as the main energy source in ischemic but viable myocardium. ^{18}F fluorodeoxyglucose (FDG), a glucose analog combined with a tracer (Rubidium-82), is used to evaluate the glucose metabolism in the myocyte. Following the detection of a fixed perfusion defect on perfusion imaging, preservation of ^{18}F-FDG uptake in that region by a PET scan is suggestive of hibernating myocardium and indicative of the beneficial effect of revascularization in patients with ventricular dysfunction.

Table 51.2 Common Myocardial Blood Flow Tracers

Tracer	PET/SPECT	Tracer production	Half-life	Scan time (min)	Positron energy (MeV)	Radiation dosage (m)*Sv
Technetium-99m sestamibi/tetrofosmin	SPECT	Generator	6 h	12–15 (dual head camera)	–	1-d: stress-rest. Sestamibi/tetrofosmin 12/10.6 (40 mCi) 2-d: stress-rest. Sestamibi 17.5 (30 mCi + 30 mCi)
Thallium-201	SPECT	Cyclotron	72.9 h	12–15 (dual head camera)		25.1 (4 mCi)
Rubidium-82	PET	Generator	76 s	6–10	3.15	16 (90 mCi)
Oxygen-15 water	PET	Cyclotron	110 s	6–10	1.72	2.5 (60 mCi)
Nitrogen-13 ammonia	PET	Cyclotron	9.97 min	2–4	1.19	2.4 (30 mCi)
F-18 FBnTP	PET	Cyclotron	110 min	12–15 (dual head camera)	0.63	Not available

From: Yoshinaga K, Manabe O, Tamaki N. Physiological assessment of myocardial perfusion using nuclear cardiology would enhance coronary artery disease patient care: which imaging modality is best for evaluation of myocardial ischemia? (SPECT-side). Circ J 2011;75(3):713-722.

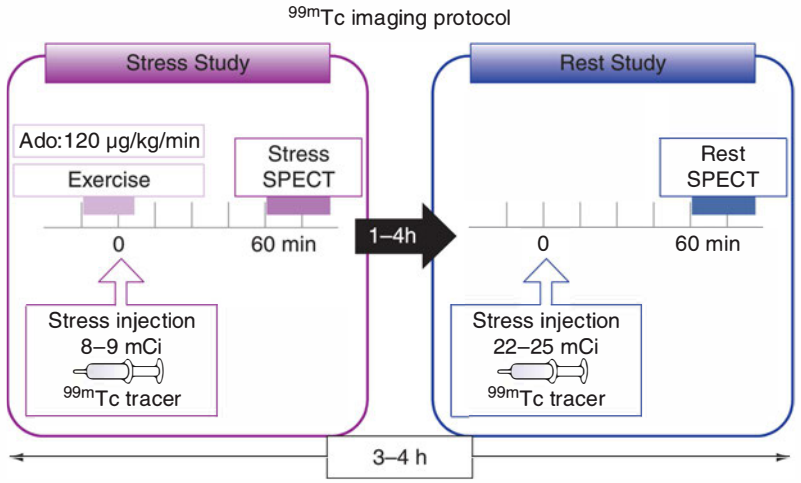


Fig. 51.1. Exercise/pharmacological stress ^{99}mTc myocardial perfusion imaging protocol. (From Yoshinaga K, Manabe O, Tamaki N. Physiological assessment of myocardial perfusion using nuclear cardiology would enhance coronary artery disease patient care: which imaging modality is best for evaluation of myocardial ischemia? (SPECT-side). Circ J 2011;75(3):713-722.)

Disturbances in fatty acid metabolism in the myocyte persist for up to 30 hours following the cessation of symptoms from a transient coronary event. This pathway has been investigated as a potential diagnostic tool for a recent coronary event by using radioiodine-labeled branched chain fatty acid β-methyl-*p*-[1231]-iodophenyl pentadeconic acid (BMIPP) SPECT scans to detect acute coronary syndromes in the emergency room with a sensitivity, specificity, and negative predictive value of 81%, 61%, and 83%, respectively.

Cardiovascular Magnetic Resonance

Over the last decade, cardiovascular magnetic resonance (CMR) has gained in popularity as a reliable diagnostic modality for cardiac structure and function and myocardial ischemia to the extent that clinical utility pathways are being developed to utilize this technology for patients presenting with acute coronary syndromes (Fig. 51.2).

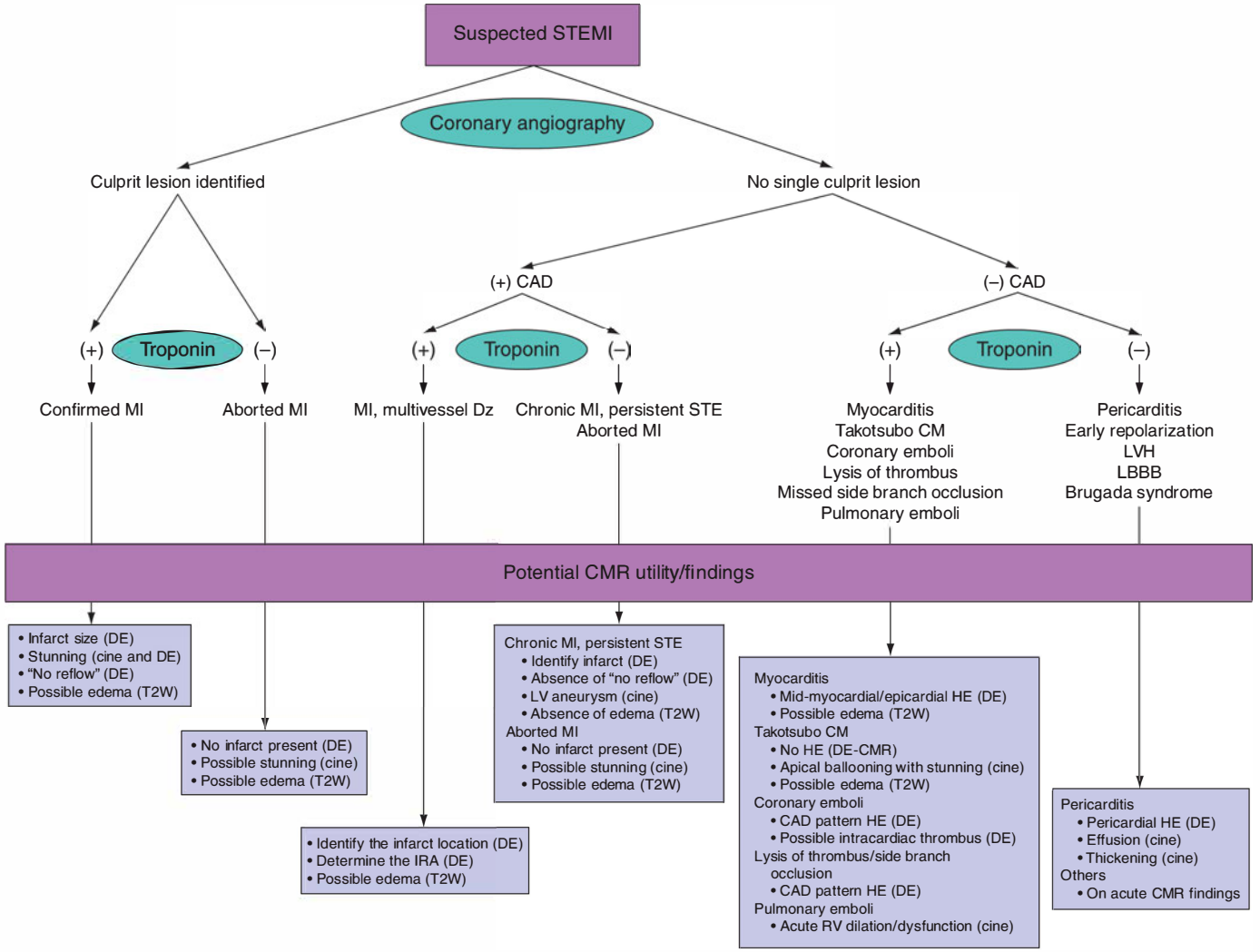


Fig. 51.2. Algorithm for potential usage of cardiovascular magnetic resonance imaging in patients with suspected ST elevation myocardial infarction. From Kim HW, Farzaneh-Far A, Kim RJ. Cardiovascular magnetic resonance in patients with myocardial infarction: current and emerging applications. J Am Coll Cardiol 2009;55(1)1-16, with permission.

Cardiovascular Magnetic Resonance Techniques

The powerful magnetic field exerted by CMR scanner with a field strength of 1.5 to T (30,000 times stronger than the earth's magnetic field) aligns the hydrogen atoms in the human body. Subsequently, radiofrequency pulses delivered by the scanner excite the hydrogen nuclei and receiver coils detect the signals as the hydrogen nuclei relax. The strength of the signal is dependant on the density of the protons in any given tissue and accounts for the contrast in the MR images. In addition, the images are also dependant on two distinct relaxation patterns of the protons. T1 is the longitudinal relaxation time and is the time required for a substance to be magnetized after being placed in a magnetic field, whereas T2, the transverse relaxation time is a measure of how long the resonating protons remain in phase following a radiofrequency pulse.

- *Spin Echo Imaging:* Blood is black and tissues are bright.
- *Gradient Echo Imaging:* Blood is bright and cardiac tissue is dark.
- *Cine CMR:* Provides important information regarding segmental and global ventricular function, wall thickness at different time points of the cardiac cycle, and structural heart defects including aneurysms, septal defects, and valve pathologies.
- *Perfusion Imaging:* For the assessment of myocardial perfusion and used in conjunction with vasodilators. Vasodilation with adenosine or dipyridamole following a bolus contrast injection causes a three- to five-fold increase in blood flow in myocardial regions supplied by normal coronary arteries resulting in bright areas in contrast to the areas supplied by stenotic coronary arteries, which appear as dark regions on imaging.
- *Delayed Contrast Imaging:* For the assessment of the ability of myocardium to clear the contrast (gadolinium diethylenetriamine penta-acetic acid). Normal myocardium clears the contrast quickly and appears dark. Ischemic or scar tissue appears bright due to delayed clearance of contrast.

ASSESSMENT OF MYOCARDIAL VIABILITY

In canine models of acute and chronic MI, contrast MRI was used to distinguish between reversible and irreversible ischemic injury independent of wall motion and infarct age. Kim et al. observed that contrast MRI in combination with cine MRI could identify acute MI (hyperenhanced with contractile dysfunction), injured but viable myocardium (not hyperenhanced but with contractile dysfunction and normal myocardium (not hyperenhanced and with normal function). Subsequently, the same group investigated the use of contrast-enhanced MRI to predict improvements following revascularization in regions of abnormal ventricular contraction. Gadolinium-enhanced MRI was performed in 50 patients with ventricular dysfunction before either surgical or percutaneous revascularization to assess the transmural extent of the hyperenhanced myocardium postulated to represent nonviable myocardium. In addition, cine MRI was performed to assess the regional contractility at the same locations before and after revascularization. Following analysis of the data, the authors demonstrated that the likelihood of improvement in regional contractility after revascularization decreased progressively as the transmural extent of the hyperenhancement before revascularization increased. The amount of left ventricular mass that was dysfunctional and not hyperenhanced before revascularization strongly related to the degree of improvement in the wall motion score and ejection fraction after revascularization. The investigators clearly demonstrated that reversible myocardial dysfunction can be identified by contrast-enhanced MRI before coronary revascularization (Fig. 51.3). Further confirmatory studies established the value of delayed enhancement MRI as a powerful predictor of myocardial viability after surgery.

HYBRID CORONARY REVASCULARIZATION

While conventional revascularization is routine in patients with triple-vessel CAD, hybrid coronary revascularization (HCR) has evolved to combine (a) surgical revascularization to the LAD coronary artery with the LIMA through a minimally invasive approach and (b) percutaneous coronary revascularization (PCI) to the circumflex and right coronary artery (RCA) territories. HCR is an alternative revascularization strategy for patients with triple-vessel CAD. HCR achieves complete revascularization and combines the advantages of bypass surgery and PCI (Fig. 51.4).

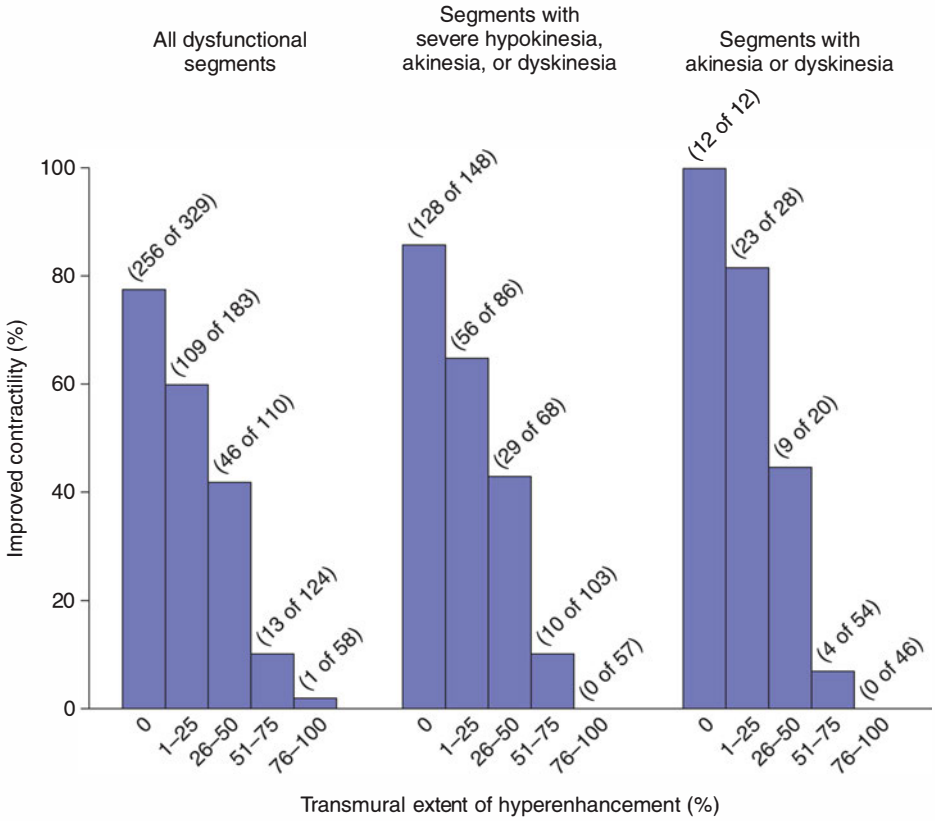


Fig. 51.3. Relationship between the transmural extent of hyperenhancement before revascularization and the likelihood of increased contractility after revascularization. (From Kim RJ, et al. The use of contrast-enhanced magnetic resonance imaging to identify reversible myocardial dysfunction. N Engl J Med 2000;343(20):1445-1453.)

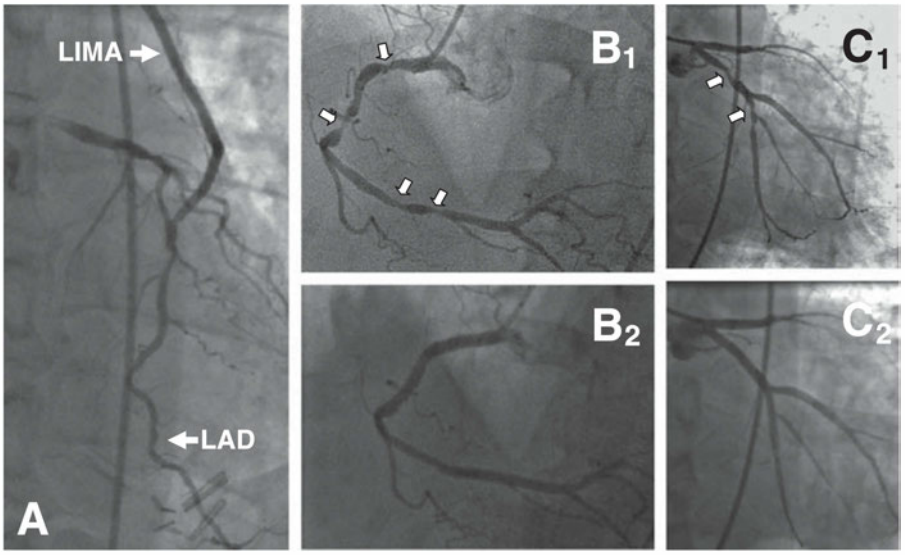


Fig. 51.4. Hybrid coronary revascularization in a patient with triple-vessel coronary artery disease. (**A**) Completion angiogram of the left internal mammary artery to left anterior descending. (**B1**) Right coronary artery (RCA) lesions that were treated by percutaneous coronary intervention (PCI). (**B2**) RCA after PCI. Lesions in the circumflex coronary artery before (**C1**) and after (**C2**) PCI.

Although traditional CABG is highly effective in improving symptoms and increasing survival, it is associated with morbidity and mortality mostly related to the invasiveness of the procedure. Revascularization of the LAD coronary artery with the LIMA provides the major clinical advantage of CABG and provides the majority of the survival advantage provided by CABG with the best long-term patency rate. Compared with PCI, LIMA bypass for isolated lesions of the LAD results in better angina relief, and lower incidence of major adverse cardiovascular and cerebrovascular events and need for repeat revascularization. Long-term patency of saphenous vein bypass grafts (SVG) is inferior to that of arterial grafts. It is unknown how the patency of SVG compares to the patency of DES in the same position.

Table 51.3 summarizes the potential advantages of HCR and compares them with PCI and CABG. HCR achieves complete revascularization potentially combining the advantages of both revascularization strategies: low procedural morbidity and mortality, low restenosis rate, and low repeated revascularization rates.

HCR builds on the concept of "heart team" approach for the management of coronary revascularization. The heart team, integrated by cardiac surgeons, interventional cardiologists, clinical cardiologists, and primary physicians, evaluate the patient's clinical and angiographic characteristics and offer the patients a revascularization strategy based on best evidence and clinical guidelines. For HCR, interventional cardiologist and surgeons should agree that the patients are candidates for both (a) PCI to the non-LAD territories and (b) minimally invasive bypass to the LAD using the LIMA.

Theoretically, all patients with triple-vessel CAD can be considered for HCR. However, until the results of randomized prospective trials exploring the safety and efficacy of HCR compared with conventional CABG are available, HCR should be reserved for:

(a) Patients with proximal LAD stenosis or chronic total occlusion of the LAD and focal lesions in the RCA or circumflex artery amenable to PCI. Patients with multisegment and diffuse disease of the RCA or circumflex artery, which would require complex PCI with multiple stents, are better candidates for conventional CABG. The SYNTAX score is helpful to select these patients. Patients with a high SYNTAX score because of complex lesions in the RCA and circumflex territory may benefit from conventional CABG rather than PCI to those targets. The LAD target should be adequate for bypass.
(b) Patients with limited or absent venous conduits.
(c) Patients with severe calcification of the ascending aorta.
(d) Patients at high risk for conventional surgery secondary to multiple comorbidities or who would tolerate a median sternotomy poorly (elderly, disabled or deconditioned patients, immunosuppressed).

Patients with double-vessel CAD may also benefit from HCR. These indications are reflected in the European Society of Cardiology and ACCF-AHA guidelines (Table 51.4).

HCR is contraindicated in patients who are hemodynamically unstable or in cardiogenic shock, or who have malignant arrhythmias. Patency of the LIMA should be evaluated preoperatively either by conventional angiogram or by CT angiography. Patients unable to tolerate single-lung ventilation due to underlying poor pulmonary function (severe COPD with FEV_1 <50% predicted, baseline hypercarbia or hypoxia on ABG, severe pulmonary hypertension) are not candidates for HCR.

Patients with contraindications to PCI are also not candidates for HCR. Those include patients with:

(a) In stent stenosis in a non-LAD territory.
(b) Intolerance to clopidogrel or inability to take double antiplatelet therapy for more than a year.
(c) Nonresponders to antiplatelet therapy.
(d) Severe renal dysfunction.

Table 51.3 Comparison of Potential Benefits of Percutaneous Coronary Intervention (PCI), Conventional Coronary Artery Bypass Grafting (CABG), Minimally Invasive CABG, and Hybrid Coronary Revascularization (HCR)

	PCI	CABG	Minimally invasive CABG	HCR (potential benefits)
Invasiveness	+	+++++	++	++
Procedural morbidity	+	++++	+	+
Procedural mortality	+	++	+	+
Long-term patency arterial graft	n/a	+++++	+++++	+++++
Repeated revascularization rate	+++	+	+	+
Completeness revascularization	+++	+++++	+	+++++

Table 51.4 American College of Cardiology Foundation (ACCF), American Heart Association (AHA), European Society of Cardiology (ESC), and European Association for Cardio-Thoracic Surgery (EACTS) Guidelines for Hybrid Coronary Revascularization

2011 American College of Cardiology Foundation/American Heart Association Guideline for Coronary Artery Bypass Graft Surgery
Class IIa
(1) Hybrid coronary revascularization is reasonable in patients with one or more of the following (level of evidence B):
(a) Limitations to traditional CABG, such as heavily calcified proximal aorta or poor target vessels for CABG (but amenable to PCI).
(b) Lack of suitable graft conduits.
(c) Unfavorable LAD artery for PCI (i.e., excessive vessel tortuosity or chronic total occlusion).
Class IIb
(1) Hybrid coronary revascularization may be reasonable as an alternative to multivessel PCI or CABG in an attempt to improve the overall risk–benefit ratio of the procedures (level of evidence C)
2010 European Society of Cardiology/European Association for Cardio-Thoracic Surgery Guidelines on myocardial revascularization
Hybrid procedure consisting of ITA to LAD and PCI of other territories appears reasonable when PCI of the LAD is not an option or unlikely to portend good results. Indications should be selected by the Heart Team and potential opportunities for using a hybrid approach are listed here (recommendation class IIb, level of evidence B):
(1) Primary PCI for posterior or inferior STEMI and severe CAD in nonculprit vessel(s), better suited for CABG.
(2) Emergent PCI prior to surgery in patients with combined valvular and coronary disease, if the patient cannot be transferred for surgery, or in the presence of acute ischemia.
(3) Patients who had previous CABG and now require valve surgery and who have at least one important patent graft (e.g., ITA to LAD) and one or two occluded grafts with a native vessel suitable for PCI.
(4) Combination of revascularization with nonsternotomy valve intervention (e.g., PCI and minimally invasive mitral valve repair, or PCI and transapical aortic valve implantation).
(5) In patients with conditions likely to prevent healing after sternotomy, surgery can be restricted to the LAD territory using minimally invasive direct coronary artery bypass (MIDCAB) excluded patients left ITA grafting. Remaining lesions in other vessels are treated by PCI.

CABG, coronary artery bypass grafting; CAD, coronary artery disease; ITA, internal thoracic artery; LAD, left anterior descending; PCI, percutaneous coronary intervention; STEMI, ST elevation myocardial infarction.

(e) Severe PVD that preclude angiographic access.
(f) Lesion at high risk of restenosis (bifurcation, trifurcation lesions).

Relative contraindications include:

(a) Previous left-sided thoracic surgery or adhesions on left pleural space.
(b) Previous cardiac surgery.
(c) Morbid obesity (BMI >40).
(d) Small left chest cavity with large cardiac silhouette and a thick chest wall due to obesity. This combination makes the trocar placement dangerous. It also compromised the movements of the robotics arms and requires a larger thoracotomy to perform the anastomosis.
(e) EF <20%.
(f) Inability to tolerate CO_2 insufflation.
(g) Intramyocardial LAD.

The LIMA harvesting can be performed either robotically, thoracoscopically, or directly via a small left anterior thoracotomy. Our preferred approach is robotically using the DaVinci system (Intuitive, Surgical, Inc. Sunny Valley, CA). The robotic system allows for very precise and reproducible harvesting of the LIMA from the first rib to its bifurcation. There is no retraction of the ribs as it is in the traditional MIDCAB approach.

Robotics ports are placed on the third, fifth, and seventh intercostal space slightly anterior to the left anterior axillary line (Figs 51.5 and 51.6A). CO_2 insufflation is established via a trocar side port. We prefer high flow at low pressure to avoid hemodynamic compromise (20 L/min at 6 to 8 mmHg). Patient hemodynamics should be carefully monitored during the time the chest is insufflated. The intrathoracic pressure may need to be adjusted and vasoconstrictors may be necessary to maintain blood pressure and cardiac output during the procedure. Excessive volume infusion should be avoided to prevent right ventricular dilatation that may compromise the performance of a totally endoscopic procedure.

The internal mammary artery is taken down in a skeletonized manner using the electrocautery and bipolar forceps. Side branches are cauterized or clipped. After heparin is administered to achieve an ACT >300 seconds, the distal end of the mammary is divided between clips. The left phrenic nerve should be identified and avoided. A pericardium window is performed robotically as a trapdoor incision with the base on the diaphragm. One incision is placed anterior and parallel to the left phrenic nerve and another in the midline. Both pericardial incisions are connected at the base of the heart. The LIMA, the target area, and its relationship to the chest wall are identified after careful release of the left chest insufflation. This reestablishes the normal anatomic relation between the heart and the chest wall and allows for identification of the intercostal space closer to the target area minimizing the size of the thoracotomy incision. The robotic instruments and ports are then removed. There is a learning curve associated with the procedure with decrease in robotic time, rate of injury to the LIMA, and conversion rate as operators became more experienced.

TECAB can also be performed. The mammary is taken down as previously described with the DaVinci system. A robotic endostabilizer, placed through an additional port under the xyphoid, is used to immobilize the target vessel. The anastomosis is performed with the robotic instruments either with sutures or with U clips (Medtronic, Minneapolis, MN). It is a more

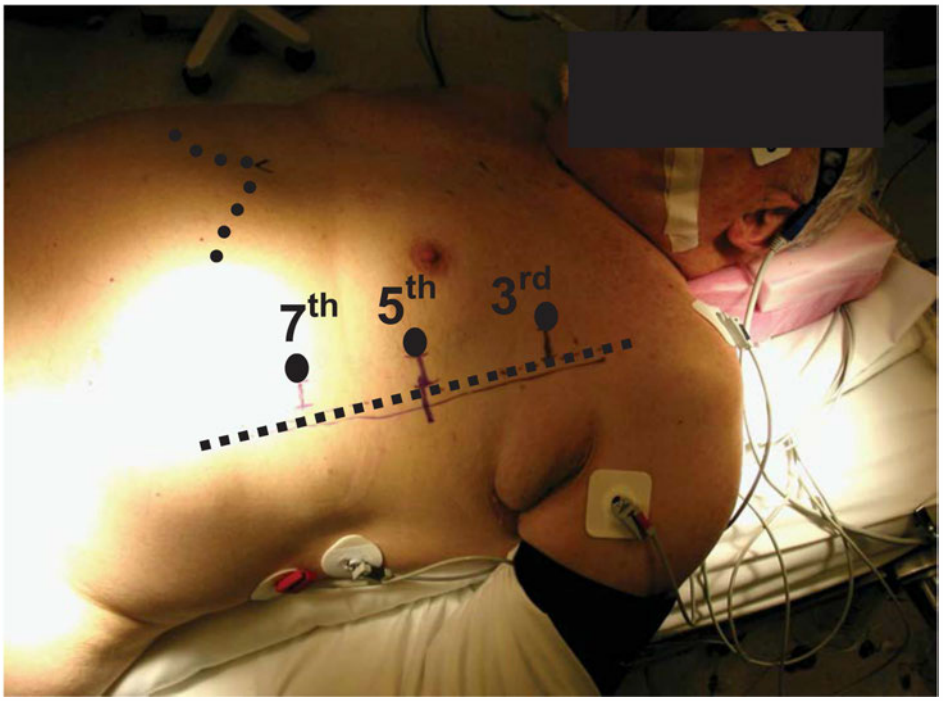


Fig. 51.5. Positioning and port location for robotic left internal mammary artery takedown. Note left shoulder retracted inferiorly to avoid conflicts with the right robotic arm. Ports are located in the third, fifth, and seventh intercostal spaces anterior to the anterior axillary line (*dotted line*). The fifth intercostal port incision is conversed to a left anterior minithoracotomy.

technically demanding and time-consuming procedure that has been mastered only by a few centers of excellence. TECAB can be performed on cardiopulmonary bypass either in the arrested heart or on a beating heart. Cardiopulmonary support can be instituted through the groin with cannulation of the left common femoral artery and vein. On-pump beating heart bypass can be used to avoid the hemodynamic and pulmonary disturbances associated with single-lung ventilation, chest insufflation, hypercarbia, cardiac manipulation, and coronary occlusion. Decompression of the heart also increases the intrathoracic space, which is especially important for TECAB procedures. TECAB can also be performed with an arrested heart occluding the aorta with an EndoClamp aortic occlusion catheter (Edwards Lifesciences, Irvine, CA). Additional bypasses to the diagonal, ramus intermedius, and obtuse marginal coronary arteries can also be performed with the MIDCAB or TECAB approach.

Revascularization of the non-LAD vessels can be staged (PCI followed by CABG or CABG followed by PCI) or performed concomitantly in the same setting as the minimally invasive CABG. There are advantages and disadvantages associated with each approach:

(a) *PCI first followed by CABG:* This approach minimizes the risk of ischemia during minimally invasive bypass to the LAD by providing collateral circulation through the PCI revascularized targets. If PCI is unsuccessful or the result is not satisfactory, the revascularization strategy can be converted to conventional CABG. This approach can also be used in the setting of and acute MI where the culprit vessel is not the LAD. PCI is initially performed to the non-LAD culprit lesion and it is later followed by minimally invasive CABG to the LAD. The disadvantages include the risk of stent thrombosis during discontinuation of antiplatelet therapy during the staged CABG. If antiplatelet therapy is not discontinued, there is an increased risk of bleeding during the staged CABG.

(b) *CABG followed by PCI:* It is the most common approach used for HCR. In our practice, we perform PCI 48 hours after the minimally invasive CABG when the chest tube has been removed. Since minimally invasive CABG can be associated with a lower early patency rate than CABG performed through a median sternotomy, this approach allows for routine angiography of the LIMA to LAD bypass. It provides quality control and confirms the patency of the graft. Dual antiplatelet therapy can be instituted once the risk of surgical bleeding has subsided and continued indefinitely. The presence of a patent LIMA to LAD

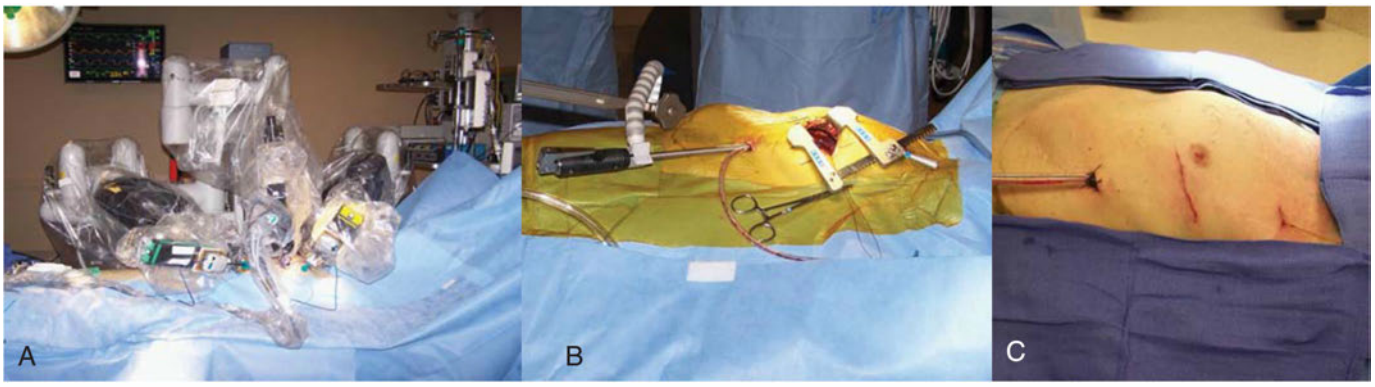


Fig. 51.6. Minimally invasive coronary artery bypass. (**A**) The left internal mammary artery is harvested robotically using the DaVinci System. (**B**) Then a left anterior minithoracotomy is performed in the fifth or fourth intercostal space and a direct hand sewn left internal mammary artery to left anterior descending anastomosis is performed. Note the use of endostabilizer that is placed through the seventh intercostal space port site. (**C**). End result with closed incision and chest tube placed through the seventh intercostal space port site. Note exiting from the third space port site the OnQ pump catheters. They are placed in the subpleural space for postoperative pain control.

graft minimizes the risk of PCI to critical coronary lesions. An example of such lesion is a left main lesion at the bifurcation requiring stenting into the circumflex. The disadvantages of this approach include (a) increased risk of ischemia in non-LAD territories during CABG. (b) If the staged PCI is unsuccessful, the patient is faced with the possibility of requiring conventional CABG or incomplete revascularization.

(c) *Concomitant CABG-PCI:* Both procedures are performed in a single setting under general anesthesia with confirmation of LIMA patency and PCI before chest closure. Even though it is the most attractive and convenient approach for the patient, it is associated with the most challenges. The most important one is the management of antiplatelet therapy with a fresh open wound that may result in increased bleeding. From the infrastructure standpoint, it requires a hybrid operating room or to transfer the patient from the operating room to the catheterization lab. It is also associated with longer operating room times, increased costs, and resource utilization. Hospital and physician reimbursement for both procedures may not be adequate. As a consequence of these challenges, this approach is only performed in a few centers.

The main theoretical advantages of HCR are considered to be shorter hospital stay, rapid rehabilitation, shorter time to return to work, and improved quality of life. To date, no randomized trial comparing HCR with either CABG or PCI has been published. The reported experience with HCR has been reviewed by Bonaros et al. It included 29 studies with a total of 774 patients. Operative mortality was 1.2%. The stenosis of the LIMA to LAD graft was 2.9% and the rate of PCI reinterventions was 14.5% (Table 51.5).

Table 51.5 Published Experience of Hybrid Coronary Revascularization (Modified from N. Bonaros et al.)

Author	Year	Patients	Mortality	LIMA stenosis@ 6 months (%)	PTCA/stent restenosis@ 6 months (%)
Angelini	1996	6	0	n.a.	n.a.
Friedrich	1997	2	0	n.a.	n.a.
Mack	1997	1	0	n.a.	n.a.
Zenati	1999	31	0	0	10
Wittwer	2000	35	0	0	7
Farhat	2000	1	0	n.a.	n.a.
De Canniere	2001	20	0	0	5
Presbitero	2001	42	2	10	n.a.
Riess	2002	57	0	3	24
Cisowski	2002	50	0	2	10
Stahl	2002	54	0	n.a.	n.a.
Bonatti	2005	1	0	n.a.	n.a.
Kiaii	2005	1	0	n.a.	n.a.
Bonatti	2005	1	0	0	0
Davidaviclus	2005	20	0	0	0
Katz	2006	27	0	4	33
Hulusi	2006	17	0	0	18
Srivastava	2006	1	0	n.a.	n.a.
Vassiliades	2006	47	0	1.9	6.6
Gilard	2007	70	1	n.a.	2.3
Bonatti	2007	5	0	0	0
Kon	2008	15	0	n.a.	n.a.
Kiaii	2008	57	0	0	11
Reicher	2008	13	0	n.a.	n.a.
Hoizhey	2008	117	2	4	22
Jansens	2009	1	0	0	0
Shim	2009	1	1	0	100
Del Giglio	2009	2	0	n.a.	n.a.
Gao	2009	10	0	0	0
Total		774	1.2%	2.9%	14.5%

n.a., not available.

Halkos et al. compared by propensity matching 147 patients with triple-vessel CAD treated with HCR patients with 588 patients who were treated with off-pump CABG. The incidence of in-hospital major adverse cardiac and cerebrovascular events (MACCE: death, stroke, and MI) was similar between groups. Length of ICU and hospital stay were similar between the groups. At a mean follow up of 3.2 years, the need for repeated revascularization was higher in the HCR group than in the off-pump CABG group (12.2% versus 3.7%; $P < 0.001$). There was also an increased need for target vessel revascularization in the HCR group (8.8% vs. 3.1%; $P < 0.002$) driven by both lesions in the LIMA to LAD graft and in-stent restenosis. Lesions in the LIMA to LAD graft were more common in the HCR that in the off-pump group (4.8% vs. 1.0%; $P < 0.001$). The 5-year survival was similar between the groups. There were no data reported on angina symptoms or acute coronary events at follow up.

A multicenter NIH observational study is being conducted to evaluate the incidence of death, stroke, MI, and repeat revascularization after HCR or PCI. In addition, to explore the effectiveness and safety of HCR, this study will establish the foundation for a randomized multicenter trial that will help to determine the role of HCR in the treatment of CAD.

OUTCOMES OF CORONARY ARTERY BYPASS GRAFTING

Advancements in percutaneous treatment strategies for CAD including angioplasty and drug-eluting stents combined with more minimally invasive open techniques for revascularization such as off-pump CABG and robotic CABG have selected a different profile of patients to undergo the traditional on-pump CABG. The landscape has shifted toward a patient population that is older and sicker with greater risk for morbidity and mortality. As an outgrowth of the changing patient demographics,

major improvements in patient risk stratification and quality improvement have taken place. Giant strides in understanding patient-specific risks can be attributed to the development of the European System for Cardiac Operative Risk Evaluation (EuroSCORE), the Society of Thoracic Surgeons (STS) online risk calculator, and the Veterans Administration risk score. These combined with institutional quality improvement programs have continued to improve our understanding of CAD and have been invaluable in discussing patient-specific expectations and risks in the perioperative setting. Indeed, despite the increased scrutiny of CABG procedures and the more challenging patient profile, on-pump CABG continues to have improving patient outcomes with a mortality rate below 2% for isolated revascularization procedures.

ON-PUMP CORONARY ARTERY BYPASS GRAFTING VS MEDICAL THERAPY

The indications for surgical revascularization over optimized medical therapy were shaped in the 1970s and 1980s by three major randomized studies. The most recent AHA/ACC guidelines for surgical revascularization are patients with left main disease, left main equivalent (>70% stenosis of proximal LAD and LCx), significant left ventricular dysfunction, intractable angina pectoris despite maximal nonsurgical therapy, and significant three-vessel disease.

More recent randomized studies show that CABG yields up to 44% decrease in total mortality, Q-wave MI, or refractory angina requiring revascularization at a 5-year follow-up in patients with multivessel CAD. A survival advantage for early coronary intervention over medical therapy has also been shown in patients ranging from single vessel to multivessel coronary arterial disease. Undoubtedly, the optimal outcome for patients suffering from coronary arterial disease is the amelioration of ischemia with revascularization combined with both medical and risk factor optimization.

ON-PUMP CORONARY ARTERY BYPASS GRAFTING VS PERCUTANEOUS CORONARY INTERVENTION

The SYNTAX trial, designed as a noninferiority of PCI to CABG, was a randomized trial comprising 1,800 patients with left main or three-vessel CAD. After a follow-up of 12 months, it was noted that MACCE were significantly higher in the PCI group as opposed to the CABG group (17.8% vs. 12.4%; $P = 0.002$). The major contributing factor for this outcome was the need for further revascularization procedures in the PCI arm. As such, the conclusions of the study were that CABG remains the standard of care for these patients. The findings of increased need for revascularization after a PCI in the SYNTAX trial mirrored concerns raised almost a decade earlier by the BARI trial (54% need for revascularization after PCI vs. 8% for CABG in a follow-up of 5 years). A subgroup of the BARI trial that deserves special mention was patients suffering from diabetes mellitus. A statistically significant 5-year survival of 80.6% versus 65.5%, $P = 0.003$ in the CABG group over the PCI group, respectively, was reported. Other studies designed at looking at the validity of PCI for unprotected left main disease confirmed the increased rate of MACCE seen with PCI versus CABG and in addition showed the detrimental additive costs of the need for repeat revascularizations in the PCI group. Recent reports from the New York State Cardiac registry using National Death Index data comparing the relative survival benefits of CABG and PCI with follow-up longer than 5 years confirm that in 7,235 matched pairs of patients, the overall 8-year survival was 78% for CABG and 71.2% for stenting ($P < 0.001$).

ON-PUMP VS OFF-PUMP CORONARY ARTERY BYPASS GRAFTING

The theoretical advantages of off-pump CABG (OPCAB) versus on-pump CABG is all related to the avoidance of the cardiopulmonary bypass circuit. There should be a decreased systemic inflammatory response, a lower degree of myocardial ischemia, and a lower risk of end-organ damage. This theoretical advantage should be seen highest in patients who are at greatest risk (i.e., older, diabetic, chronic renal insufficiency, porcelain aorta). However, there is great controversy and debate in the literature over the two techniques with no clear cut advantage. Initial review of the multitude of prospective, retrospective, and meta-analyses studies conducted on the merits of off-pump (OPCAB) versus on-pump coronary arterial bypass showed no advantage to off-pump revascularization. Subsequently, the proponents of OPCAB have argued that there is a less inflammatory response, myocardial injury, neurologic morbidity, renal insult, equal graft patency, and economic advantage of using off-pump techniques. The ROOBY trial aimed at answering the controversy was conducted as a randomized study of over 2,200 patients receiving either an elective or urgent CABG in the Veterans Affairs population. The primary long-term end point was death from any cause, a repeat revascularization procedure, or a nonfatal MI within 1 year of surgery with secondary end points of completeness of revascularization, graft patency and neurological outcomes. The conclusions of the study showed no differences in survival, use of resources or neurological outcomes. It did, however, report a decreased graft patency rate and worse composite outcome (death or major adverse event) in the OPCAB group. The shortcomings of the trial were quickly pointed out, however, by those prominent in the surgical community (surgeon inexperience with the OPCAB technique, selection bias of low-risk male patients, condition of the aorta—among others) thus adding further fuel to the debate. Despite the evergrowing amount of literature on the topic, the controversy continues to exist. Traditional on-pump CABG continues to have excellent results, but the technique of OPCAB is a tool that all cardiac surgeons should have in their armamentarium.

TIMING OF CORONARY ARTERY BYPASS GRAFTING AFTER AN ACUTE MYOCARDIAL INFARCTION

Although early PCI after an acute MI remains the goal for patients with coronary ischemia, the timing of surgical revascularization after an MI remains controversial. Patients who undergo early (<48 hours) CABG after an acute MI have a higher mortality. This is undoubtedly a direct result of the higher acuity and hemodynamic decompensation of patients in this group. For patients with stable CAD who have had an acute MI, it is commonplace to perform CABG in an elective, outpatient setting. The subset of patients for which the timing of CABG after an MI is debatable is the patients with a high grade lesion not amenable to PCI, the ever increasing population of patients who have been loaded with an irreversible antiplatelet medication prior to a cardiac catheterization, patients with unstable angina, and lastly patients with poor ventricular function. Although there is no consensus statement to timing of CABG after an MI, it is common practice to wait

at least 5 days when possible after a loading dose of irreversible, antiplatelet therapy to reduce the chance of postoperative bleeding. An argument has been made for the use of on-pump but beating-heart CABG strategy to reduce the morbidity and mortality associated with cardioplegic arrest of the myocardium in the acute infarction period.

CHOICE OF BYPASS CONDUITS

There is little doubt about the benefits of using the left internal mammary in situ as a bypass conduit to the LAD artery whenever possible. The use of the right internal mammary artery and the use of both internal mammary arteries in improving long-term survival, especially in the young, nondiabetic patient population have been extensively highlighted. The benefits of total arterial revascularization using the radial artery as the third bypass conduit have been shown to have excellent 4-year patency, above 90% in target-specific arteries and in the experienced surgeons' hands. The availability, resistance to spasm, and relative ease of handling the saphenous vein graft have kept it as the most commonly used conduit following the LIMA. Groups have reported that no difference exists between the 1-year patency of radial arterial grafts and grafts using the saphenous vein. In addition to the above-named conduits, the gastroepiploic arteries, the lesser saphenous vein, and the inferior epigastric artery and others have been described with some success.

CORONARY ARTERY BYPASS GRAFTING IN PATIENTS WITH ISCHEMIC HEART FAILURE

Patients who are often the sickest with the poorest left ventricular function are often the ones with the most to gain from surgical revascularization. The assessment of left ventricular dysfunction and myocardial viability in the previous discussion has identified advanced diagnostic modalities to evaluate these patients.

The STICH trial was conducted as a randomized trial comparing optimal medical therapy alone with medical therapy with the addition of CABG in patients with significant left ventricular dysfunction (ejection fraction ≤35%). Accruing more than 600 patients in each arm, the results showed that the addition of CABG decreased rates of death from cardiovascular causes as well as rate of hospitalization for cardiovascular causes. Specifically, the medical therapy group had a 68% occurrence of the above compared with a 58% occurrence in the CABG group (hazard ratio of 0.81, confidence interval 0.66 to 1.00; $P = 0.05$) in a median follow-up period of 56 months. This may be an underestimation of the true effect, given the 17% crossover rate from those assigned to the medical therapy alone group into the CABG group.



COMPLICATIONS

According to the STS risk adjustment database, the mortality rate for all isolated CABG is 1.9% with the risk of a major complication at 15%. The risk of bleeding requiring reoperation is 1.8%, the risk of a perioperative MI is 0.9%, chance of any neurologic outcome (including coma, stroke, paralysis, transient ischemic attack) is 1.6% with a 1.1% risk of a permanent stroke. A chance of any infection is 1.1%, deep sternal wound infection is 0.2%, and a leg infection is 0.3%. The possibility of renal failure is 3.8% and the possibility of any need for reintervention (operation for valvular dysfunction, graft occlusion, bleeding, and other cardiac/noncardiac operations) is 4.2%. Finally, the chance of postoperative atrial fibrillation is 23%.

FUTURE DIRECTIONS

As surgeons continue to push the borders of technology and minimally invasive surgery with both hybrid procedures (minimally invasive CABG combined with PCI) and totally robotic coronary arterial revascularization, the landscape of patients undergoing on-pump CABG will continue to evolve. Increasingly, patients are being referred for surgery with multiple previous stents and failed angioplasties allowing for minimal room for error in the perioperative period to maintain superb results. In a period of heavy scrutiny on outcomes and large databases with surgeon-specific data available, it is incumbent on the cardiac surgeon to plan out a safe, expeditious, and efficacious operation in a patient-specific manner.

SUGGESTED READINGS

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EDITOR'S COMM NTS

This chapter by Kilic and his colleagues at Ohio State University is a outstanding chapter and discusses multiple controversial areas in cardiac surgery. The section on imaging is comprehensive and certainly demonstrates how coronary disease imaging will change in the future. There are three controversial areas that are mentioned in the chapter that are worth discussing. The first relates to viability. We are stuck as surgeons operating on patients with very poor ventricular function. This is certainly a major change in the last two decades. The big issue is deciding to whom to operate on. Our basic rule here is with ischemic cardiomyopathy is there needs to be evidences of viability and the arteries must be graftable. One can operate on very diffuse disease in patients with decent function but in those patients with major dysfunction one must have excellent distal targets.

The second area that is discussed is hybrid revascularization. This is an important area and certainly is being practiced in multiple institutions. There is now a trial being worked out for the CTSN sponsored by NHLBI related to hybrid revascularization. I think we are going to have to learn a great deal more about this area and determine which patients will benefit from these technologies. Finally, the area of off-pump versus on-pump is a continuing area of controversy. I believe the bottom line relates to graft patency. If one can get outstanding graft patency with off-pump technology, then that is an appropriate technique. If one cannot guarantee it, then their on-pump technology has certainly compared favorably the off-pump approach. Certainly off-pump has less bleeding and perhaps a shorter hospital stay but there are really no other major safety differences in most of the randomized trials. I should mention these areas, latter cases will be highlighted by the authors.

ILK

52 Off-Pump Coronary Artery Bypass Surgery

W. Brent Keeling, Michael E. Halkos, and John D. Puskas

INTRODUCTION

Over the past decade, there has been increasing interest in performing coronary artery bypass grafting (CABG) without the use of cardiopulmonary bypass. The growth in off-pump coronary artery bypass surgery (OPCAB) has been largely driven by increasing recognition of the deleterious effects of cardiopulmonary bypass and the desire to avoid the diffuse inflammatory response, multiorgan dysfunction, and neurocognitive complications that may follow. Increasing clinical experience with OPCAB has allowed analysis of outcomes following the procedure and demonstrated improved clinical outcomes in both prospective and large, risk-adjusted, retrospective comparisons among various patient populations.

Approximately 20% of CABG cases performed annually in the United States are performed off-pump and some centers report a significantly higher percentage of OPCAB cases. Increasing technical proficiency has been facilitated in large part by continued improvements in exposure and retraction techniques and the development of specialized stabilizers and positioners, which allow experienced off-pump surgeons to conduct complex multivessel coronary revascularizations that were not previously feasible without the use of cardiopulmonary bypass. With current positioners and stabilizers, OPCAB can be performed in the vast majority of patients needing coronary revascularization.

Even among cardiovascular surgeons not routinely performing coronary revascularization off-pump, there are newly recognized clinical scenarios, such as patients with severe atherosclerosis of the ascending aorta, for whom the use of OPCAB techniques may be particularly beneficial. OPCAB continues to be a facilitating technology for surgeons developing and promoting minimally invasive approaches to coronary revascularization. OPCAB has, therefore, evolved into a component of the armamentarium of the modern cardiovascular surgeon.

PREOPERATIVE CONSIDERATIONS

Although OPCAB is now performed routinely in some clinical practices, the tendency to automatically or blindly assign patients to OPCAB should be resisted. Preoperative factors favoring OPCAB must be weighed carefully against relative or absolute contraindications for the procedure. Rational and systematic consideration of these factors maximizes the likelihood of a technically successful procedure while limiting the chance for adverse events.

SURGEON

The individual surgeon's training, experience, and attitudes toward OPCAB are the starting point from which any preoperative decision making should begin. OPCAB presents a unique set of technical challenges to the cardiovascular surgeon, who otherwise would be operating in a motionless and bloodless field. It has been estimated that OPCAB is feasible in up to 95% of patients presenting for primary CABG.

Individual surgeon experience in OPCAB is an important determinant in patient selection for OPCAB. The unique technical challenges of OPCAB grafting and its relative unfamiliarity have raised concern that adoption of OPCAB may lead to poorer outcomes during each surgeon's "learning curve." With careful patient selection, OPCAB surgery can be gradually assimilated into clinical practice while preserving and ultimately improving clinical outcomes. Very early in a surgeon's experience, it is reasonable to exclude patients with depressed left ventricular function, left main disease, and those requiring multiple lateral wall grafts. With experience, more complex and technically challenging cases can be performed safely off-pump. Over time, OPCAB can be applied to a broad spectrum of clinical settings, including patients with advanced age, multivessel disease, depressed left ventricular function, left main disease, and complete arterial revascularization. Gradual assimilation of OPCAB thereby develops surgeon familiarity and comfort with the technique, allowing its broader application to an increasing pool of patients who derive benefit from avoiding cardiopulmonary bypass.

Patient

Patients for whom OPCAB is inappropriate are those in cardiogenic shock, those suffering from recurrent ischemic arrhythmias, and those with thoracic anatomy that severely limits the ability to rotate the heart, for instance, those with pectus excavatum or previous left pneumonectomy (see Table 52.1). Relative contraindications include intramyocardial coronary arteries, and small or diffusely calcified distal target vessels. These targets may be safely bypassed off-pump only with the benefit of considerable experience. On the other hand, patients with left main coronary lesions and recent myocardial infarction can safely have coronary revascularization performed off-pump and should be considered candidates for the procedure.

The preoperative evaluation of the patient proceeds with a complete history and physical exam. If radial artery harvest is contemplated, patients with inconclusive Allen's tests undergo radial and ulnar artery duplex examinations. Criteria for preoperative carotid duplex examination include left main disease, peripheral vascular disease, carotid bruits, history of CVA, history of heavy tobacco use, and age >65 years. If significant carotid disease is detected, further work-up is pursued and staged or concomitant carotid endarterectomy is performed.

The role of atherosclerotic lesions of the ascending aorta in perioperative stroke has become increasingly evident over the past decade, and it is in this group of patients where OPCAB may be of greatest benefit. OPCAB surgery combined with clampless proximal anastomosis techniques is a uniquely valuable alternative for patients with severe atherosclerosis of the ascending

Table 52.1 Contraindications to Off-Pump Coronary Artery Bypass

Absolute contraindications
Cardiogenic shock
Ischemic arrhythmias
Anatomic factors preventing rotation of the heart
Previous left pneumonectomy
Severe pectus excavatum
Relative contraindications
Intramyocardial coronary arteries
Small or calcified coronary arteries

aorta in which aortic clamping may pose a significant risk for atheroembolic stroke. Routine intraoperative use of epiaortic ultrasound is an invaluable tool for identifying patients with severe atherosclerosis of the ascending aorta so that their stroke risk can be effectively managed.

INTRAOPERATIVE MANAGEMENT

The planned conduct of OPCAB introduces a number of new patient care issues to the staff of an operating room. An important underlying concept to ensuring patient safety and the smooth conduct of the procedure is communication between the surgeon, operating room staff, and anesthesiologists. Excellent communication between the surgeon and the anesthesia provider is paramount in order to ensure intraoperative hemodynamic stability and to coordinate any departure from the planned order of distal or proximal anastomoses.

ANESTHESIA

Several aspects of anesthetic management of patients during OPCAB are common to the management of patients having conventional CABG. All patients undergoing OPCAB require invasive monitoring. At a minimum, an arterial line and central venous line are required. Pulmonary artery catheters are used routinely. Monitoring of pulmonary artery pressures can be particularly useful during retraction of the heart for the construction of distal anastomoses, as pulmonary artery pressure elevations are frequently the first sign of hemodynamic compromise prior to ischemic arrhythmias or cardiovascular collapse. Transesophageal echocardiogram is utilized sparingly but may be of use in order to identify areas of hypokinesis or significant mitral regurgitation following periods of regional myocardial ischemia.

In addition to the routine monitoring and safe anesthetic induction desired in all CABG procedures, there are a number of anesthesia management issues that are specific to OPCAB. In contrast to conventional CABG procedures, maintenance of normothermia is critically important throughout the case, as the ability to actively rewarm the patient by cardiopulmonary bypass is forfeited. Significant hypothermia adversely affects coagulation, is arrhythmogenic, and delays postoperative extubation. Efforts to maintain normothermia should therefore begin prior to induction. At our institution, a convective air system cycles warm air continuously to warm the patient throughout the procedure (Bair Hugger; Arizant Healthcare, Eden Prairie, MN).

The major challenge in the intraoperative anesthetic management of patients undergoing OPCAB is maintenance of hemodynamic stability during the elevation and retraction of the heart necessary to obtain exposure of coronary targets. Significant alterations in blood pressure and cardiac output may occur if preparatory steps are not taken. This can occur with rightward retraction of the heart for lateral wall exposure. Acutely, this is typically related to decreases in preload and left ventricular filling that occur as the vena cavae, right atrium, right ventricular outflow tract, and pulmonary veins may be compressed with this maneuver. An effective first-line treatment for this response is the administration of intravenous fluids. An assessment of the patient's intravascular volume status is made prior to manipulation of the heart and preload is subsequently optimized. In order to compensate for the acute changes in preload that occur during cardiac manipulation, placement of the patient in steep Trendelenburg and reverse Trendelenburg positions can rapidly alter preload conditions to favor different hemodynamic states. Patient positioning is particularly useful when moment-to-moment changes in blood pressure are desired, for instance when rightward displacement of the heart is anticipated (Trendelenburg position) or when moderate hypotension is desired prior to placement of a partial occluding vascular clamp on the ascending aorta (reverse Trendelenburg position). Often, significant pharmacologic manipulation of systemic blood pressure can be avoided with bed positioning alone. Releasing the right-sided pericardial traction sutures and elevating the right limb of the sternal retractor on rolled towels are routine and valuable maneuvers to facilitate exposure and hemodynamic stability. Opening the right pleural cavity or releasing pericardial attachments along the diaphragm can also prove useful during rightward displacement of the heart.

As experience with OPCAB continues to evolve, the use of inotropes and vasopressors to maintain hemodynamic stability has been liberalized. Early in our experience, adjustment of preload conditions with intravenous fluid was the primary means by which stable blood pressures were maintained. In order to limit the intravenous fluid load given to patients intraoperatively, low-to-moderate doses of alpha agents such as norepinephrine are now routinely used. This practice has led to more favorable volume status in the postoperative period and has not had untoward effects on myocardial protection. It is unusual for patients to have a significant inotrope or vasopressor requirement beyond the immediate perioperative period.

Familiarity with the OPCAB procedure on the part of the anesthesia team is critical to the safe conduct of the operation. Patient care issues specific to OPCAB are thereby anticipated and addressed before they adversely affect the outcome of the procedure. Communication between the anesthesia team and the surgeon prior to and during the operation is important, particularly when changes in the surgeon's operative plan occur.

SURGERY

The surgeon should come to the operating room with an operative plan that optimizes the likelihood for a successful outcome but also should remain flexible enough to change the operation as intraoperative findings and events dictate. This is one area where OPCAB and conventional CABG procedures differ significantly. OPCAB procedures frequently require significant intraoperative decision making that may result in departure from the routine. The following describes how a typical OPCAB case is performed in a patient requiring multivessel bypass.

Preparation

All patients for whom OPCAB is planned receive an aspirin suppository following

induction of anesthesia and prior to being prepped for surgery. In our practice, we administer aspirin perioperatively and dual antiplatelet therapy (aspirin and clopidogrel) in the postoperative period for platelet inhibition. Clopidogrel is administered within 4 hours after surgery if chest tube output is <100 cm^3/hour for 4 hours.

The chest is opened in the usual manner through a midline sternotomy incision. Left and/or right internal mammary arteries (LIMA/RIMA) are harvested with the use of an upward-lifting retractor. Other radial artery or saphenous vein conduits are harvested simultaneously by endoscopic techniques as needed. Prior to vein or radial artery harvest, patients receive 5,000 units of heparin; this is important to avoid fibrin strands in endoscopically harvested conduits. Prior to division of IMA conduits from the chest wall, heparin is given (180 units/kg) to achieve a target activated clotting time of >350 seconds. Heparin is re-dosed every 30 minutes to maintain this level of anticoagulation. A sternal retractor (OctoBase; Medtronic, Inc., Minneapolis, MN) designed to act as a platform for OPCAB stabilizers and positioners (Octopus Evolution AS and Starfish EVO, Medtronic, Inc.; ACROBAT and EXPOSE, Maquet, Inc., Wayne, NJ) is placed in the chest. A wide inverted "T"-shaped pericardiotomy is performed, dividing the pericardium along the diaphragm. The phrenic nerves should be identified and protected during pericardiotomy. It is important to divide the pericardium to the apex to facilitate cardiac displacement. The left pleural space is opened widely; opening the right pleural space will facilitate cardiac displacement, but has been less necessary since the introduction of cardiac positioning devices and is infrequently performed in the absence of RIMA harvest. Care is taken during the dissection to clip any large vessels encountered and to avoid the phrenic nerves. It is also important to divide the diaphragmatic muscle slips, which insert on the right side of the xyphoid in order to allow elevation of the right sternal border, creating space for rightward cardiac displacement. Placement of two rolled towels under the right limb of the retractor elevates the right sternal edge, allowing the heart to be positioned toward the right without compression against the sternum or retractor.

The most important traction suture is a deep posterior pericardial suture placed approximately two-third of the way between the inferior vena cava and left pulmonary vein at the point where the pericardium reflects over the left atrium. Care should be taken with superficial placement of this suture to avoid the underlying descending thoracic aorta, esophagus, left lung, and pulmonary veins. The suture is covered with a soft rubber catheter to prevent laceration of the epicardial surface of the heart. The purpose of this deep traction suture is to elevate the heart up and out of the pericardial well to facilitate exposure of the coronary targets. When this suture is retracted toward the patient's left hip, it elevates the base of the heart toward the ceiling and points the apex vertically with remarkably little change in hemodynamics. When the deep pericardial traction suture is retracted toward the left shoulder, the heart rotates from left to right. This exposure can occasionally be obtained without the deep stitch by placing a warm laparotomy pad underneath the heart.

Epiaortic ultrasound is performed on all patients prior to manipulation of the aorta. This procedure adds only 1 to 2 minutes to the operative time and is the most sensitive means to detect atherosclerotic lesions of the ascending aorta intraoperatively. More importantly, it allows the surgeon to individualize placement of aortic clamps and proximal anastomosis devices to minimize the risk of atheroembolism. Its use has led to the reduction of postoperative stroke in patients at high risk for atheroembolism. A finding of grade IV or grade V atherosclerosis precludes application of a clamp to the ascending aorta (Table 52.2). The use of a clampless facilitating device that allows construction of a handsewn proximal anastomosis (Heartstring; Guidant Corporation, Indianapolis, IN) is possible when relatively uninvolved areas of the ascending aorta are detected by epiaortic ultrasound. When atherosclerotic involvement is very diffuse, a "no-touch" technique is employed with the innominate artery, LIMA, or RIMA utilized for proximal bypass graft in-flow. It is often necessary in these circumstances to construct a "Y" or "T" graft off either IMA for the purposes of arterial inflow, thus obviating aortic manipulation (Fig. 52.1).

Table 52.2 Epiaortic Ultrasound Grading System

Grade 1—Intimal thickening <2 mm
Grade 2—Intimal thickening >2 mm but <3 mm
Grade 3—Intimal thickening >3mm but <5 mm
Grade 4—Intimal thickening >5 mm
Grade 5—Presence of mobile disease

The heart is allowed to roll with gravity into the left or right chest, facilitated by table rotation or tension on traction sutures. With proper exposure and retraction, the heart should never be compressed against the sternum or pericardium. Right pericardial traction sutures are released when exposing the left side of the heart, and similarly left pericardial traction sutures are released when exposing the right coronary artery. Pericardial sutures on both the right and left sides are never under tension simultaneously when displacing the heart to expose coronary targets. Gentle application of these techniques maintains stable hemodynamics while providing excellent exposure.

When grafting of the lateral wall or inferior wall is planned, a cardiac positioning device (Starfish EVO; Medtronic, Inc.; ACROBAT and EXPOSE, Maquet, Inc.) is also used (Figs. 52.2 and 52.3). This device uses suction to attach to the epicardial surface of the heart and elevates and displaces the heart to provide exposure of coronary targets with little hemodynamic compromise. Its hemodynamic advantage comes from the fact that it rotates the heart along the axis of the vena cavae while elevating it out of the pericardial well, thereby avoiding compression or kinking of the atria, vena cavae, and pulmonary veins. This device aids in exposure and presentation, especially in cases of cardiomegaly and depressed left ventricular function.

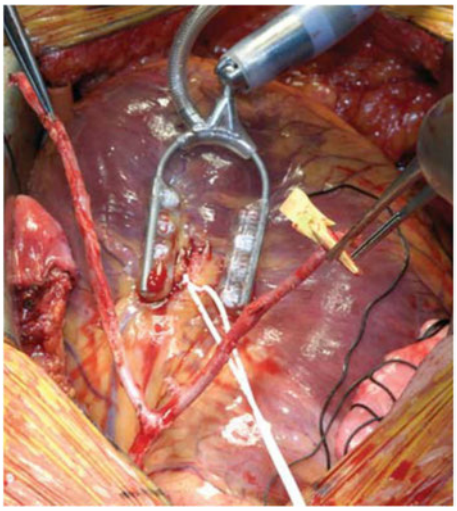


Fig. 52.1. Construction of a "Y" arterial graft using the "no touch" technique for arterial inflow.

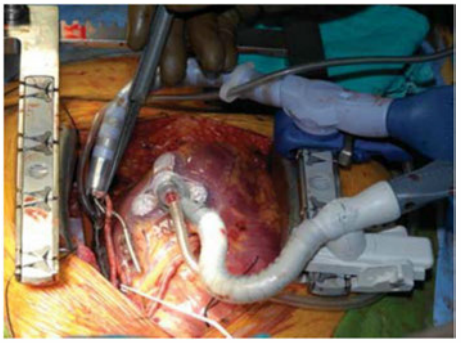


Fig. 52.2. Cardiac displacement and presentation of diagonal branches of the left anterior descending artery. The cardiac positioner is applied to the apex or left lateral wall of the heart and lifts the heart out of the pericardial well and rolls it to the right allowing the heart to roll under the right sternal border. *Left*-sided pericardial traction sutures and the deep stitch are placed under tension. The coronary stabilizer reaches across the heart from the right side of the sternal retractor, aiding in both the presentation and stabilization of the lateral wall and diagonal coronary targets.

It may be applied anywhere on the epicardial surface of the heart and is frequently moved away from the apex to facilitate exposure of various coronary artery targets.

On rare occasions when bradycardia and cardiomegaly coexist, ventricular distension hinders effective cardiac displacement. In this situation, temporary epicardial atrial pacing may significantly reduce cardiac size by decreasing diastolic filling time and improving target vessel exposure.

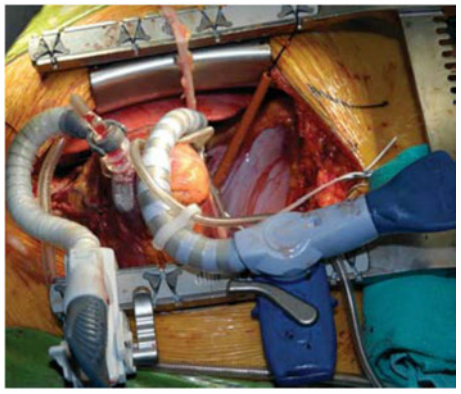


Fig. 52.3. Cardiac displacement and presentation of inferior wall coronary targets. The cardiac positioner is applied to the apex of the heart and elevates the heart out of the pericardial well, exposing the inferior wall of the heart. The deep traction stitch is pulled toward the patient's feet to assist in lifting the base of the heart out of the pericardial well.

Sequence of Grafting

In OPCAB surgery, the chosen sequence of grafting to maintain hemodynamic stability and avoid critical ischemia is one of the most important factors in the success of the operation. As a general rule, the collateralized vessel or vessels are grafted first and then reperfused by performing the proximal anastomoses or releasing flow through the IMA. The last coronary target grafted is the collateralizing vessel(s). This strategy avoids interrupting vital flow from the collateralizing vessel to the collateralized territory until after the collateralized vessel has been grafted.

At times, the proximal anastomoses may be performed early in the operative sequence to aid in early reperfusion of a collateralized vessel. Performing the proximal anastomoses first may make estimation of graft length more difficult. The anticipated length is measured with a silk tie so that conduit can be appropriately cut in this circumstance. If the LIMA to left anterior descending (LAD) artery graft must be performed first, it is necessary to leave a long mammary pedicle to avoid tension on the LIMA anastomosis during subsequent displacement of the heart to expose other target vessels.

A preferred sequence of grafting is as follows:

1. Perform the anastomosis to the completely occluded or most collateralized vessel first. The collateralizing vessel can then be safely grafted, thus minimizing myocardial ischemia.
2. The LIMA to LAD artery anastomosis should be performed first if the LAD is collateralized or in cases of tight left main stenosis. This anastomosis is performed last when the LAD is the collateralizing vessel.
3. The proximal anastomosis can be performed first or early after the distal anastomosis if the target is a critical, collateralized vessel. This allows simultaneous perfusion during subsequent occlusion of the collateralizing vessel and minimizes overall myocardial ischemia.
4. Beware of a large right coronary artery with moderate stenosis. The right coronary artery, particularly if large and dominant, can cause significant problems when occluded during OPCAB. Acute occlusion of a moderately stenotic right coronary artery may lead to severe hemodynamic compromise secondary to bradycardia. The surgeon must be prepared to use an intracoronary shunt or epicardial pacing promptly to correct bradycardia and interrupt events leading to cardiovascular collapse.
5. Beware of mitral regurgitation during OPCAB. Prolonged displacement of the heart especially when grafting lateral wall vessels in the setting of mild-to-moderate mitral regurgitation can lead to severe hemodynamic compromise. Attempts should be made to address acute ischemic mitral regurgitation early in the operative sequence by grafting and perfusing the culprit vessel responsible for presumed papillary muscle dysfunction or utilizing intracoronary shunts to allow for continuous distal perfusion.
6. Most importantly, graft sequence should be individualized, depending on anatomic patterns of coronary occlusion and collateralization, myocardial contractility, atherosclerosis of the ascending aorta, conduit availability, and graft geometry.

Cardiac Displacement and Presentation of Coronary Targets

It is important to understand that the cardiac displacement techniques for exposure of the inferior and lateral coronary arteries are different. The lateral wall vessels are approached by rolling the apex of the heart toward/under the right sternal border. As previously described, the traction sutures on the right pericardium are released and the right pleural cavity can be opened. The left-sided traction sutures are pulled up taut on the sternal retractor and the table is rotated sharply to the right to assist in rolling the heart under the right sternal border. The deep stitch is pulled toward the patient's left shoulder and secured to the sternal retractor. The coronary stabilizer is mounted on the right side of the sternal retractor and its arm reaches across the heart, aiding in both the presentation and stabilization of the obtuse marginal coronary arteries (Fig. 52.2). The cardiac positioner may be applied to the left lateral wall of the heart rather than the apex, as needed.

For the inferior wall vessels, such as the posterior descending artery, left ventricular branch of the right coronary artery, or posterolateral obtuse marginal branch, the deep traction stitch is pulled toward the patient's left hip or directly caudad and clamped to the drapes (Fig. 52.3). The coronary stabilizer is attached to the left limb of the sternal retractor. The patient is placed in Trendelenburg position with the bed tilted to the patient's right. The base of the

heart is elevated and the apex is oriented vertically toward the ceiling. The positioner may be applied to the apex of the heart and helps to elevate the heart.

In contrast to targets on the lateral and inferior walls, the anterior wall vessels (LAD and diagonals) are exposed with very little manipulation of the heart. The deep traction stitch is secured to the drapes on the patient's left side, and the coronary stabilizer is brought over the anterior wall from the caudal or left side of the sternal retractor. An apical positioner is usually not necessary for grafting of the anterior wall. Care is taken to divide the pericardium to allow the LIMA pedicle to fall posteriorly into the left chest, medial and posterior to the apex of the left lung.

Coronary Stabilization and Grafting

The current generation of coronary stabilization devices rely on suction rather than compression to maintain epicardial tissue capture. This characteristic allows the device to achieve coronary stabilization at the mechanical median of the cardiac cycle, rather than compressing the cardiac chambers excessively. Thus, stabilization is maintained while mechanical interference with ventricular function is minimized. Once the device is applied, a few seconds may be needed to ensure hemodynamic stability. If hemodynamics are compromised, the degree of compression should be reduced and the mechanical median of the cardiac cycle should be more clearly identified by allowing the stabilizer arm to become flexible while still maintaining suction. The suction is maintained to avoid losing tissue capture. After the appropriate position for the stabilizer arm is determined, it is tightened once more. The malleable pods of the stabilizer allow the surgeon to spread the epicardium adjacent to the coronary targets, significantly improving visualization of the coronary artery. Epicardial fat retractors are rarely necessary.

After optimal exposure is obtained, a soft silastic vessel loop (Quest Medical, Allen, TX) is placed proximally around the target vessel for occlusion (Fig. 52.4). Care should be taken when the vessel loop is placed to avoid entering the ventricle or damaging epicardial veins. When this occurs, a superficial epicardial suture generally stops bleeding. The vessel loop may be directed out of the surgeon's field of view with the aid of a loose pericardial suture acting as a pulley (Fig. 52.5).

When the target vessel is poorly collateralized, an interval of test occlusion lasting 2 to 5 minutes followed by reperfusion for a similar interval can be used to determine how regional myocardial ischemia will be tolerated once the anastomosis is underway. This gives the surgeon some measure of assurance prior to creating an arteriotomy. Once the distal anastomosis is underway, it is critical for the anesthesia team to continuously communicate with the surgeon. Changes in hemodynamics should be quickly addressed and arrhythmias promptly treated with epicardial pacing or an intracoronary shunt.

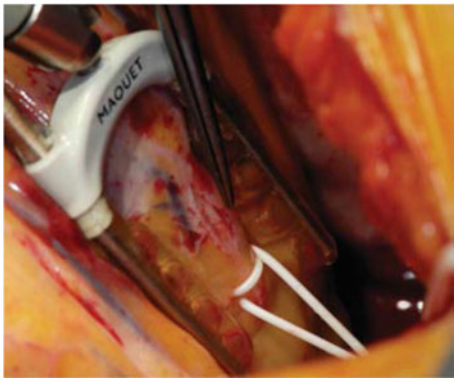


Fig. 52.4. Dissected coronary artery with the silastic stitch and coronary stabilizer in position.

The target vessel is occluded by applying the minimum necessary tension on the vessel loop. This tension is directed in such a manner that the vessel is lifted out of surrounding epicardial fat, further improving exposure of the target. The target vessel is opened with a coronary knife and the arteriotomy is extended with coronary scissors. The field is kept free of blood by dispersing retrograde bleeding from the distal end of the arteriotomy with a humidified CO_2 blower (ClearView Blower/Mister; Medtronic, Inc.). It is important that the surgeon's assistant blow on the target only when the surgeon is placing the needle through the tissue of the conduit or target vessel so as to minimize the potential trauma to the intima of the target vessel that can occur with excessive use of the blower. Excellent visualization is critical for a precise anastomosis. Magnification of 3.5×, a headlight, and Castro viejo needle drivers are used for all anastomoses. 8-0 monofilament suture is used for all distal anastomoses to optimize precision, unless severe calcification or thickened conduit mandates use of a heavier needle.

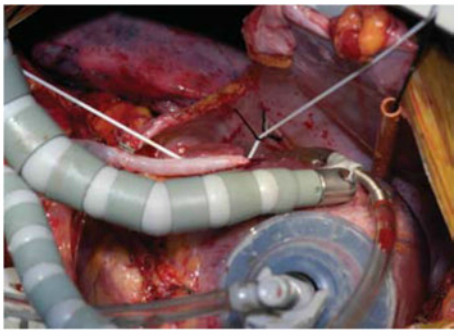


Fig. 52.5. Loose pericardial suture that allows for displacement and correct tension for retraction on the silastic stitch.

Myocardial Protection

Myocardial protection during OPCAB has evolved as techniques for performing the procedure have been refined. Early on, before the development of suction-based stabilizers, intermittent pharmacologic arrest and profound bradycardia were induced during the procedure with adenosine and short-acting β-blockers. In addition to reducing the motion of the target vessel, this strategy achieved some degree of myocardial protection by reducing myocardial oxygen demand. These maneuvers have largely passed from clinical practice with the widespread application of modern suction-based coronary stabilizers and positioning devices.

A number of other myocardial protection strategies developed early in the experience of OPCAB remain in routine use. Maintenance of systemic blood pressure by optimizing preload conditions and the use of vasopressors are part of careful anesthetic management. During target vessel occlusion, however, this practice is an important component of myocardial protection as perfusion to ischemic myocardium via collaterals is dependent on this perfusion pressure. Another myocardial protection strategy that may be taken for granted is the careful use of traction sutures, apical heart positioners, and coronary stabilizers that provide adequate exposure of the target vessel without excessively compressing the cardiac chambers and causing undue hemodynamic compromise. When ideally placed, these devices do not interfere with the cardiac cycle, vasopressor requirements are minimized, and overall myocardial oxygen demand is minimized. This careful positioning of the heart should be considered a routine strategy for myocardial protection. Finally, selection of the order in which distal anastomoses are constructed during a multivessel OPCAB procedure is another strategy by which the surgeon can limit the degree of regional ischemia to which the heart is subjected. Occlusion of a collateralized target vessel

first in the operative sequence allows this territory to be perfused via collaterals during construction of the anastomosis. Subsequently, occluded territories can then be perfused via reversed flow in the collaterals from completed grafts.

Construction of one or more proximal anastomoses early in the operative sequence is an adjunct to the thoughtful selection of the order in which distal anastomoses are constructed. Construction of a proximal anastomosis prior to the first distal anastomosis allows immediate reperfusion of the ischemic territory following target vessel occlusion and construction of the distal anastomosis. If this target vessel is collateralized, then a subsequently constructed distal anastomosis to the collateralizing target vessel can be performed with the benefit of reversed collateral flow originating from the first bypass graft. An advantage of grafting the LAD with an in situ LIMA pedicle first in the operative sequence is that this anastomosis requires minimal lifting of the heart, and subsequent target vessels can be exposed and grafted with the benefit of collateral flow from the LIMA to LAD anastomosis without interrupting the operative sequence to perform a proximal anastomosis.

An intracoronary shunt may be placed if significant hemodynamic compromise occurs following target vessel occlusion despite the use of the routine measures described above. The shunts (ClearView Intracoronary Shunts; Medtronic, Inc.) range in size from 1.0 to 3.0 mm. When appropriately sized for the target vessel, they are easily placed and removed and provide significant flow and a reasonably bloodless field. Intracoronary shunts may be particularly useful with large right coronary arteries and bradyarrhythmias, intramyocardial vessels where the placement of an occlusive vessel loop may be hazardous, and with critical anatomy where occlusion of an important collateralizing vessel may lead to global myocardial ischemia and hemodynamic collapse. The shunt is removed and the coronary artery is de-aired prior to tying the suture on the distal anastomosis and flow is re-established.

Proximal Anastomoses

Proximal anastomoses to the disease-free aorta are routinely performed with an aortic partial occlusion clamp. The systolic blood pressure is lowered to <95 mmHg prior to application of the clamp. Once the clamp is applied, aortotomies are made with a 4.0-mm aortic punch. Vein graft anastomoses are created with 6-0 monofilament suture and arterial grafts with 7-0 monofilament suture. Any graft taken as a "T" off an IMA is anastomosed with 8-0 monofilament. The aortic root is de-aired through the most anterior anastomosis after the clamp is removed, prior to tying this suture. The vein grafts are kept occluded until they are de-aired with a 25-gauge needle. Arterial grafts are not punctured but are allowed to backbleed prior to clamp removal.

As described previously, placement of the partial occluding aortic clamp is guided by the results of routine epiaortic ultrasound scanning. Clamping is not performed in the presence of diffuse grade III or any grade IV or V atherosclerotic involvement of the ascending aorta. When an uninvolved segment of the ascending aorta can be identified that would yield acceptable graft geometry, we typically use a clampless proximal aortotomy system (Heartstring; Maquet, Inc.) that allows creation of a beveled, hand-sewn anastomosis. If atherosclerotic involvement of the ascending aorta is diffuse, precluding use of the Heartstring device, the proximal anastomosis is taken off of an IMA pedicle. Alternatively, a proximal device system can be utilized but many current iterations requires creation of the proximal anastomosis prior to the distal anastomosis, so graft length must be judged accordingly.

After completion and reperfusion of all grafts, protamine is administered (0.75 to 1.0 mg/kg) to partially correct the activated clotting time to approximately 150 seconds. As hemostasis is being achieved, chest drains are placed, one in each open pleural space and one in the mediastinum. Temporary epicardial pacing wires are placed only if the patient requires epicardial pacing immediately prior to chest closure and are rarely used. The chest is closed in the standard manner with sternal wires and running absorbable sutures in the fascia, subcutaneous, and subcuticular layers.



POSTOPERATIVE MANAGEMENT

The postoperative care of patients who have undergone OPCAB surgery is similar in many respects to that of patients undergoing conventional CABG surgery. The requirement for close monitoring of patient cardiopulmonary status, renal function, and chest tube drainage is similar. However, there are a number of important differences that caregivers must be aware of in order to take advantage of the opportunity for expedited care that OPCAB surgery can offer.

Patients who have undergone OPCAB surgery have a decreased need for inotropic support in the postoperative period, most likely because of avoidance of global ischemia and reduced myocardial stunning. Postoperative hemorrhage and transfusion requirements are reduced in OPCAB patients because of reduced fibrinolytic pathway activation and coagulation factor and platelet consumption. Patients who have undergone OPCAB surgery are in a relatively hypercoagulable state compared with the coagulopathy that is typical following cardiopulmonary bypass. This state has the potential to adversely affect graft patency in the postoperative period. As described earlier, we address this preoperatively with the administration of aspirin following induction of anesthesia. Following surgery, we continue aspirin administration daily as with patients undergoing conventional CABG surgery. Dual antiplatelet therapy is continued for 6 months (albeit in the absence of a strong evidence base for this practice); aspirin therapy is continued for life, as per guidelines.

OPCAB patients have a decreased need for mechanical ventilation postoperatively. To realize this benefit for the patient and the health system, caregivers should be immediately prepared to wean and extubate patients as their need for mechanical assistance is diminished. With appropriate anesthesia planning and staffing, patients can generally be extubated in the operating room following an OPCAB procedure or within 30 to 90 minutes of arrival in the intensive care unit. When this is not feasible, clinical pathways that set objective criteria and goals facilitate the timely progression of ventilator weaning and extubation that minimizes patient exposure to ventilator-related complications and maximizes efficiency and cost-effectiveness.

CLINICAL OUTCOMES FOLLOWING OFF-PUMP CORONARY ARTERY BYPASS SURGERY

Patient outcomes following OPCAB have been studied by several prospective randomized studies. A consistent finding among most of these studies has been that OPCAB in experienced hands is a safe and efficacious method to perform coronary revascularization. The magnitude of the overall clinical benefit to the patient from avoiding cardiopulmonary bypass remains a point of contention.

Exposure of circulating blood to the large foreign surface of a cardiopulmonary

bypass circuit leads to the broad activation of plasma protein systems and cellular blood components, which triggers a whole body inflammatory response, coagulation cascade activation, and fibrinolytic activity. As expected, avoidance of cardiopulmonary bypass by OPCAB is associated with a decreased inflammatory response following surgery as measured by neutrophil activity, cytokine levels, and complement activation.

OPCAB presents unique technical challenges to the surgeon who must master the exposure and stabilization techniques that make beating heart surgery possible. Because of this, graft patency in patients undergoing OPCAB has been an area of concern throughout the development of the field. In the largest single-center randomized study in which graft patency was systematically studied, there were no differences in patency rates in any coronary distribution for patients undergoing OPCAB or conventional CABG. A smaller study subsequently demonstrated reduced patency rates for patients undergoing OPCAB 3 months following surgery. More recent data from a multi-institutional trial showed decreased patency rates for patients who underwent OPCAB at 1-year follow-up. Individual operator variability may explain the differences between these studies, as surgeon experience and expertise with off-pump techniques have been questioned in the latter trial. Though there is a significant learning curve for OPCAB, through careful patient selection, this period can be managed and safely traversed while maintaining excellent clinical outcomes.

OPCAB offers certain benefits in postoperative outcomes, specifically shorter hospitalizations and periods of mechanical ventilation and potentially fewer blood transfusions without compromising long-term survival. Accordingly, outcomes following OPCAB for high-risk subpopulations have demonstrated the greatest improvement over on-pump surgery. For example, OPCAB patients with poor left ventricular function have been observed in several large observational studies to have lower morbidity and mortality. Patients with advanced age comprise another high-risk subgroup, which has increasingly been found to benefit from off-pump coronary revascularization. Other high-risk patients potentially benefiting from OPCAB include female patients and patients with obesity, low body mass index, diabetes mellitus, and renal insufficiency.

FUTURE DIRECTIONS FOR OFF-PUMP CORONARY ARTERY BYPASS SURGERY

At its inception, OPCAB surgery was performed rarely and only by a few surgeons and only on patients with certain limited coronary anatomies.Since that time, OPCAB has evolved into a routinely performed operation that can be applied to virtually the entire spectrum of patients requiring coronary revascularization. OPCAB surgery is now well accepted to have specific advantages to the patient compared with conventional CABG and is becoming the gold standard of care for certain clinical situations, such as patients with heavy atherosclerotic disease of the ascending aorta. For many surgeons, OPCAB surgery has become an important component of the armamentarium with which to manage a wide array of clinical challenges.

OPCAB surgery has matured and been accepted by the clinical community to the extent that it has itself become a facilitating technology for further advancements in cardiovascular surgery. Minimally invasive approaches to surgical coronary revascularization are now by definition OPCAB procedures. These evolving procedures will continue to be dependent on OPCAB techniques to construct anastomoses. At our institution, robotic techniques for LIMA harvesting are being combined with nonrib spreading mini-thoracotomies through which minimally invasive coronary revascularization is performed to the anterior wall. This procedure is potentially extending the benefits of LIMA to LAD grafting to patients who would otherwise not be referred for CABG.

Application of OPCAB to patients with multivessel disease is also performed in combination with percutaneous coronary intervention (PCI) of other territories by combining minimally invasive LIMA-LAD grafting with PCI of non-LAD vessels. Hybrid procedures are being evaluated rigorously to determine what subpopulation of patients with multivessel disease may benefit from this approach. Patients with multivessel disease currently being treated with percutaneous techniques alone represent a group for whom hybrid procedures may increasingly be used.

Off-pump coronary artery bypass is a useful strategy for coronary revascularization for the modern cardiothoracic surgeon. Certain at-risk groups of patients demonstrate improved short-term and equivalent long-term outcomes following OPCAB. OPCAB is a requisite tool for the practitioner wishing to pursue hybrid or minimally invasive coronary revascularization.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Dr. Puskas and colleagues at Emory University have developed a thoughtful approach for off-pump coronary artery bypass surgery. I would like to compliment Dr. Puskas for doing coronary angiography on his initial cases to determine the technique was worthwhile. The description by these authors on techniques and indications are critical to the safe use of this modality. At our institution, we do not perform off-pump as routine. However, as the authors have stated it must be part of every surgeon's armamentarium. There certainly are cases from which the patient would benefit by using off-pump approach. These include calcified aortas, minimally invasive coronary artery bypass grafting, and in some patients where one must truly limit the amount of blood utilized. Therefore, I think this chapter is a primer for these situations. I do not think the question is whether one should do off-pump versus on-pump but rather in which cases one modality is more suitable than the other. Dr. Puskas and colleagues have stated some true and relative contraindications and I think these should be followed. By the same token, there are some specific indications for off-pump surgery and I believe we now have a better understanding when that should be done.

ILK

53 Hybrid Coronary Artery Bypass Grafting Surgery

Daniel G. Cuadrado, Marzia Leacche, Ramanan Umakanthan, and John G. Byrne

INTRODUCTION

Coronary artery bypass grafting (CABG) surgery and percutaneous coronary intervention (PCI) are established treatment for coronary artery disease (CAD). Both have been shown to be effective in both alleviating symptoms and increasing long-term survival.

CABG surgery has been shown to have superior outcomes in the high-risk patient cohorts (diabetics, left main disease, three vessel disease, high SYNTAX scores, and patients with reduced cardiac function). The extent of benefit in low and intermediate risk patients is not as well defined. The main advantage of CABG arises from the use of the left internal mammary artery (LIMA) to left anterior descending (LAD) artery graft, which has an unparalleled long-term patency. Saphenous vein graft (SVG) failure remains one of the weaknesses of CABG with early failure rates, ranging from 6.2% to 30%.

Conversely, multivessel PCI with drug-eluting stents (DES) has the distinct advantage of being a less invasive form of therapy, with faster recovery and lower stroke rates, compared with CABG. DES have lower failure rates at 12 to 18 months compared with SVG (defined as stent restenosis and thrombosis compared with SVG occlusion). One of the main disadvantages of PCI is the higher rates of target vessel reintervention when compared with CABG, especially with respect to PCI in the LAD territory.

The potential of hybrid coronary revascularization (HCR) strategy is the ability to offer patients a tailored approach to the treatment of CAD, while minimizing the risks of the operation. The LIMA to LAD graft is performed in tandem with PCI to non-LAD vessels, combining the superior patency of the LIMA to LAD graft with that of DES to non-LAD vessels. As less surgical grafts will be required with HCR, this provides the additional advantage of enabling the LIMA to LAD graft to be performed through a nonsternotomy approach. Several observational studies have shown that HCR is a feasible and safe technique with short-term outcomes that are noninferior to standard CABG.

TIMING

Hybrid revascularization can be performed as either a "one-" or "two-staged" procedure. HCR performed in a single setting requires a specially designed hybrid operating room combining the facilities of the cardiovascular surgery operating room with that of a catheterization laboratory (Fig. 53.1).

The "one-stage" approach has the distinct advantages of allowing for completion angiography to assess graft patency, allows for more aggressive multivessel PCI, and facilitates completion of the procedure during a single operation while entailing a single technical fee (Table 53.1). The main disadvantage is the increased risk of bleeding given that the surgery must be performed under the influence of the potent antiplatelet agents used for PCI (Table 53.1).

On the other hand, for the "two-staged" approach, PCI and CABG are performed in their respective procedural environments separated by hours or days, although they are routinely performed during the same admission. When PCI is performed prior to CABG, lesions that cannot be successfully addressed by PCI can later be grafted at the time of CABG (Table 53.2).

The disadvantage to this strategy is that CABG will have to be performed under the effect of dual anti-platelet therapy such as aspirin and Clopidogrel®, thereby increasing the rate of major bleeding and bleeding-related complications. Also, especially when minimally invasive approaches are utilized, completion angiogram to ensure LIMA to LAD integrity requires the performance of an additional procedure. Performing the LIMA to LAD graft prior to PCI allows for more aggressive interventions to be performed with the LAD territory revascularized (Table 53.3). The integrity of the LIMA to LAD can be assessed at the time of PCI in this approach.

Performance of CABG prior to PCI provides the benefits of a LIMA to LAD graft prior to intervention on the subsequent vessels, which avoid the risks associated with anti-platelet agents. This approach also allows for evaluation of the LIMA to LAD patency during PCI via angiography. The disadvantage of this approach is that CABG will be performed with incomplete revascularization due to residual coronary artery stenosis that could, potentially, increase perioperative morbidity. Additionally, in the event of unsuccessful PCI or procedure-related coronary complications, a repeat and higher risk CABG procedure may be required.



SURGICAL TECHNIQUES

Revascularization of non-LAD vessels is performed during the PCI portion of the HCR procedure. The femoral or radial approaches are utilized and the procedure is performed as per the standard routine protocols for stent implantation.

The CABG component of HCR constitutes the surgical revascularization of the LAD utilizing the LIMA to LAD graft. Multiple techniques exist to perform this surgical portion of the procedure and are discussed below.

Minimally Invasive Direct Coronary Artery Bypass (MIDCAB) or Minimally Invasive Cardiac Surgery (MICS) refers to the technique in which the LIMA mobilization is performed through a limited anterior or left thoracotomy through the fourth or fifth intercostal space utilizing a special retractor. A hand-sewn LIMA to LAD anastomosis is then performed on the beating heart using specialized stabilizers, retractors, and a coronary shunt. Single-lung ventilation with a double-lumen



Fig. 53.1. View of the hybrid operating room at the Vanderbilt Heart & Vascular Institute.

Table 53.1 "One-Staged" Approach to Hybrid Coronary Revascularization

Advantages

1. Immediate completion angiography to access graft patency
2. Allows for more aggressive PCI
3. Single operation and single technical fee

Disadvantages

1. Requires operating under potent anti-platelet therapy required for PCI
2. Requires a hybrid operating room

PCI, percutaneous coronary intervention.

Table 53.2 "Two-Staged" Approach to Hybrid Coronary Artery Revascularization (Percutaneous Coronary Intervention [PCI] before Coronary Artery Bypass Grafting [CABG])

Advantages

1. Allows for multivessel PCI without risk to bypass grafts
2. Lesions not successfully treated with PCI can be later bypassed
3. Does not require hybrid operating room

Disadvantages

1. CABG must be performed under dual anti-platelet therapy
2. PCI is performed without the protection of the LIMA–LAD graft
3. Completion angiography requires an additional contrast load

LAD, left anterior descending; LIMA, left internal mammary artery; PCI, percutaneous coronary intervention.

Table 53.3 "Two-Staged" Approach to Hybrid Coronary Artery Revascularization (Coronary Artery Bypass Grafting before Percutaneous Coronary Intervention)

Advantages

1. PCI can be performed with the protection of the LIMA–LAD graft
2. CABG can be performed prior to dual anti-platelet therapy
3. Allows for evaluation of the LIMA–LAD graft

Disadvantages

1. CABG is performed with incomplete revascularization
2. Unsuccessful PCI or procedure-related complication may require high risk reoperative CABG

CABG, coronary artery bypass grafting; PCI, percutaneous coronary intervention; LAD, left anterior descending; LIMA, left internal mammary artery.

endotracheal tube is preferred to facilitate optimal anatomical exposure.

Thoracoscopic Endoscopic Atraumatic Coronary Artery Bypass (Endo-ACAB) refers to the technique in which the LIMA mobilization is performed via thoracoscopy through port access incisions. The LIMA to LAD anastomosis is then performed on the beating heart similar to the MIDCAB. Dissection of the LIMA is facilitated in this technique with single-lung ventilation and the insufflation of CO_2 at pressures of 8 to 15 mmHg creating a controlled pneumothorax.

Robotically Assisted Coronary Artery Bypass Graft refers to a technique in which robotic assistance is utilized for the LIMA takedown. This is followed by a hand-sewn LIMA to LAD anastomosis on the beating heart utilizing a 4 to 5 cm anterior thoracotomy while an endoscopic stabilizer, placed through a separate port incision, stabilizes the LAD.

Beating Heart Totally Endoscopic Coronary Artery Bypass Graft (TECAB) refers to a technique in which the LIMA takedown and the anastomosis are performed endoscopically utilizing a robot. The LIMA to LAD anastomosis can be performed on the beating heart or on an arrested heart using cardiopulmonary bypass (CPB).

Trans-sternal approach refers to the classic approach in which the LIMA mobilization and anastomosis are performed

utilizing a conventional sternotomy incision. While this approach is not as minimally invasive as the previously described approaches, it may be preferable in circumstances where there is concern that a minimally invasive approach could impede exposure or if there is a concern that the patient may have difficulty tolerating the requirements of a minimally invasive approach.

PATIENT SELECTION

The ideal candidates for HCR are patients with multivessel CAD comprising a high-grade lesion in the LAD and lesions amenable to PCI in the left circumflex and right coronary artery territories. There are other scenarios in which minimizing the extent of surgical intervention may be desirable. These include lack of conduits, the inability to graft a vessel but with lesions amenable to PCI, a repeat cardiac operation, or high-risk patients with concomitant pre-existing organ dysfunction, recent history of myocardial infarction, and severe atherosclerotic aortic disease (Table 53.4).

Several important contraindications must be considered. The presence of concomitant valve disease, an intramyocardial LAD, pulmonary conditions that prohibit single-lung ventilation, and subclavian artery stenosis are absolute contraindications to MIDCAB and TECAB. Morbid obesity and prior chest irradiation are relative contraindications to MIDCAB. Any conditions that preclude peripheral cannulation for CPB are contraindications to TECAB with CPB.

Table 53.4 Patient Selection for Hybrid Coronary Artery Bypass

Indications

1. High-grade lesions in LAD ± lesions amenable to PCI in the left circumflex or right coronary artery
2. Lack of acceptable conduits
3. Reoperative coronary artery bypass
4. High-risk patients (recent MI, comorbidities, atherosclerotic aortic disease)

Contraindications

1. Concomitant valvular disease
2. Intramyocardial LAD
3. Severe pulmonary disease (inability to tolerate single-lung ventilation)
4. Subclavian artery stenosis

LAD, left anterior descending; PCI, percutaneous coronary intervention.

CURRENT DATA

The current data on HCR are based on single-institutional experience. At present, there are no randomized clinical trials on HCR. The National Heart, Lung, and Blood Institute (NHLBI) have initiated the first observational trial to attempt to define the ideal patient population for HCR.

Several retrospective studies have published their results on HCR since its initial implementation in 1996. Since then, approximately 918 patients have undergone HCR. The data from these studies have shown low 30-day mortality (0% to 2%) and in-hospital morbidity (0% to 21%, average of 4.4%). The rate of percutaneous transluminal coronary angioplasty (PTCA) and stent-restenosis at mean follow-up of 1 to 44 months was 0% to 30%. It is important to note, however, that several of the earlier studies were conducted using bare metal stents (BMS) and many of the earlier studies utilized PTCA alone and not the more durable DES, which were introduced in 2003.

In evaluating more recent studies of HCR using only DES far better outcomes have been observed. Thirty-day mortality in these studies is only 0% to 1.4% and in-hospital morbidity is only 0% to 4.2%, with an average of 1.0%. The rate of reported stent–restenosis at mean follow-up of 6 to 33 months is only 0% to 6.6%. Hence, it is evident that the advent of DES technology has substantially enhanced the potential benefits HCR has to offer. Table 53.5 provides data from the recent studies that used primarily DES or all DES.

ANTI-PLATELET REGIMEN

The optimal anti-platelet strategy for use during single-staged HCR has yet to be defined. The challenge is to find the appropriate balance to achieve the appropriate levels of anti-platelet activity to prevent stent thrombosis, while simultaneously obviating the need for platelet transfusion due to bleeding and associated complications. Various anti-platelet strategies have been reported. In our series at Vanderbilt, we reported on 112 patients who underwent CABG with concomitant PCI. We administered a 300 mg loading dose of Clopidogrel® preoperatively in the holding area for electively planned hybrid procedures. For unplanned hybrid procedures, where the decision to perform PCI was made after surgery, the Clopidogrel® was given via nasogastric tube at the time the decision was made. Our choice to use a 300 mg loading dose of Clopidogrel® rather than the standard 600 mg dose used for conventional PCI was to balance the risks of significant bleeding complications against the risks of stent thrombosis. Only three patients (3%) required re-operation for bleeding and only one patient (1%) developed in-stent thrombosis during their hospital stay.

LIMITATIONS

The limitations of HCR arise from the respective limitations of minimally invasive CABG and PCI. Although the data on DES currently appear encouraging, their efficacy in certain complex situations is not as well defined. Complete total occlusions, bifurcated coronary lesions, diffuse disease or extensive lesions requiring multiple stents, ostial stenosis, and small or calcified vessels do not fair as well. Also certain high-risk subgroups, such as diabetics and those with high SYNTAX scores, have higher DES restenosis rates, which may limit the benefits of HCR in a given patient.

One of the main concern with minimally invasive CABG is the long-term patency of the LIMA to LAD graft which may be lower than those performed through a conventional median sternotomy. Limited surgical exposure and the technical challenges facing a minimally invasive approach must be taken into account and all such procedures carry a steep learning curve. In addition, the previously described contraindications to the performance of certain minimally invasive techniques need to be taken into consideration. For these reasons, completion angiography of the LIMA to LAD graft after MIDCAB seems prudent.

Table 53.5 Outcomes of Hybrid Coronary Revascularization using Drug-Eluting Stents

Author	*N*	In-hospital mortality (%)	Thirty-day mortality (%)	Mean follow-up period (months)	Stent re-stenosis (%)
Gilard	70	4.2	1.4	33	2.3
Kon	15	0	0	12	3
Vassiliades	47	0	0	7	6.6
Bonatti	5	0	0	6	0

CONCLUSIONS

The feasibility and efficacy of HCR, based on the results from individual institutions, appears to be encouraging. In selected patients, HCR appears to be an effective therapeutic modality with good clinical outcomes. As our ability to develop hybrid strategies utilizing the tools of cardiovascular surgeons and interventional cardiologists improve, the potential to tailor therapies to a wider selection of patients seems limitless. However, randomized control trials will be required to determine the full scope of HCR and gauge its comparative efficaciousness to standard CABG and PCI. The data from the currently ongoing NHLBI study will be pivotal in answering these questions and advancing this field.

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EDITOR'S COMMENTS

Dr. Byrne and colleagues at Vanderbilt University have developed a template for hybrid coronary artery revascularization. This is an important new technology and will be used in some form in most institutions. Certainly, if one plans to do true minimally invasive coronary artery bypass grafting, a hybrid approach is the way to go. In addition, early angiography to make sure grafts are patent certainly would be an advantage.

The NHLBI through the Cardiothoracic Surgical Network is planning a randomized trial on this technology. We will soon learn the best circumstances to perform this. Finally, there are going to be situations that hybrid revascularization is ideal for the given patient. The bottom line is the use of the left internal mammary artery to a large viable LAD distribution clearly prolongs survival. I think if this principle is used, then there will be many patients who would be available for this kind of technology.

ILK

54 Left Ventricular Reconstruction

Lorenzo Menicanti and Serenella Castelvecchio

OUTLINE

The increase in left ventricular (LV) volume after a myocardial infarction (MI) is a component of the remodeling process and it is associated with a poor clinical outcome. Hence, the current management strategy for ischemic LV dysfunction has been aimed to reverse the remodeling process (i.e., reduction of LV volume and improved ejection fraction [EF]) by medical therapy, devices, and/or surgical strategies. Surgical left ventricular reconstruction (LVR) has been introduced as an optional therapeutic strategy aimed to reduce LV volumes through the exclusion of the scar tissue, thereby restoring the physiological volume and shape and improving LV function and clinical status. Later, the technique has been widely adopted by skilled surgeons and refined in an effort to standardize the procedure and to optimize the results. Until recently, several studies have shown that surgical LVR is effective and relatively safe with a favorable 5-year outcome. However, in spite of the large amount of reports drawn on various data sets, the additional benefit of LVR to CABG remains debated.

This chapter briefly discusses the rationale to surgically reverse LV remodeling through LVR, and, more extensively, the technique and the indications to the best of our knowledge.

INTRODUCTION

Despite many breakthroughs in cardiovascular medicine, MI and heart failure (HF) are still among the most major public health challenges in the developed countries. The 5-year survival rate of patients diagnosed with HF is still <50% and might even be underestimated. HF is associated with ischemic heart disease in a percentage of patients ranging from 46% to 68%. Research has been very effective in delivering major advances in therapy of ischemic HF patients, including drugs, device therapy, and surgery. However, despite advances in different therapeutic strategies, the prognosis for patients with chronic ischemic HF remains poor. Indeed, HF is a syndrome with a broad spectrum of heterogeneous symptoms and signs caused by cardiac dysfunction and resulting in a wide range of clinical expressions. Treating generically, HF syndrome is reductive and misleading: to be really successful, the underlying disease, named LV remodeling, should be addressed and treated.

LEFT VENTRICULAR REMODELING: MECHANISMS AND CHARACTERISTICS

LV remodeling is a complex, dynamic, and time-dependent process that may occur in different clinical conditions, including MI, leading to chamber dilatation, altered configuration, and increased wall stress. MI, particularly large, transmural infarctions, results in a complex of structural changes involving both the infarcted and noninfarcted zones. LV remodeling usually begins within the first few hours after an MI and results from fibrotic repair of the necrotic area with scar formation, elongation, and thinning of the infarcted zone. LV volumes increase, a response that is sometimes considered adaptive, associated with stroke volume augmentation in an effort to maintain a normal cardiac output as the EF declines. However, beyond this early stage, the remodeling process is driven predominantly by eccentric hypertrophy of the noninfarcted remote regions, resulting in increased wall mass, chamber enlargement, and geometric distortion (Fig. 54.1). These changes, along with a decline in performance of hypertrophied myocyte, increased neurohormonal activation, collagen synthesis, fibrosis, and remodeling of the extracellular matrix within the noninfarcted zone, result in a progressive decline in ventricular performance. Left untreated, LV hypertrophy, dilatation, and contractile dysfunction may progress indefinitely as evidenced by progressive increases in LV volumes.

Furthermore, as part the complex process of LV remodeling, functional mitral regurgitation (MR) may occur to adversely affect the prognosis (Fig. 54.2). Chronic ischemic MR occurs in approximately 20% to 25% of patients followed up after MI and in 50% of those with postinfarct congestive HF. The papillary muscle displacement, which may occur as a consequence of the LV dilatation, results in tenting of the mitral valve at closure with lack of a proper coaptation, in turn leading to secondary MR. In addition, ventricular dilatation results in annular enlargement, which further increases valve incompetence. It is well known that functional MR, causing LV volume overload, worsens prognosis. Until recently, it was unclear if the volume overload created by MR adds a greater pathologic burden to an already adverse condition or, simply, the worse prognosis is related to a poorer LV function, and functional MR is merely an indicator of this bad condition. To this regard, a major contribution has recently been provided by Rossi and coworkers in a large population of patients affected by dilated cardiomyopathy and HF. The authors found that functional MR was strongly associated with the outcome after adjustment of left ventricular ejection fraction (LVEF) and restrictive mitral filling pattern, either in patients with ischemic dilated cardiomyopathy or nonischemic.

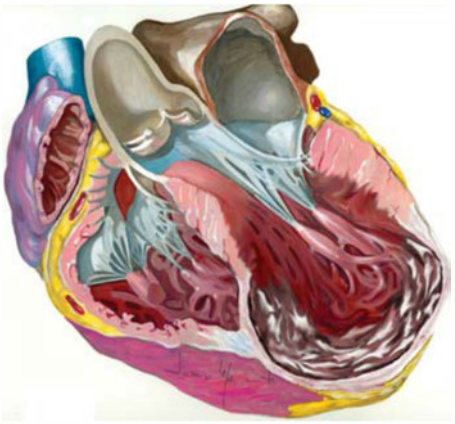


Fig. 54.1. Left ventricular remodeling. The left ventricular chamber appears dilated and distorted.

The Rationale to Perform Left Ventricular Reconstruction in Reversing Left Ventricular Remodeling

Surgical LVR has been introduced as an optional therapeutic strategy aimed to reduce LV volumes through the exclusion of the scar tissue, thereby reducing the ventricle to a more physiological volume, reshaping the distorted chamber, and improving cardiac function through a reduction of LV wall stress in accordance with the principle of Laplace's law. Since LV wall stress is directly proportional to LV internal radius and pressure and inversely proportional to wall thickness, any intervention to optimize this relationship would be beneficial either in terms of improving wall compliance and reducing filling pressure or, as wall stress is a crucial determinant of afterload, in terms of enhancing contractile performance of LV by increasing the extent and velocity of systolic fiber shortening. Furthermore, LVR of failing ventricles is usually combined with myocardial revascularization with the aim to treat the underlying coronary artery disease. Finally, although the matter of functional chronic ischemic MR, in terms of whether, when, and how it should be corrected is still considerably controversial, it should be pointed out that LVR offers either the possibility to repair the mitral valve through the LV opening or the potential of improving mitral functioning by reducing LV volumes, papillary muscles distance (which is a main determinant of functional MR), and hence rebuilding a more normal geometry.

Left Ventricular Reconstruction Technique

After the first description of the linear suture by Cooley in 1958 and the circular external suture described by Jatène in 1984, Dor started to use a circular patch to reconstruct LV cavity ("endoventricular circular patchplasty"—EVCPP), addressing anatomic situations different from the classical LV aneurysm. The technique, performed under total cardiac arrest, involved the opening of the ventricle in the center of the depressed area, thrombectomy when indicated, and exclusion of dyskinetic or akinetic LV free wall through an endoventricular circular suture passed in the tissue above the transitional zone. A Dacron patch was secured at the junction of the endocardial muscle and scarred tissue, thereby excluding noncontractile portions of the LV and septum. In 1998, Dor further refined the technique by placing a volume-measuring device, in the form of a saline-filled balloon, into the ventricle before tightening the suture, thereby standardizing the conduct of the operation and ensuring that the ventricle was left neither too large nor too small. Later, the procedure has been adopted by many surgeons leaving, however, the technique far from a real standardization and making the results difficult to compare. McCarthy described a double purse-string suture no-patch technique. Mickleborough described a tailored scar excision along with septoplasty, when indicated (dyskinetic septum), and modified linear closure. We adopted a technique that does not differ substantially from the Dor procedure except for the use, since July 2001, of a preshaped mannequin (TRISVR™, Chase Medical Inc., Richardson, TX; Fig. 54.3). The operation is performed under total cardiac arrest, with antegrade cold blood cardioplegia. According to the potential of this surgery to address "ischemia, ventricle and valve," we perform the procedure as follows.

Ischemia

Complete myocardial revascularization is performed first with particular attention to revascularize the proximal left anterior descending segment, to preserve the

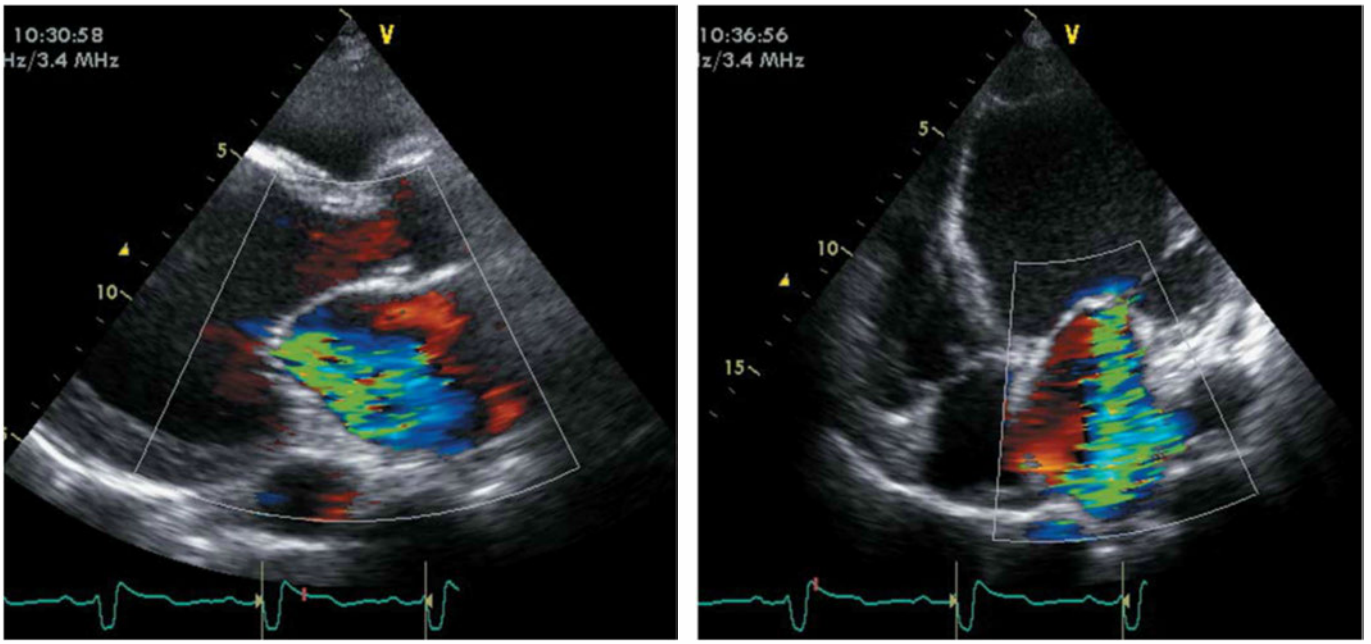


Fig. 54.2. Ischemic mitral regurgitation. Parasternal long-axis and four-chamber view showing ischemic mitral regurgitation. Left ventricular internal diameter is increased and the shape is spherical due to the presence of mitral regurgitation.

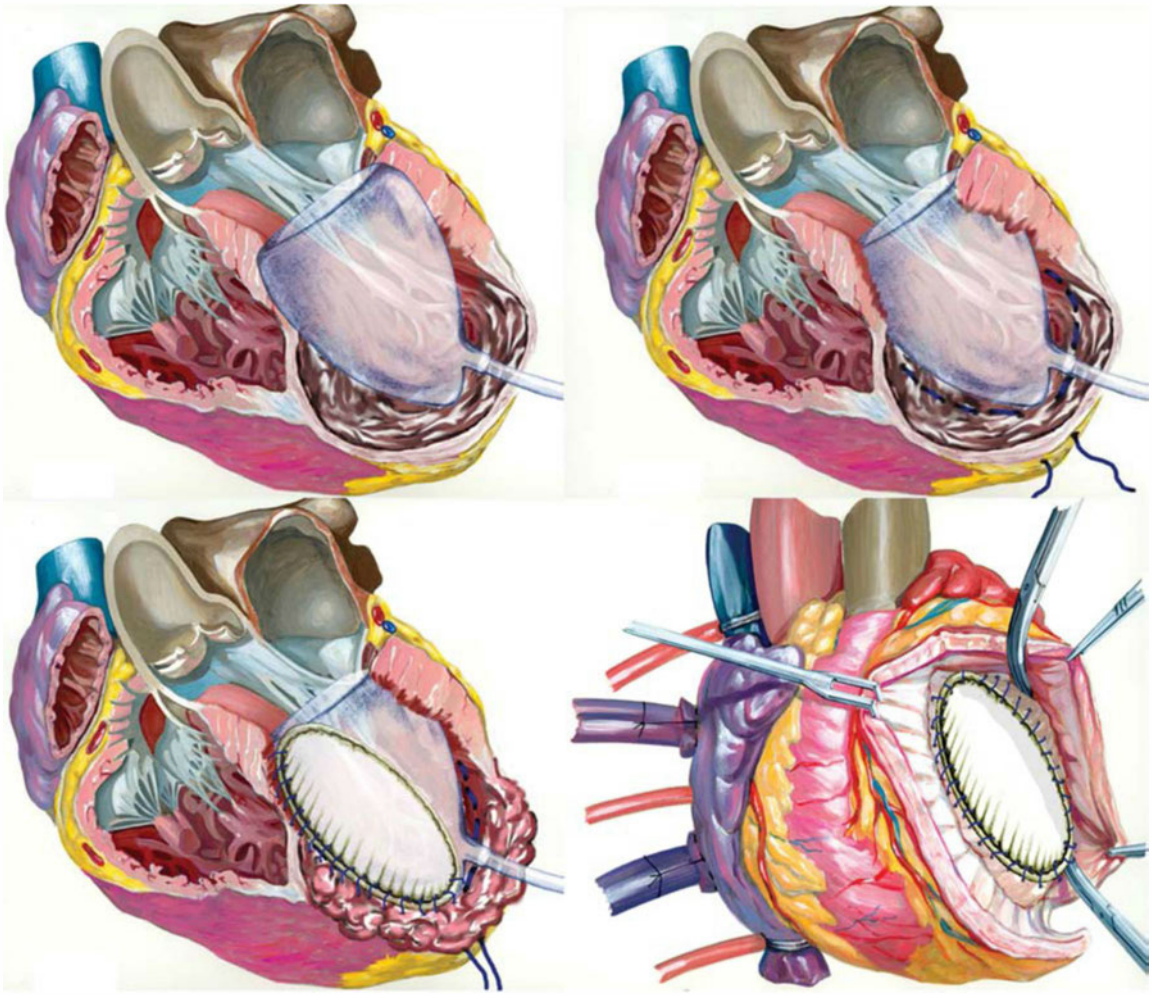


Fig. 54.3. Left ventricular reconstruction procedure for anterior remodeling (schematic). *Upper panel:* The mannequin is inside the ventricle (*on the left*); the circular suture follows the curvature of the mannequin to re-shape the ventricle in elliptical way (*on the right*). *Lower panel:* The patch is used to close the ventricular opening.

upper part of the septum. For this purpose, a left internal mammary artery is almost always placed on the left anterior descending artery. Revascularization is completed, when indicated, through sequential saphenous vein coronary bypass grafting on right and circumflex arteries.

Ventricle

After completion of coronary grafting, the ventricle is opened with an incision parallel to the left anterior descending artery, starting at the middle scarred region and ending at the apex. The cavity is carefully inspected and any thrombus is removed if present. The surgeon must be careful to identify the transitional zone between scarred and nonscarred tissue, driven by cardiac magnetic resonance (CMR) with late gadolinium enhancement (LGE), when previously performed, or alternatively by echocardiographic analysis. After that, a preshaped mannequin is inserted into the LV chamber and inflated with saline (Fig. 54.3, *upper panel on the left*). The mannequin is useful to optimize the size and shape of the new LV, particularly when the ventricle is not very enlarged (to reduce the risk of a too small residual cavity), or when the transitional zone between scarred and nonscarred tissue is not clearly demarcated, as occurs in akinetic aneurysms. Furthermore, the mannequin is useful in giving the surgeon the correct position of the apex and in maintaining the long axis of the ventricle in a physiologic range (7.5/8.5), reducing thereby the risk of sphericalization of the new ventricle. The size of the device is defined by multiplying the body surface area of the patient by 50 or 60 ml. The exclusion of dyskinetic or akinetic LV free wall is performed through an endoventricular circular suture passed in the tissue of the transitional zone (Fig. 54.3, *upper panel on the right*). The conical shape of the mannequin guides the orientation of the plane of the endoventricular circular suture at the transitional zone, obliquely toward the aortic flow tract, mainly in rebuilding the new apex. The reconstruction of the apex may be difficult when the apical and inferior regions are severely dilated and scarred; in this case, we apply a modification of the Dor procedure that involves plication of the distal inferior wall before patch placement, thus placing the apex in a more superior position and leaving a small portion of scar (Fig. 54.4). The mannequin is removed before the closure of the ventricle and the opening is closed with a direct suture (simple stitches tangential to the mannequin) if it is <3 cm large or with an elliptical, synthetic patch if >3 cm to avoid distortion of the cavity (Fig. 54.3, *lower panel on the left*). In the first case, a second stratum with the excluded tissue is sutured on the first suture to avoid bleeding. If the closure is performed by using a patch, a few millimeters of border is left sewing the patch in the everting way (Fig. 54.3, *lower panel on the right*). This technique assures a good hemostasis and makes easier to

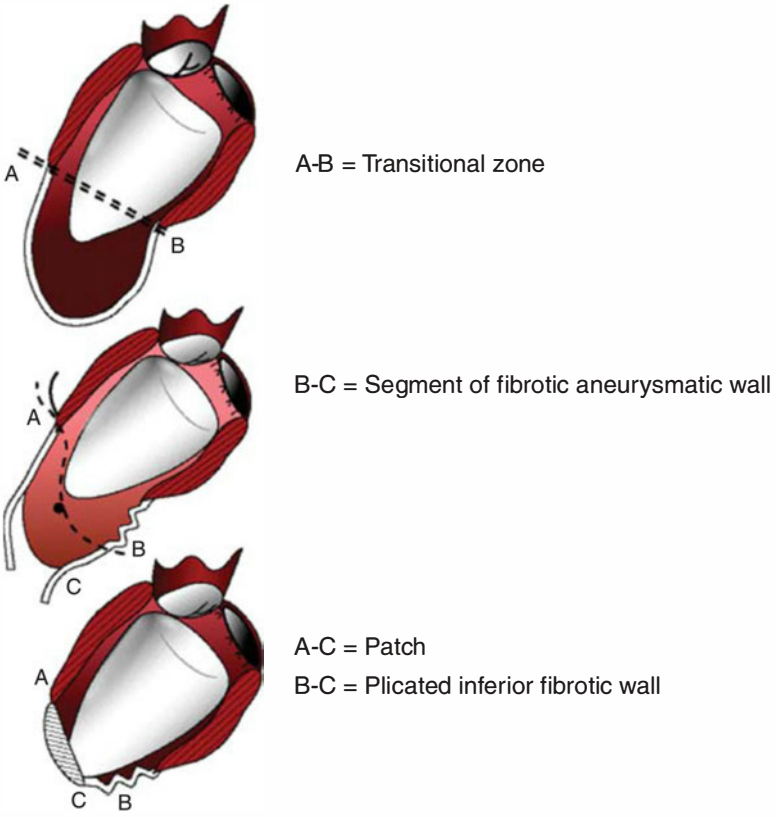


Fig. 54.4. The plication at the inferolateral portion of the ventricle is useful to lift up the new apex.

put some stitches, if needed. The positioning of the patch, which follows in some way the endoventricular circular suture, is crucial in determining the residual shape of the new ventricle. To this aim, we pay attention to positioning the patch with an oblique orientation, toward the aortic outflow tract. In this way, we avoid to obtain a box-like shape of the ventricle that may occur when the orientation of the patch is almost parallel to the mitral valve. Usually, a Dacron patch is used; the pericardial patch has been abandoned because of the risk of heavy calcification within the heart. More recently, research is focused on the use of a patch that should be able to promote an ingrowing of stem cells; however, data are not available yet.

Valve

When indicated, mitral valve is repaired through the ventricular opening with a double-arm stitch running from one trigone to the other one, embedding the two arms in the posterior annulus of the mitral valve (Fig. 54.5). To avoid tears of the posterior left of the mitral valve, we reinforce the suture with a Teflon strip. After that, the suture is tied to undersize the mitral orifice. A Hegar sizer No. 26 is used to trim the mitral annulus. Alternatively, a restrictive mitral annuloplasty with a ring implantation has been recently introduced for selected patients, when the LV opening is not big enough to have a good exposition of the mitral valve.

Special Conditions

The surgical procedure as described above is usually performed to reverse LV remodeling after an anterior MI. However, beside the LV remodeling following an anterior MI, other pattern of postinfarction LV remodeling may occur, according to the site of coronary occlusion and varying from the classic posterior aneurysm with a bulging of the inferior wall and good contraction of the remaining cavity to a global dilatation of the LV chamber with regional wall dysfunction at the inferior and posterobasal region. In these situations, the same concept used for anteroseptal dilatation can be applied, excluding the scar tissue and reducing the volume. Surgery for the posterior aneurysm generally involves a patch to close the neck of dilatation. However, the treatment of global dilatation of the inferoposterior wall is more complex, especially for the relationship between the scar and the dilatation with respect to the papillary muscles. After an inferior MI,

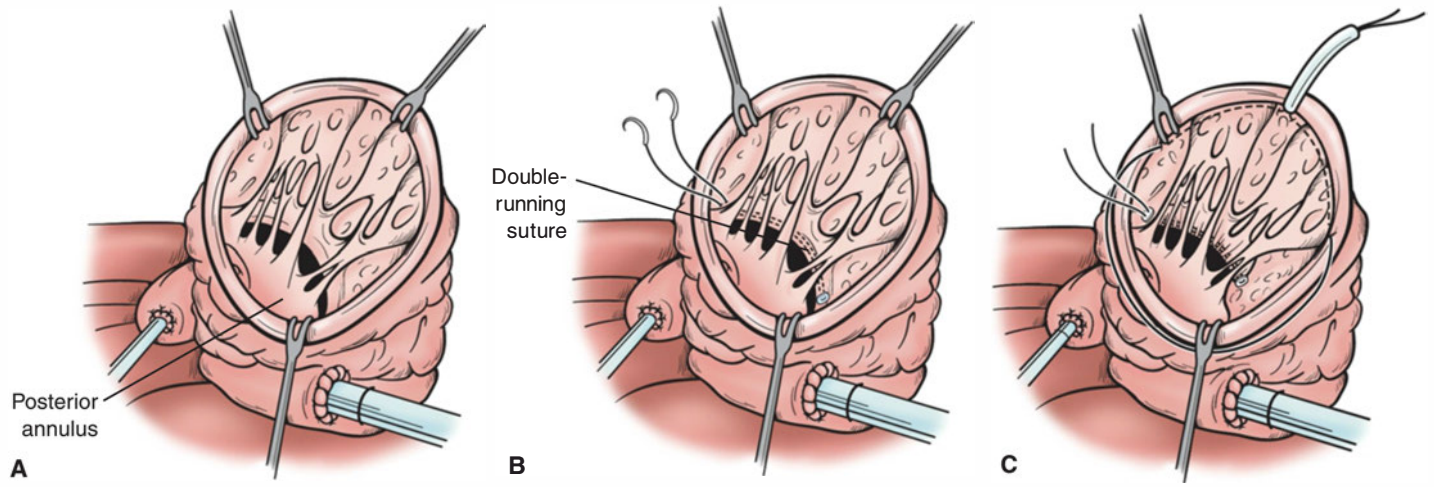


Fig. 54.5. Mitral valve repair. Mitral valve is repaired through the ventricular opening with a double-arm stitch running from one trigone to the other one, embedding the two arms in the posterior annulus of the mitral valve.

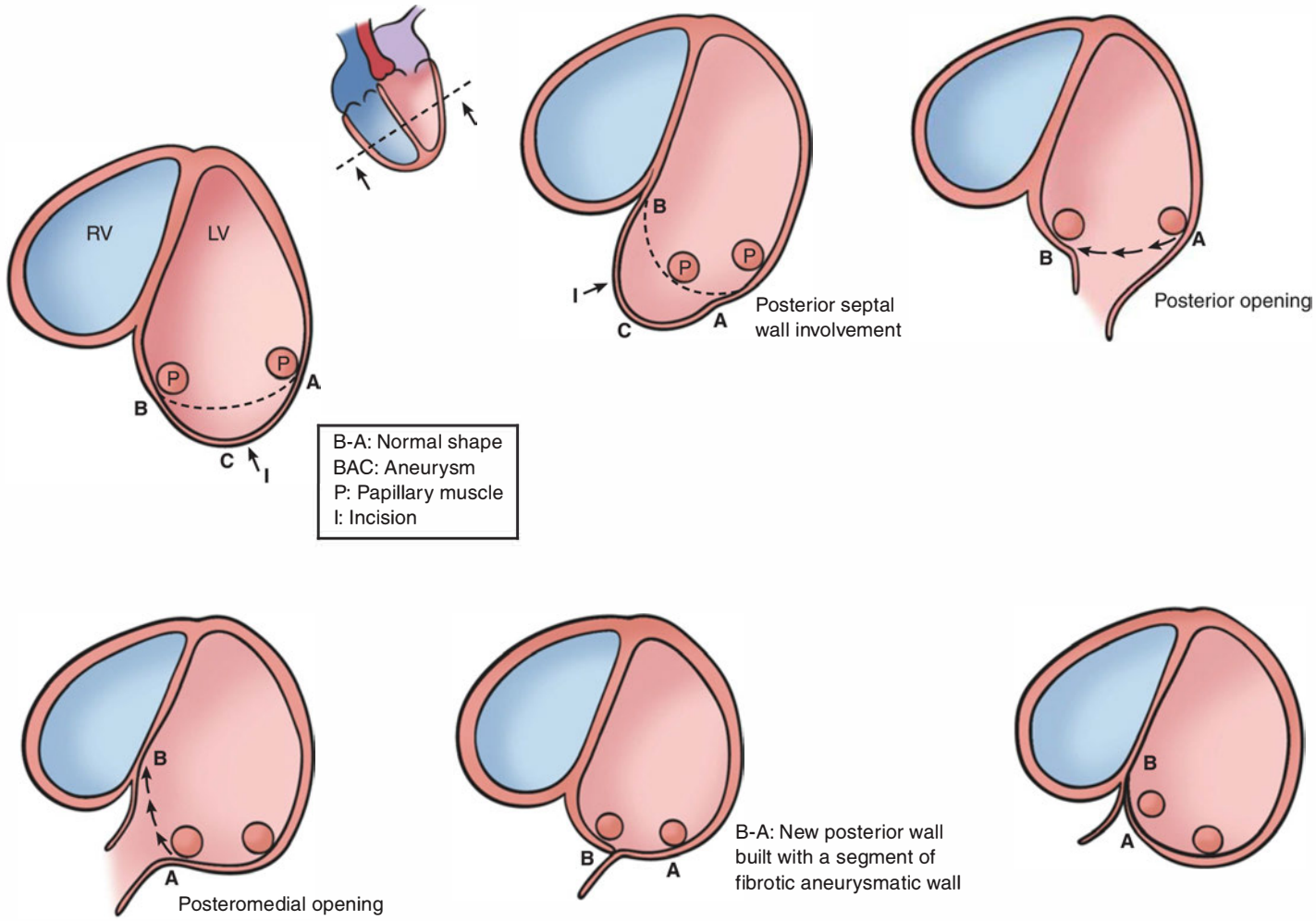


Fig. 54.6. Left ventricular reconstruction procedure for posterior remodeling (schematic). The picture shows the two different techniques, from top to bottom on *the left* (which is applied when the dilatation occurs mainly between the two papillary muscles—see text) and from top to bottom on *the right* (which is applied when the dilatation occurs between the posteromedial papillary muscle and the septum—see text).

there are two possibilities: (a) the dilatation is mainly between the two papillary muscles or (b) the dilatation is between the posteromedial papillary muscle and the septum, which is deeply involved. We use two techniques for LV dilatation after an inferior MI. The first (Fig. 54.6, *on the left panel*) involves the opening of the scarred wall at the level of the scar or at the level of the collapsed area, parallel to the posterior descending artery. A continuous 2-0 Prolene suture is performed to obtain the reapproximation of the two papillary muscles and the exclusion of the entire dilated zone. The suture is started at the beginning of the dilatation (sometimes just at the level of the mitral annulus) and continues toward the apex. According to the second technique (Fig. 54.6, *on the right panel*), the wall is opened and the continuous suture is brought behind the posteromedial papillary muscle, bringing the posterior wall against the septum.

DRAWBACKS AND SUGGESTIONS

Inside the Ventricle

Despite the use of the sizing device, the residual volume is not always congruent with the expected volume, since the operation depends not only on the preoperative volume but also on the preoperative shape, on the papillary muscles size and position, on the presence of trabeculae, or on the LV compliance. Despite any efforts to describe different LV shape abnormalities (Type 1, Type 2, and Type 3, according to Di Donato's classification, or using old or new indexes, including sphericity index, eccentricity index, and/or conicity index), the true anatomy of the ventricle and the extension of the damage might not be extensively clear before the opening of the cavity. This is especially true when the anterior septum is deeply involved (Fig. 54.7). In this case, noninvasive imaging testing is mandatory to evaluate the feasibility of the procedure and to tailor eventually the operation for a specific patient.

Regarding the impact of LVR on LV compliance, our group reported for the first time that the likelihood for diastolic function worsening after LVR is higher when the LV cavity is globally dilated (i.e., akinetic aneurysm) and relatively too *small* at the preoperative evaluation (leading to a residual volume even smaller), suggesting a careful preliminary echo-Doppler evaluation. Furthermore, we reported that diastolic dysfunction (early-to-late diastolic filling pressure >2) increases the operative risk of mortality when associated with MR and a New York

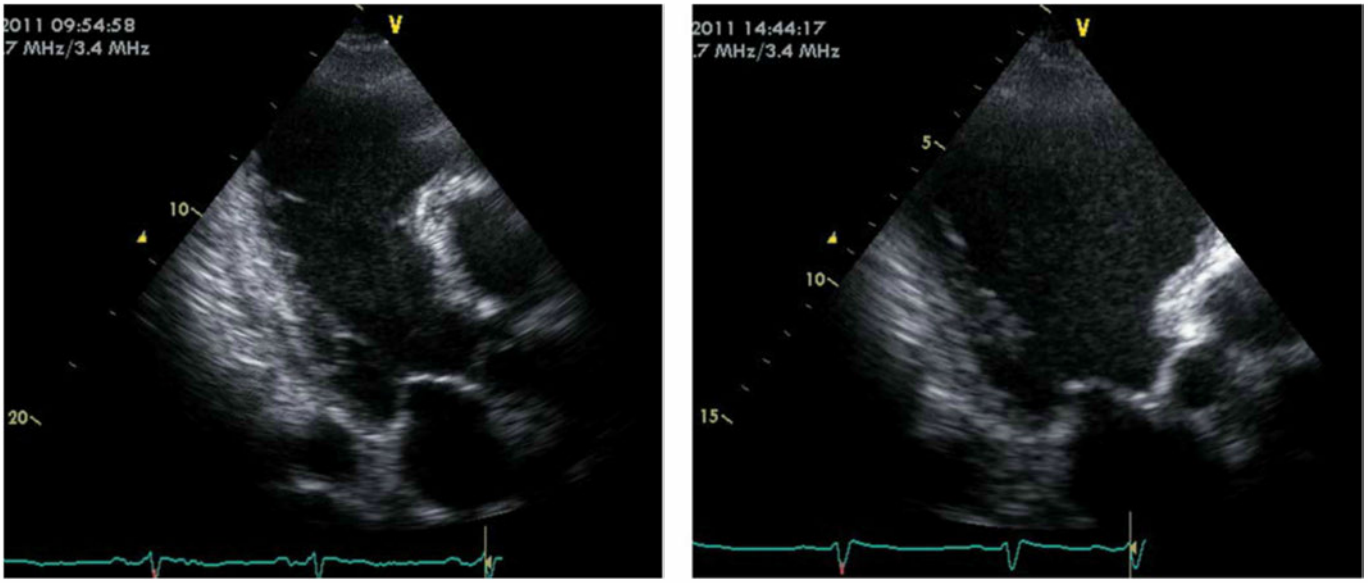


Fig. 54.7. Anterior left ventricular remodeling. Apical three-chamber view showing an extensive involvement of the anterior septum.

Heart Association class >II, reinforcing the need for a more comprehensive and accurate preoperative evaluation of diastolic function, if any possible.

Inside the Valve

In agreement with the data from the literature, showing a recurrence of MR up to 30% in patients undergoing undersized mitral ring annuloplasty, we observed a recurrence of moderate or severe MR (grade 3 or 4+) of 29% at 6 months follow-up in a subgroup of patients with previous anterior MI undergoing CABG plus LVR combined with MV repair performed according to the above-mentioned technique. Among geometric factors associated with recurrence, the postoperative long axis was the most sensitive and specific, in the meaning that a residual too short long-axis (<7.5) was associated with an increase in the sphericity index, which in turn plays a central role in the recurrence of MR. Therefore, surgeons must be aware to preserve the length of the ventricle. However, other preliminary remarks should be taken into account when we decide to repair the mitral valve in patients with chronic ischemic MR. In addition to having to distinguish between MR after inferior MI (which is the most common) and MR after anterior MI, it should be reinforced the concept that, in any case, chronic ischemic MR is a "complex and dynamic disease," involving coronary arteries, mitral annulus, subvalvular apparatus, and ventricle, in which the large number of geometric and hemodynamic variables carries the risk of a suboptimal result at follow-up. Recently, Gelsomino and coworkers showed that anterior mitral leaflet tethering is a powerful predictor of MR recurrence after undersized mitral ring annuloplasty, suggesting the need for concomitant or alternative surgery addressing the leaflet tethering (i.e., papillary muscle repositioning). In this regard, LVR associated with MV repair takes the advantage to address both ventricle and mitral annulus, but it does not seem to be enough in saving the result, as in our cohort. It should be emphasized that LV *adverse* remodeling is a dynamic process as well as LV *reverse* remodeling, both evolving over time, the latter depending on the completeness of revascularization, the residual shape of the LV (in case of performing LVR), the technique to reduce the mitral annulus (with or without a prosthetic ring and type of ring), and the complex interaction between each other. Finally, it should be outlined that, although the term *functional mitral regurgitation* is commonly used to refer to ischemic MR without "organic lesions" of the mitral valve, pathologic studies have shown that, in patients with *heart failure* (therefore, potentially suitable for LVR), the mitral leaflets are thinned out with altered collagen composition compared with normal autopsy control. It could be hypothesized that in HF patients MR might not be purely functional, arising implications for alternative, extreme strategies including mitral valve replacement.

HOW TO MAKE THE DECISION

The choice to add LVR to CABG should be based on a careful evaluation of patients, including symptoms, measurements of the LV volumes, careful assessment of mitral valve, including geometry and MR severity, assessment of the transmural extent of myocardial scar tissue and viability of regions remote from the scar, and should be performed only in centers with a high level of surgical expertise.

Symptoms

In the last two decades, the indications for revascularization in patients with ischemic HF were limited to patients with angina and significant coronary artery disease. The management of patients with ischemic HF without angina has been a challenge because of the lack of randomized data with patients who predominantly had HF symptoms. Until recently, coronary revascularization has been supported for this population. However, only approximately 40% of patients with ischemic HF show improvement in LVEF after revascularization. The possibility to combine myocardial revascularization with surgical LVR has been addressed in the STICH (Surgical Treatment for Ischemic Heart Failure) trial that compared CABG alone with the combined procedure of CABG with LVR in 1,000 patients with CAD amenable to CABG, an LVEF of 35% or less, and a dominant anterior region of myocardial akinesia or dyskinesia amenable for LVR. The results show no difference in the occurrence of the primary outcome (a composite outcome of death from any cause or hospitalization for cardiac causes) between the two groups. However, only 49% of the STICH patients were in NYHA class III/IV and the same percentage

were in CCS III/IV, indicating a population more representative of the real world of ischemic patients independently by the HF. We believe that in patients selected for LVR, HF symptoms should be predominant over angina.

DIAGNOSTIC IMAGING TOOLS

Echocardiography

Echocardiography (M-mode and two-dimensional transthoracic approach—TTE) is the first-choice diagnostic imaging tool, providing accurate information about LV dimensions (internal diameters and volumes) and systolic function (Fig. 54.8). The feasibility of a reliable echocardiographic examination is sometime limited by poor acoustic windows, an inadequate endocardial border definition or, when the ventricle is particularly enlarged, by incomplete visualization of the apex. The assessment of LV volumes plays a central role in the decision making. The LV dilatation is the "conditio sine qua non," the volume reduction should not be performed, first to avoid deterioration in diastolic function, as we have previously reported. Furthermore, echocardiography provides information about regional wall motion abnormalities and diastolic function, according to the left atrium volume and, when combined with Color- and Tissue Doppler, transmitral flow velocity and TDI measurements. In addition, two-dimensional TTE is a reliable tool to assess the MV apparatus in terms of geometry (leaflet tethering, annulus dilatation, tenting area—*the area between the tented leaflets and the annular plane in systole*—and tenting height—*the distance between the point of leaflet coaptation and the mitral annular plane in systole*—and interpapillary muscle distance, Fig. 54.9) and MR severity, using a semiquantitative or quantitative methods. Recently, Rossi and coworkers showed that a quantitatively defined functional MR [using the vena contracta (VC) or effective regurgitant orifice (ERO) or regurgitant volume (RV)] was strongly associated with the outcome of patients with HF, independently of LV function, suggesting the following criteria to define severe MR: an ERO >0.2 cm^2, an RV >30 ml, and a VC >0.4 cm. However, resting examination may underestimate the full severity of ischemic MR especially in this subgroup of HF patients. Exercise echocardiography (using a semisupine

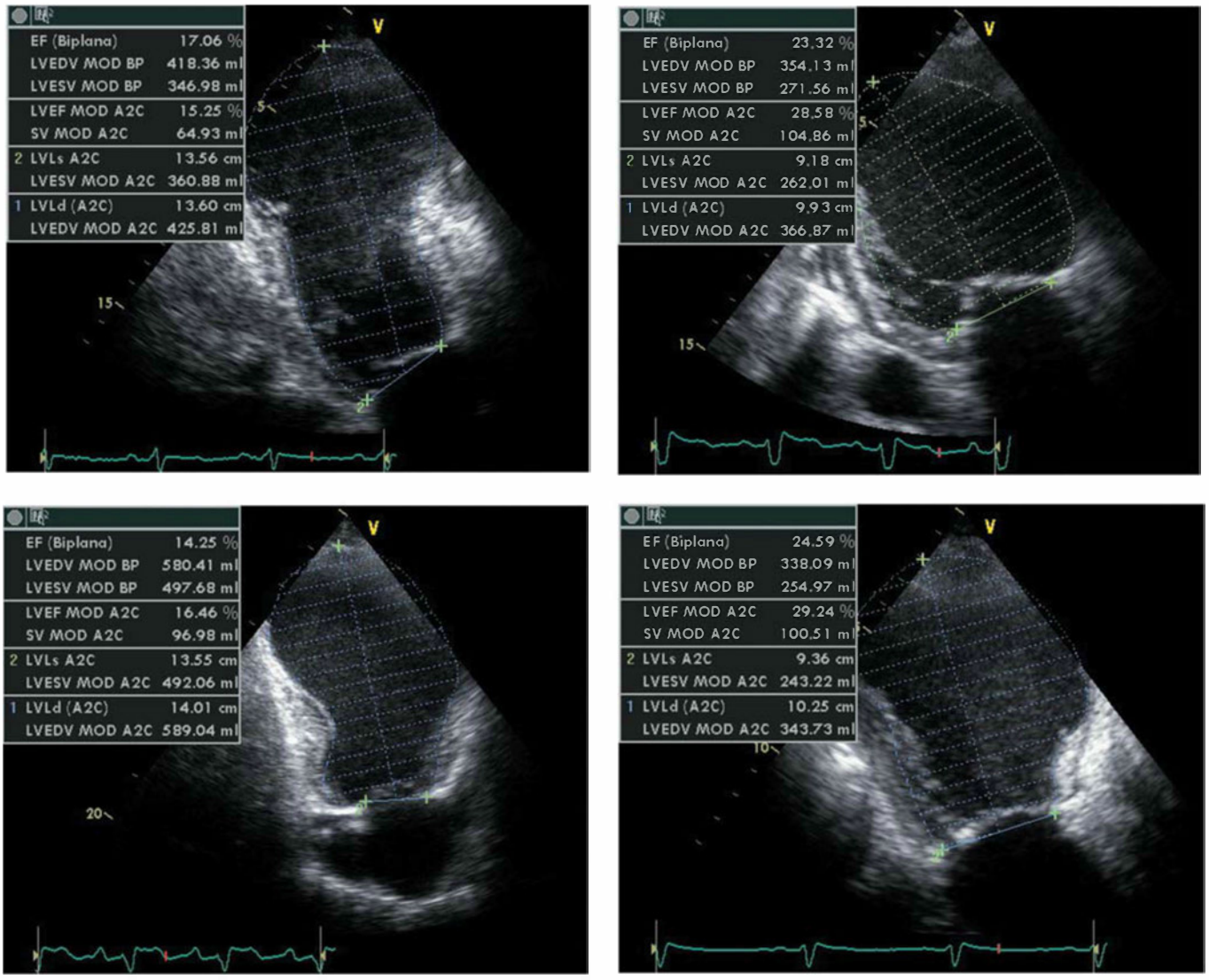


Fig. 54.8. Left ventricular remodeling. Echocardiographic two-chamber view showing marked increase in left ventricular volumes, chamber distortion, and low ejection fraction.

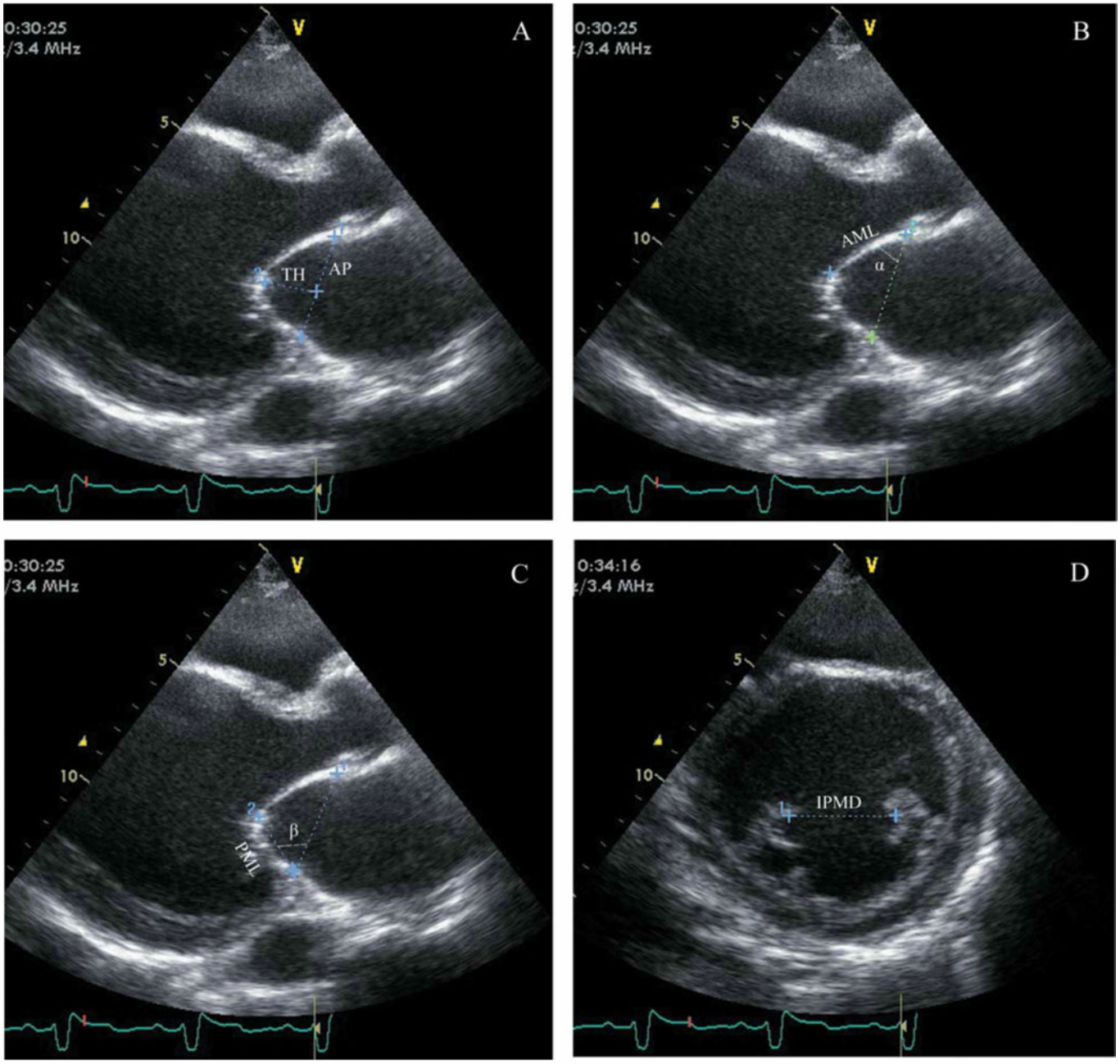


Fig. 54.9. Two-dimensional echocardiographic assessment of MV geometry. (**A**) Parasternal long-axis view showing annular plane and tenting height; (**B**) tethering of the anterior mitral leaflet was assessed by measuring the angle between the annular plane and the anterior mitral leaflet (α-); (**C**) tethering of the posterior mitral leaflet was assessed by measuring the angle between the annular plane and the posterior mitral leaflet (β-); (**D**) parasternal short-axis showing the interpapillary muscle distance.

bicycle) can be helpful in clinical decision making, unmasking higher degrees of MR.

Cardiac Magnetic Resonance

Cardiac magnetic resonance (CMR) is increasingly being used for the noninvasive imaging of the HF population and it is nowadays the gold-standard imaging technique to assess myocardial anatomy, regional and global function, and the extension of the scar. Unlike echocardiography, CMR has the ability to image in any desired plane and with a nearly unrestricted field of view, allowing unprecedented flexibility to evaluate abnormal cardiac and extracardiac structures. The functional information derived from cine CMR includes global LV and RV volumes and mass, without the need to make any geometrical assumptions, and therefore applies to ventricles of all sizes and shapes, even to those that have been extensively remodeled. A basic CMR protocol includes the assessment of LV volumes and global and regional function based on contiguous short-axis cine images. The greatest usefulness of CMR is in the detection of myocardial scar with LGE. LGE imaging visualizes irreversible damage (myocardial scar or fibrosis) due to an accumulation of contrast agent in areas with increased extracellular space. In LGE images, viable myocardium appears dark, whereas necrotic or fibrotic myocardial tissue appears bright (Fig. 54.10). LGE has unprecedented spatial resolution and can determine the transmural extent of scar, which is not possible with other imaging modalities. At the same time, CMR offers

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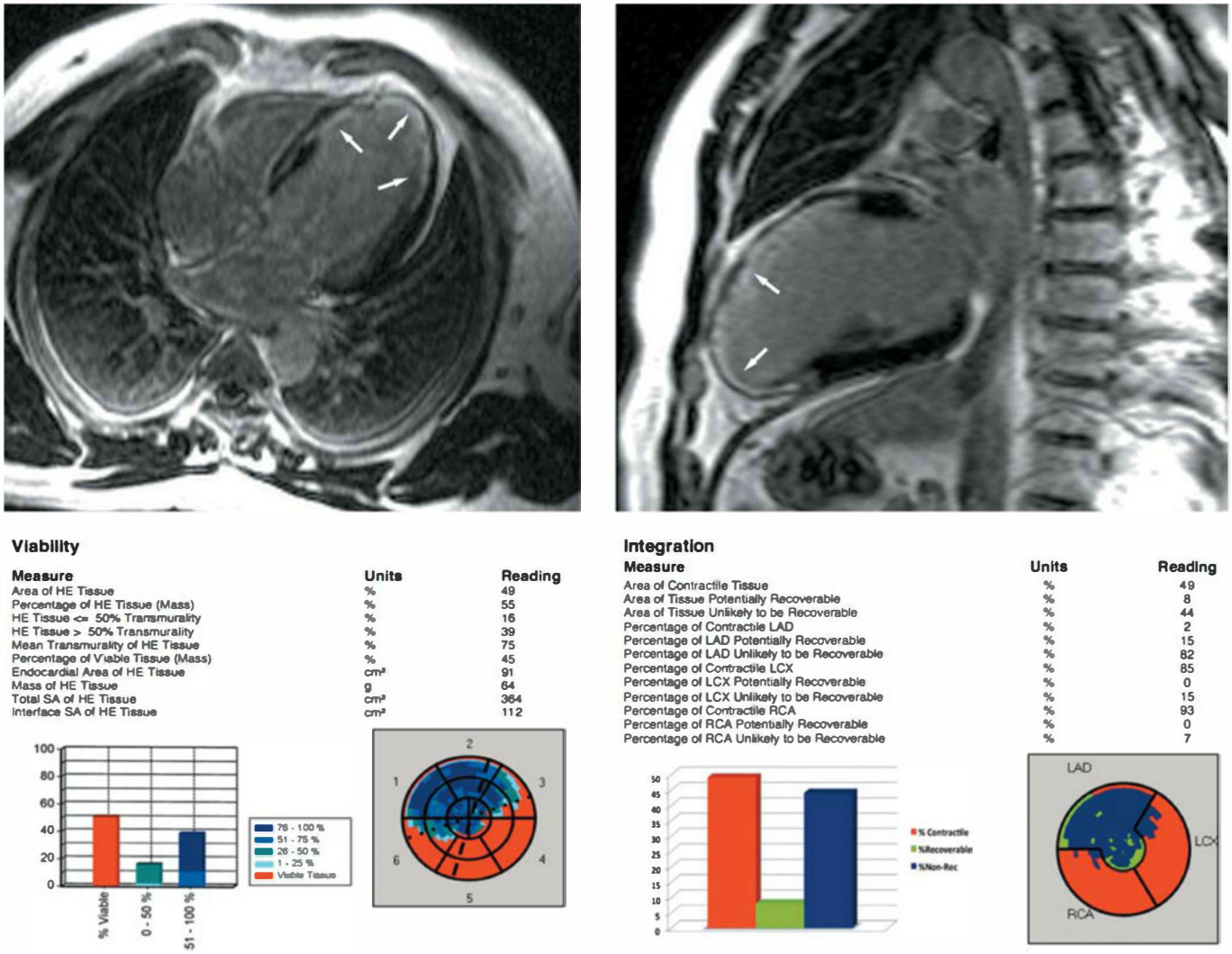


Fig. 54.10. Contrast-enhanced cardiac magnetic resonance. Preoperative viability analysis was performed with a dedicated software (Compass—Chase Medical™). *Upper panel:* Late enhancement IR-GE images showing four-chamber and two-chamber projections. Note the extended anteroapical ventricular remodeling surrounded by transmural fibrosis. *Lower panel:* Viability report showing localization, transmurality, and amount of fibrotic tissue. Dotted line represents four-chamber, while dashed line indicates two-chamber view (*on the left*). *Lower panel:* Integration report showing localization and amount of tissue potentially and unlikely to be recoverable. The polar map is obtained by merging functional information with scar distribution, allowing identification of hypokinetic but viable tissue (*on the right*).

the opportunity to assess thickness and function of the remaining nonenhanced viable myocardial tissue ("the remote regions"), which may be hibernating (ischemic but viable myocardium likely for functional recovering after CABG) or nonischemic but dysfunctional because of the high local tension that reduces shortening and likely for functional improvement after volume reduction obtained through LVR, as previously demonstrated. On the other hand, the detection of scar by LGE in the remote regions, especially at the level of basal segments, may predict an unsatisfactory LV systolic and diastolic functional recovery and adverse clinical outcomes after LVR, as recently reported. Commercially available software dedicated to LV analysis now provides a semi-automated assessment of thickening, global and regional function, and the percentage and extension of scarred tissue, which can potentially be combined in predicting myocardial viability without a need for pharmacologic stress (Fig. 54.10).

The main limitations, at this time, are the exclusion of patients with pacemakers or devices for cardiac resynchronization therapy and the potential reduction of image quality in patients with significant arrhythmia or severe shortness of breath. Therefore, both echocardiography and CMR, either for respective limitations or because of the fact that they provide different information, must be recommended in the surgical decision making.

Suggested Indications

According to our experience, we consider the following to be the indications for LVR:

- Previous anterior MI, as evaluated by electrocardiogram or CMR
  - CMR should be preferred, when available and not contraindicated.
- LVESVI > 60 ml/m²
  - Preoperative LVESV should be careful evaluated to avoid the selection of patients with small ventricles for which the likelihood for diastolic function worsening is high.
- LV dysfunction with regional asynergy, either dyskinetic or akinetic; when

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LV asynergy is severe and diffuse, SVR should be performed only if regions remote from the scar show some degree of detectable contraction.

- To this aim, CMR should be mandatory.

- Predominant HF symptoms [New York Heart Association (NYHA)—class III/IV]
  - The indication can also be expanded to patients presenting with ventricular arrhythmias and/or angina who need surgical revascularization if the previous conditions are present, to avoid further remodeling.

Suggested Contraindications

- Severe right ventricular dysfunction (biventricular dilated cardiomyopathy) (absolute).
- Severe pulmonary hypertension not associated with MR (relative).
- Severe regional asynergy without LV dilatation (absolute).
- Restrictive diastolic pattern associated with high functional class and MR (absolute).

LEFT VENTRICULAR RECONSTRUCTION AND OUTCOME: INSIGHTS INTO THE LITERATURE

The first consistent results on SVR have been reported by Dor and coworkers showing that the procedure improves LV systolic function, NYHA functional class and survival by a reduction in ventricular volumes, and an increase in EF not only in patients with classic dyskinetic aneurysm but also in dilated ischemic cardiomyopathy and severe LV dysfunction. After that, a large amount of reports drawn on various data sets from registries and mainly observational studies have shown that SVR is effective and relatively safe with a favorable 5-year outcome. The RESTORE Group (The Reconstructive Endoventricular Surgery returning Torsion Original Radius Elliptical shape to the left ventricle), the first international registry, including 1,198 patients undergone SVR, showed that the LVEF increased from 29.6 ± 11.0% preoperatively to 39.5 ± 12.3% postoperatively ($P < 0.001$) and the LV end-systolic volume index (ESVI) decreased from 80.4 ± 51.4 ml/m² preoperatively to 56.6 ± 34.3 ml/m² postoperatively ($P < 0.001$). Thirty-day mortality after SVR was 5.3%, and it was higher among patients in whom mitral valve repair was combined to SVR (8.7%) versus patients in whom no mitral valve procedure was required (4.0%; $P < 0.001$). Overall, the 5-year survival was 68.6 ± 2.8% and 5-year freedom from hospital readmission for CHF was 78%. In 2007, we published the largest single-center experience with SVR (1,161 patients) reporting a 30-day cardiac mortality of 4.7%. Patients requiring mitral valve repair/replacement (18%) had a significantly higher (13% vs. 3.0%; $P < 0.001$) operative mortality rate in agreement with the results from the RESTORE. In a subgroup of 254 patients, we reported that MR alone does not significantly increase operative mortality risk; conversely, if associated with NYHA class III/IV, it determines a significant increase in the mortality risk; and if a severe diastolic dysfunction is also present (E/A > 2), the risk is further increased, providing, for the first time, that severe diastolic dysfunction may be a risk factor for SVR. Furthermore, beneficial effects from SVR include an improvement in LV mechanical synchrony, resulting in more efficient myocardial pump function.

Along with the above-mentioned reports, other contributions have been published by many centers performing SVR with a good surgical outcome. However, all these studies were not randomized to compare the potential additional benefit of SVR with CABG alone. Only one single-center study randomized a small number of patients ($n = 74$) with dyssynergic myocardium to CABG with or without SVR and reported that outcome of CABG + SVR was better than that of CABG alone.

The STICH trial has been the first, multicenter, international, randomized controlled trial (RCT), designed to assess the potential superiority of CABG over intensive medical therapy in improving long-term survival ("the revascularization hypothesis 1"), and the benefit of SVR combined with CABG in improving hospitalization rates for cardiac cause compared with CABG alone in patients with LV dysfunction (EF ≤ 35%) and coronary artery disease suitable for surgical revascularization ("the reconstruction hypothesis 2"). The results of the Hypothesis 2 STICH trial showed no difference in the occurrence of the primary outcome (a composite outcome of death from any cause or hospitalization for cardiac causes) between the CABG group and the combined-procedure group. The 30-day mortality was similar (5% and 6% for CABG alone and for CABG plus SVR, respectively), and no difference in the rate of death from any cause was observed in a median follow-up period of 48 months. Both CABG alone and the combined procedure were equally successful in improving the postoperative CCS angina class and NYHA functional class as well as had similar improvements in the 6-minute walk test and similar reductions in symptoms. However, there was a greater reduction in the ESVI with the combined procedure (16 ml/m², a reduction of 19%, lower than the percentage of reduction reported in previous observational series, ranging from 30% to 50%; Fig. 54.11), as compared with CABG alone (5 ml/m², a reduction of 6%); difference between the two groups in the change from baseline was significant ($P < 0.001$). This improvement in ventricular volume did not translate into a measurable benefit for the patients in terms of survival; indeed, the postoperative LVESVI still remained large (>60 ml/m²), in both arms. This observation, along with the relative small percentage of ESVI reduction observed in the combined group, raised concerns on the extent of the SVR procedure that was applied in this trial.

As a matter of fact, the role of LV end systolic volume in patients with previous MI and LV remodeling has been known for years. In 1987, White and coworkers

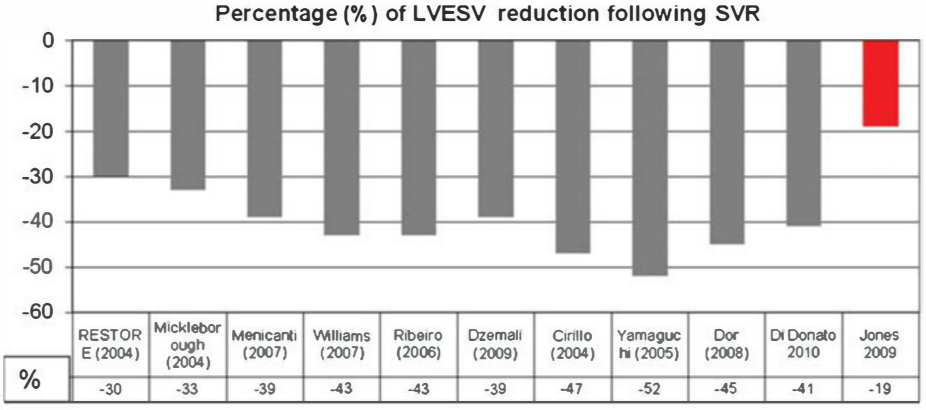


Fig. 54.11. Left ventricular (LV) end systolic volume reduction after left ventricular reconstruction (LVR) in different populations.

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showed that patients with LVESVI >60 ml/m² had approximately a fivefold increase in mortality compared with those with normal volumes after an MI. Ten years later, the GUSTO-I trial confirmed that an ESVI of 40 ml/m² or more was an independent predictor of early and late mortality after a reperfused MI. Until recently, coronary revascularization has been strongly recommended for this population. However, it has been reported that patients with severely dilated left ventricles have a low likelihood of showing improvement in EF despite the presence of substantial viability. Bax and coworkers showed that the change in EF after revascularization was linearly related to the baseline LVESV, with a higher end-systolic volume being associated with a low likelihood of functional recovery after revascularization. In addition, patients with a large LVESV had a worse long-term prognosis as compared with patients with a smaller LVESV. Based on that, to better understand the STICH results, our group reported the impact on survival of a residual LVESVI of ≥ or <60 ml/m². We showed that LVESVI following SVR impacts on the probability of death, being significantly higher in patients who remain with a postoperative LVESVI of ≥60 ml/m². We hypothesized that the lack of additional improvement in terms of survival in the SVR group observed in the STICH trial might be due to the inadequate volume reduction, which left the patients in the two arms at identical risk. This observation has been recently confirmed by Witkowski and colleagues showing that a residual postsurgical LVESVI of at least 60 ml/m² was independently associated with a fivefold increase in death and HF rehospitalization at 2 years follow-up after SVR. On the other hand, in the same report the authors showed a significant increase in LV filling pressures after SVR, as determined by an increase in E/A ratio (from 0.9 to 1.5; $P < 0.001$) and in E/E′ ratio (from 17 to 31; $P < 0.001$), supporting the hypothesis advanced by the STICH Investigators that benefits anticipated from LV volume surgical reduction (reduced wall stress and improvement in systolic function) are counterbalanced by a reduction in diastolic compliance leading, in turn, to a worsening in diastolic function. Our group had previously showed that diastolic dysfunction occur in the majority of patients affected by ischemic HF and suitable for LVR. After surgery, diastolic function remains unchanged or improved in the great majority of patients, whereas it worsens in a minority of cases (18% at discharge up to 21% at mid-term follow-up). Nevertheless, it should be pointed out that in a study by Witkowski and colleagues, patients with MR were not excluded (as we did in our series, because of the fact that MR increases E wave velocity by itself) and, even more questionable, up to 58% of patients have had concomitant restrictive mitral annuloplasty, which may induce a more pronounced increase in LV filling pressure.

The STICH trial has also been strongly criticized because there were too many expectations from these long-awaited results. Several limitations have led to substantial clinical uncertainty in making such results widely generalizable. Generally, it is admitted that randomized trials enroll a relatively small percentage of the eligible population (20% in the STICH trial), not fully representative of the daily clinical practice: only 49% of the STICH patients were in NYHA class III/IV and the same percentage were in CCS III/IV, indicating a population more representative of the real world of *ischemic patients* independently of the HF; in other words, in spite of the acronym, the study shifted from a HF population (which was also missing in the previous studies) to a broad horizon of ischemic patients. Furthermore, in the STICH trial there was no registry to follow eligible patients who were not randomized. Furthermore, the design of the STICH trial initially excluded patients with a LVESVI <60 ml/m². As the STICH study evolved, due to the empirical nature of the entry criteria, it was decided to liberalize inclusion criteria to include patients amenable to SVR surgery in the opinion of the investigators. This led to the inclusion of patients with a broad range of baseline LVESVI in the STICH population (ranging from 22 to 231 ml/m²) reinforcing the question on which ventricles have been randomized. Changing the inclusion criteria was an effort to apply the concept of SVR to a larger population with low EF in which SVR has unknown effects. Moreover, viability assessment to optimize the patient selection was not required. As mentioned above, LGE-CMR is gaining widespread use because of the information provided on the amount of myocardial scarring and its relationship with the outcome. Consequently, LGE-CMR should be considered an important diagnostic tool to identify a subset of ischemic HF patients likely to benefit from surgical therapies.

RCTs are the gold standard for building evidence, whereas observational studies are potentially more susceptible to both known and unknown confounding factors. However, the latter include a broad spectrum of patients, enroll very large samples, and extend over long periods, making the consistency of their findings nevertheless remarkable. Thus, the Task Force on Myocardial Revascularization of the European Society of Cardiology and the European Association for Cardio-Thoracic Surgery recognized the merit of SVR, which has been included as a surgical option combined with CABG in selected HF patients with a scar in the LAD territory and a baseline LVESVI ≥60 ml/m² (Class of Recommendation IIb; level of evidence B). Recently, the strength of recommendations issued in clinical practice guidelines has been questioned because of the fact that they are largely developed from lower levels of evidence or expert opinion. However, we believe that the problem is not how guidelines are drawn up but rather in the growing difficulties in developing rigorous RCTs, in which, first of all, the randomized population should reflect the patients who will ultimately receive the study treatment in practice. That having been said, it seems that the daily surgical practice is not likely to change based on the STICH trial.

Beyond the debate that will probably continue for a long time, the choice to add SVR to CABG should be based on a careful selection of patients, coming from a tight collaboration between surgeons, cardiologists, and radiologists. In the meantime, it is desirable that a more detailed analysis of the STICH data, if any, will solve long-standing unanswered questions.

SUGGESTED READINGS

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EDITOR'S COMMENTS

I asked Dr. Menicanti to do this chapter because he is one of the true international experts on left ventricular reconstruction. He took Dr. Dor's original techniques and has continued to expand its application. I also had the opportunity to scrub in on a case with him here at The University of Virginia and indeed we found our techniques were remarkably similar. The surgical aspects of this procedure are well described and are certainly safe. The big issues are application. A large randomized trial of this procedure versus coronary artery bypass grafting alone demonstrated no advantage to left ventricular reconstruction. There are a lot of issues about the STICH trial that have been discussed very carefully by Dr. Menicanti and his colleagues, but the fact reminds that the data noted in this investigation were not positive for routine left ventricular reconstruction. Having said that, this type of reconstruction for the treatment of left ventricular aneurysms certainly has been well described and is clearly better than other approaches of aneurysm reconstruction. For now, I believe this is where this procedure will have its greatest benefit. Further reviews of the STICH trial may show some other places where the technology works.

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55 Ventricular Assist

Daniel P. Mulloy and John A. Kern

BACKGROUND

Since 2000, two major developments have ushered in the modern era of left ventricular assist devices (LVADs): the 2001 Randomized Evaluation of Mechanical Assistance for the Treatment of Congestive Heart Failure (REMATCH) trial, which resulted in the Food and Drug Administration (FDA) approval of LVADs for long-term destination therapy and the emergence of smaller continuous-flow devices starting in 2005. It is now indisputable that implantable LVADs are an important therapeutic modality for patients with end-stage heart failure and will remain so for the foreseeable future.

Approximately 5 million Americans suffer from heart failure with over 550,000 new cases diagnosed each year and an estimated 287,000 deaths resulting from heart failure in the United States every year. In addition to the loss of life, heart failure poses a significant financial burden with estimated annual direct costs of $35 billion dollars per year in the United States alone. Despite advances in the management of heart failure, there may be as many as 100,000 people who have been treated with guidelines-based therapy but have remained relatively unresponsive in New York Heart Association (NYHA) Class IIIb or IV. In these end-stage heart failure patients with recurrent hospitalizations, few options exist. Orthotopic heart transplantation, the gold standard of therapy for end-stage heart failure, has been shown to definitively improve outcomes with median survival of 13 years but this is restricted to a select relatively young population. In addition, a critical shortage of suitable grafts limits the number of heart transplants performed in the United States to about 2,000 per year, only 2% to 4% of patients who need definitive therapy. With a growing population of end-stage heart failure patients and a static donor pool, additional treatment modalities must be pursued.

To date, the most promising new treatment to emerge for end-stage heart failure has been mechanical circulatory support with ventricular assist devices (VADs). The development of VADs began in earnest in 1964 with the National Institutes of Health establishment of the Artificial Heart Program, whose stated goal was putting a man-made heart into a human being by the end of the decade. Unlike the moon landing, this lofty goal was not met by the end of the decade; nevertheless, progress proceeded slowly and clinical use of VADs on a more routine basis began in the mid-1980s. The majority of implantable VADs are designed for the support of the left ventricle (LVADs) and our discussion will focus on these devices. Implantable LVADs function by removing oxygenated blood from the apex of the left ventricle, passing it through a mechanical pump, and returning pressurized blood to the ascending aorta. Despite the predominance of implantable LVADs, other temporary nonimplantable configurations do exist and the use of VADs designed for right ventricular assist devices (RVADs) failure, or biventricular failure is also well established.

Currently, implantable LVADs can be classified into two main categories: volume-displacement (pulsatile) and continuous-flow (nonpulsatile) pumps. Implantable LVADs are typically used for either bridge to heart transplantation (BTT) or long-term destination therapy for those patients not eligible for transplant. Initial clinical success was achieved with the so-called first-generation devices, or pulsatile pumps, which include the HeartMate XVE and its predecessors the HeartMate IP1000 and HeartMate VE (Thoratec Corp., Pleasanton, CA), the Thoratec PVAD (Paracorporeal Ventricular Assist Device) and IVAD (Implantable Ventricular Assist Device; Thoratec Corp.), and the Novacor LVAS (World Heart Corp., Oakland, CA). These first-generation LVADs came into clinical use in the mid-1980s and all of the above are FDA-approved for the BTT indication. Of these first-generation devices, only the HeartMate XVE is FDA-approved for destination therapy. In the landmark 2001 REMATCH trial, patients with irreversible heart failure who were ineligible for transplantation were randomized to either maximal medical therapy or HeartMate XVE implantation. LVAD implantation doubled the 1-year survival rate of these patients from 25% to 52%. The FDA subsequently approved the HeartMate XVE for permanent destination therapy, ushering in the modern era of LVAD therapy.

In recent years, the pulsatile first-generation LVADs have been replaced in clinical practice by second-generation, continuous flow LVADs. These devices, which include the HeartMate II (Thoratec Corp.), Jarvik 2000 FlowMaker (Jarvik Heart, Inc., New York), and MicroMed–DeBakey (MicroMed Cardiovascular, Inc., Houston, TX), have an internal rotor within the blood flow path that is suspended by contact, blood-immersed bearings. These continuous-flow, rotary pumps were introduced to overcome many of the shortcomings of the first-generation devices, and they are simpler in design with only a single moving part: the internal rotor. Advantages over first-generation devices include smaller size requiring less extensive surgical dissection for implantation, the absence of valves that are a primary site of wear, higher efficiency with less energy requirement, and a smaller percutaneous lead. With these improvements, the second-generation devices have demonstrated improved reliability with device support of more than 6 years reported. The most extensive clinical experience is with the HeartMate II.

Currently, the HeartMate II is the only second-generation device approved for both BTT and destination therapy in the United States. In a recently published randomized clinical trial comparing the HeartMate II versus the first-generation HeartMate XVE for destination therapy, the HeartMate II was shown to be significantly better than the HeartMate XVE in achieving the primary end point of survival free from device failure or disabling stroke at 2 years. Moreover, patients with HeartMate

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II support also had significantly superior actuarial survival rates at 1 (68% vs. 55%) and 2 years (58% vs. 24%). More recent evidence shows that the actuarial 1-year survival rate for those patients receiving a HeartMate II is now almost 80%, a staggering improvement from the 52% seen in the REMATCH trial barely 10 years ago, and a number which approaches the accepted survival rate for the gold-standard of heart transplantation.

PATIENT SELECTION AND PREOPERATIVE EVALUATION

Despite recent developments in device technology and patient care, there is no substitute for proper patient selection in the achievement of optimal outcomes. Whenever possible, LVAD implantation should be performed under elective and not emergent circumstances. Appropriate patient selection and proper timing of implantation are likely more important than any other aspect of a successful LVAD program. Each patient's comorbidity profile, ability to survive surgery, social support, and expected functional level after implantation should be carefully assessed prior to surgery. Optimal timing of LVAD implantation should occur prior to the development of irreversible end-organ damage but after correction of reversible comorbidities. Risk assessment of medical and surgical issues for both BTT and destination therapy are virtually identical and close collaboration with heart-failure cardiologists is essential for the identification of those patients who will benefit most from LVAD implantation.

Numerous factors are known to influence outcomes and most risk factors are additive. Several evidence-based predictive models, composite risk scores, and defined patient profiles are available, which are useful in selecting appropriate patients for LVAD implantation. For example, the Seattle Heart Failure model is a useful tool to identify those heart-failure patients who will derive maximum benefit from LVAD. Using easily obtained clinical parameters, a heart failure patient's probability of 1- and 2-year survival with and without LVAD can be calculated using this validated model. In addition, the Lietz and Miller 90-day predictive model identifies nine weighted risk factors affecting mortality. This model has proven useful but must be applied with care as it was developed by analysis of 222 patients undergoing HeartMate XVE implantation for BTT and is not validated for continuous-flow LVADs or destination therapy. The Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) has defined seven patient profiles and has correlated survival after LVAD with these patient profiles (Table 55.1). For example, those in profile 3, stable but inotrope dependent, have the best survival while patients in profile 1, cardiogenic shock, have high mortality and may be too sick for implantable LVAD placement. We find the combination of the INTERMACS profile and Seattle Heart Failure model most useful and believe that these tools support the trend in practice toward LVAD implantation earlier in the progression of heart failure.

One key factor for optimal patient outcome after LVAD implantation is RV function. RV failure is a leading cause of morbidity and mortality due to inability of the RV to pump sufficient blood through the pulmonary circuit. The LVAD unloads the LV allowing a decrease in pulmonary artery pressures and a corresponding reduction in RV afterload. On the other hand, LVADs increase systemic vascular return to the RV and reduced LV pressure can cause displacement of the interventricular septum into the left side, thereby reducing efficient RV function. In our practice, postimplant RV failure can be anticipated preoperatively and can be avoided through a combination of careful patient selection, prompt pharmacologic treatment of at-risk patients, and careful protection of the RV intraoperatively.

Other key factors that should be specifically addressed as part of a comprehensive preoperative assessment include nutritional status, renal function, hepatic function, gastrointestinal (GI) bleeding, pulmonary function, coagulation status, infection, neurologic function, and perhaps most importantly, psychosocial and psychiatric function. One of our key exclusion criteria for LVAD is the lack of a well-established social support system. Patients will require extensive assistance after leaving the hospital and must demonstrate the presence of a supportive network of family and friends who will be available to assist

Table 55.1 INTERMACS (Interagency Registry for Mechanically Assisted Circulatory Support) Profiles and Timing to Initiation of Mechanical Circulatory Support

INTERMACS profile		Description	Time frame for mechanical circulatory intervention
Profile 1	"Crash and burn"	Critical cardiogenic shock—life-threatening hypotension despite rapidly escalating inotrope support with critical end-organ hypoperfusion	Within hours
Profile 2	"Sliding on inotropes"	Inotrope dependence with continued deterioration—declining function despite intravenous inotrope support	Within a few days
Profile 3	"Dependent stability"	Stable but inotrope dependent—stable blood pressure and end-organ function but demonstrating repeated failure to wean from support	Within a few weeks
Profile 4	"Recurrent advanced heart failure"	Resting symptoms—can be stabilized but experiences daily symptoms of congestion at rest and diuretic doses fluctuate at high levels	Within weeks to months
Profile 5	"Exertion intolerant"	Comfortable at rest—unable to engage in any other activity without congestive symptoms	Variable
Profile 6	"Exertion limited"	Able to perform mild activity—fatigues after the first few minutes of any meaningful physical exertion	Variable
Profile 7	"Advanced NYHA III"	Clinically stable—reasonable level of comfortable activity but limited to mild exertion	Not a candidate

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with care. Given the complexity of management concerns, comorbidity profiles, and social issues associated with LVAD implantation, the importance of a multidisciplinary team approach to patient care cannot be overemphasized. All social and medical risk factors are assessed and addressed by our team of LVAD professionals including the surgeon, heart failure cardiologist, LVAD coordinator, nurse practitioners, social workers, pharmacists, dieticians, and many others. Table 55.2 lists our minimum goals with regard to standard metabolic markers associated with several patient risk factors. Nutritional status, as an example, demonstrates the importance of the multidisciplinary approach to optimizing patient outcomes. Malnutrition is common in patients with heart failure and if not addressed prior to implantation, it leads to increased infection risk, decreased functional recovery, and generally poor outcomes. For patients with a prealbumin <15, enteral feeding should be initiated preoperatively and should continue through the postoperative period until the patient is taking adequate oral nutrition. Caloric intake, nutritional supplementation, and the metabolic response to changes in nutritional management must all be monitored carefully throughout the preoperative, postoperative, and outpatient course of care. This is best achieved with the input of multiple specialists. Similar proactive approaches must be taken toward the other mentioned patient comorbidities and directed therapy should begin prior to LVAD implantation and continue throughout the postoperative stay and outpatient course of therapy.

Table 55.2 Minimal Preferred Preimplantation Goals for Various Metabolic Markers

Parameter	Minimum preferred value
Preimplantation goals	
Hemodynamic	
Central venous pressure	<15 mmHg
PCWP	<25 mmHg
Renal	
Serum creatinine	<2.5 mg/dl
Estimated GFR	>50 ml/kg/min
Hematologic	
Hematocrit	>30%
INR	<1.3
Platelets	>120,000/ml
Nutritional	
Albumin	>3 g/dl
Prealbumin	>15 g/dl
Transferrin	>240 mg/dl
Hepatic	
Total bilirubin	<2.5 mg/dl
ALT, AST	<2 times normal limit

Implantation of a long-term LVAD should not be considered in patients with irreversible major end-organ dysfunction, severe hemodynamic instability, unknown neurologic status, major coagulopathy, irreversible respiratory failure requiring mechanical ventilation, bacteremia, or right heart failure. In patients who present in cardiogenic shock and accompanied end-organ dysfunction, we have found success with temporary mechanical support in the form of the TandemHeart® Percutaneous Ventricular Assist Device (pVAD) system (Cardiac Assist, Inc., Pittsburgh, PA). This device, which can be inserted under fluoroscopic guidance in the catheterization laboratory, employs a transeptally placed inflow cannula to pump oxygenated blood from the left atrium, through an external centripetal blood pump, and back into the outflow cannula which is secured in the femoral artery. Flow rates of up to 4.0 L/min can be achieved and we have successfully used this pVAD system in multiple patients who presented in cardiogenic shock, in whom the reversibility of their condition was unknown. In our experience, patients who demonstrate significant reversal of end-organ failure within several days after initiation of pVAD support are excellent candidates for subsequent implantable LVAD therapy. To date, we have successfully placed implantable HeartMate II LVADs in four patients who were bridged temporarily with a Tandem Heart pVAD.

HEMODYNAMICS AND VALVULAR DISEASE

The physiologic goal of LVAD implantation remains the same regardless of the device implanted: mechanical unloading of the failing left ventricle along with an increase in aortic perfusion. The decompression of the LV and increase in systemic perfusion in turn lead to improved RV efficiency—via an increase in venous return to the RV and a reduction in pulmonary pressures by unloading the LV and decreasing pulmonary pressure afterload. All of these changes reduce cardiac work, improve myocardial perfusion, and allow for reversal of the neurohormonal activation and catabolic state present in patients with progressing heart failure. The combination of improved perfusion along with suspension of the heart failure milieu allows for recovery of nonreversible end-organ damage and, in rare cases, significant myocyte recovery allowing for subsequent device removal.

While the physiologic changes that accompany LVAD implantation have been proven to provide relief from heart failure for properly selected patients, the re-routing of blood flow and concomitant flow and pressure changes in the cardiac chambers and great vessels must be carefully considered in each patient. After LVAD placement, the majority of systemic blood flow bypasses the aortic valve, and the structural integrity and valvular function of the heart must be carefully evaluated in light of these anticipated changes. Echocardiography both prior to LVAD implantation and at the time of surgery is essential for the evaluation of cardiac function and identification of anatomic abnormalities that may warrant correction at the time of surgery. All atrial and ventricular septal defects should be repaired during LVAD implantation and a dedicated bubble study is an important part of the echocardiographic evaluation. Even the smallest patent foramen ovale (PFO) must be closed during the LVAD procedure as any PFO may become hemodynamically significant after LVAD placement. For this reason, it is imperative to perform careful echocardiographic evaluation for the presence of any atrial or ventricular septal defects both before and after initiation of LVAD support. In addition, the functional capacity of any valve may change after LVAD implantation so all valves should be carefully evaluated intraoperatively and monitored for changes in valve function after initiation of LVAD support. Particular attention should be paid to valvular lesions that impact forward flow, and therefore require surgical correction including aortic insufficiency, mitral stenosis, and tricuspid insufficiency.

In the case of aortic insufficiency (AI), we opt to replace the aortic valve with a tissue bioprosthesis or surgically close the aortic valve in all patients with moderate or severe AI, and most patients with mild AI. With the increased aortic root pressure and decreased LV pressure that accompany LVAD support, even mild or moderate AI can become hemodynamically significant after device placement. In our experience,

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trace AI does not require treatment and does not tend to significantly progress during LVAD support but again we stress the importance of intraoperative postimplantation echocardiography to confirm this finding. A number of aortic valve fusion procedures are available (complete oversewing of valve leaflets, partial fusion, patch covering, etc.) and fusion has proven to be a successful method for addressing AI both at our institution and others. Nevertheless, fusion procedures render the patient completely dependent on the LVAD, such that even a short disruption in device support could prove fatal, and our preference is to perform a bioprosthetic valve replacement when significant AI is present. In most patients supported by an LVAD, the aortic valve does not open with every cardiac cycle and this decrease in valve movement and stasis in perivalvular flow puts those with a mechanical valve at a high risk for thromboembolic events. For this reason, we replace any existing mechanical aortic valve with a bioprosthetic valve at the time of surgery. In contrast to AI, it is generally agreed upon that no degree of aortic stenosis requires treatment.

With regard to the mitral valve, stenosis may require correction but mitral insufficiency does not cause significant problems after LVAD placement. In most cases, mitral insufficiency improves with decompression of the LV and resolution of ventricular dilatation. Furthermore, even severe mitral insufficiency does not impair forward flow through the LVAD because the passive pressure gradients in the supported ventricle prevent significant backflow across the valve. On the other hand, moderate or severe mitral stenosis is an indication for valve replacement with a bioprosthetic valve, as this lesion reduces effective inflow into LVAD pump and may prevent achievement of optimal flow rates. In patients with a mechanical valve at the mitral position, we again opt to replace the mechanical valve with a bioprosthetic valve given the increased risk of thromboembolic complications associated with the mechanical valve.

For the right heart valves, we agree with the general consensus that only severe tricuspid or pulmonary insufficiency should be repaired or treated with valvular repair or replacement. Efficient RV function is essential for successful patient outcomes after LVAD and in most cases, mild-to-moderate insufficiency of either the tricuspid or pulmonary valve will improve after LVAD placement, secondary to LV decompression and reduced pulmonary pressures and RV afterload. Again, we emphasize the importance of intraoperative postimplantation echocardiography to confirm appropriate valvular function in every patient and to rule out new pathologic blood flow caused by the changing pressure and flow gradients associated with LVAD support.

HEARTMATE II PLACEMENT

Specific aspects of the surgical procedure vary according to the device-type being implanted but the principles of placement are similar for every device. We will describe the operative procedure for the HeartMate II LVAD, as this device is the most commonly used at the time of writing. The device and its components are prepared on the back table according to the Thoratec directions. Briefly, the HeartMate II pump is removed from the sterile packaging, fully submerged in a basin of normal saline, and run at the minimum pump speed of 6,000 RPM for at least 5 minutes to lubricate the bearings. Previously, the inflow and outflow conduits required preclotting but with the newer sealed conduits, this is no longer necessary. After priming of the pump, the tunneling bullet is attached to the distal end of the percutaneous lead and the inflow conduit is attached to the pump housing, filled with sterile saline, and any air bubbles are removed by gently tapping the side of the pump. The fingertip of a powderless glove is used to cover the inlet extension of the inflow graft and the pump/inlet extension is covered with antibiotic-soaked sterile laparotomy pads until the time of implantation. The bend relief is placed over the sealed outflow graft but remains disengaged for the deairing procedure and is also put aside until needed.

Meanwhile, the patient is intubated and standard hemodynamic access and monitoring for major cardiac surgery is initiated. The transesophageal echocardiography probe is inserted and assessment of cardiac anatomy and function is performed. Careful attention is paid to RV function. A detailed assessment is made of cardiac valve function, in particular looking for the valvular lesions described above that may require surgical correction. Attention is also paid to cardiac anatomy looking for ventricular or atrial septal defects (ASDs) that require surgical closure prior to LVAD placement (i.e., PFO). In the case of patients already supported with the Tandem Heart, an iatrogenic ASD is present and this requires correction prior to implantable LVAD placement.

After the standard prep and drape, the surgical timeout is performed and checklist reviewed. While minimally invasive LVAD implantation techniques have been described, we opt to perform a median sternotomy in all patients. Access sites for cardiopulmonary bypass (CPB) are carefully considered preoperatively and in patients undergoing repeat cardiac surgery, a preoperative computed tomography (CT) is routinely performed to assist with decisions on appropriate cannulation sites. In the uncomplicated patient without additional anatomic concerns, we perform the LVAD implantation on the beating decompressed heart. However, if septal defects or valve pathology are present, we perform the necessary aortic cross-clamp with antegrade and retrograde cardioplegia. After completion of the sternotomy, the sternal retractor is placed and the pericardium is opened. Prior to LVAD pocket formation, it is important to expose the heart and any additional vessels to allow for the rapid initiation of CPB in the event of hemodynamic instability. Any pericardial adhesions are carefully taken down and pericardium is suspended to create the appropriate cardiac well. Next, attention is turned toward creation of the preperitoneal LVAD pocket.

The HeartMate II is designed for preperitoneal placement and many centers place the device between the left rectus muscle and the posterior rectus sheath. We opt to place the device directly between the posterior rectus sheath and the peritoneum as we feel creation of an appropriately sized hemostatic pocket is easier in this location and requires less extensive dissection. The pocket is created by electrocautery dissection below the suspended pericardium superiorly. The anterior border of the pocket is formed by the posterior rectus sheath and transversalis fascia, the posterior border by the diaphragm superiorly, and the preperitoneal fat and peritoneum inferiorly. The medial diaphragmatic attachments to the left ribs are taken down to allow for creation of an appropriately sized pocket, and dissection along the posterior rectus sheath is continued under the costal margin on both sides (space of Larrei). The plastic sizing device is used frequently to determine adequacy of the pocket and to guide further dissection, especially with regard to the necessary lateral diaphragmatic exposure. Care is exercised to create a completely hemostatic pocket as patients have varying degrees of coagulopathy postoperatively and a pump pocket hematoma increases infection risk and contributes to other wound-healing problems. If the peritoneal cavity is entered, 4-0 Vicryl sutures are used to close the entry site. The pleural

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spaces are often violated during lateral dissection, especially on the left side. Pleural openings do not require closure but do necessitate pleural drain placement at the conclusion of the operation. Once an appropriately sized cavity is created and the sizing device can be easily inserted into the appropriate position, the patient is prepared for the initiation of CPB.

In patients who are undergoing their first cardiac operation, we prefer central aortic and caval cannulation. For reoperative surgeries, preoperative planning is key and a CT scan with or without contrast is routinely performed preoperatively for anatomic assessment. If the aortic anatomy is not optimal for central cannulation, we prefer an axillary approach by placing the arterial cannula into an 8-mm Dacron graft sewn onto the axillary artery. In any patient with significant atherosclerotic disease as documented by CT, we avoid femoral cannulation due to the elevated thromboembolic stroke risk. For patients in whom the LVAD is the first cardiac operation, care is taken not to elevate the pericardium off the aorta so that a virgin plane will be present at the time of potential subsequent orthotopic heart transplant. With regard to venous cannulation, we prefer a standard dual-stage venous cannula for uncomplicated cases. When additional procedures are planned that require access to the right atrium (i.e., correction of PFO, repair of ASD, Tandem Heart removal with septal repair, tricuspid valve repair), bicaval cannulation is performed. For single cannulation, a 3-0 Prolene purse-string suture is placed in the right atrial appendage and for bicaval cannulation, an additional 3-0 Prolene purse-string is placed at the junction of the IVC and right atrium. In both cases, we attempt to minimize caval dissection and employ vacuum-assisted venous drainage without caval tapes in order to maintain virgin tissue planes in the event of subsequent transplantation. In patients who have had previous cardiac surgery, we prefer percutaneous femoral venous cannulation again to minimize dissection and maintain tissue planes for subsequent transplantation.

Following preparation of CPB access sites, systemic heparinization is performed and the standard arterial cannula is placed according to patient size and secured. Next, the dual-stage venous cannula or bicaval venous cannulae are placed and secured according to standard procedure. If aortic cross-clamp and cardioplegia are necessary, a retrograde cardioplegia cannula is placed via a right atrial stab incision and is directed into the coronary sinus and the DLP cannula is placed into the ascending aorta. Following the placement of appropriate cannulae, CPB is initiated.

Attention is next turned to the ventricular apex and the chest is flooded with carbon dioxide at 2 to 4 L/min to minimize the risk of air embolism or entrapment. The carbon dioxide flow continues for the remainder of the procedure. Laparotomy pads are placed posterior to the LV to elevate the apex and with the ventricle distended, the characteristic LV apical dimple is identified. In our experience, the ideal coring site is at the anterior aspect of and slightly lateral to this dimple. Once the coring site is chosen, a 0.5-cm cruciate incision is made using a No. 11 blade scalpel. A 14-F Foley catheter is inserted through this incision into the LV cavity and is filled with 30 ml of sterile normal saline. The catheter acts as a centering device and the circular coring knife is advanced over the distal end and gentle traction is placed on the catheter. The coring knife is aimed away from the interventricular septum toward the mitral valve posteriorly, and laterally toward the lateral LV wall. When aiming the coring knife, it is important to consider the anticipated changes in ventricular size resulting from LVAD decompression, as obstruction of the inflow cannula must be avoided to achieve optimal postoperative flow rates. The cutting edge of the coring knife is applied to the endocardium and gently rotated back and forth until the LV cavity is entered (Fig. 55.1). The coring knife is removed, Foley catheter deflated and removed, and attention is turned to examination and completion of the ventriculotomy. Metzenbaum scissors are often necessary to excise residual endocardial muscle at the coring site and care should be taken to preserve the subvalvular apparatus of the mitral valve as this is done. Once the ventriculotomy is satisfactorily completed, the ventricular cavity is inspected for mural thrombi or crossing trabeculae. Loose thrombi must be thoroughly removed as well as any crossing trabeculae. If significant left ventricle thrombi are present, aortic cross-clamp and cardioplegia should be performed to allow adequate removal of all thrombi and minimize the chances of distal embolus.

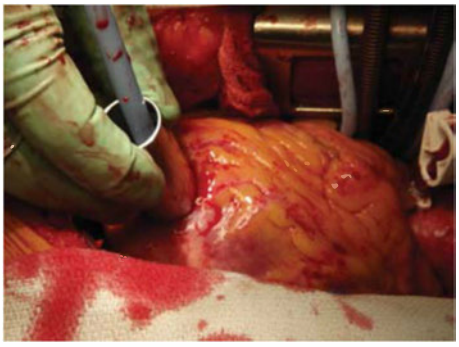


Fig. 55.1. Coring of the left ventricular apex.

After completion of the coring ventriculotomy, 12 to 16 circumferential double-armed 2-0 Ethibond sutures with 10 mm × 12 mm pledgets are placed at that site. These sutures are placed approximately 15 mm from the epicardial perimeter in a partial thickness horizontal mattress manner with minimal travel between adjacent sutures (Fig. 55.2). Next, the sewing ring is brought into the field and polar coordinates are marked on the ring felt to assure proper alignment. The Ethibond apical sutures are passed through the sewing ring in corresponding position, taking care to penetrate the sewing ring only and not the attached sleeve. The ring is then advanced to the apical surface and each suture is tied with six or seven knots snugly enough to cause slight dimpling of the pledgeted myocardial cuff (Fig. 55.3). Adequate positioning of the sewing ring is again verified, and the device is then brought into the operative field for attachment of the inflow cannula. Some have reported the use of BioGlue® (CryoLife, Kennesaw, GA) or other hemostatic agents at the apical cannulation site, but we have found that this is a low-pressure site and does not require any additional hemostatic agents to maintain an adequate and hemostatic seal.

The HeartMate II LVAD is placed into its preperitoneal pocket and the inflow

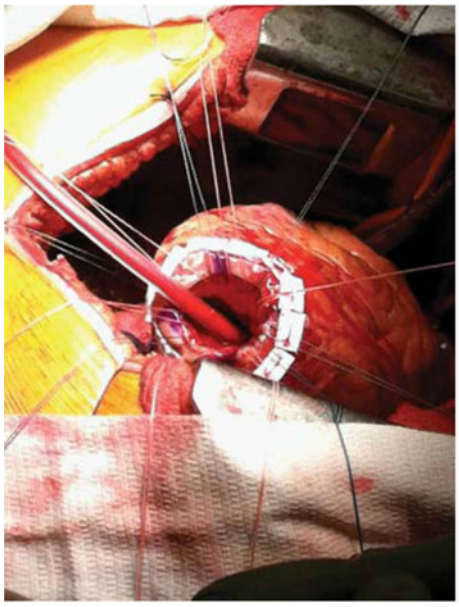


Fig. 55.2. Placement of the apical ventricular sutures.

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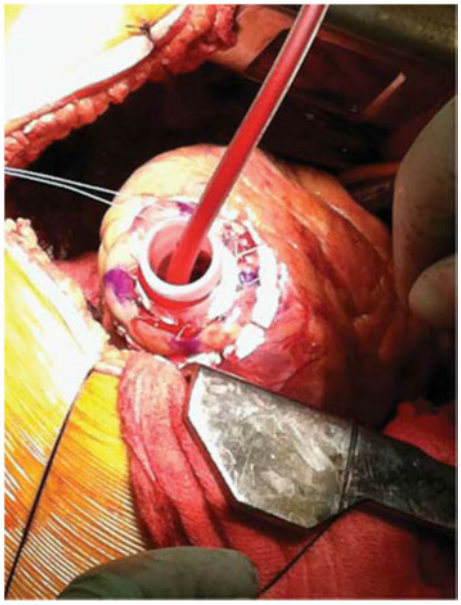


Fig. 55.3. Completion of the apical sewing ring anastomosis.

cannula is positioned toward the sewing ring. Once alignment is satisfactory, the inlet extension is inserted into the sewing ring and secured in position by tying the attached green nonabsorbable suture. Early in our experience, we opted to further secure the inlet extension using a standard zip-tie (such as those used to secure CBP tubing) deployed proximal to the green suture but on reoperation for subsequent transplantation, we encountered loose zip-ties on more than one occasion. Currently, we tie a single No. 5 silk suture distal to the green suture and have found this to be adequate. Once the inflow cannula placement is complete, exposed portions of the device are covered with antibiotic-soaked sterile laparotomy pads and attention is turned toward the outflow graft anastomosis.

The ascending aorta is exposed and a site for anastomosis on the right lateral aspect of the proximal aorta is selected. The aortic anastomosis should be performed as proximal as possible on the ascending aorta to provide for adequate distal length for potential future aortic cross-clamp application and aorto-aortic transplantation anastomosis. Moderate tension is then placed on the outflow Dacron graft extension of the device, and the Dacron graft is cut to appropriate length. Some describe cutting the graft at a 45-degree angle to facilitate anastomosis but we observe superior results when it is cut at almost 90 degrees, as this allows use of a shorter section of aorta. Before cutting, ensure that the outflow graft is not twisted by checking the position of the black line on the graft above and below the bend relief. The DLP cannula is removed, a partial occlusion clamp is placed along the right lateral aspect of the aorta, and a longitudinal aortotomy is performed. If the patient's aorta is too small to allow appropriate placement of the partial occlusion clamp, aortic cross-clamp, and cardioplegia should be performed. The ends of the aortotomy are circularized using a 4.8-mm punch and the aortotomy is enlarged by trimming the intervening edges of aortic tissue. The end-to-side anastomosis is then performed using two separate 4-0 or 5-0 Prolene sutures with 2 mm × 3 mm pledgets running from each end of the anastomosis (Fig. 55.4). Following completion, we place Omnex™ (Ethicon, Somerville, NJ) surgical sealant along the anastomosis to help ensure adequate hemostasis. The partial occlusion clamp is then slowly released and after allowing momentary filling of the outflow graft, a cross-clamp is placed on the proximal portion of the outflow graft. A pledgeted purse-string is placed in the outflow graft distal to the cross-clamp followed by creation of a 3-mm graftotomy using a No. 11 blade. The DLP cannula is then placed and secured in the outflow graft for subsequent deairing.

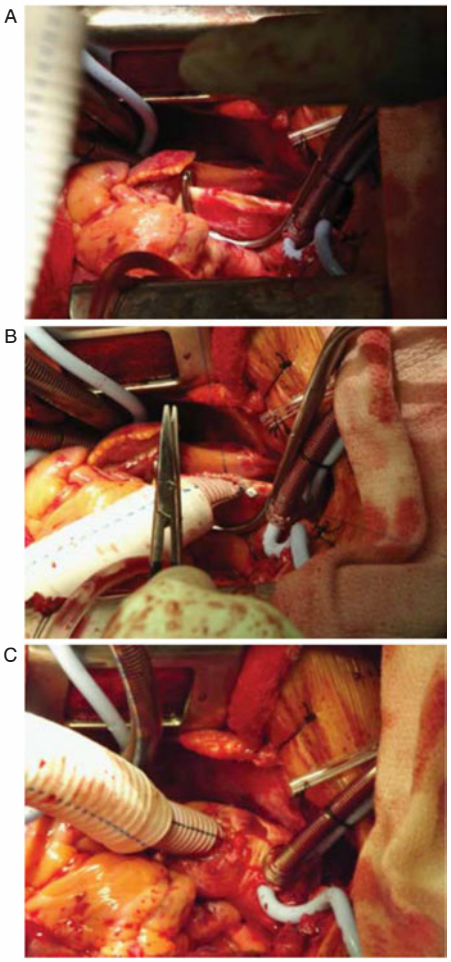


Fig. 55.4. Outflow graft to aortic anastomosis. (**A**) Placement of the partial occlusion clamp and aortotomy. (**B**) Outflow graft being sewn to the proximal aorta. (**C**) Completed outflow graft to ascending aortic anastomosis.

The next step is tunneling of the percutaneous lead or driveline. The patient should be evaluated for lead site placement prior to the procedure and marked. The selected exit site should be two finger breadths below the right costal margin, midclavicular line, or slightly lateral, in a position that will allow for freedom from tethering on clothing while standing or sitting. The tunneling device is bent into a gentle curve and starting from the right inferior aspect of the pump pocket, the tunneler is slowly advanced to the skin surface at the marked site. The tunnel should take a long curved route to maximize the amount of velour-coated percutaneous lead in the subcutaneous space. This will promote optimal ingrowth of the percutaneous lead and help prevent postoperative driveline or pump pocket infection. We opt to make a small 1.5-cm latitudinal skin incision at the exit site rather than using the enclosed 8-mm skin punch due to improved experience with wound healing using this technique. The bullet on the percutaneous lead is then thread onto the end of the tunneling device and the tunneler followed by percutaneous lead is pulled through the skin incision such that the velour coating ends approximately 2 cm below the skin surface and the rubber coating is in contact with the dermis and epidermis. While the Thoratec instructions recommend velour contact at the skin interface, we and others have noted a decreased infection rate when the entire velour coating remains below the skin surface. The percutaneous driveline is secured at the skin level with two 0 Prolene sutures, and the remaining skin incision is closed with a running subcuticular 4-0 Monocryl (Fig. 55.5). Excess percutaneous lead is shaped into a gentle loop in the LVAD pocket, and the loop is secured to the posterior rectus fascia with a 2-0 Prolene suture (Fig. 55.6). Once lead placement is completed, the lead is connected to the system power-based unit (PBU) and the patient is ready for deairing, weaning from CPB, and initiation of device support.

During deairing and device initiation, continuous echocardiographic monitoring is essential. The patient is placed in steep Trendelenburg position and the chest continues to be flooded with carbon dioxide. The LV and attached inflow graft

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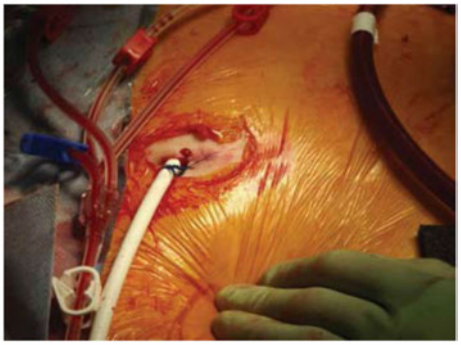


Fig. 55.5. Driveline exit site with the driveline-skin interface.

and pump are deaired through the Luer-lock connection on the outflow cap; the outflow graft itself is deaired by allowing aortic backflow via partial release of the graft cross-clamp. Once initial deairing using these techniques is complete, the outflow graft and overlapping bend relief are attached to the screw ring on the pump. With the outflow graft cross-clamp still in place, the HeartMate II is turned on at the lowest speed of 6,000 RPM to initiate flow through the pump. At this time, deairing is continued through the DLP cannula, which was previously placed at the highest point in the outflow graft, between the screw ring and graft cross-clamp. Close communication with the perfusionist is essential as CPB flow is now slowly decreased to allow for partial filling of the LV without overdistension. Once adequate deairing of the LV and aortic root is confirmed by both echocardiography and visual inspection of the DLP cannula showing no further egress of air bubbles, the outflow graft cross-clamp is removed. While monitoring the LV continuously with visual inspection and echocardiography, the device RPMs are slowly increased while CBP is weaned and then stopped. Before titration up to higher device RPMs (>8,000), CBP should be stopped and the LV should be filled to avoid air entrapment.

Selection of an optimal pump speed should begin in the OR while continuous echocardiographic guidance is available. An optimum speed is reached when the cardiac index and LV size are within normal limits, and there is no rightward or leftward deflection of the interventricular septum. A ramped speed study can be performed to determine the lower limit speed (when the aortic valve opens with every cardiac cycle and the septum begins to deflect rightward) and the upper limit speed (when the aortic valve ceases to open and the septum begins to deflect into the LV). The optimal pump speed will be somewhere between these two and should allow some pulsatility with an ideal aortic valve opening of once every two to three cardiac cycles. The usual speed fitting these parameters varies slightly for each patient but generally falls between 8,400 and 9,600 RPM for the HeartMate II. Notably with the HeartMate II, the most efficient mechanical speed for this device is approximately 9,000 RPM and one may observe slight decreases in pump power at this speed; these power decreases should not be confused with suction events.

While the speed is being adjusted in the OR under echocardiographic guidance, attention is turned toward drain placement and chest closure. The DLP catheter is removed from the outflow graft, the purse-string suture is tied down, and Omnex sealant is applied to this site. The aortic and venous CPB cannulae are removed and purse-string sutures tied. Hemostasis is confirmed at all anastomosis and cannulation sites, and anticoagulation is fully reversed with protamine sulfate. We typically place three 24-F Blake mediastinal drains: one posterior, one left lateral, and one right lateral. If either pleural space was entered, an additional 24-F Blake drain is placed in the pleural space. Patients are given 24 hours of appropriate perioperative antibiotics, cefazolin in most cases, and the mediastinal and pleural drains are left in place until they produce <100 ml per 24-hour period. Once drain placement is complete, hemostasis is adequate, and the appropriate initial speed setting has been determined, the chest is closed in standard fashion using six stainless steel sternal sutures. To minimize infection risk, we perform sternal closure in all patients. With this routine sternal closure, we have observed no mortality occurring from sudden cardiac tamponade and our reexploration rate is approximately 15%.



Fig. 55.6. X-ray of the completed HeartMate II LVAD with a gentle loop of the intracorporeal driveline.

POSTOPERATIVE MANAGEMENT

In most instances, the postoperative management of LVAD patients is similar to the care of any cardiac surgery patient. The continuous-flow LVAD does present some unique care considerations including blood pressure monitoring, anticoagulation, and management of RV function. In the immediate postoperative period, while invasive monitors are still in place, blood pressure monitoring is not a significant issue as mean pressure should be titrated to the same goals as for any other patient. Once arterial pressure lines are removed, the most effective way to monitor blood pressure is by using a standard sphygmomanometer and a Doppler probe typically placed over the radial artery. The goal at this point is to maintain the mean arterial pressure in the 70 to 80 mmHg range.

Anticoagulation is required for all currently FDA-approved continuous-flow LVADs to avoid thromboembolic complications. Experience from the HeartMate II BTT and destination therapy trials, along with subsequent reports, demonstrates that the level of anticoagulation needed is less than initially believed and that systemic heparinization prior to warfarin initiation is not necessary. In our institution, we hold all anticoagulant medications during the initial postoperative period until any coagulopathy or hemorrhage is definitively addressed. Usually, by postoperative day 2 to 3 the patient is started on 81 mg aspirin per day. A day or two later, typically postoperative days 3 to 5, oral warfarin is initiated and will be continued indefinitely with a target INR of 1.5 to 2.5. We no longer routinely use systemic heparinization prior to warfarin due to several hemorrhagic

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complications along with recent evidence confirming this practice.

Postoperative right heart failure is a feared complication after LVAD placement but in our experience (91 HeartMate II patients at the time of writing), only one patient has required postoperative mechanical circulatory support of the right ventricle. That patient required temporary right ventricle support with the AB5000™ external VAD (Abiomed, Danvers, MA) and was successfully weaned from this device after several days of support. In the initial HeartMate II BTT trial, 20% of patients experienced some degree of RV failure with 6% requiring RVAD placement. In the subsequent HeartMate II destination therapy trial, RV failure requiring extended inotrope use was again 20%, but the requirement for RVAD placement decreased to 4%. We have found the management and prevention of RV failure after LVAD to be similar to management and prevention of RV failure after any major cardiac operation. RV failure after implant can be anticipated preoperatively via echocardiography and assessment of other clinical parameters (CVP, RV stroke work index, hepatic congestion) and an important aspect of prevention is appropriate patient selection. Patients with RV failure preoperatively are not appropriate for LVAD support. In addition, intraoperative protection of the RV is critical especially when additional procedures are being performed that require aortic cross-clamp and cardioplegia. In patients deemed preoperatively to be at a higher risk for RV failure, intra- and postoperative use of milrinone, epinephrine, and isoproterenol are important and vasopressor agents should be kept to a minimum whenever possible. Inhaled epoprostenol sodium (Flolan—GlaxoSmithKline, London) or nitric oxide have proven useful in several cases to decrease pulmonary vascular resistance but we have observed increased bleeding rates with the use of inhaled Flolan and now reserve its use for very select cases. Inhaled nitric oxide is an additional tool that may be useful to decrease pulmonary vascular resistance. We rely heavily on intraoperative echocardiography and frequent postoperative echocardiography to assess RV function and titrate pump speed to avoid deflection of the interventricular septum either into or away from the RV. Consideration of temporary mechanical RV support should occur if the cardiac index is persistently below 2.0 L/min/m^2, CVP is >20 mmHg, and all other management strategies have been exhausted.

POTENTIAL COMPLICATIONS

The most common complications associated with LVAD therapy include perioperative hemorrhage, infection, right-sided heart failure, acute renal failure, GI bleeding, and ventricular arrhythmias. Again, patient management for the majority of these complications is equivalent to the management of any patient after major cardiac surgery. Perioperative coagulopathy and hemorrhage is common with 80% of patients requiring postoperative packed red blood cell transfusion and about 15% requiring chest reexploration. Acute management of coagulopathy and hemorrhage is no different in the LVAD patient; however, LVAD recipients are more likely to experience coagulopathy and should be treated aggressively. Patients should have all coagulation factors checked preoperatively and repleted as necessary before surgery. Standard intraoperative strategies are useful to limit postoperative bleeding including careful hemostatic dissection, shorter CPB times, maintenance of normothermia, and close monitoring of platelets and coagulation factors intraoperatively with appropriate early replacement. When diffuse coagulopathy occurs and patients require large amounts of blood products, aminocaproic acid, recombinant Factor VII, and Factor IX complex should be considered. All anticoagulants should be held postoperatively until full resolution of coagulopathy and/or hemorrhage is achieved.

With regard to infection, LVAD recipients are at an increased risk for infection for multiple reasons. As mentioned previously, heart failure induces a catabolic state that depletes nutritional reserves, suppresses immune function, and increases infection risk. Whenever possible, nutrition should be optimized prior to LVAD placement and postoperative nutritional supplementation is crucial for minimizing infectious complications. In addition, the extensive dissection required for LVAD placement along with implantation of a large foreign body imparts a high risk for infection. The percutaneous driveline provides a site for pathogen entry and meticulous care of this site is essential for the prevention of driveline infection, pump pocket infection, and sepsis. The driveline exit site must be carefully immobilized, covered with a sterile dressing, meticulously cleaned on a regular basis, and we recommend continuous use of an abdominal binder to minimize driveline movement. As always, the mainstay of treatment for any infection that does develop is judicious use of appropriate antibiotics, surgical drainage, debridement as needed, and ultimately removal of the infected foreign body if necessary. If a pump pocket infection occurs, the only durable means for resolving the infection is transplantation. In the meantime, washout, drainage, and use of antibiotic beads may be used for infection control until a transplant graft is available. With regard to driveline infections, we have a very low threshold for drainage and debridement as we have had good success in treating these infections early and minimizing the development of the more serious pump pocket infection that could require device removal. When a driveline infection is present, the driveline exit site can be cored out and moved using the pictured technique with demonstrated success in achieving durable resolution of infection (Fig. 55.7).

Postoperative GI bleeding after LVAD implantation seems to be a particularly common problem after HeartMate II implantation with up to 20% of patients experiencing GI bleeding that requires transfusion. The cause of this bleeding is thought to be related to the axial-flow pump. No definitive therapy for this syndrome is available and anticoagulation should be stopped in the event of bleeding and GI endoscopy colleagues consulted for assistance with the management of these patients. GI endoscopists should be aware that in a significant percentage of patients, bleeding may come from a small bowel source and a capsule endoscopy or push enteroscopy may be needed for definitive diagnosis. Some have reported success in controlling GI bleeding by decreasing pump speed and there are reports of extended cessation of warfarin and/or aspirin therapy without adverse events. In the case of intractable GI bleeding, lowering of anticoagulant goals or even complete cessation may be necessary and the risks of this approach should be discussed with patients prior to initiation.

Finally, ventricular arrhythmias are common before and after LVAD placement with one-quarter to one-third of all patients experiencing at least one episode of sustained (>30 seconds) ventricular tachycardia (VT) or fibrillation (VF) postoperatively. While these episodes are much better tolerated in the LVAD recipient than in the unsupported patient, and some may even be asymptomatic, VT/VF usually results in reduced pump flows and contributes to morbidity, mortality, and prolonged hospitalization after LVAD. Most of our patients have a defibrillator in place prior to surgery and in those who do not, we seriously consider postoperative defibrillator placement prior to discharge

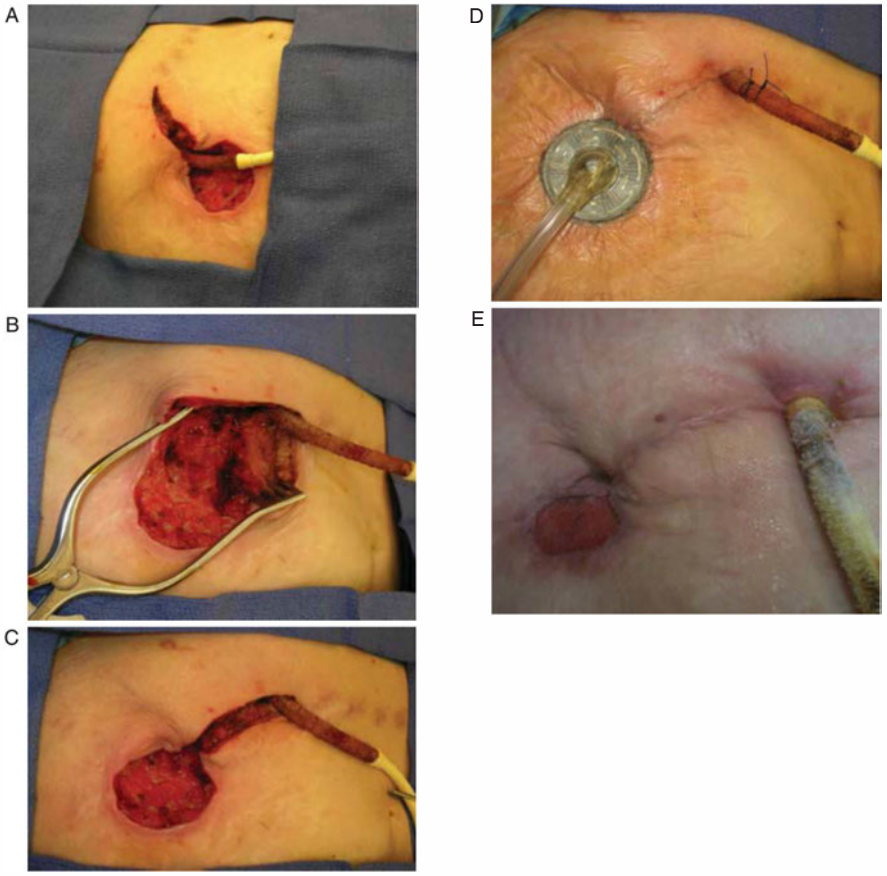


Fig. 55.7. Technique for driveline repositioning and vacuum dressing placement due to driveline infection. (**A**) Coring out of the peridriveline infected tissue. (**B** and **C**) Medial relocation of the driveline out of the infected tract. (**D**) Tract closure and vacuum dressing placement. (**E**) Healing relocated driveline without evidence of further infection.

from the hospital. In patients with recurrent preoperative VT/VF, we have recently experienced success performing intraoperative epi- and endocardial cryoablation at the time of LVAD implantation (Fig. 55.8). We have found this procedure to be useful in reducing the frequency of postoperative VT/VF and now consider this approach in every patient with a history of preoperative ventricular arrhythmias.

One aspect in the postoperative care of the LVAD patient that is worth reiterating is the importance of a multidisciplinary approach to care. Any successful LVAD program is reliant on a large team of cardiac surgeons, heart failure cardiologists, invasive and electrophysiology cardiologists, nurse practitioners, VAD coordinators, nutritionists, pharmacists, nurses, social workers, transplant coordinators, and many others. LVAD candidates present with complex physiologic and social comorbidities and the seamless interaction of all team members is necessary to help these patients through successful LVAD implantation, hospital discharge, potential transplantation, and beyond.

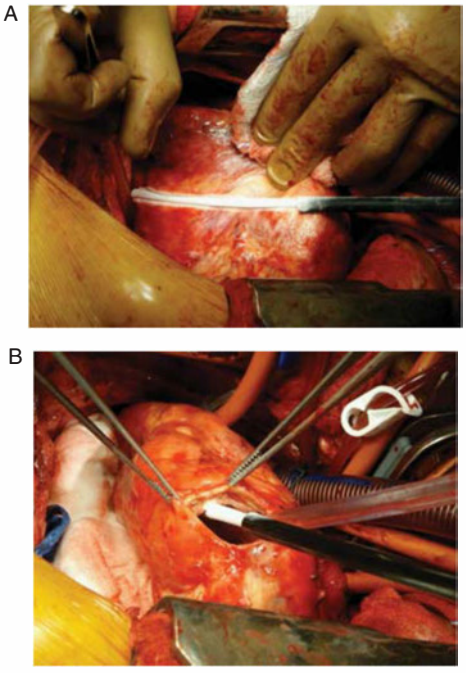


Fig. 55.8. (**A**) Epicardial and (**B**) endocardial cryoablation for ventricular arrhythmia treatment.

CONCLUSION

With the prevalence of congestive heart failure continually increasing worldwide and the frequency of cardiac transplantation remaining stable, it is clear that other options must remain available for effective care of the heart failure patient. With the dramatic improvements witnessed in post-LVAD survival and quality of life just in the last decade, LVAD implantation has now firmly crossed the cusp from experimental rescue surgery to an accepted and essential reality in the care of patients with end-stage heart failure. As LVAD outcomes and technology continue to improve, it is now increasingly apparent that the importance of LVAD implantation as a tool for the successful treatment of heart failure will only increase in the coming years. The progress witnessed in the field of ventricular assist should serve as another demonstration of the important interdependence between cardiac surgeons and mechanical circulatory support technology. With third-generation noncontact centripetal flow pumps available in Europe and in varying degrees of clinical testing here in the United States, further pump miniaturization in development, transcutaneous battery charging on the horizon, and wholly percutaneously inserted devices coming down the pipeline, the future of LVAD implantation has never been brighter. The pace of change in this rapidly developing field will continue to accelerate and decreased patient morbidity will allow for expansion into a broader class of less severe heart failure patients. The immediate and long-term future of heart failure therapies will be shaped by ventricular assist technologies, and the cardiac surgeon will play an essential role in guiding this exciting future development.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Drs. Mulloy and Kern have presented the UVA approach of ventricular assistance. Dr. Kern has done a great deal to make this procedure safe and predictable. It is his treatment regimens that have allowed outstanding results with the use of LVAD. I think the proper focus by both authors has been on RV function. The authors have a great approach, pharmacological support, and some contraindications to LVAD placement.

An important area of focus is that elective left ventricular assist should not be done on acutely ill patients. There are other forms of support that can be used until the patient is stabilized. Someone with gross end-organ failure will not be helped by the present support devices. These patients need to be optimized with more temporary support prior to permanent LVAD implantation.

ILK

56 Surgery for Complications of Myocardial Infarction

Philip W. Carrott, Timothy J. Gardner, and Irving L. Kron

INTRODUCTION

Surgical intervention is often required to manage acute mechanical complications of myocardial infarctions. These mechanical complications, which are responsible for 15% to 20% of deaths after acute myocardial infarction, have historically been considered to include free ventricular wall rupture, acute ventricular septal defects (VSDs), and acute ischemic mitral regurgitation (MR). An acute myocardial infarction complication increasingly considered to be mechanical and amenable to surgical intervention is "pump failure," or cardiogenic shock refractory to maximal medical treatment and revascularization. Surgical techniques used for this complication include ventricular assist device implantation and transplantation.

FREE VENTRICULAR WALL RUPTURE

Incidence and Pathogenesis

Free ventricular wall rupture is found in approximately one-fourth of patients who die within 3 weeks of an acute myocardial infarction. Overall, free wall rupture occurs in up to 11% of patients after acute infarction. Rupture location depends on the site of the infarct, with the rupture tract most commonly occurring between viable and necrotic myocardium. Rupture usually occurs after acute expansion of a transmural infarction. Infarct hemorrhage may play a role because revascularization with thrombolytic therapy but not angioplasty has been found to be independently associated with rupture of both the free wall and the septum.

Clinical Presentation and Diagnosis

Free wall rupture has acute, subacute, and chronic presentations. Acute rupture usually results in death within minutes. Subacute rupture represents 20% to 40% of the cases of free wall rupture and presents as cardiac tamponade progressing to shock. Echocardiography evaluating for a pericardial effusion in the presence of ventricular wall defects in a patient with tamponade physiology after acute infarction is used for diagnosis. Chronic rupture is rare and involves a contained ventricular leak that results in a pseudoaneurysm. Most patients have symptoms of congestive heart failure (CHF), chest pain, or dyspnea, although 12% to 23% are asymptomatic. More than two-thirds of patients have a murmur, and virtually all patients have nonspecific electrocardiogram (ECG) abnormalities. Most patients have cardiomegaly on chest X-ray. Angiography, echocardiography, computed tomography scans, radionuclide scans, and magnetic resonance imaging can be used to identify the pseudoaneurysm.

Natural History

Acute rupture rarely allows time for intervention and is invariably fatal. Patients with subacute rupture have a median survival of 8 hours after symptom onset, with a range from 45 minutes to 6.5 weeks. Only 17 cases of survival without surgery for subacute rupture have been reported. The natural history of chronic rupture is not well defined. Frances et al. identified 290 patients reported in the literature as having a left ventricular pseudoaneurysm, 139 of which had resulted from a myocardial infarction. Of 31 patients treated conservatively, 15 died at <1 week from infarction and the other 16 survived long term, which suggests that chronic pseudoaneurysms are relatively stable.

Preoperative Treatment and Operative Timing

Acute rupture is generally not amenable to any attempts at repair. Emergent operative repair is needed for any patient with a subacute rupture, and the patient must be brought immediately to the operating room. Pericardiocentesis at the time of echocardiography can provide short-term hemodynamic improvement in some patients, allowing some degree of stability while preparing for surgery. Inotropic agents, fluid infusion, vasoconstrictors, and an intra-aortic balloon pump (IABP) can also be used to maintain hemodynamics during transfer to the operating room.

Timing of surgical repair for chronic rupture depends on the interval between infarction and diagnosis. Patients who are within a few months of infarction are thought to have a high risk for rupture and should undergo urgent cardiac catheterization to evaluate the extent of coronary artery disease followed by operative repair of the pseudoaneurysm. The need for surgery in patients who present after an extended period after infarction depends on whether the pseudoaneurysm is >3 cm, expanding, or symptomatic, or if the patient has MR and/or severe coronary artery disease that requires intervention.

Operative Technique

Patients with subacute rupture are rapidly prepared for a standard median sternotomy, with close monitoring of hemodynamics during anesthesia induction because significant hypotension can occur. Close hemodynamic monitoring is also required when the pericardium is decompressed because hypertension can occur and possibly worsen the rupture size and the amount of bleeding. The tear location and status and the patient status determine the need for cardiopulmonary bypass. Cardiopulmonary bypass is instituted in all hemodynamically unstable patients. Bypass is also used if the rupture cannot be exposed adequately without excessive compromise of circulation or if the bleeding from the rupture cannot be controlled or prevent adequate repair.

Subacute rupture can be repaired by various techniques, which should be kept as simple as possible. Sutures should not be placed into friable myocardium. Epicardial repair can be performed by the placement of a patch of pericardium, Dacron, or polytetrafluoroethylene felt over the ventricular defect; the patch is sutured to healthy myocardium along the periphery of the infarct (Fig. 56.1A). The rupture can also be closed directly with horizontal mattress sutures buttressed with Teflon felt strips and a patch secured over the repair (Fig. 56.1B). Biocompatible glue can also be used to further reinforce the patch over the defect (Fig. 56.1C). Simple epicardial repair is not appropriate when an intraventricular defect such as septal rupture or papillary muscle rupture also exists. Operative options in these cases require cardiopulmonary bypass and aortic cross-clamping with infarct excision and defect closure using a patch or infarct exclusion, which are described in detail in the following sections.

A new product, Tachosil (Takeda Pharmaceuticals International GmbH, Zurich, Switzerland), which is a white collagen patch with a fibrinogen and thrombin coating on one side, has been used to buttress free wall rupture in leaking and blown out myocardium without need for further operative patching. The technique was used in six patients with good operative and long-term outcomes. For selected patients with small defects or oozing rupture, this simple repair offers a good option with minimal operative trauma to the myocardium.

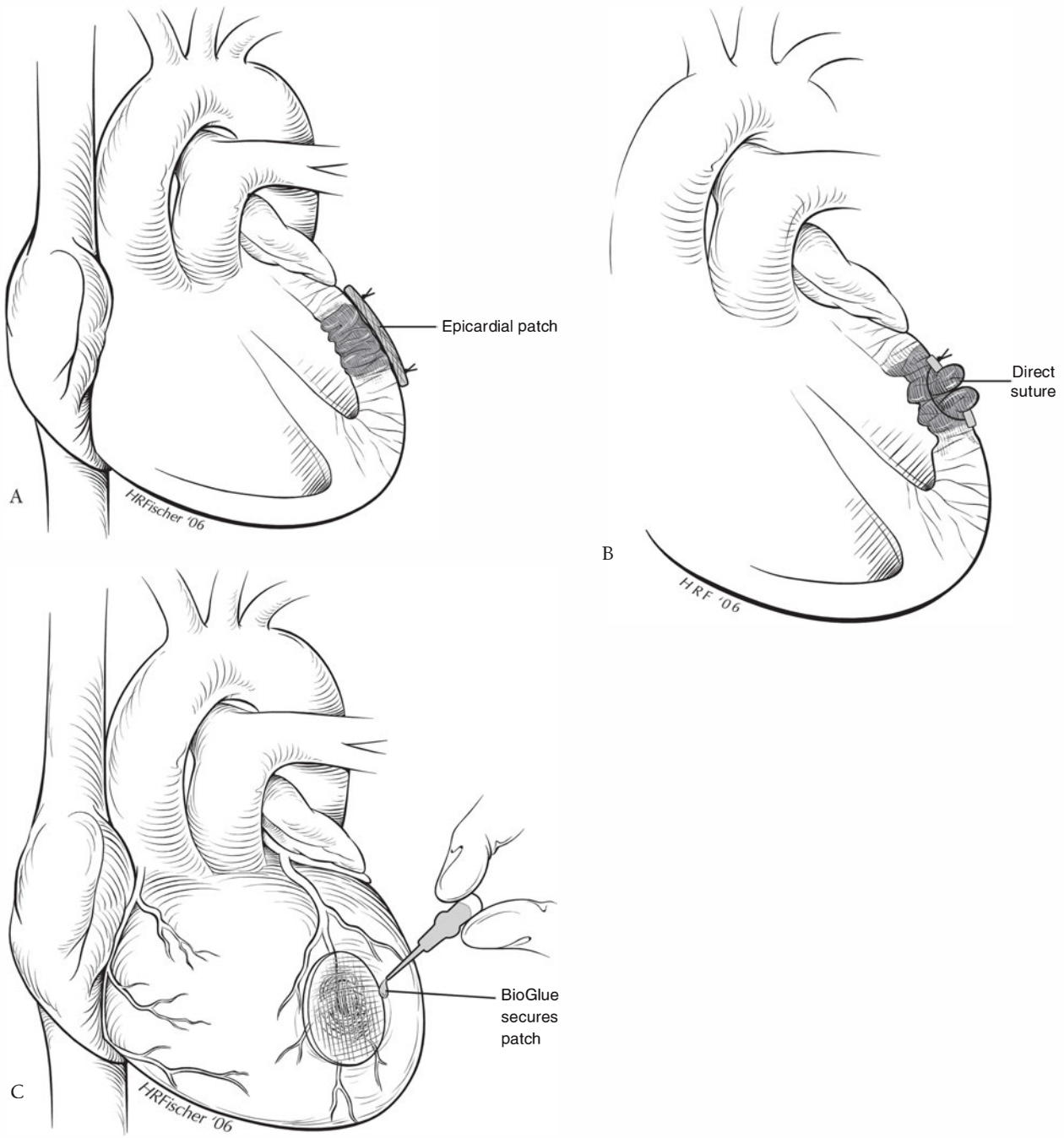


Fig. 56.1. Repair of left ventricular free wall rupture with (**A**) epicardial patching, (**B**) direct suture, and (**C**) application of patch secured to ventricle with BioGlue.

The decision to repair a chronic ventricular rupture is partially based on the presence of either mitral dysfunction or coronary disease. Cardiopulmonary bypass is instituted, and the chronic rupture site is repaired similar to an acute rupture. Anterior pseudoaneurysms can usually be closed directly because of the presence of fibrotic edges. Posterior pseudoaneurysms are closed with a patch so ventricular geometry is not distorted. Even for cases in which the primary purpose of the operation is to repair a large or expanding pseudoaneurysm, cardiopulmonary bypass is indicated to allow adequate exposure for repair and to also prevent the systemic embolization of thrombotic material from within the pseudoaneurysm cavity.

Survival

Most reports of surgical intervention for subacute rupture involve a limited number of patients. These reports, however, indicate that a significant number of patients can be saved from this lethal condition with prompt diagnosis and intervention. In one series, two of five patients who were extremely unstable and underwent emergent repair of a subacute rupture survived long term. In another series of five patients treated similarly, there were four survivors. In a series of six patients treated with a sutureless patch over an 8-year period, five patients survived to leave the hospital. The experience with surgical management of chronic rupture is also limited and largely anecdotal. In one series, 8 of 12 patients treated for a chronic rupture were long-term survivors. All deaths occurred in patients with poor ventricular function or mitral valve disease.

VENTRICULAR SEPTAL DEFECT

Incidence and Pathogenesis

An acute VSD is an uncommon but lethal complication of infarction. A VSD that occurs within 4 to 6 weeks of infarction is considered acute. Even though the septum is involved in up to 70% of all infarcts, acute VSD historically has complicated only 1% to 2% of all infarctions, with recent evidence suggesting that the incidence is now <1%. This decreased incidence may reflect the success of interventions that limit muscle necrosis and prevent transmural infarctions. However, the time between infarction and the VSD occurrence appears to be becoming shorter. Because infarct hemorrhage may play a role in septal rupture, thrombolysis may accelerate the time course of VSDs in those patients in which they occur.

Patients with acute postinfarction VSD have an average age of 62.5 years; the male-to-female ratio is 3:2, although the incidence may be rising in women. Acute VSD usually occurs after occlusion of a coronary artery that results in a full-thickness infarction that is on average larger (25% of the total left ventricular wall mass) than an infarction that is not complicated by a VSD (15%). The infarcts in patients who develop a postinfarction VSD more commonly involve both ventricles than the infarcts in those patients who do not develop a VSD. The typical patient has single-vessel disease with poor collateral flow and presents with an anterior infarct. Although this complication may occur as early as a few hours and as late as a few weeks after infarction, the diagnosis is most commonly made within 4 days of the onset of infarction symptoms. The time of septal rupture correlates with the period when necrotic muscle from the infarction has not been adequately replaced by fibrous connective tissue. Defect size ranges from 0.3 to 4.0 cm, with an average of 1.7 cm.

Historically, acute septal defects were described primarily in the anteroapical septum as a result of left anterior descending artery occlusion. Recent data, however, suggest that the proportion of posterior VSDs is rising, and these now represent one-third to one-half of VSDs. Posterior VSDs result from occlusion of a dominant right coronary artery or a dominant circumflex artery. Defects are classified as either simple or complex. Simple defects are usually located anteriorly. Complex defects are usually located inferiorly and have a worse prognosis. Multiple VSDs occur in 5% to 11% of cases. One-third of cases involve mitral valve regurgitation, which results from either papillary muscle infarction (15% of cases) or left ventricular dysfunction and mitral annular dilation. The MR seen due to left ventricular dysfunction generally resolves with repair of the VSD, whereas papillary muscle rupture requires valve replacement.

Clinical Presentation and Diagnosis

An acute VSD typically presents with recurrent chest pain, a new holosystolic murmur and palpable left sternal thrill, and hemodynamic deterioration a few days after acute infarction. Septal rupture causes left-to-right shunting that can result in heart failure. Clinical presentation varies from an asymptomatic murmur to cardiogenic shock. Symptoms occur due to both ventricular dysfunction and the shunting caused by the VSD. Failure occurring with an anterior VSD is usually the result of both the VSD and extensive left ventricular infarction, whereas failure occurring with a posterior VSD is usually due to extensive right ventricular infarction. Infarct location on the ECG correlates highly with VSD location. The ECG often shows a rightward QRS axis shift and a right-bundle-branch block, and approximately one-third of patients get a transient atrioventricular conduction block before septal rupture.

The clinical appearance of an acute VSD is very similar to that of acute MR. Physical examination and the ECG are used to distinguish between the two entities. The murmur associated with a VSD is most prominent at the left sternal border and is often accompanied by a thrill, whereas the murmur from acute MR is best heard at the apex and has no thrill. In addition, acute MR is more commonly associated with inferior infarctions and no conduction abnormalities. Echocardiography has a very high sensitivity and specificity in identifying the presence, size, and location of a VSD and shows a diagnostic transseptal flow jet, as well as an echo-free area of the septum.

Demonstrating a 9% step-up in oxygen saturation from the right atrium to the pulmonary artery during right heart catheterization with pulmonary artery placement also confirms shunt presence due to VSD.

Natural History

Patients with a postinfarction VSD treated medically have a 1-year mortality as high as 97%. One-fourth of patients die within 24 hours, and 80% die within 4 weeks. Death usually occurs as a result of end-organ failure due to shock.

Preoperative Therapy and Operative Timing

Because early surgical repair of acute VSDs had a very high mortality, surgical repair was previously delayed for 4 to 6 weeks to allow hemodynamic stabilization and fibrosis of the infarct to facilitate suturing around the VSD. Although some patients survived this delayed management, it became clear that only low-risk patients survived to surgical repair. Surgical intervention is now recognized as required before cardiogenic shock results in irreversible end-organ damage, and operative timing for repair is dictated by the patient's hemodynamic status. Completely stable patients, comprising only a

small proportion of patients, should have elective repair sometime during their hospitalization for the acute infarction.

Patients requiring pharmacologic support for heart failure require intervention within 12 to 24 hours of diagnosis. A patient in cardiogenic shock is a true surgical emergency and needs immediate repair. Patients who have already developed multisystem failure or sepsis are extremely at high risk for emergency surgery and likely require further attempts at stabilization, with antibiotics as needed, before attempted repair. As more experience with ventricular assist devices evolves, their use in this complex population as a temporizing measure is expanding. The Impella 5.0 catheter (Abiomed, Inc., Danvers, MA) is most useful since it can be placed through a femoral artery cut-down and does not require opening the chest for cannulation. As the name implies, the maximum flow is 5 l/min, which should help to off-load the right ventricle and allow the myocardium to heal. A delayed repair may then be undertaken with stronger myocardial tissue. Case reports describe deferring operative repair for around 14 days with a mortality rate of 40%.

The preoperative treatment goal is to divert blood systemically and maintain cardiac output, blood pressure, and coronary flow. Diuretics, inotropes, and vasodilators can minimize the left-to-right shunt; however, vasodilators are often not tolerated due to systemic hypotension. Vasoconstrictors increase afterload and can worsen left-to-right shunting and should be avoided if possible. An IABP may improve cardiac output by reducing afterload and decreasing shunting and is mandatory in management of VSD patients who present in shock. However, IABP use has peak improvement within 24 hours and then no further benefit, so surgery should not be delayed beyond this time.

The role of preoperative coronary angiography is not completely clear; delaying surgical repair to perform cardiac catheterization during a period of patient instability is risky. In addition, the use of coronary angiography and coronary artery bypass grafting did not show improved short-term or long-term survival in a study of 179 patients with postinfarction VSD. However, routine coronary angiography detected significant coronary artery disease beyond the vessel that resulted in the acute infarction in 28 patients in another study of 54 patients with acute postinfarction VSDs. These patients underwent concomitant revascularization and experienced similar early and late survival compared with patients without associated coronary disease, suggesting that revascularization controlled the added risk of associated coronary disease. In another recent multi-institutional review, 42 of 65 patients who had coronary bypass grafting at the same time as VSD repair had significantly better survival both in the short-term and at 4 years. Given that coronary angiography can be performed very rapidly, a reasonable policy is to perform angiography with a limited amount of contrast in all patients who can be temporarily stabilized. Patients who are in severe shock should proceed directly to surgery for VSD repair.

Operative Technique

Several techniques can be used to repair acute postinfarction VSDs. Regardless of the specific technique utilized, a number of technical guidelines have been developed. A median sternotomy is performed with expeditious establishment of cardiopulmonary bypass using bicaval venous drainage. Cardioplegia should be first administered antegrade followed by retrograde via the coronary sinus to achieve optimal myocardial protection. In addition, patients with critical coronary stenoses identified preoperatively should undergo revascularization prior to VSD repair in order to maximize myocardial protection. A transinfarct approach to the VSD should be used. Because surgical success is completely dependent on adequate VSD closure, thorough evaluation for multiple defects must be performed both preoperatively and intraoperatively. In patients with associated significant MR, mitral valve replacement is needed only if frank papillary muscle rupture has occurred. The septal defect and the infarctectomy must be closed without tension, which generally necessitates the use of prosthetic material. Finally, all suture lines must be buttressed with pledgets or Teflon felt.

Apical septal ruptures can be repaired using the technique of apical amputation described by Daggett (Fig. 56.2). An incision through the infarct is made, and all necrotic muscle of both ventricles and the septum is debrided. The remaining apical parts of the ventricles and septum are reapproximated by a row of interrupted mattress sutures using buttressing strips of felt.

Operative procedures to repair acute anterior VSDs include both infarctectomy and infarct exclusion. For the infarctectomy technique, a transinfarct incision is made, and infarct debridement with thorough trimming of left ventricular margins to viable muscle with more conservative trimming of the right ventricle is performed to simply allow adequate visualization of the septal defect margins (Fig. 56.3A). Small defects can be closed using a plication technique (Fig. 56.3B). The VSD is closed by approximating the anterior edge of the septum to the right ventricular wall with mattress sutures buttressed with felt strips. The ventricular incision is also closed with mattress sutures over felt. Larger defects require reconstruction of the septum and ventricular walls with prosthetic material to reduce postrepair tension (Fig. 56.3C). Sutures are first passed from right to left through the septum, and then additional sutures are passed from the epicardial surface to the endocardial surface of the right ventricle. The anchored sutures are then passed through the patch and tied to close the VSD with the patch secured on the left side of the septum. The debrided infarct of the free ventricular wall is similarly closed with a patch as needed.

Infarct excision can distort ventricular geometry and has high potential for right heart dysfunction. David developed a procedure involving infarct exclusion rather than excision. A patch sutured to healthy myocardium excludes both the infarct and the VSD, preserving left ventricular geometry and function (Fig. 56.4). A transinfarct incision is made parallel to the left anterior descending coronary artery in the apex of the left ventricle. A patch tailored to the shape of the left ventricular infarction is sutured first to the endocardium of a noninfarcted portion of the septum and then to the noninfarcted endocardium of the anterolateral ventricular wall. The ventriculotomy is then closed over strips of felt.

The closure of posterior septal defects is technically challenging. The left side of the heart should be vented via the right superior pulmonary vein and the heart retracted as if preparing for a posterior bypass. As with anterior defects, repair can be performed with infarctectomy or infarct exclusion techniques. For the infarctectomy technique, a transinfarct incision is made, and the left ventricular portion of the infarct is thoroughly debrided to expose the septal defect. The papillary muscles should be inspected, and mitral valve replacement should be performed if papillary muscle rupture has occurred. As with anterior VSD repair, the portion of the right ventricle involved in the infarct is more conservatively trimmed to simply allow adequate visualization of the septal defect margins (Fig. 56.5A). Septal defects close to the adjacent ventricular free wall and not associated with significant loss of septal tissue can be repaired by approximating the septal rim of the posterior defect to the

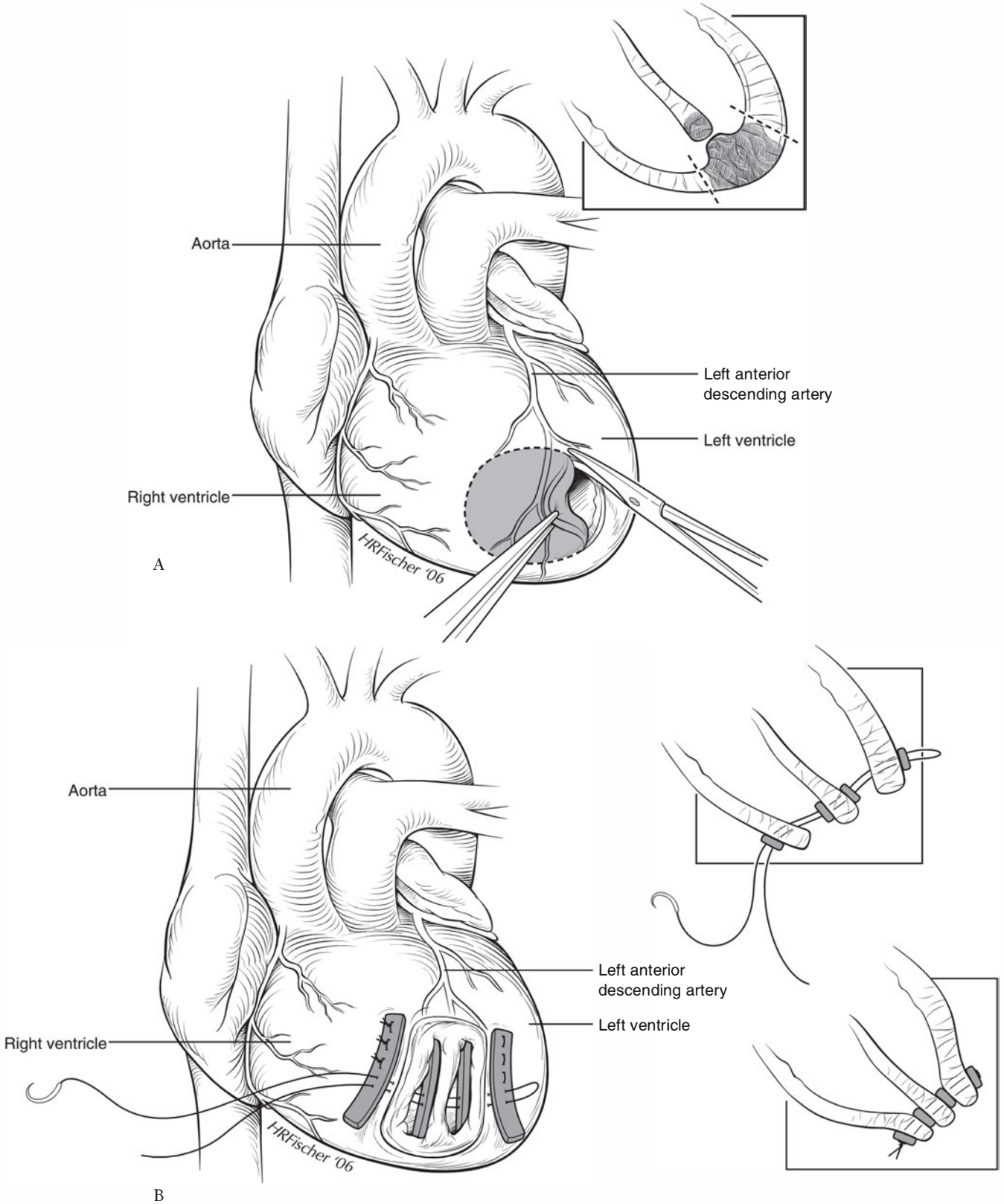


Fig. 56.2. Repair of apical ventricular septal defect. (**A**) The infarcted apex of the left and right ventricles is amputated to expose the septal defect. (**B**) The left ventricle, septal apex, and right ventricle are reapproximated with interrupted, buttressed mattress sutures.

right ventricular free wall using buttressed mattress sutures (Fig. 56.5B). Larger defects require patch closure of both the septal defect and the infarct (Fig. 56.5C). Sutures are first passed from right to left through the septum, and then additional sutures are passed from the epicardial surface to the endocardial surface of the right ventricle. All placed sutures are then passed through the patch and tied to close the VSD.

David's infarct exclusion method can also be used to repair posterior defects (Fig. 56.6).

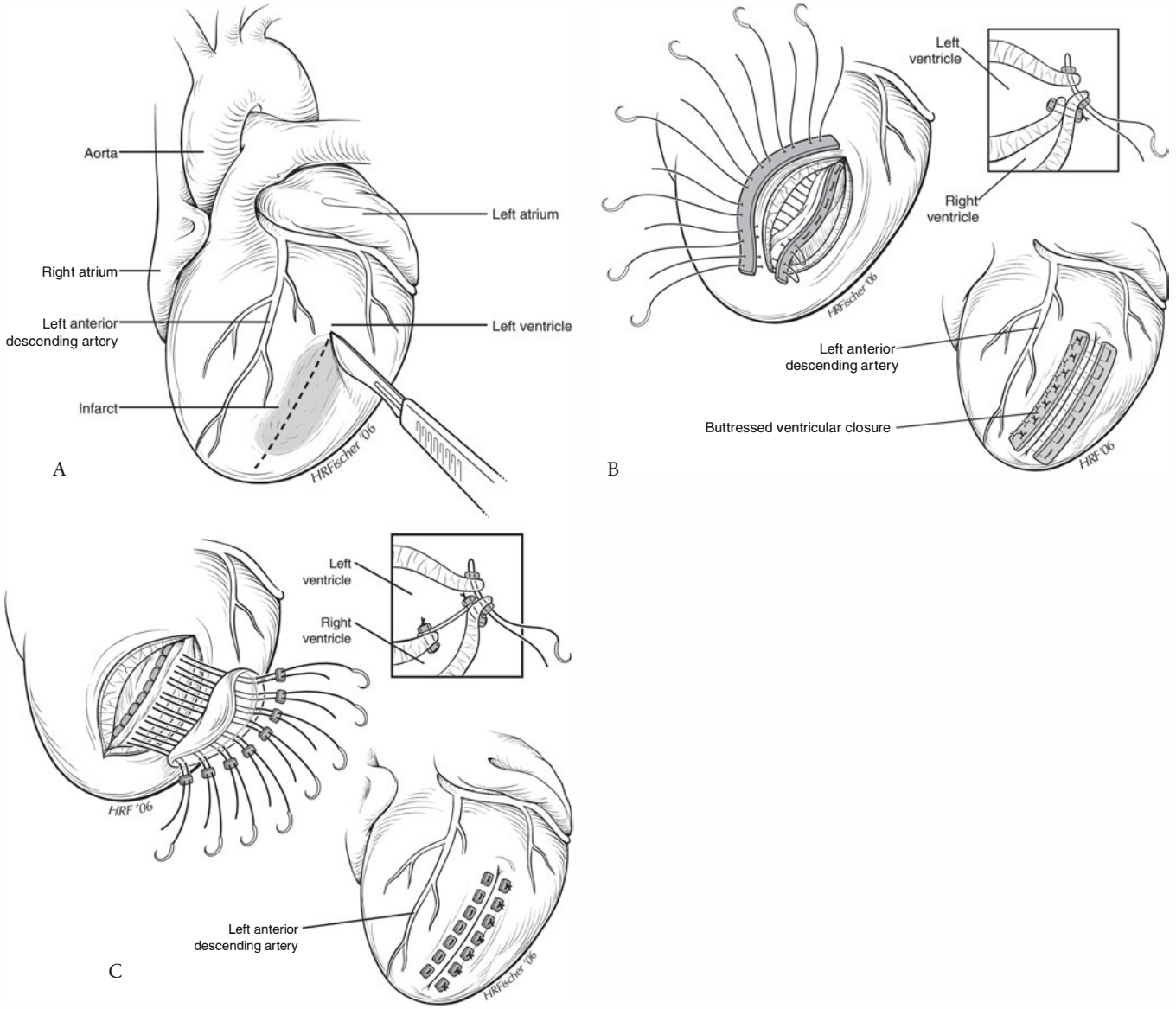


Fig. 56.3. (**A**) A transinfarct incision is made in the left ventricle parallel to the left anterior descending coronary artery to repair an anterior postinfarction ventricular septal defect. (**B**) A small defect is repaired using the plication technique. The septal defect is closed by first approximating the anterior edge of the septum to the right ventricular wall with mattress sutures buttressed with felt strips, followed by buttressed closure of the ventricular incision. (**C**) Prosthetic material is used for the reconstruction of the septum to reduce postrepair tension in repair of a large defect. The patch is first secured to the left side of the ventricular septum using pledgeted sutures passed from right to left through the septum and then the patch. Sutures are then passed from the epicardial surface to the endocardial surface of the right ventricle and then through the patch and tied. The debrided infarct of the free ventricular wall is closed with buttressed sutures.

An incision through the midportion of the inferior wall a few millimeters from the posterior descending artery is extended proximally toward the mitral annulus and distally toward the apex of the ventricle. The base of a patch tailored in a triangular shape is sutured to the fibrous annulus of the mitral valve with a continuous suture starting at a point corresponding to the level of the posteromedial papillary muscle and moving medially toward the septum until reaching noninfarcted endocardium. The suture is then interrupted with trimming of any excess patch material. The medial margin of the triangular patch is sewn to healthy septal endocardium with a continuous suture. The lateral side of the patch is sutured to the posterior wall of the left ventricle along a line corresponding to the medial margin of the base of the posteromedial papillary muscle. Sutures placed into infarcted myocardium require full-thickness bites buttressed with felt on the epicardial surface. The infarct incision is closed in two layers of buttressed sutures, leaving the infarcted right ventricular wall undisturbed.

Another approach for closing small posterior defects is through the right atrium. The ventriculotomy is avoided, but the exposure and repair necessitate limiting this approach to smaller defects. In addition, there are multiple reports of, which we have utilized as well, the use of a percutaneous closure device for small postinfarction VSDs. This should be regarded as a temporizing measure for an extremely unstable patient, as the friable nature of the necrotic tissue does not hold the closure device well. Despite reports in the literature,

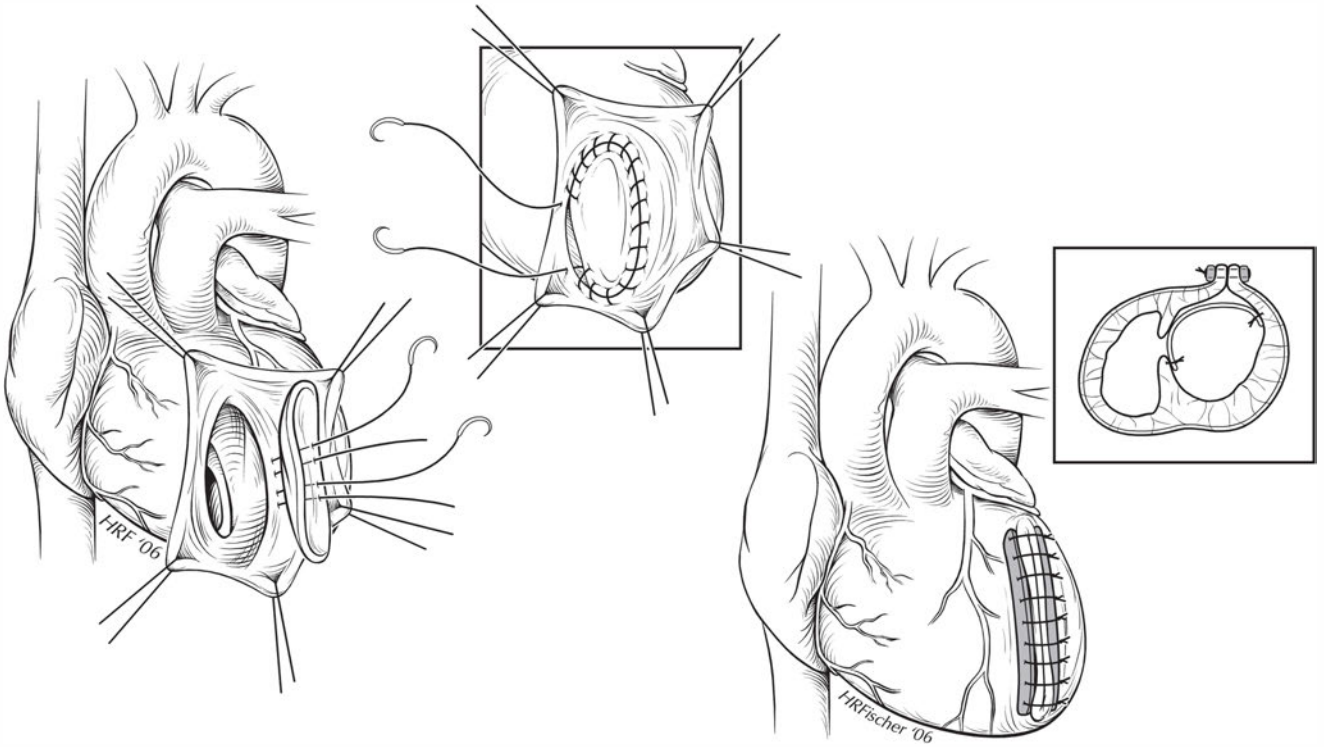


Fig. 56.4. Repair of an anterior postinfarction ventricular septal defect using the infarct exclusion technique. After a transinfarct incision is made to expose the defect, a patch that is tailored to the shape of the left ventricular infarction is sutured to the endocardium of the noninfarcted portion of the septum and then sutured to the noninfarcted endocardium of the anterolateral ventricular wall. The ventriculotomy is then closed with buttressed sutures.

we have seen very limited success with this minimally invasive approach.

Survival

Although survival for the treatment of an acute postinfarction VSD has improved significantly over time, operative mortality in most large series is between 30% and 50%. In the largest published series, involving 179 patients, survival at 1, 5, and 10 years was found to be 60%, 49%, and 31%, respectively. The observation that mortality is not significant between 1 and 5 years indicates that those patients who survive the perioperative period have reasonable long-term survival. The technique of infarct exclusion has been associated with both a low operative mortality of 14% and an actuarial survival of 66% at 6 years, which is significantly better than all previous series.

ACUTE ISCHEMIC MITRAL REGURGITATION

Incidence and Pathogenesis

Ischemic MR is mitral insufficiency caused by myocardial infarction. From 17% to 55% of patients have either echocardiographic evidence of MR or a new mitral systolic murmur early after acute infarction. The MR is mild or disappears in many patients but is moderate to severe or severe in as many as 4% of patients. Overall, acute severe MR complicates 0.4% to 0.9% of acute myocardial infarctions.

Acute ischemic MR is actually a myocardial problem, not an intrinsic valve problem. Of the six components of the mitral valve (the leaflets, chordae tendineae, annulus, papillary muscles, left ventricle, and left atrium), the leaflets, chordae, and annulus are not acutely affected by a myocardial infarction. Myocardial injury involving papillary muscle rupture or displacement causes acute severe MR. The posterior papillary muscle, which has a single blood supply from the circumflex coronary artery, is involved 6 to 12 times more frequently in acute severe MR than the anterior papillary muscle, which is supplied by both the left anterior descending and circumflex coronary arteries. The MR is also usually more severe when the posterior papillary muscle is affected by the infarction.

Papillary muscle rupture results in a flail valve leaflet and generally occurs 2 to 7 days after infarction, with a mean of 4 days. Trunk rupture leads to complete dehiscence of the papillary muscle and the rapid onset of severe MR, pulmonary edema, and shock. Tip rupture of smaller papillary muscle heads also causes severe MR but results in less severe clinical deterioration initially, although the subsequent course is unpredictable. Complete trunk rupture usually occurs within the first week after the acute infarction, whereas partial rupture can be delayed for as long as a few months. Both types of rupture are highly lethal conditions without surgical intervention.

Acute ischemic MR due to papillary muscle displacement results from a number of small geometric changes in left ventricular shape, size, and wall motion that lead to incomplete coaptation of intrinsically normal leaflets. Papillary muscle ischemia alone usually causes a murmur but not severe MR with significant hemodynamic compromise. In the absence of papillary muscle rupture, left ventricular wall ischemia in conjunction with papillary muscle ischemia is required to cause severe MR. Severe MR resulting from papillary muscle displacement, although generally more compatible with short-term survival than severe MR resulting from rupture, can cause a impairment of cardiac function similar to papillary muscle rupture and also almost always requires surgical intervention to ensure long-term survival.

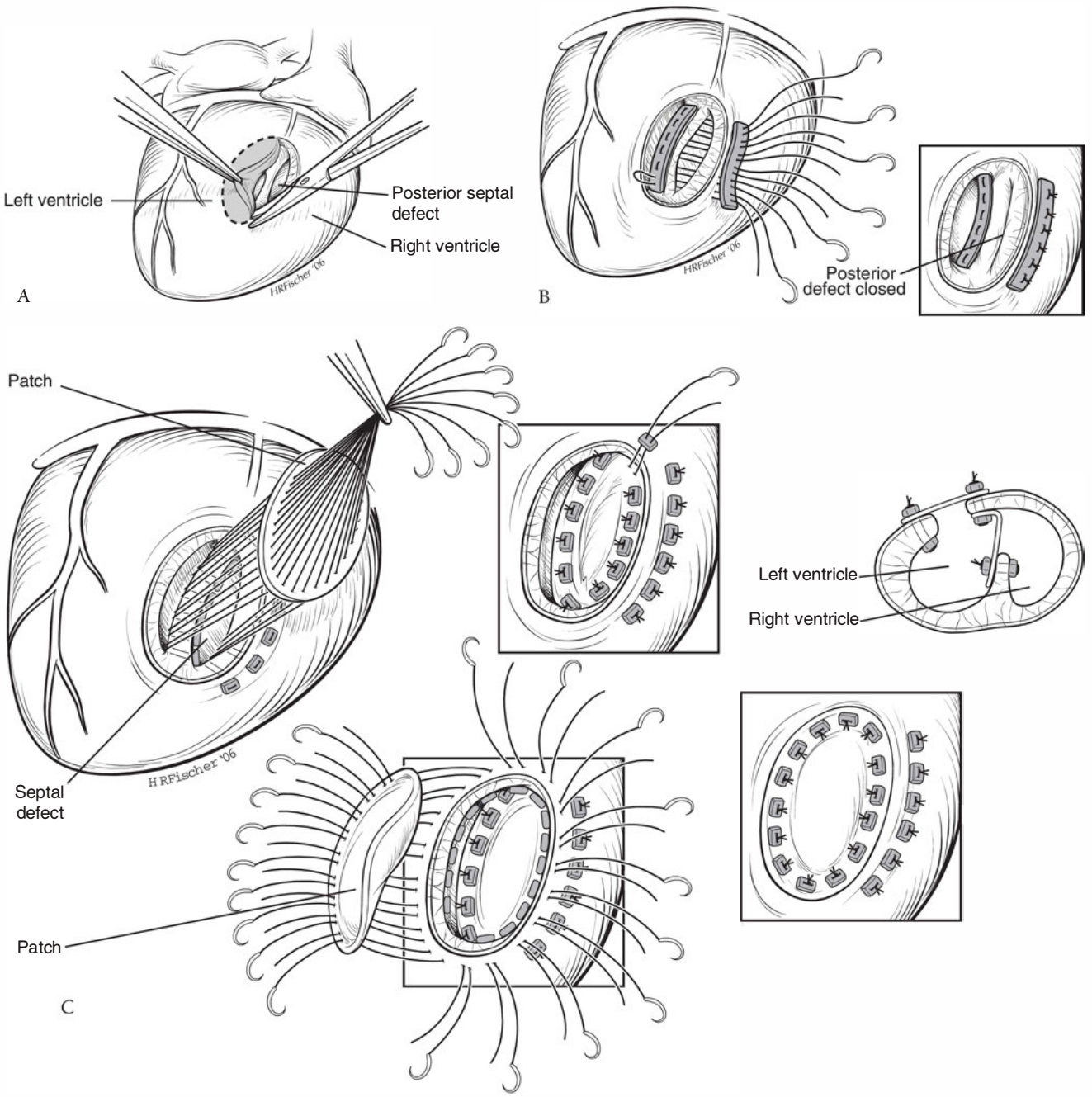


Fig. 56.5. (**A**) A transinfarct incision is made, and the left ventricular portion of the infarct is debrided to expose the septal defect. (**B**) The infarctectomy technique is used to repair small posterior septal defects. The septal rim of the posterior defect is approximated to the right ventricular free wall using buttressed mattress sutures. (**C**) Large posterior defects are repaired using patch closure with the infarctectomy technique. Sutures are first passed from right to left through the septum, and then additional sutures are passed from the epicardial surface to the endocardial surface of the right ventricle. All placed sutures are then passed through the patch and tied to close the ventricular septal defect. The debrided infarct of the free ventricular wall is then closed with a patch and buttressed sutures.

Both papillary muscle rupture and displacement result in regurgitant flow into the left atrium during systole, with volume overload of the left atrium during systole and the left ventricle during diastole. The regurgitant flow causes an increase in stroke volume and a decrease in cardiac output. In acute MR, there is usually a marked increase in pressure in the noncompliant left atrium. This pressure increase actually helps to decrease the left ventricular–left atrial pressure gradient and may decrease the regurgitant volume.

Clinical Presentation and Diagnosis

Patients who develop acute MR after acute infarction typically have acute chest pain or shortness of breath with the appearance of a systolic murmur 2 to 7 days after the initial infarction. Acute severe MR due to either papillary muscle rupture or displacement leads to pulmonary edema and cardiogenic shock that is usually more abrupt and severe than that resulting from an acute VSD. Patients have a mean age of 60 years, usually have a history of hypertension, and

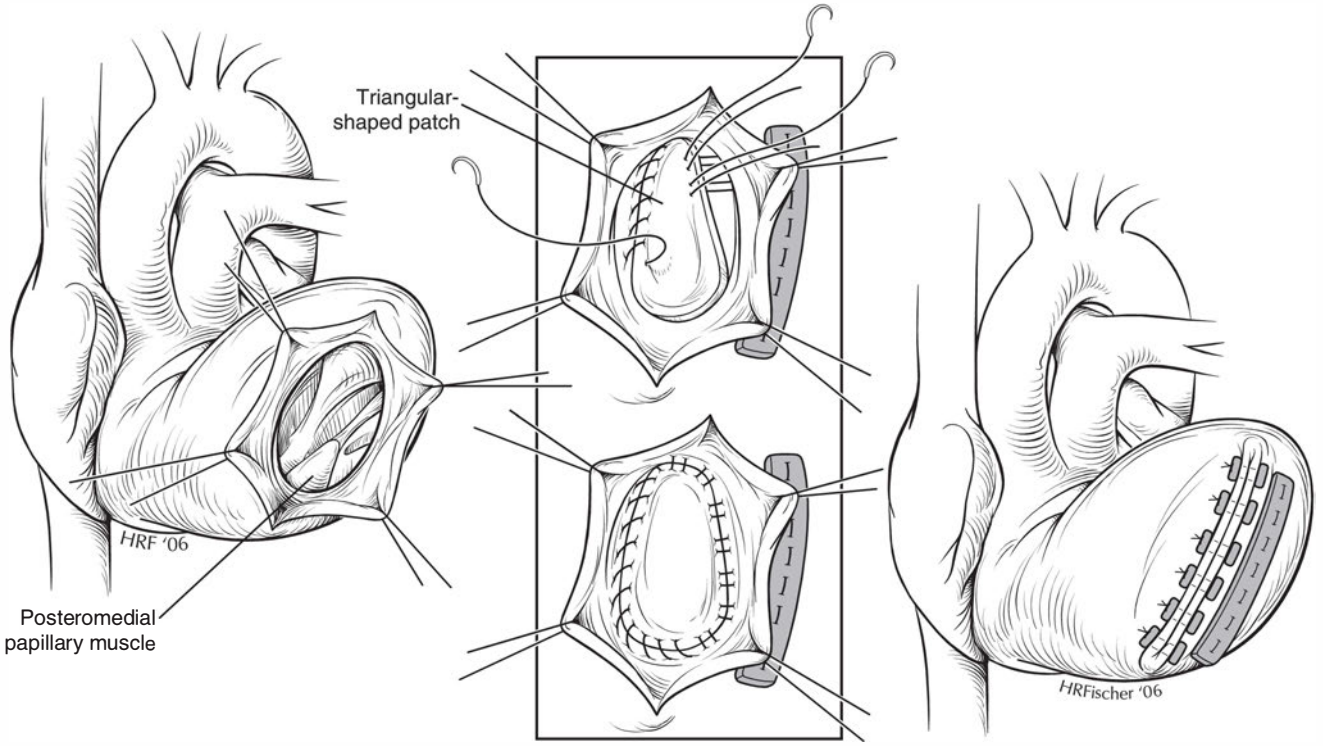


Fig. 56.6. Repair of a posterior postinfarction ventricular septal defect using the infarct exclusion technique. An incision is made in the inferior wall of the left ventricle a few millimeters from the posterior descending artery. The base of a triangular patch is sutured to the fibrous annulus of the mitral valve with a continuous suture from the level of the posteromedial papillary muscle toward the septum until one reaches noninfarcted endocardium. The suture is then interrupted with trimming of any excess patch material. The medial margin of the triangular patch is sewn to healthy septal endocardium with a continuous suture. The lateral side of the patch is sutured to the posterior wall of the left ventricle along a line corresponding to the medial margin of the base of the posteromedial papillary muscle. The infarct incision is then closed in two layers of buttressed sutures.

are more commonly male. Chest X-rays usually show a normal-sized heart and pulmonary edema. Right heart catheterization generally shows elevated pulmonary artery pressures and tall V waves, but there is no oxygen step-up in the pulmonary artery.

As described, the clinical appearance of acute MR is very similar to that of an acute VSD. Again, physical examination and the ECG can distinguish acute MR from an acute VSD. The holosystolic murmur associated with acute MR is best heard at the apex, has no thrill, and is associated with an atrial gallop (S_4), whereas the murmur associated with a VSD is most prominent at the left sternal border and is commonly accompanied by a thrill. Acute MR is more commonly associated with an inferior infarction and the absence of conduction abnormalities than an acute VSD.

Echocardiography with Doppler color flow mapping is the preferred diagnostic tool for acute severe MR. Transthoracic echocardiography is useful for assessing the degree of MR and can confirm wall motion abnormalities. In the case of papillary muscle rupture, echo can demonstrate flail mitral leaflets and also possibly a mass attached to the chordae, which is the ruptured portion of the papillary muscle. Even if a flail leaflet is not seen, the diagnosis of acute papillary muscle should be suspected if intact systolic function is seen in the clinical setting of acute pulmonary edema and shock. Transthoracic echocardiography can be a reliable diagnostic test for direct transfer to surgery when there is no question in the diagnosis. Transesophageal echocardiography is the tool of choice when the diagnosis is in question because it more definitively assesses the degree of MR and wall motion abnormalities while also assessing the posterior papillary muscle. Transthoracic echocardiography also does not always reliably distinguish papillary muscle rupture versus displacement.

Natural History

Acute severe ischemic MR is associated with a mortality of 24% at 30 days, 42% at 6 months, and 52% at 1 year. For an acute papillary muscle rupture, approximately 50% of patients die within 24 to 48 hours without surgical intervention.

Preoperative Therapy and Operative Timing

Acute ischemic MR that is not severe requires no specific treatment other than that used to treat the infarction. Immediate surgery is indicated for patients with severe MR and cardiogenic shock or CHF. Attempts are made to stabilize hemodynamics while surgery is arranged. As the size of the regurgitant orifice is increased by increased afterload and volume overload, inotropes, vasodilators, and an IABP can decrease the regurgitant flow and increase cardiac output.

As with acute VSDs, the role of preoperative coronary angiography for acute severe MR is not entirely clear. Delaying surgical repair to perform cardiac catheterization in a potentially unstable patient can be risky. Revascularization also prolongs both surgery and the duration of cardiopulmonary bypass and likely has no impact on immediate survival, particularly in the case of a ruptured papillary muscle, although it may be beneficial in those patients who cannot be weaned off bypass. However, given the etiology of myocardial infarction as the cause of MR and the fact that approximately

half of the patients with acute ischemic MR have three-vessel coronary disease, identifying and bypassing associated coronary lesions can improve long-term prognosis. Given that coronary angiography can now be performed rapidly so that surgical intervention is not delayed, a reasonable policy, as with acute VSDs, is to perform angiography with a limited amount of contrast in all patients who can be temporarily stabilized medically or with the use of a balloon pump. Patients who cannot be stabilized must go directly to surgery.

Operative Technique

Transesophageal echocardiography must be used in the operating room to guide and assess surgical results. A standard median sternotomy is performed with expeditious establishment of cardiopulmonary bypass. Bicaval cannulation with caval tourniquets is used to minimize systemic venous return, which could warm the heart and decrease myocardial protection during aortic cross-clamping. An alternative approach such as via a right thoracotomy with bypass established by peripheral cannulation is not appropriate in these emergency situations, except possibly in cases in which a patient has had a previous sternotomy and is known to have patent midline bypass grafts. The patient is cooled systemically to 28°C. The heart is arrested with cold cardioplegia solution delivered retrograde via the coronary sinus after the aorta is cross-clamped, and cold blood cardioplegia is delivered both antegrade and retrograde intermittently to maximize myocardial protection. The left atrium is opened to decompress the heart, and patients with associated critical coronary stenoses should then have the distal anastomoses completed.

Mitral valve exposure must allow full evaluation of all components of the valve and the subvalvular apparatus so that a decision to either repair or replace the valve can be made. The sternotomy must be well centered, with the right side of the pericardium suspended to sternal fascia. An incision is made in the left atrium near the right superior pulmonary vein, parallel to the interatrial groove, extended behind the superior vena cava and below the inferior vena cava, which must be fully dissected (Fig. 56.7). The superior vena cava is dissected to the level of the innominate vein to maximize mitral exposure. The azygos vein is divided as needed during dissection of the superior vena cava to prevent tearing due to traction during valve exposure.

Although the feasibility of valve repair should be assessed, repair in the setting of acute severe MR is very difficult. Given that these patients are usually in extremis, the possible benefits of repair instead of replacement are generally not worth the risk that surgery and bypass time could be significantly prolonged if revising or even replacing the initial repair is ultimately required. In the case of a ruptured papillary muscle, repair by reimplantation of the ruptured muscle with the placement of sutures into freshly ischemic, friable tissue is quite precarious and generally not advisable, although sometimes for distal partial papillary muscles the disrupted fibrous tip of the papillary segment can be reimplanted into an adjacent noninfarcted papillary muscle or adjacent noninfarcted ventricular myocardium (Fig. 56.8).

Because of the difficulties in performing mitral valve repair, mitral valve replacement is almost always required for patients in the setting of acute severe ischemic MR. As many chordal attachments to the annulus as possible should be preserved when replacing the valve to maintain the mitral subvalvular apparatus and optimize postoperative left ventricular geometry and function (Fig. 56.9A). Although the need for chronic anticoagulation and questions of valve durability may not be particularly great concerns in the setting of emergency surgery, the surgeon must choose between a mechanical valve and a bioprosthetic valve. Some recommend that a mechanical valve be used in patients already chronically anticoagulated. Otherwise, the surgeon must consider the patient's age and comorbid conditions to evaluate whether the patient is likely to outlive a bioprosthetic valve if he/she survives his/her current state. The valve chosen is sutured to the annulus with interrupted sutures (Fig. 56.9B and 56.9C). For a bioprosthetic valve, the sutures can be passed from the ventricular to the atrial side within the annulus. For a mechanical valve, the sutures are placed from the atrial to the ventricular side. The atrium is closed with a running suture, and proximal coronary anastomoses are completed.

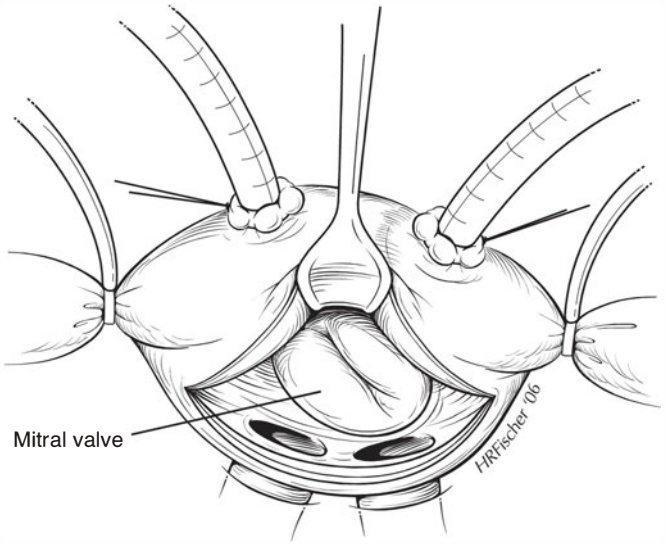


Fig. 56.7. A transverse left atrial incision made at the level of the right superior pulmonary vein and extended behind the superior vena cava and below the inferior vena cava fully exposes the mitral valve.

Survival

The 30-day mortality for surgical repair for acute severe MR is 18% to 27%. The 1-, 5-, and 10-year survival rates are 75% to 81%, 65% to 68%, and 32% to 56%, respectively. The relatively stable long-term mortality observed in these series indicates that although this condition is highly lethal, left ventricular function is usually maintained, particularly in the case of a ruptured papillary muscle, and surgical correction can indeed be life-saving. Operative risk factors for mortality include advanced age, preoperative shock, other comorbidities, infarction size, and operative delay.

PUMP FAILURE

Incidence and Pathogenesis

Cardiogenic shock occurs in 6.7% of patients after acute myocardial infarction, and predominant left ventricular failure is the cause of shock in >75% of these cases. Cardiogenic shock results from a cycle of progressive myocardial damage and necrosis beyond the initial infarct. The overall incidence is decreasing due to better medical and interventional

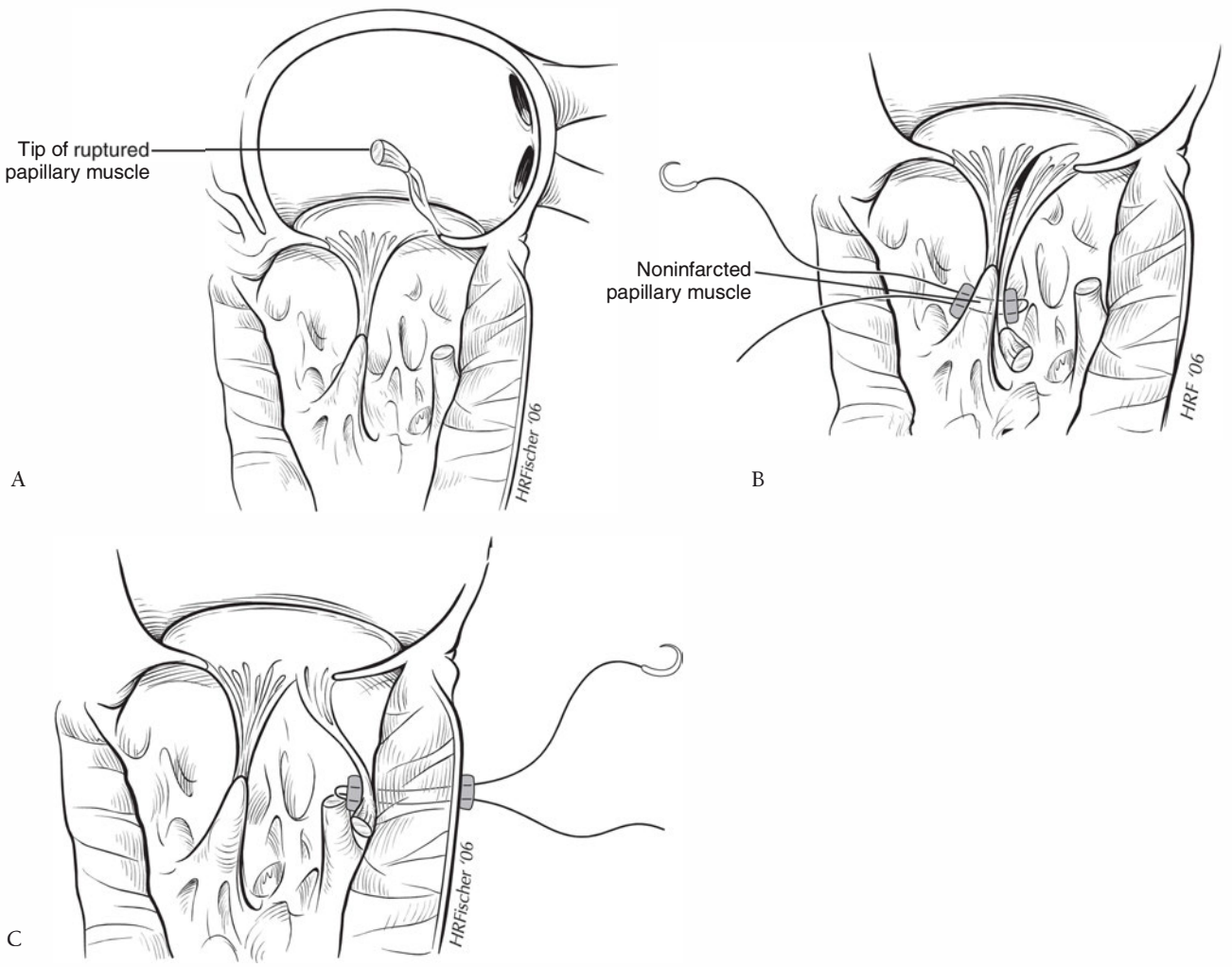


Fig. 56.8. The fibrous tip from (**A**) a ruptured papillary muscle head can be reimplanted into either (**B**) an adjacent noninfarcted papillary muscle or (**C**) adjacent noninfarcted left ventricular free wall.

treatments for an acute myocardial infarction. Failure occurs when left ventricular function is <30% of normal or when the abnormally contracting segment is >25% of the left ventricular circumference. Complete regional myocardial ischemia usually results in only 60% to 70% necrosis of the cardiomyocytes in the ischemic area, which probably explains why the decrease in function seen in these instances is only transient for 24 to 72 hours in two-thirds of cases. When the damage involves <35% to 40% of the left ventricle, the usual clinical scenario is left ventricular dysfunction and pulmonary congestion but no evidence of end-organ hypoperfusion. Cardiogenic shock occurs when >35% to 40% of the left ventricle is involved in the infarct.

Clinical Presentation, Diagnosis, and Natural History

Patients in cardiogenic shock have left ventricular dysfunction and end-organ hypoperfusion manifested by oliguria, decreased mental status, and cold, clammy skin. Pulmonary congestion may or may not be present. Cardiac index is <1.8 l/min/m^2, systolic blood pressure is generally <80 to 90 mmHg, and pulmonary capillary wedge pressure is >18 mmHg. Echocardiography shows severe left ventricular dysfunction without evidence for any of the mechanical complications described. Patients with persistent cardiogenic shock after acute myocardial infarction despite maximal therapy have mortalities of 35% to 80%.

Preoperative Treatment and Operative Timing

The treatment goal of a patient in cardiogenic shock is to increase both coronary and end-organ perfusion and maintain oxygenation. Volume is used to keep the pulmonary capillary wedge pressure >15 mmHg. Inotropes and vasopressors are used as needed to maintain blood pressure. If tolerated, vasodilators are useful in reducing afterload. An IABP reduces afterload to decrease left ventricular work and myocardial oxygen demand while diastolic aortic pressure is increased to improve coronary blood flow to the myocardium. Any patient who does not respond to these measures must be taken for cardiac catheterization and restoration of coronary blood flow with either angioplasty and/or stenting or surgically with coronary artery bypass grafting. Revascularization does not significantly reduce mortality at 30 days but does have a significant survival benefit at 6 months. Patients who remain in cardiogenic shock refractory to these maximal measures require additional circulatory assistance to prevent otherwise certain death. A new treatment paradigm in suitable patients with "pump failure" is implantation of a ventricular assist device, which unloads the left ventricular to greatly reduce myocardial oxygen consumption and may salvage critical myocardial mass and act as a bridge to

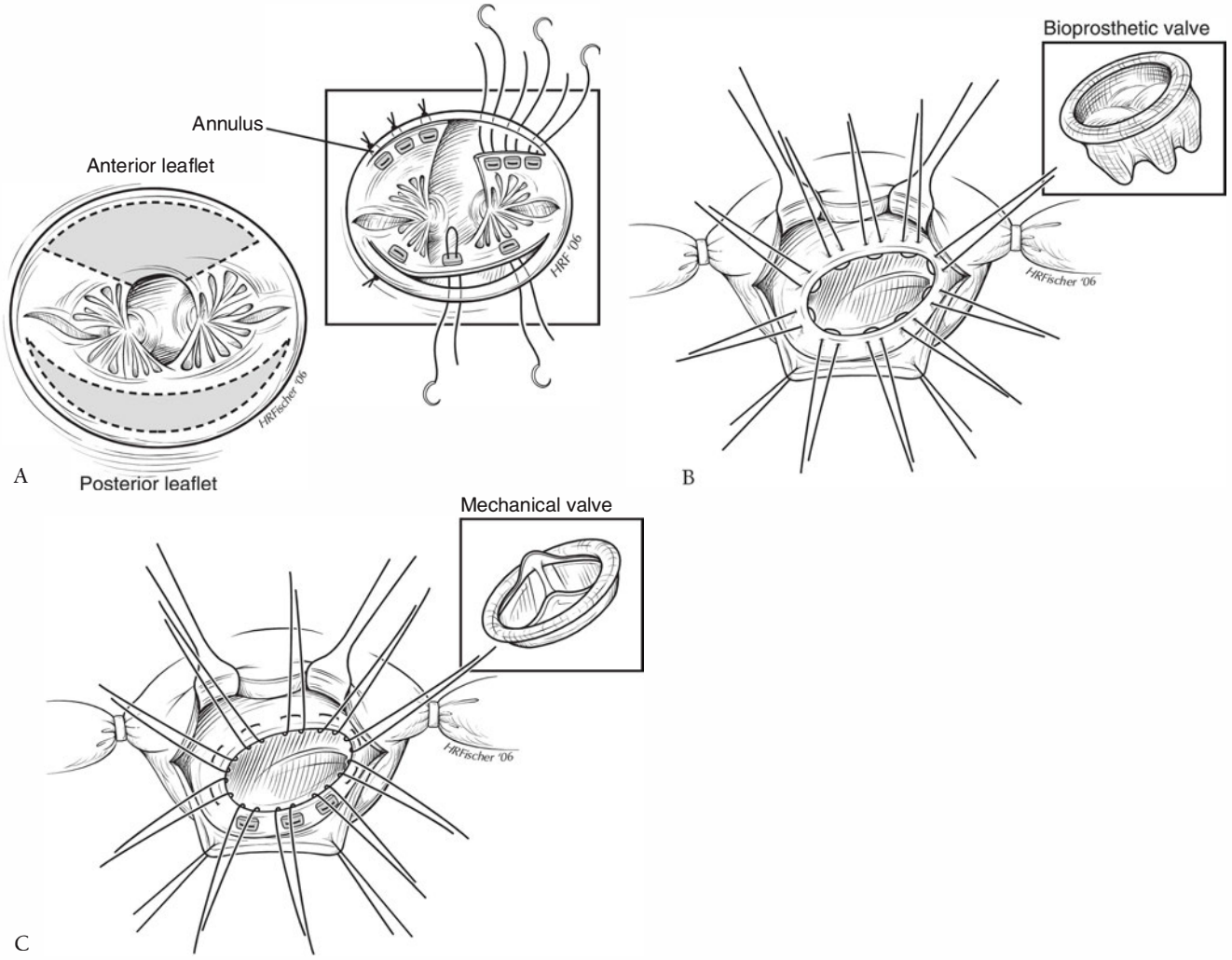


Fig. 56.9. (**A**) Preservation of chordal attachments during mitral valve replacement. The central triangular segment of the anterior leaflet is excised, and residual leaflet is resuspended to the annulus using sutures that are securing the prosthetic valve to the annulus. An ellipse is excised from the posterior leaflet, and the free edge of the leaflet is reattached to the annulus, also with the sutures securing the valve. (**B**) Sutures are placed from ventricle to atrium to secure bioprosthetic valves to the mitral annulus. (**C**) Sutures are placed from atrium to ventricle to secure mechanical valves to the mitral annulus.

recovery (Fig. 56.10A). For those patients who do not recover adequate myocardial function, this technique can act as a bridge to transplantation. The ventricular assist device is implanted as quickly as possible when it becomes clear that the patient's status is not improving with maximal conventional therapy; otherwise, univentricular failure can progress to biventricular failure, and end-organ dysfunction may become permanent.

Operative Technique and Survival

A median sternotomy is performed, and the patient is prepared for bypass and heparinized. Device preparation is performed according to the manufacturer's protocols. The outflow tract for the assist device is sewn into the aorta; this can either be done before institution of bypass using a side-biting aortic clamp to reduce bypass time or after instituting bypass if partial clamping off bypass is not tolerated by the patient. The assist device inflow tract is sewn into the heart while on bypass. The inflow tract can be sewn into either the left atrium or the left ventricle. Left ventricular cannulation may prevent left ventricular stasis and therefore reduce the risk of subsequent thrombus formation. If device flows are not adequate, a right ventricular assist device should also be implanted. Anticoagulation is reversed at the end of the case, and heparin is started 24 hours later if significant post-device insertion bleeding has not occurred.

Short-term (days to weeks), intermediate-term (weeks to months), and long-term (months to years) ventricular assist devices are available. Patients suffering from refractory postinfarction cardiogenic shock generally should have a short-term device such as the Abiomed AB5000 (Abiomed, Inc., Danvers, MA) or CentriMag (Thoratec, Pleasanton, CA) initially implanted. These devices can be implanted at virtually any cardiac center, although a tertiary care institute that performs heart transplantations should be available for patient transfer if indicated. Assist-device weaning is begun after the patient begins to demonstrate end-organ recovery. Device flows are reduced with minimal-to-moderate pharmacologic support while hemodynamics is closely monitored. Consideration can be given for explanting the device if the patient's hemodynamic status is stable on moderate amounts of pharmacologic support; otherwise, the device is maintained. If the patient is not weaned from assistance within 1 week and is a transplant candidate, then the short-term device should be exchanged in the operating room

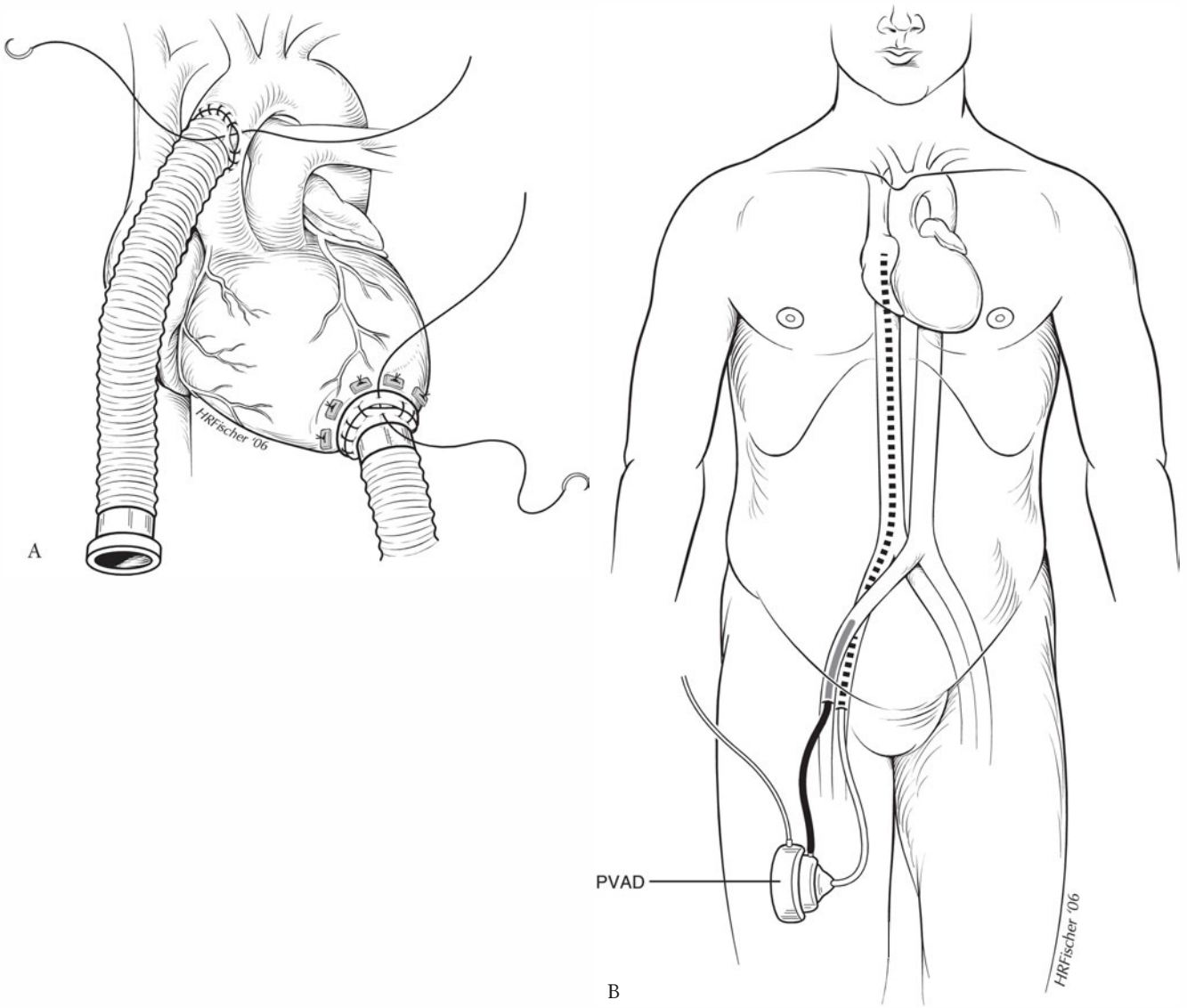


Fig. 56.10. (**A**) Ventricular assist device with outflow tract sutured to the aorta and the inflow tract being sutured to the left ventricular apex. (**B**) Percutaneous ventricular assist device (PVAD).

for a longer term implantable device to act as a bridge to transplantation. If additional myocardial recovery does not occur, transfer to a transplant center is indicated. If the patient is not a transplant candidate, then the original device should be maintained until a decision is made that further support is futile or patient death or recovery occurs.

Multiple series show success in treating patients with acute infarction and refractory cardiogenic shock with a left ventricular assist device. In one series, 22 of 25 patients were successfully bridged to transplant, with six early posttransplant mortalities and 1-, 2-, and 5-year survival rates of 71%, 71%, and 51%, respectively. In another series, 10 of 15 patients were successfully bridged to transplant with this strategy, with 1 patient having the device explanted without requiring transplant. Six of seven patients in another series were successfully bridged to transplant, with five patients well at a median follow-up of nearly 900 days.

A percutaneous ventricular assist device as described by Thiele has also been used for patients with acute myocardial infarction and delayed recovery of myocardial function and continued shock after revascularization. This method uses percutaneous left atrial-to-femoral artery bypass, with the venous cannula introduced via the femoral vein and a trans-septal puncture and the arterial cannula introduced via the femoral artery and advanced into the iliac artery (Fig. 56.10B). This device, which is deployed in the cardiac catheterization laboratory, provides up to 4.0 l/min of assisted cardiac output to allow unloading of the left ventricle. In one study of 18 patients, support was maintained for an average of 4 days, with a 30-day mortality of 44%.

Double-bridge mechanical resuscitation has also been used to treat acute cardiogenic shock. With this technique, extracorporeal membrane oxygenation (ECMO) is used acutely for the treatment of profound cardiogenic shock with an early bridge to a ventricular assist device before transplantation. In one study, 9 of 23 patients were transferred to ventricular assist device, with 7 transplanted, 3 of 23 weaned off ECMO, and 11 of 23 withdrawn from support due to severe neurologic injury or multiorgan failure. In a similar study, 7 of 14 patients were transferred to ventricular assist device, with 1 directly transplanted.

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EDITOR'S COMMENTS

Drs. Carrott, Gardner, and Kron have contributed an extensive and detailed review of the mechanical complications seen after transmural myocardial infarction, including frank rupture, ventricular septal defect, acute ischemic mitral regurgitation, and "pump failure" requiring mechanical support. The detail with which the chapter addresses these uncommon but potentially life-threatening problems confronted by the practicing cardiac surgeon makes this chapter extremely valuable for those on the front line.

The pathogenesis and natural history of each complication are detailed along with "how to fix it" and the optimal clinical outcome. The section on mechanical support is particularly useful as it emphasizes the use of this technology to address ventricular failure when it presents as a complication of myocardial infarction and specifically urges early institution of this type of support to best manage the patient.

Another outstanding aspect of this chapter is the operative technical details for management of these uncommon problems. For instance, ischemic ventricular septal defects represent a daunting technical challenge, especially for a surgeon who has little experience in dealing with this complication. A quick review of the outstanding description of the operative techniques presented in this chapter will prove extremely helpful to the surgeon in alleviating some of the anxiety in approaching this problem.

For each of the pathologic entities discussed in this chapter, the authors emphasize the importance of rapid intervention. In the current era of delayed surgical revascularization following a completed transmural infarction, it is helpful to be reminded that there are still indications for emergency surgery in this setting, including frank rupture, ischemic ventricular septal defect, acute ischemic mitral regurgitation, and pump failure! In such instances, surgical repair often cannot be delayed as the primary problem is a mechanical one that may rapidly be fatal without timely intervention. Only a mechanical solution can solve a mechanical problem and hopefully prevent a patient's death.

The authors are to be congratulated for having produced such a comprehensive review of this subject and in particular making it of tremendous practical value for cardiac surgeons.

LRK and TSG

57 Annuloaortic Ectasia

Tirone E. David

The term "annuloaortic ectasia" was first used by Denton Cooley in 1961 to describe aortic root aneurysm due to Erdheim's cystic medial necrosis in a patient without the stigmata of Marfan syndrome. Although annuloaortic ectasia is often associated with aortic root aneurysm, it may also occur in patients without aneurysm but with aortic insufficiency due to bicuspid or tricuspid aortic valve disease and in those with subaortic ventricular septal defect. Conversely, not all patients with aortic root aneurysm have annuloaortic ectasia. Annuloaortic ectasia is now used to describe a dilated aortic annulus, which is often found in patients with connective disorders of the aortic root. This chapter is dedicated to surgery for aortic root aneurysm with or without annuloaortic ectasia.

The aortic root has four distinct anatomic components: aortic annulus, aortic cusps, aortic sinuses, and sinotubular junction. In addition, the triangles beneath the commissures of the aortic valve are also affected in annuloaortic ectasia, although anatomically they are part of the left ventricular outflow tract. The aortic annulus is the structure that attaches the aortic root to the left ventricle. The aortic annulus is attached to the ventricular myocardium (interventricular septum) in approximately 45% of its circumference and to fibrous structures (anterior leaflet of the mitral valve and membranous septum) in the remaining 55%. Dilation of the aortic annulus alters this relationship and occurs along the fibrous insertion of the aortic annulus. The subcommissural triangles, particularly those beneath the noncoronary cusp, tend to become more obtuse as the annulus dilates. Annuloaortic ectasia usually occurs in patients with aortic root aneurysm due to inherited connective tissue disorders such as in Marfan syndrome and others but it is also often associated with incompetent bicuspid aortic valves.

Cardiac surgery in patients with aortic root aneurysm may be necessary because of aortic insufficiency, dilation of the aortic sinuses, acute type A aortic dissection, or a combination of these problems. The first effective surgical treatment of aortic root aneurysm was described by Bentall and DeBono in 1968. It consisted in replacing the aortic valve and ascending aorta with a Dacron conduit containing a mechanical valve, and the coronary arteries were reimplanted into the graft in a side-to-side manner and the aneurysm wall wrapped around the graft to control hemorrhage. False aneurysm of this graft-to-coronary artery anastomosis was common. Reimplantation of the coronary arteries using the button technique largely resolved the problem. Composite replacement of the aortic valve and ascending aorta with a valved conduit (mechanical, biological, or bioprosthetic) became the standard treatment for patients with aortic root aneurysms for several decades. In 1992, David and Feindel and 1 year later Sarsam and Yacoub pointed out that patients with aortic root aneurysm often had normal aortic cusps and a conservative procedure, whereby the aneurysmal aortic sinuses could be excised and the aortic root reconstructed with a tubular Dacron graft was feasible. We coined the term aortic valve sparing to describe these operations to distinguish them from aortic valve repair, which largely addresses problems with the aortic cusps. There are basically two types of aortic valve–sparing operations to treat patients with aortic root aneurysms: remodeling of the aortic root and reimplantation of the aortic valve. Several modifications of these two types of aortic valve sparing have been described and although the early outcomes appear satisfactory, there are long-term data only on the two original procedures, which will be described in detail in this chapter.

AORTIC VALVE–SPARING OPERATIONS FOR AORTIC ROOT ANEURYSM

Aneurysms often alter the anatomic relationships of the various components of the aortic root (subcommissural triangles, aortic annulus, aortic cusps, aortic sinuses, and sinotubular junction) and aortic valve–sparing operations were developed to correct these abnormalities, which can be quite challenging if all components are abnormal. A sound knowledge of functional anatomy of the aortic valve and how the various components of the aortic root interplay is indispensable to perform these operations. The feasibility of an aortic valve–sparing operation is largely dependent on the quality of the aortic cusps. If the cusps are grossly abnormal, replacement of the entire aortic root is necessary.

Remodeling of the Aortic Root

This procedure should be reserved for older patients with normal aortic annulus (e.g., ≤25 mm for women and 27 mm for men). Most of these patients have primarily ascending aorta aneurysm and the aortic sinuses become secondarily involved by a degenerative process. Remodeling of the aortic root is performed by excising the aneurysmal aortic sinuses and leaving approximately 5 mm of aortic wall attached to the annulus as illustrated in Figure 57.1. The three commissures are pulled upward and approximated until



Fig. 57.1. Aortic root aneurysm. The aortic sinuses are excised, and coronary arteries are detached from the aortic root leaving 5 to 6 mm of arterial wall attached to the aortic annulus and around each of the coronary arteries.

the cusps touch each other centrally, and the diameter of the imaginary circle that includes all three commissures is probably the ideal diameter of tubular Dacron graft if the cusps are not elongated. Three neoaortic sinuses (two for bicuspid aortic valves) are tailored in one of the ends of the graft. The height of the sinuses should be approximately equal to the diameter of the graft. The width of each neoaortic sinus should be proportional to the intercommissural distance. That is, larger cusps should have proportionally larger neoaortic sinuses. Next, the three commissures are sutured on the outside of the graft, immediately above the neoaortic sinuses as illustrated in Figure 57.2. The neoaortic sinuses are sutured to the remnants of arterial wall and aortic annulus with continuous 4-0 polypropylene sutures. This suture line must be carefully done with bites close together to avoid bleeding, a common complication in this operation. We do not use Teflon felt on this suture line. If the arterial wall is paper-thin, we use 5-0 polypropylene sutures in a fine needle, otherwise a 4-0 polypropylene suture in a fine needle. The coronary arteries are reimplanted in their respective neoaortic sinus. Following that, the cusps are inspected for the level of coaptation. The three cusps should coapt at the same level and several millimeters above the level of the aortic annulus. If one or more cusps appear to be coapting at a lower level than the other two, its free margin should be shortened by plication on the central portion along the nodule of Arantius as illustrated in Figure 57.3. If large stress fenestration is present, we routinely reinforce the free margin of the cusp with a double layer of a fine Gore-Tex suture (W.L. Gore & Associates, Langstaff, AZ) as illustrated in Figure 57.4. Clamping the distal end of the graft and injecting cardioplegia into the reconstructed aortic root is a reliable method to test for valve competence. Finally,

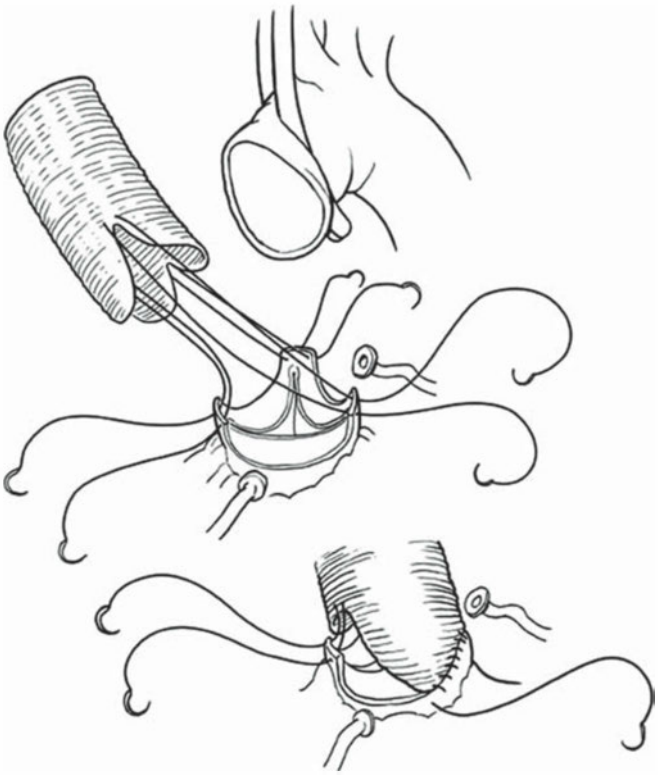


Fig. 57.2. Remodeling of the aortic root. The three commissures are resuspended into a tailored tubular Dacron graft with neoaortic sinuses. The neoaortic sinuses of Dacron are sutured to the junction between remnants of the aortic sinuses and aortic annulus.

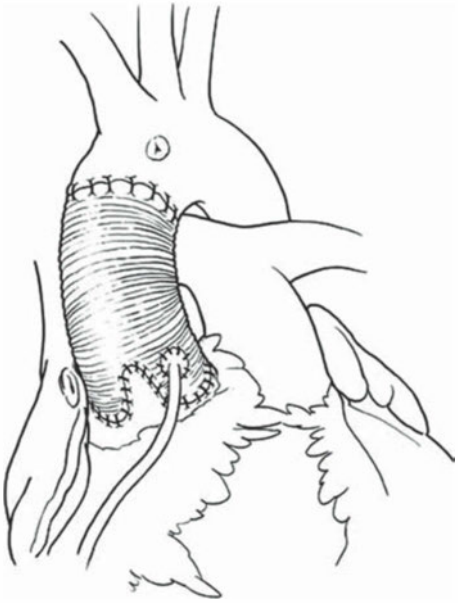


Fig. 57.3. Remodeling of the aortic root. The coronary arteries are reimplanted and the distal anastomosis is performed.

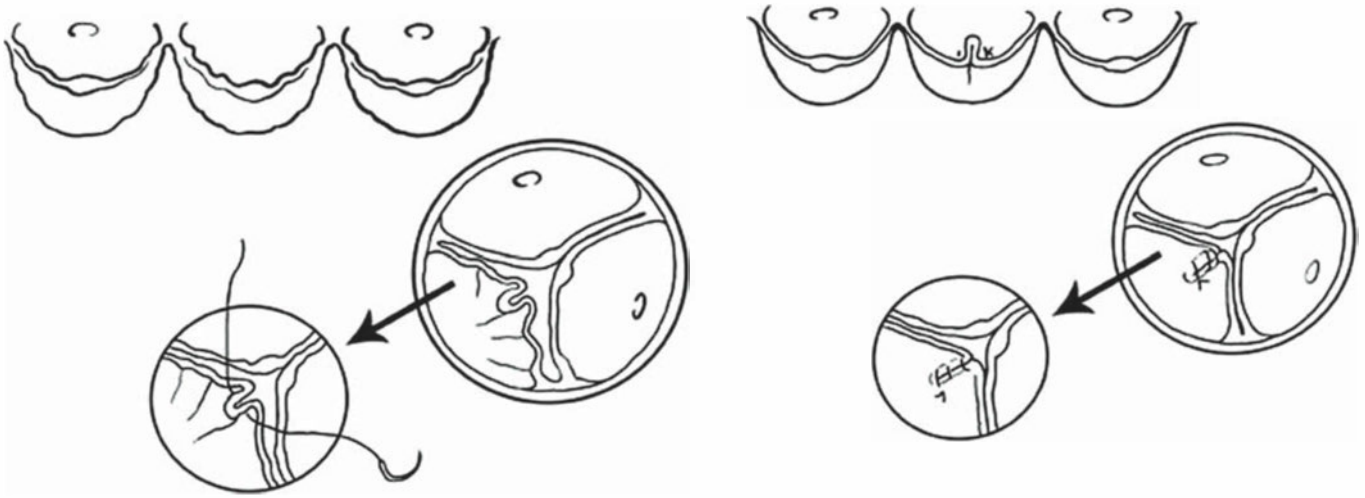


Fig. 57.4. Repair of cusp prolapse. The free margin of the prolapsing cusps is plicated along the nodule of Arantius with fine polypropylene sutures.

the graft is sutured to the distal ascending aorta or transverse arch graft if it also was replaced as illustrated in Figure 57.5. Valve competence is assessed by intraoperative echocardiography. In addition to a competent valve (no more than trace aortic insufficiency should be accepted), the coaptation level and coaptation length should be measured. Ideally, the coaptation height should be at least 9 mm from the nadir of the aortic annulus and the coaptation length at least 4 mm.

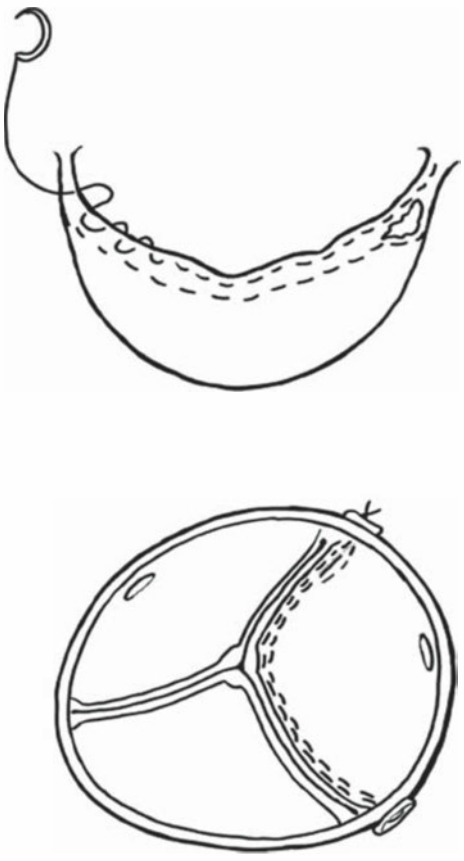


Fig. 57.5. Reinforcement of the free margin of the aortic cusp. A double layer of 6-0 expanded polytetrafluoroethylene suture is woven along the free margin. If there is a large fenestration, the first layer runs along the upper and lower rims of the fenestration.

Reimplantation of the Aortic Valve

This type of aortic valve sparing is complicated because it addresses every component of the aortic root. The aortic root has to be dissected circumferentially down to just below the level of the nadir of the aortic annulus. This is not always possible along the membranous septum because the right ventricular is often attached a couple of millimeters higher than the nadir of the cusps. The aortic sinuses are excised as described above (Fig. 57.1). Selection of the size of the graft is based largely on the ideal diameter of the sinotubular junction as described above. A tubular Dacron graft of diameter of the estimated sinotubular junction is selected (usually 26 to 30 mm) and secured to the outside of the aortic root at a level below the nadir of the aortic annulus. A small triangular segment is cut from one of the ends of the Dacron graft to correspond to the commissure between the left and right aortic cusps as illustrated in Figure 57.6. This tailored end of the graft is secured on the outside of the left ventricular outflow tract with multiple horizontal mattress (9 to 12) 2-0 or 3-0 polyester sutures. This suture line follows the scalloped shape of the aortic annulus along its muscular portion and it is as horizontal as possible along the fibrous portion of the left ventricular outflow tract. In the area of the membranous septum, this suture line may have to be slightly higher than the level of the nadir of the annulus because of the insertion of the right ventricle. If one suture is higher than the other, the same thing must be done when passing it through the Dacron graft. Teflon-felt pledgets are used along the membranous septum and subcommissural triangle between the left and noncoronary cusps because the tissue is often very thin. We believe this suture line is crucial in patients with annuloaortic ectasia and the reduction in annular diameter must be beneath the subcommissural triangles of the noncoronary cusp. The three commissures are suspended inside the graft and fixed to the wall with transfixing 4-0 polypropylene sutures with Teflon-felt pledgets. The spatial relationship of the three commissures is very important for proper cusp coaptation. Once the proper level and orientation are determined, the remnants of aortic sinuses and aortic annulus are secured to the tubular Dacron graft with a full thickness, transfixing sutures along the scalloped shape annulus, creating a crescent shape aortic annulus for each cusp. The coronary arteries are reimplanted into their respective neoaortic sinus as illustrated in Figure 57.7. This technique of aortic valve implantation creates an aortic root without aortic sinuses. It is possible to create an aortic root with neoaortic sinuses by using a Dacron graft 4 to 6 mm larger than the diameter of the sinotubular junction (usually 30 to 34 mm in diameter). The diameter of the aortic annulus is also measured. If the diameter of the aortic annulus is less than the sinotubular junction, the graft is plicated to reduce its diameter to that of the aortic annulus plus 6 mm in the end, which will be sutured to the left ventricular outflow tract as illustrated in Figure 57.8. The graft

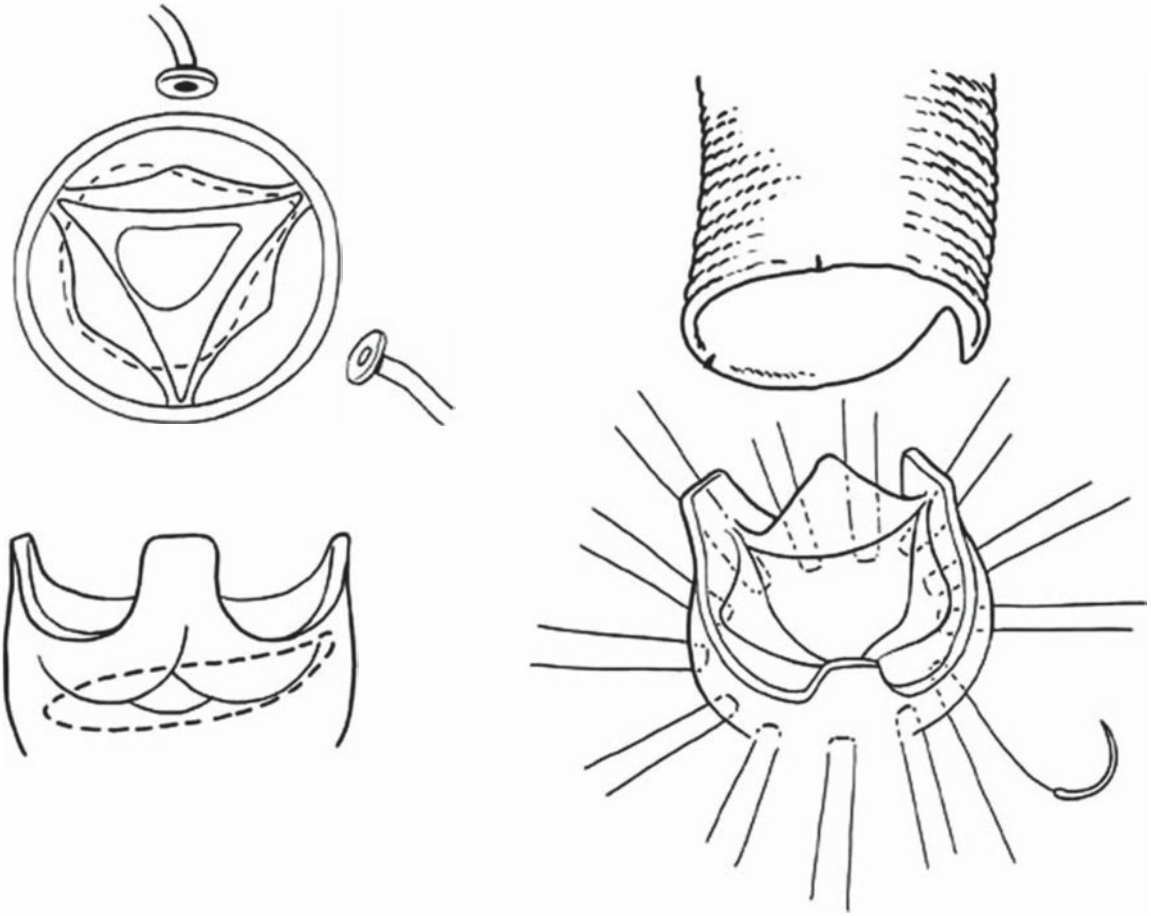


Fig. 57.6. Reimplantation of the aortic valve. (*Right*) A small triangular segment is cut from one of the ends of the Dacron graft to correspond to the commissure between the left and right aortic cusps. (*Left*) Multiple horizontal mattress sutures are passed from the inside to the outside of the left ventricular outflow tract along a single horizontal plane immediately below the aortic annulus along its fibrous components and following the scalloped shape of the annulus along the muscular interventricular septum.

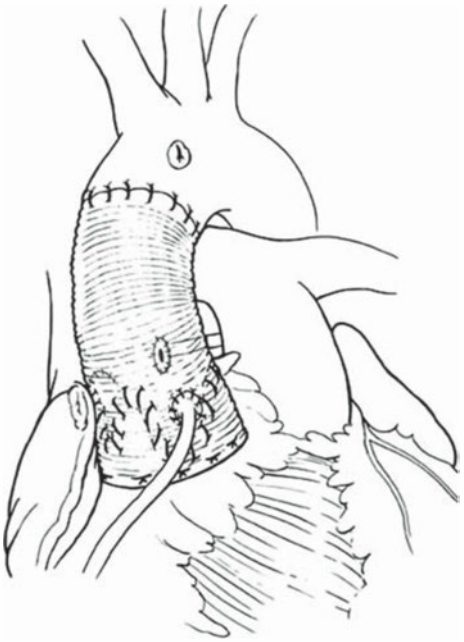


Fig. 57.7. Reimplantation of the aortic valve. The remnants of the aortic sinuses are sutured to the Dacron graft recreating a scalloped shape aortic annulus. The coronary arteries are reimplanted into their respective aortic sinuses.

is secured to the left ventricular outflow tract as described above and the commissures resuspended inside the graft and the scalloped shape of the aortic annulus recreated by suturing the remnants of aortic sinuses with the graft. The coronary arteries are reimplanted. Darts are placed in the graft in spaces between commissures to create neoaortic sinuses as illustrated in Figure 57.9. This maneuver reduces the diameter of the sinotubular junction at the rate of 1 mm for each 3 mm of graft plication. Since a graft larger than needed was selected, creation of neoaortic sinuses should not cause cusp prolapse. Whether neoaortic sinuses are created or not, at the completion of the procedure the cusps must coapt at the same level as described for remodeling of the aortic root. Valve competence and hemostasis along the coronary artery buttons can be assessed by injecting cardioplegia in graft with the distal end occluded. Finally, the graft is sutured to the distal ascending aorta or transverse arch graft if it also was replaced.

In a small proportion of patients, particularly those with bicuspid aortic valve, one coronary artery orifice may be too close to a commissure to be safely detached and reimplanted. In this case, the coronary artery is left in situ, the graft incised vertically in that area and after being secured in the annulus, an opening is created and the tissue around the artery is secured to the graft.

Aortic valve function and cusps coaptation level and coaptation length should be assessed after discontinuation of cardiopulmonary bypass as described above.

REPLACEMENT OF THE AORTIC ROOT AND ASCENDING AORTA

In patients with aortic root aneurysm and grossly abnormal aortic cusps, the best treatment is replacement of the entire aortic root and ascending aorta. A valved conduit is needed to replace the aortic root and there are several choices of valves and conduits. There are commercially available conduits of Dacron graft containing mechanical valves as well as glutaraldehyde-fixed porcine aortic root. Some valve manufacturers provide the choice of having mechanical valves into a straight tube or

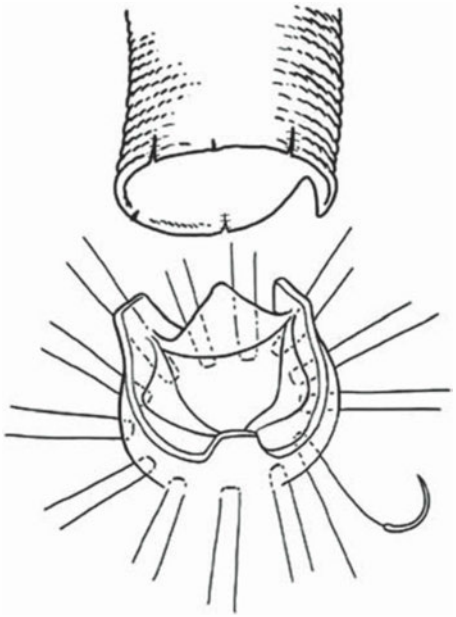


Fig. 57.8. Reimplantation of the aortic valve with neoaortic sinuses. A graft larger than needed based on the ideal sinotubular junction diameter. If the annulus is not dilated, the proximal end of the graft is plicated to reduce its diameter before being secured to the outside of the left ventricular outflow tract. This plication should be in the areas of the nadir of the aortic annulus.

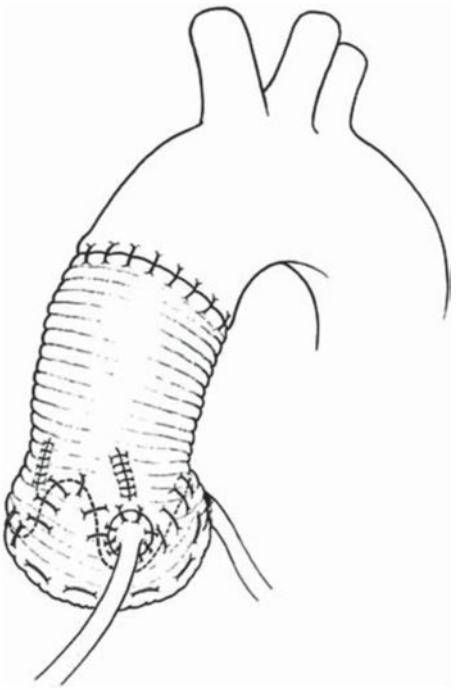


Fig. 57.9. Reimplantation of the aortic valve. After reimplantation of the coronary arteries, darts are placed in the graft in the spaces in between commissures to create neoaortic sinuses.

inside a Valsalva graft (Vascutek Ltd, Renfreshsire, Scotland). There is no evidence that spherical sinuses above a mechanical valve are beneficial to valve function or blood flow. Valved conduits can also be custom-made during surgery by securing a heart valve (bioprosthetic or mechanical) inside a Dacron graft. This approach is useful when a skirt of Dacron graft is desirable beneath the level of the sewing of the valve or whenever a bioprosthetic valve is used to facilitate reoperations should it become necessary. Aortic root homograft can also be used to replace the aortic root. The Ross procedure is not advisable in patients with aortic root aneurysm because inherited connective tissue disorders are the most common cause of aortic root aneurysm, and the genetic abnormality is also probably present in the native pulmonary root. The choice of one over other types of valves is largely made in consultation with the patient. It is important to consider that reoperative aortic root replacement is a more complicated procedure than isolated aortic valve re-replacement and it is probably associated with higher operative mortality and morbidity. Thus, patients who are likely to outlive their biological or bioprosthetic valve, the risk of reoperation must be weighed against the risk of life-long anticoagulation. Aortic root homograft is probably the best conduit for patients with aortic root abscess that required extensive resection and reconstruction of the left ventricular outflow tract.

Aortic root replacement is performed by clamping the ascending aorta close to the origin of the innominate artery and transecting the aorta just above the sinotubular junction. The aortic cusps are excised and the annulus is debrided when calcified. The coronary arteries are detached from the root leaving 4 to 5 mm of aortic sinus wall around their orifices. When the coronary arteries are not displaced, they should be mobilized to avoid tension after their reimplantation into the valved conduit. If the aneurysm is limited to the aortic root, the ascending aorta is resected up to 8 to10 mm to the aortic clamp to allow enough tissue for the anastomosis. If the entire ascending aorta and/or the transverse arch are aneurysmal, hypothermic circulatory arrest is needed for replacement. We use systemic hypothermia of 25 to 28 C and antegrade cerebral perfusion at 10 ml/kg/min (see also Chapter XX on aortic arch surgery).

The valved conduit is then secured to the aortic annulus. When the annulus is dilated, the issue of valve size is unimportant and a valve of appropriate size is used to match the patient's body surface area. In this case, we use multiple inverting interrupted horizontal mattress sutures of 2-0 polyesters with pledgets. These sutures are passed through the sewing ring of the mechanical valve and tied down as illustrated in Figure 57.10. Because the aortic annulus may be larger than the prosthetic valve in the conduit, some plication of the native aortic annulus is necessary. This plication should be performed along the fibrous portion of the aortic annulus. Thus, the sutures should be evenly spaced in the sewing ring of the valve and in the aortic annulus along its muscular component and closer together in the sewing ring than in the aortic annulus along its fibrous portion, mostly in the subcommissural triangles. Round openings 2 to 3 times larger than the diameter of the coronary arteries are made in the graft, and the remnants of the arterial walls around the coronary arteries are sutured to the graft with continuous 5-0 polypropylene sutures. It is imperative that the coronary arteries be implanted without tension in any direction to prevent kinking and/or bleeding when the ventricles distend and the heart begins beating. The distal end of the conduit is anastomosed to the distal ascending aorta or Dacron graft used to replace the arch. It is important to remember that the length of a normal ascending aorta is no more than 5 or 6 cm. Excessively long grafts can kink and cause obstruction and hemolysis. Long grafts can also make the mitral/aortic annuli more acute and increase turbulence along the left ventricular outflow tract.

If the aortic annulus is normal in diameter, we implant the largest possible mechanical valve and secured it to the annulus with multiple (20 to 24) simple sutures of 2-0 polyester without pledget. If the annulus is too small for the patient's body surface area and age, it should be enlarged to accommodate a valve of appropriate size.

We no longer use xenograft porcine aortic root because they do not appear to be more durable than stented bioprosthetic valves, and reoperations can be troublesome because of the inflammatory reaction with surrounding tissues. If a bioprosthetic aortic valve is desirable, we suture it in a Dacron graft approximately 1 cm from one of its ends as illustrated in Figure 57.11. This is easily done by inverting the graft and using a continuous suture between the Dacron graft and sewing ring of the bioprosthetic valve. The skirt of Dacron beneath the valve can be sutured to the aortic annulus with continuous polypropylene sutures. This technique has allowed for re-replacement of the aortic valve without taking down the graft from the aortic root and coronary arteries.

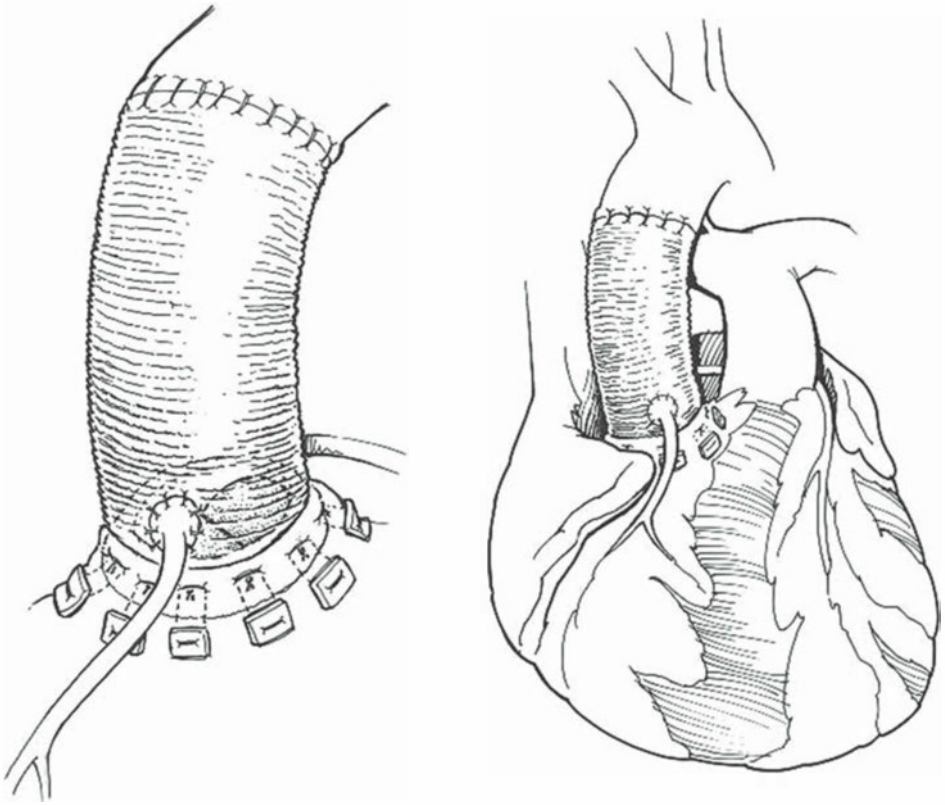


Fig. 57.10. Aortic root replacement with commercially available conduit containing a mechanical valve.

POSTOPERATIVE COMPLICATIONS

Bleeding is probably the most common complication of aortic root surgery. Bleeding after remodeling of the aortic root is particularly troublesome along the suture line between graft and the remnants of aortic sinuses. That anastomosis must be meticulously performed and care must be exercised to avoid tears in the aortic wall. Bleeding from the coronary artery buttons can also be a problem but largely preventable by injecting blood cardioplegia into the graft under pressure before performance of the distal anastomosis. Finally, these operations usually require long cardiopulmonary bypass time and a degree of coagulopathy is common. Thus, transfusion of clotting factors, including recombinant factor VII, may be necessary.

Myocardial ischemia due to poorly aligned coronary artery buttons in the Dacron graft is probably the second most common intraoperative complication of aortic root replacement. If the right ventricle is hypokinetic, careful inspection of the right coronary artery button may disclose a kinked artery because of incorrect orientation or reimplantation level. A common problem in patients with bicuspid aortic valve is the nondominant right coronary artery, which is sometimes

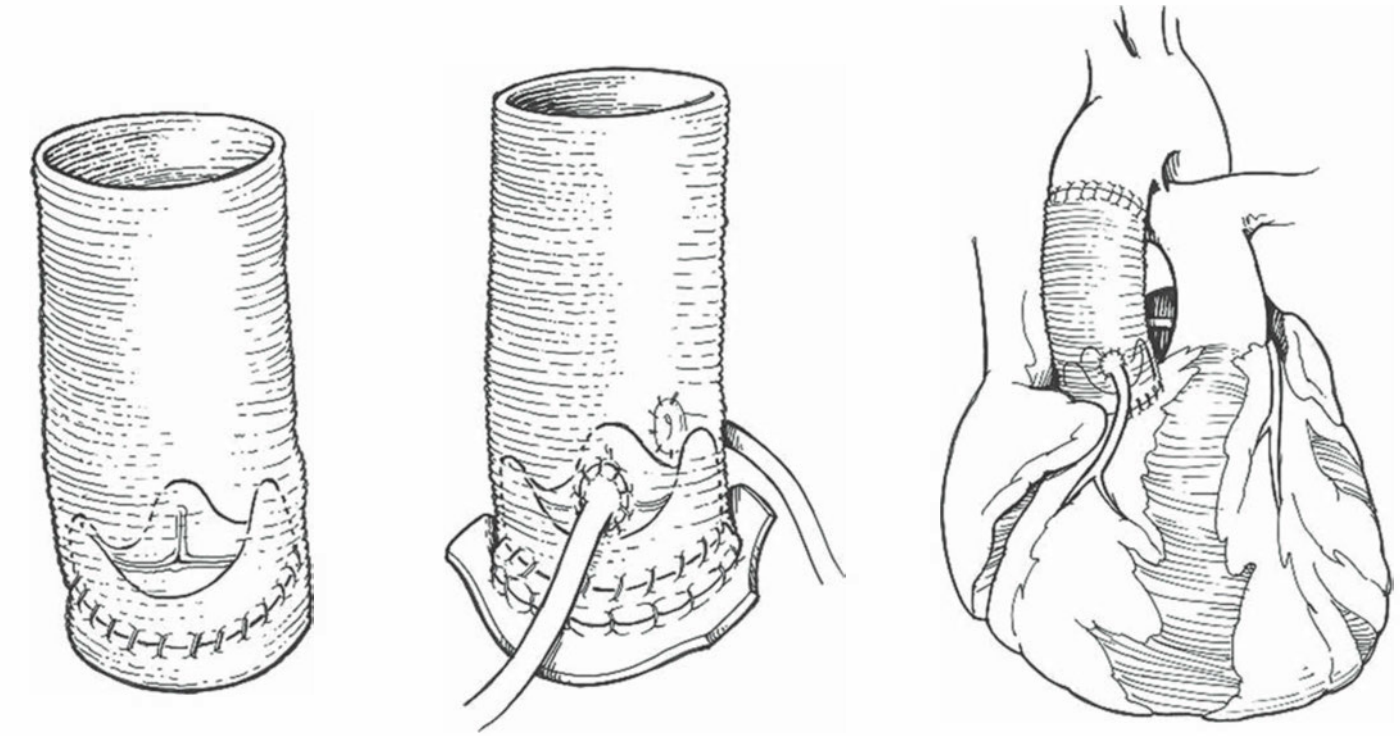


Fig. 57.11. Aortic root replacement with a stented bioprosthetic heart valve. Note that the bioprosthetic valve is sutured a few millimeters above the graft-annulus anastomosis. The rim of graft beneath the bioprosthetic valve is secured to the aortic annulus.

suboptimally perfused during aortic clamping, and the right ventricle may become ischemic and hypokinetic after discontinuation of cardiopulmonary bypass.

Temporary heart block is common after long operations when cold blood cardioplegia is used for myocardial protection but permanent block is rare. We routinely place temporary pacer wires in the atria and ventricle in all heart valve operations and often use atrial or atrioventricular pacing to optimize heart rate and cardiac output.

Stroke is rare in these patients because most of them are relatively young and free of arch atherosclerotic disease. Postoperative bacteremia is extremely serious in these patients and must be promptly treated to prevent endocarditis. We routinely do a transthoracic echocardiogram prior to discharge to evaluate valve and ventricular function and to make sure the patient is not developing pericardial effusion, which can cause delayed tamponade.

CLINICAL OUTCOMES

First time, elective aortic root surgery should be associated with very low operative mortality, probably no higher than 1% or 2%. Surgery in patients who require urgent or emergency surgery because of acute type A aortic dissection, infective endocarditis with annular abscess, or advanced functional class for any reason is associated with increased operative mortality. The operative mortality and morbidity also increase when additional procedures such as aortic arch replacement, mitral valve surgery, and myocardial revascularization are needed.

In our latest report on outcomes of aortic valve–sparing operations for aortic root aneurysm, there were 289 patients whose mean age was 47 years. They were prospectively followed for a mean of 7.2 years (ranging from 0 to 20 years). The operative mortality was 1.7% in spite of the fact that 25 patients had acute and 19 had chronic type A aortic dissection, 20 required mitral valve surgery, 56 arch surgery, and 35 needed myocardial revascularization. The survival at 12 years was 82.9%. The freedom from reoperation on the aortic valve was 94% at 12 years, and the freedom from aortic insufficiency greater than mild was 86.8%. Reimplantation of the aortic valve provided more stable aortic valve function than remodeling of the aortic root.

The long-term results of aortic root replacement with mechanical valves are also excellent and similar to those of aortic valve–sparing operations. Several studies attempted to determine the superiority of one versus the other procedure, and the general consensus was that they provided similar outcomes. Aortic valve–sparing operations are associated with risk of aortic insufficiency that may need reoperation, whereas aortic root replacement with mechanical valves is associated with risk of hemorrhage and thromboembolic events.

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EDITOR'S COMMENT

Dr. David is responsible for a major chapter in cardiac surgery. The valve-sparing aortic root replacement by implantation is a huge step and we practice this procedure and we believe this procedure is the treatment of choice for annuloaortic ectasia. He gives a very thorough approach to this procedure and it certainly works. I would like to mention just a couple of technical points that relate to this procedure as well as to the Bentall procedure. The reimplantation tends not to have the bleeding issues that one might have with some of the other procedures. However, once the left coronary anastomosis is done, it is possible to have bleeding that is difficult to get to after this anastomosis. We, therefore, pressurized the aorta with a needle before the distal anastomosis is complete to be certain that there is no bleeding for that left coronary. We also tend to do the distal anastomosis before we reimplant the right. Often the right should be reimplanted tighter than one thinks, and avoiding coronary ischemia is critical in this procedure. Finally, with the Bentall procedure we try to use some of the remaining aortic root to cover the proximal suture line. Again this is a difficult area to get to. This is not an issue with reimplantation since basically the internal suture line causes hemostasis.

ILK

58 Aortic Dissection

Nimesh D. Desai and Joseph E. Bavaria

INTRODUCTION

Thoracic aortic dissection is often a catastrophic disease with an incidence estimated to range from 2.9 to 3.5 per 100,000 person-years in the United States. Appropriate treatment is best managed by a team approach, as the aortic dissection process may affect any part of the circulation leading to potential organ malperfusion of the heart, the brain, the spinal cord, the gastrointestinal tract, the kidneys, and the extremities. A more focused understanding of the causes of death and morbidity of aortic dissection has allowed the development of operations specifically targeted to improve clinical outcomes. Further refinements in management such as the use of the operating room as the diagnostic suite have also contributed to improved survival.

ETIOLOGY

Aortic dissection develops from a tear within the intima of the aortic wall. Blood flows across this "entry point" into a weakened media, splitting the medial layer along the direction of flow, thus creating a new "false" channel, within the aortic media. This new channel progresses downstream and significant pressure/mechanical stress is exerted by the advancing column of blood on the aortic branches encountered in its path. An individual branch will either tear, leading to a communication from the false lumen into the original, "true" lumen of the aorta, or close off causing "malperfusion" of the organ supplied by the given arterial branch. The multiple torn branches down the path of the false lumen become known as reentry points or "fenestrations." The false lumen, thus, does not usually become a blind pouch with the potential for thrombosis but is kept patent by the many exit and reentry points of variable sizes. If the reentry points are, in aggregate, small or restrictive, a pressure gradient will develop between the false lumen and the true lumen. This pressure differential will cause the false lumen to expand in an amount commensurate to the gradient, causing the true lumen to contract. A new equilibrium is eventually reached with equalization of pressure within the two lumens, typically leaving a small true lumen and a larger false channel. This final equilibrium point is reached within minutes to hours from the time of the original tear, such that the dissection is established and relatively stable by the time the patient reaches medical attention, with a false lumen extending from the entry site most often all the way down to the aortic bifurcation.

Although the final initiating event in an aortic dissection is an intimal disruption, the disease is not an intimal disease. Endothelial cells and their basement membrane do not possess significant intrinsic tensile strength, and their disruption is due to a lack of appropriate mechanical support from the medial layer. The media can be abnormal secondary to a genetic structural abnormality as in Marfan syndrome, where abnormal fibrillin causes loss of elasticity in the medial layer. Marfan syndrome is an autosomal-dominant syndrome with complete penetrance with an overall incidence of 1 per 3,000 to 10,000 live births. It is caused by alterations in the gene coding the aortic wall protein fibrillin-1, leading to elastin derangement. Increased TGF-β activity that negatively affects smooth muscle development and the extracellular matrix has also been implicated. More often other less clearly identified genetic conditions can predispose to abnormalities in the media which lead to dissection. These less well-defined heterogeneous abnormalities are often grouped in the category of annuloaortic ectasia where different abnormalities in the media lead to the common end pathway of dilatation of the aortic diameter with thinning of the aortic wall. Atherosclerotic disease will also damage the media in a variable pattern. Wall weakness in atherosclerotic disease is more often combined with sustained high blood pressure leading to increased stress on the abnormal aortic wall. Within this hypertensive background, a paroxysm of increased arterial blood pressure will incite the initial tear.

CLASSIFICATION

Anatomic classifications of aortic dissection are based on the location of the initial tear and are important in terms of treatment modality and prognosis (Fig. 58.1). The first classification scheme was designed by DeBakey, who classified dissections into the following:

Type I. The process involves the ascending aorta, the aortic arch, and the descending thoracic and often the abdominal aorta.

Type II. The dissection involves the ascending aorta, but stops at the level of the aortic arch, and does not involve the aorta beyond the subclavian artery takeoff.

Type III. The dissection starts distal to the subclavian artery and extends to the entire thoracic aorta (IIIa) or the thoracic and abdominal aorta (IIIb).

The following Stanford classification has became more commonly used as it readily translates to clinical decision-making:

Type A. The dissection involves the ascending aorta, regardless of tear site, and will most often extend into the descending thoracic as well as abdominal aorta. The subsets of DeBakey types I and II are included in this class.

Type B. The dissection starts beyond the aortic arch usually at the subclavian artery and progresses distal, thus not involving the ascending aorta or the aortic arch. The subsets of DeBakey type IIIa and IIIb are included in this class.

The power and popularity of the Stanford classification is due primarily to the very different natural history of the two types of dissection and the requirement of very different surgical approaches to successfully treat them. A type A dissection

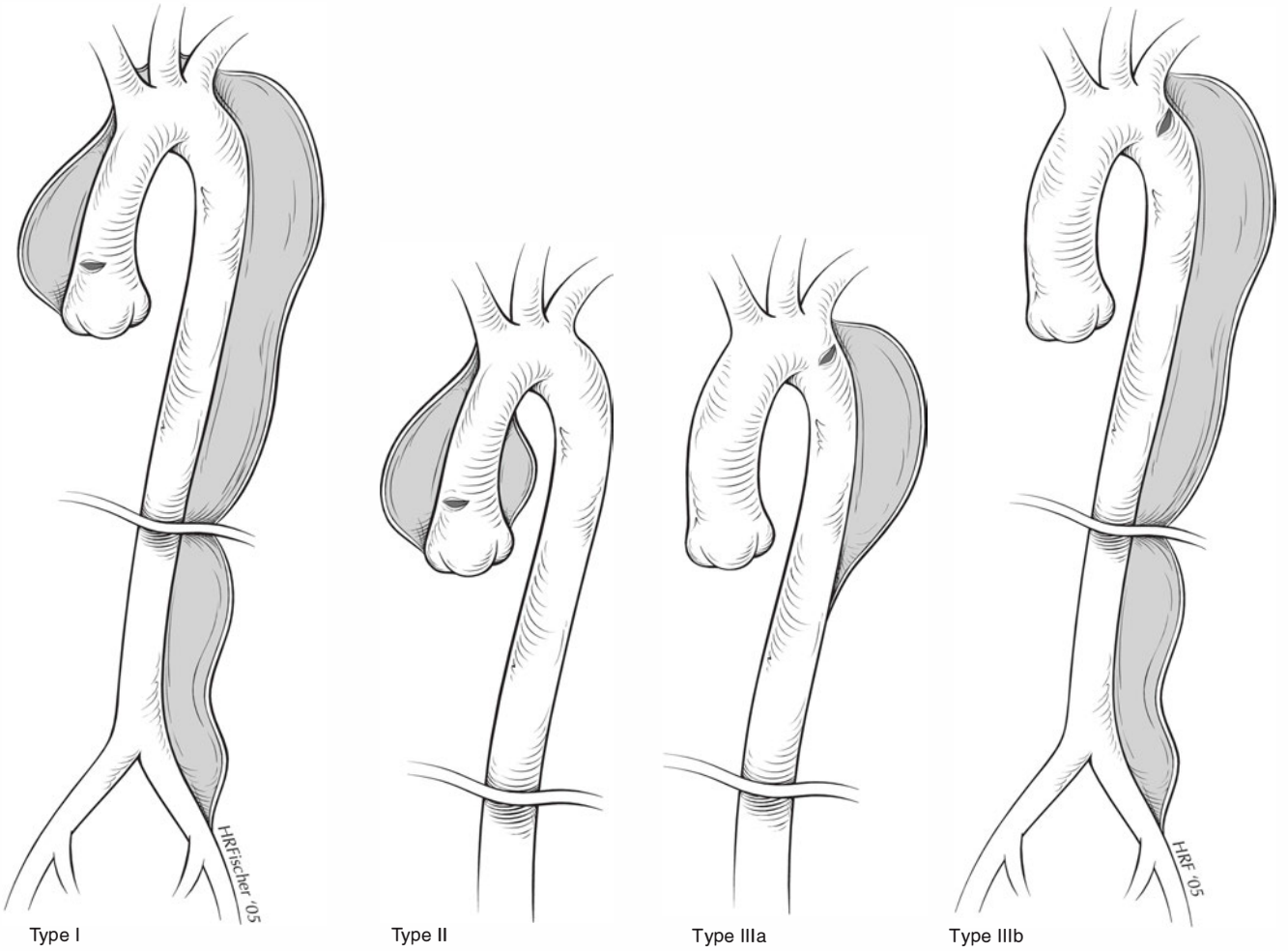


Fig. 58.1. Classification.

carries a high risk of early mortality and morbidity if not treated surgically. A type B dissection has a much lower early mortality, with early surgical treatment providing no clear advantage or even a higher mortality over observation alone except in cases of rupture or severe malperfusion. If the prognosis of type A dissection is so ominous early in the process, this represents a surgical emergency. Type B dissection is a much lower risk aortic process with a relatively low risk of rupture. The primary acute problems caused by type B dissections are malperfusion syndromes of those organs in the path of the descending thoracic or abdominal aorta or the iliac arteries.

CLINICAL PRESENTATION AND DIAGNOSTIC EVALUATION

The aortic media have a high density of nerve endings, so when medial disruption takes place, the acute onset of sharp pain is a nearly uniform presenting symptom, occurring in over 80% of patients. Atherosclerotic degeneration of the media may result in less acute and sharp pain during the dissection progression. In an acute type A dissection, the pain is classically described by the acute onset of sharp, severe chest pain which then migrates to the upper back and is often described as extending/progressing down to the lower back and even the groins depending on the dynamics of growth of the false lumen. Once the false lumen is fully established and has stabilized, the acute pain may change into a more persistent and dull pain, which is often not as easy to localize and may involve the chest and back in variable patterns with additional elements of nausea, abdominal pain, diaphoresis, and shortness of breath. The presence of new focal neurologic deficit suggests aortic arch involvement. Flank pain may also be described in patients who are suffering an acute malperfusion of one of the kidneys. Signs and symptoms of venous hypertension suggest that pericardial tamponade are usually accompanied by poorly perfused extremities, sweaty and clammy appearance, and presyncopal symptoms, consistent with a low cardiac output state. Additional presenting symptoms can be the sudden onset of a cold and pulseless extremity, most commonly the right lower extremity. A right upper extremity vascular impairment is often associated with a neurologic event, especially in a patient with an incomplete circle of Willis. These patients can have a catastrophic hemispheric infarct with a dense contralateral hemiplegia. Rarely, paraplegia can be a presenting symptom. This may occur in cases where a false lumen supplying blood to the spinal cord thromboses or the spinal blood supply is sheared off by an intramural hematoma. Acute paralysis usually associated with signs of spinal shock, which, compounded to other cardiovascular effects of the acute dissection, can

further complicate the early assessment of the patient. Similar paralysis symptoms may occur in the setting of total occlusion of the thoracic or abdominal aorta with no blood flow to the extremities, although this is also associated with pulselessness. Abdominal pain can occur in either type A or type B dissection, and it is often associated with stenosis or occlusion of either the celiac or the superior mesenteric arteries.

In a relatively young patient without significant risk factors for atherosclerotic disease, the index of suspicion for aortic dissection should be very high when any of these symptoms are described. Screening tests should include a chest X-ray and an electrocardiogram (ECG). ECG is often nondiagnostic, and occasionally it may show inferior wall changes suggestive of right coronary artery compromise by the dissection. Proximal dissection extension into the left main coronary artery causing occlusion is typically a fatal event, and such patients rarely survive long enough to reach medical care. Chest X-ray is a very nonspecific exam and it may show a dilated mediastinum or a large cardiac silhouette possibly due to an acute pericardial effusion.

Patients with chest or back pain that is not keeping with typical anginal symptoms, particularly in the setting of an abnormal chest X-ray, should undergo emergent contrast computed tomographic (CT) angiography. In most emergency departments today, a CT scan with contrast can be obtained within a few minutes of admission. Most modern multidetector CT scans with contrast will make the diagnosis of aortic dissection with exceptional accuracy and often will be the only test required prior to proceeding to definitive treatment. Rarely in a CT scan, particularly when the scan was done to search for a pulmonary embolism and the intravenous contrast is not well timed, the ascending aorta can show artifactual lines that may be interpreted as a type A dissection. These scans represent the few false-positive studies.

Transthoracic echocardiography occasionally is used to make the diagnosis of aortic dissection by visualizing either the ascending aorta or abdominal aorta. A transthoracic echo may be difficult to carry out and poor windows may limit its usefulness. If the aortic root is clearly visualized, a variable degree of aortic valve insufficiency with a dilated root or the suggestion of an intimal flap will suggest the diagnosis of type A dissection. Acute dissection flaps are not always visible with this technique, and transthoracic echo cannot definitively rule out aortic dissection. Pericardial effusion may also be seen.

After a screening transthoracic echo is suggestive but not diagnostic of an aortic dissection, a transesophageal echocardiogram (TEE) can nearly always make an accurate diagnosis of the type of dissection present. While there is no need to delay definitive treatment by adding transesophageal echo to a diagnostic contrast CT scan, in our recent experience it is as accurate as any other diagnostic study and allows rapid evaluation of an unstable patient. This is usually done in the operating room with a full cardiac surgical team available.

Contrast magnetic resonance imaging (MRI) provides excellent images of any dissection; it is not often readily available and it usually requires long acquisition times with the patient not as closely monitored because of the technical requirement of the high magnetic field. It should not be used as a primary modality except when it is more rapidly available than TEE, and the patient has evidence of renal dysfunction, making contrast CT angiography less desirable. In cases where the index of suspicion for acute aortic dissection is high, even in cases of renal dysfunction, CT angiography with contrast provides the most rapid and definitive diagnosis and should be used to avoid unnecessary delays in treatment. MRI can be extremely valuable in following a type B dissection after the initial diagnosis or in evaluating any chronic dissection.

Diagnostic catheter-based aortography has become an uncommon primary diagnostic evaluation. Typically, diagnosis is made by aortography only when the patient presented with an acute coronary syndrome with coronary malperfusion or diagnosed at the time of another cardiovascular intervention causing iatrogenic aortic dissection. Taking a patient with acute aortic dissection to the cardiac catheterization lab carries a very high mortality. Suspecting coronary disease leads to the use of platelet-inhibiting agents, anticoagulants, and occasionally fibrinolytics, all of which can transform a contained rupture into a free disruption or can increase the diffuse blood extravasation occasionally noted in a dissected, extremely thin aorta. Catheter manipulations performed can cause dangerous direct damage to the freshly dissected aorta. When obtained, aortography will only show the presence of a dissection flap and does not provide the anatomic detail a standard CT scan with contrast, a TEE or an MRI can provide today.

The question occasionally arises over the value of elective coronary evaluation after making the diagnosis of type A dissection. Retrospective reviews suggest that the incidence of coronary disease in this population is sufficiently low that more patients are placed at risk by the delay and technical risks of a cardiac catheter than are saved by the discovery of occult coronary disease. The only exception may be in patients who have undergone previous coronary bypass surgery, where the knowledge of coronary disease, graft position, and patency can contribute to a safer operation. When the technology of intraoperative coronary angiogram is available in a dedicated hybrid operating room suite, that may become an option for patients after completion of the proximal repair.

Once the diagnosis of a type A dissection is suggested by any screening test, the patient should not be delayed in the emergency department by further testing but should be transferred to an appropriate institution that can proceed with expeditious surgical repair. Over the last several years, our policy has been to transfer any patient with a documented or highly suspected type A dissection immediately to the operating room to minimize any delay. As soon as the patient arrives to the operation room, if sufficient suspicion or good studies are available, the patient is placed under general anesthesia and a TEE is obtained. If dissection is demonstrated by transesophageal echo in the ascending aorta, the patient undergoes emergency surgery. With this policy, 5% to 10% of patients admitted directly to the operating room are found to have a negative transesophageal echo and are then transferred to the intensive care unit for further evaluation. Most commonly, motion artifact in younger patients, type B dissections with some degree of arch involvement but no flap in the ascending aorta, or intramural hematomas are present in these patients.

ACUTE TYPE A DISSECTION

Patients with an acute type A dissection have a risk of death traditionally estimated at 2% per hour in the first 48 hours from their presentation. More recent data suggest that the early mortality may be as low as 60% over the first 2 to 5 days in patients without a concomitant root aneurysm and with aggressive use of antihypertensive medications, especially beta blockers.

Either way, medical therapy generally has no role in acute type A dissection except to allow a safer transfer to a center with experience in complex aortic repair or in inoperable patients. Historically, type A dissection surgery was focused on placing a short segment of Dacron graft in the ascending aorta in the hope of eliminating the entry tear and obliterating the false lumen both proximally into the aortic root and distally into the arch and beyond. This was frequently done by sewing the Dacron graft proximally to the aorta just proximal to the tear with no stabilization of the aortic root or valve and sewing distally to clamped aorta and not utilizing an open arch anastomosis. That principle often led to recurrent malperfusion proximally (myocardial infarctions and recurrent aortic insufficiency) and distally (stroke, organ, limb malperfusion) as well as a moderate distal early rupture rate. Furthermore, a high incidence of long-term failure was seen with aneurysmal degeneration of both the root and the distal ascending aorta and aortic arch.

As better understanding of the etiology for early complications and death was gained, a better operation could be designed to address those factors. There are four primary causes of early death from type A dissection, and the modern approach to surgical correction involves an operation addressing these four problems appropriately and directly.

1. *Aortic rupture*. The ascending aorta is subjected to more stress than any other portion of the aorta. It is the site of the majority of medial degeneration and thus intimal tears. Once dissection has occurred, the ascending aorta is at very high risk of early rupture. This risk is magnified when the aortic root and ascending aorta are dilated as the tension exerted on the remaining media and adventitia increases according to the Laplace law by the square of the diameter. To prevent this lethal complication, the ascending aorta needs to be completely replaced.
2. *Congestive heart failure due to acute onset of aortic valve insufficiency*. Aortic valve support is lost due to prolapse of the intimal layer at the level of one or more commissures. The most commonly affected is the noncoronary to right coronary sinus commissure, followed by the noncoronary to left coronary sinus and least common is the right coronary to left coronary commissure. To eliminate aortic valve regurgitation, the operation should include either a repair or a replacement strategy as necessary. The most common repair technique consists of aortic valve resuspension to reestablish normal commissural support (Fig. 58.2). Furthermore, the sinotubular junction may be dilated and may need to be brought back to a geometrically normal size. The ascending aortic graft is sized to best return the sinotubular dimension to its normal state.

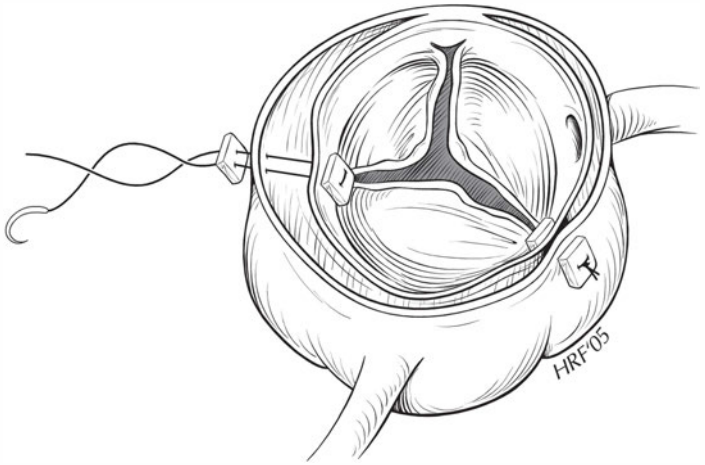


Fig. 58.2. Aortic valve resuspension.

3. *Acute myocardial infarction due to malperfusion of the coronary arteries*. Very few patients with left main coronary compromise survive to reach medical attention. Right coronary malperfusion is much more common. At surgery, the root needs to be repaired definitively to treat ongoing coronary stenosis/occlusion or to prevent future coronary compromise or late root dilatation. The incidence of aortic root dissection is quite high, especially in the noncoronary sinus where the great majority of aortic root will often be dissected down to the annulus. The second most commonly dissected sinus is the right coronary sinus with often some element of coronary malperfusion. Occasionally, the left coronary sinus of Valsalva will be also dissected and while in the past that was considered to be an indication for root replacement, as long as the intima within the root is intact and the valve leaflets are otherwise normal, dissection of the left coronary sinus can be also managed by repair techniques. Our preferred technique consists of reinforcing any dissected component of the root with an appropriately fashioned Teflon-felt sheet, placed between the intima and the adventitia becoming essentially a neomedia (Fig. 58.3). The Teflon neomedia is fashioned to allow an appropriate opening for the coronary ostia and is secured in place either with a running monofilament fine suture of either 4.0 or 5.0 at the level of the sinotubular junction or by utilizing a very small amount of biologic glue. It is important to utilize minimal amount of glue as any of them has the potential of causing long-term wall damage, especially of the adventitial layer. Over the last 10 years, repair of a nondilated dissected root with Teflon neomedia has been safe and effective with excellent and highly reproducible physiologic results leading to sparing of the aortic root in over 80% acute type A dissection, with no incidence of redissection or dilatation of the repaired root and no postrepair coronary malperfusions. Alternative procedures include complete valve-sparing root replacement where the valve is placed within an appropriately fashioned Dacron graft, or aortic root replacement with a biologic or mechanical composite graft.
4. *Stroke due to malperfusion of the aortic arch vessels*. The innominate artery is the most commonly dissected of the arch vessels, followed by the left common carotid and at a much lower rate by the left subclavian artery. Replacement of most or all of the aortic arch with reinforcement of the arch vessels insures true lumen patency and prevents most perioperative strokes, frequently reversing preoperative focal neurological deficits.

We usually resect all of the lesser curvature of the aortic arch leaving a

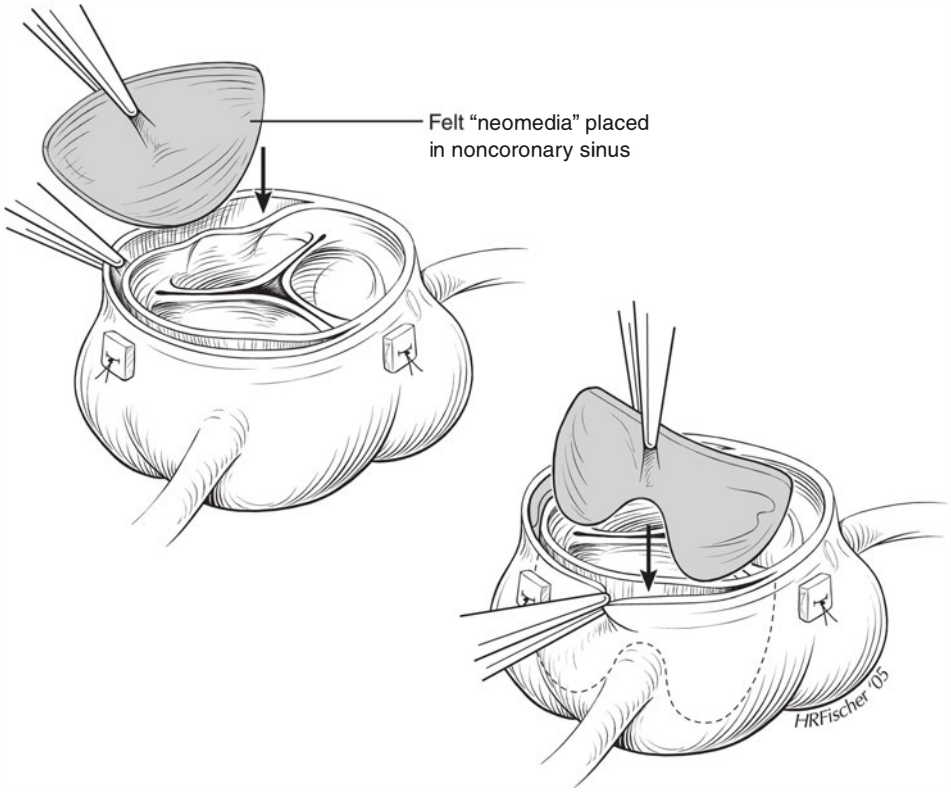


Fig. 58.3. Aortic root repair.

strip of arch tissue comprising the three-arch vessel connected to the descending thoracic aorta. Teflon-felt strips can be placed between the intima and the adventitia as a neomedia to achieve restoration of the true lumen perfusion within the arch vessels to prevent postoperative malperfusion syndromes and reinforce the arch anastomosis suture line, preventing distal disruption (Fig. 58.4). Between 80% and 90% of false lumen beyond the arch will be widely patent after the complete hemiarch repair has been achieved. That does not represent the failure of the arch reconstruction. The reentry fenestrations at the time of surgery are in aggregate so large that it is unrealistic to hope that closing the proximal entry site will ubiquitously result in thrombosis of the false lumen. Our observation is that closure of the false lumen only occurs in limited dissections as DeBakey type 2 or when dissections occur in the setting of atherosclerotic disease where variably fibrosed and calcified media may prevent the usual extension of the dissection and limit reentry sites; in these patients, obliterating the entry point in the presence of low flow from the few exit sites may cause false lumen thrombosis. Total arch replacement may be technically challenging in acute dissection and is only necessary when there are large tears in the greater curve of the arch.

A successful operation for acute type A dissection will leave the patient with a reconstructed or replaced root, a well-functioning aortic valve, a completely replaced ascending aorta, a partially or completely replaced aortic arch with true lumen patency in the arch vessels, and a distal "Type B like" residual aorta (Figs. 58.5 and 58.6). The patient in most instances will have been shifted from a dissection with a high likelihood of death (type A) to a dissection with a very low risk of early rupture and only a moderate risk of long-term aneurysm formation in the residual dissected aorta (type B).

SURGICAL TECHNIQUE

Once the diagnosis of an acute type A dissection is established, the patient is anesthetized under endotracheal general anesthesia. Minimal arterial monitoring consists of a right radial artery catheter and if feasible also a left radial artery line. The most common arch vessel to suffer malperfusion is the innominate artery; thus, the right radial artery would provide early warning of the most likely malperfusion syndrome affecting the brain. If time allows, a left radial artery catheter can give further information about the presence of downstream less common arch malperfusions. Left radial artery pressure monitoring is highly advised if the right axillary artery is being cannulated for bypass. A pulmonary artery (PA) catheter is routinely placed to optimize preoperative hemodynamics, but more importantly

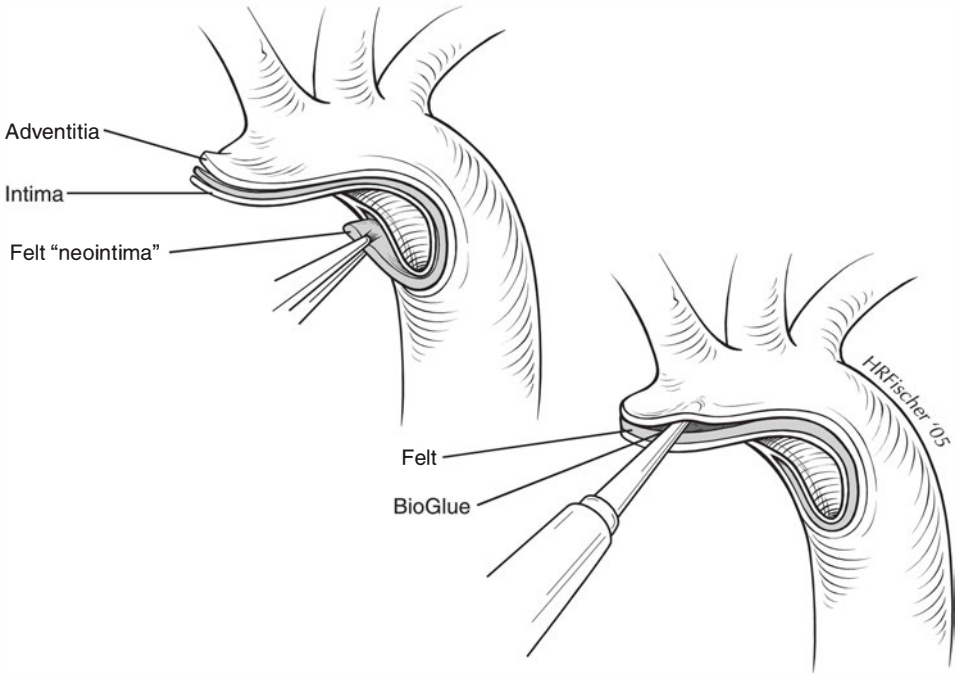


Fig. 58.4. Obliteration of distal false lumen.

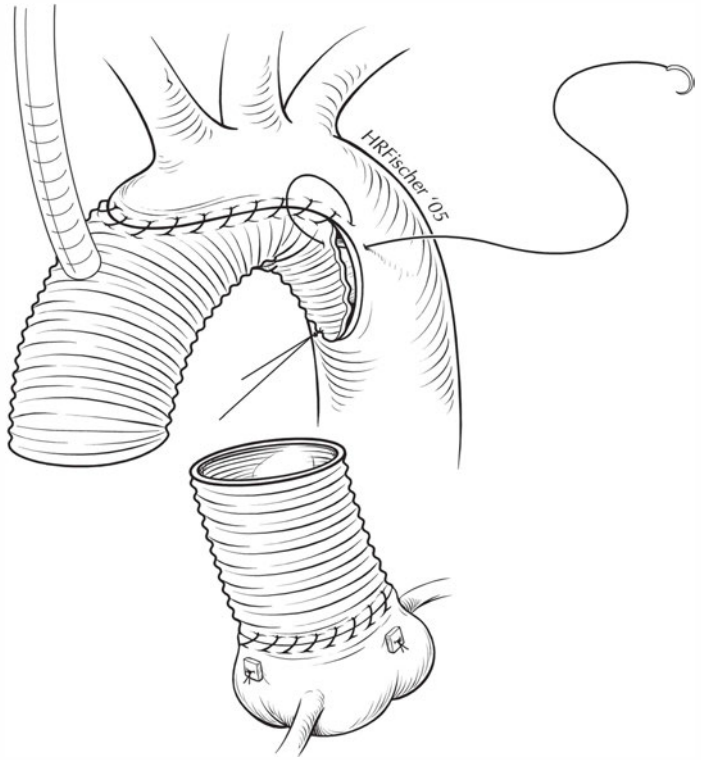


Fig. 58.5. Distal graft anastomosis.

for postoperative management. High filling pressures correlate with bloody pericardial effusion and tamponade. High central venous pressure with low left-sided filling pressures and low PA pressures are highly suggestive of right ventricular ischemia secondary to right coronary artery malperfusion. If a patient is unstable, large bore IV and arterial lines are placed, deferring the PA catheter to a more controlled time. Whenever possible, electroencephalography (EEG) monitoring should be placed both to monitor for any asymmetry of perfusion at the level of the cerebral cortex and to utilize the EEG monitoring as a criterion for achieving adequate cooling to maximize safety to the brain and improve surgical efficiency. Near-infrared cerebral oximetry monitoring is an essential tool to monitor adequacy of brain perfusion during the various phases of the operation. The patient's chest, abdomen, and both lower extremities are then prepped and draped. We currently favor either axillary artery cannulation or in experienced hands, direct over the wire ascending aortic cannulation. For axillary cannulation, an 8- to 10-mm Dacron graft is anastomosed in an end-to-side manner to the vessel. The arterial cannula is then secured to the Dacron graft. This allows for antegrade cerebral perfusion during the arch repair. At the end of the procedure, the graft is oversewn close to the anastomosis. Direct ascending aortic cannulation is performed after sternotomy and heparinization. An 18-gauge needle is placed into the dissected aorta angulated toward the arch, usually initially into the false lumen and advanced through the intimal flap and into the true lumen. A soft wire is then passed into the descending thoracic aorta in the true lumen, and the cannula is passed over the wire after serial dilatation. In a hemodynamically unstable patient without significant atherosclerotic disease, the most expeditious method to obtain arterial access for bypass is the common femoral artery, although malperfusion syndromes are more common with femoral artery cannulation. Malperfusion syndromes of the lower extremities will affect as many as 20% of patients, most commonly the right leg. If both femoral arteries have good pulses, either side can be used. In an unstable patient, the femoral artery has the clear advantage over the axillary artery of allowing two surgical teams to work simultaneously without having to share the same limited thoracic space.

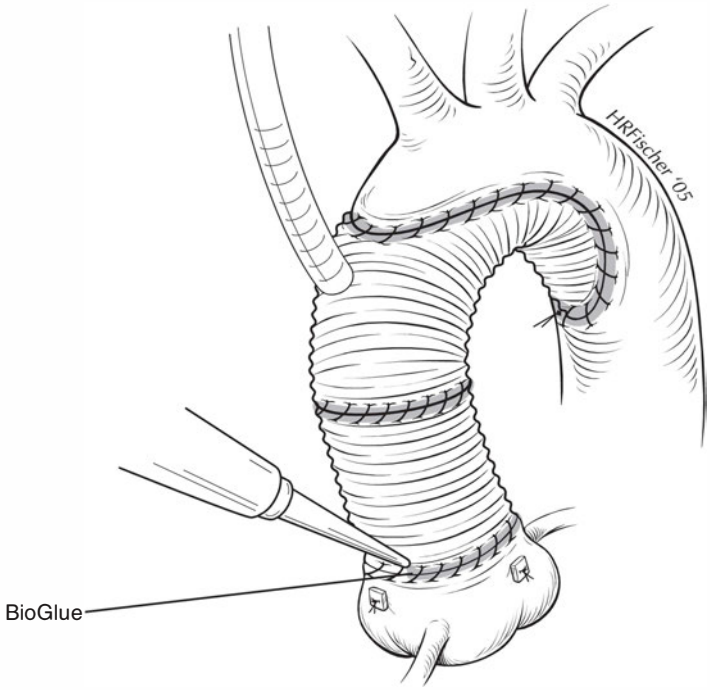


Fig. 58.6. Completed repair.

Once the sternotomy is performed, the pericardium is opened. It is common to find a variable amount of blood-tinged pericardial fluid or even moderate amount of blood under tension. Often the ascending aorta will be moderately dilated, as aneurysm is a major risk factor leading to an ascending aortic dissection. The most common area of ascending aortic leak we found is the mid-ascending aorta. Most often blood enters the areolar tissue between the main PA and the ascending aorta, sometimes dissecting down onto the right ventricular outflow tract which may look quite discolored by blood and thrombus extending often all the way down to the RV to RA groove. It is important early in the process not to

disturb this discoloration or dissected tissue, as it is easy to convert this process into a free rupture. With arterial access obtained, the right atrium is cannulated in the usual manner. We routinely place a retrograde cardioplegia cannula at this time into the coronary sinus, and if needed, an SVC cannula that we will utilize during the circulatory arrest period for retrograde cerebral perfusion either alone or as an adjunct to antegrade cerebral perfusion. The patient is then placed on full cardiopulmonary bypass. During institution of cardiopulmonary bypass, very close monitoring of arterial waves from the right radial and if available the left radial artery are monitored to detect an arch malperfusion syndrome potentially caused by placement on cardiopulmonary bypass. When available, EEG is also closely monitored for hemispheric asymmetry. When EEG monitoring is not available, carotid artery interrogation can be performed by a hand-held probe connected to the same echo machine used for TEE. Anesthesiologists should be proficient with this simple technique of common carotid artery evaluation. The detection of poor or no perfusion in one of the carotid arteries will prompt appropriate action by the surgical team. If a malperfusion syndrome is documented by ultrasound, EEG or a persistent radial artery pressure differential, the patient is weaned off bypass and a different arterial cannulation site is often necessary to resolve the problem. Occasionally, two cannulation sites may be necessary, presumably one supplying the true lumen and the other the false lumen. As soon as adequate cardiopulmonary bypass is noted and no malperfusion syndrome is documented by EEG, arterial pressures, carotid ultrasound, as well as monitoring of the back pressure of the arterial cannulas, the left ventricle is vented via the right superior pulmonary vein. In many of these patients significant aortic valve insufficiency will be present. Core cooling is then begun with the goal of achieving flatline EEG if available or, based on our experience with several hundred monitored circulatory arrest cases, cooling is continued to either achieve a nasopharyngeal temperature of 16°C to 18°C, or for a total period of 50 minutes, whichever one is reached first. Our data suggest that at this level, the vast majority of patients will have a flatline EEG. Moderate hypothermia (22°C to 26°C) may be performed safely if antegrade cerebral perfusion is used. Patients with more severe atherosclerotic disease will likely require longer periods of cooling to achieve uniform end-organ cooling, and adjunctive techniques of retrograde cerebral perfusion or selective antegrade perfusion may be less effective in these situations.

Shortly after beginning the cooling phase, the heart will usually fibrillate. Aortic cross-clamp is then applied to the distal ascending aorta. In reoperative cases, it is critical to develop a cross-clamp site prior to cooling as wide open aortic insufficiency in combination with fibrillation will lead to catastrophic ventricular dilatation. While in the past, there may have been reluctance to cross-clamp a dissected ascending aorta, over the last 15 years, no aortic disruption has resulted by cross-clamping, even in patients with severe collagen vascular disease such as Marfan and Ehlers–Danlos syndrome. Clearly, the aortic cross-clamp should be placed well below the innominate artery takeoff to allow for more margin of safety as sometime more than one cross-clamp may be necessary to fully control this friable, diseased, and often-dilated ascending aorta. At the time of ascending aortic clamping, it is again very important to assess for malperfusion syndromes. It is possible, especially when perfusing from the femoral artery, that the application of the cross-clamp may close the largest communication between the true and false lumen in the ascending aorta, precipitating an aortic arch malperfusion. Monitoring lines in the right and left radial arteries, EEG monitoring of both hemispheres, and common carotid artery Doppler interrogation may show significant asymmetry. If a malperfusion syndrome is documented, expeditious surgical correction is necessary. Cardiopulmonary bypass is turned off with the patient in deep Trendelenburg position. The aortic cross-clamp is removed and the ascending aorta is opened proximal to the clamp site to visualize the dissection flap extending into the aortic arch. The dissection flap is then cut deeply into the aortic arch, thus creating a wide communication between the true and false lumen at the arch. The cross-clamp is then reapplied to the distal ascending aorta as cardiopulmonary bypass is restarted slowly, making sure to avoid cerebral air emboli. While such episodes are rare, occurring in 3% to 4% of our dissection experience, the maneuvers described have been successful in avoiding neurologic injuries that may have lead to ultimate mortality if unrecognized and left untreated. In the era of axillary or direct aortic cannulation, such occurrences have become exceedingly rare.

Once the ascending aorta is cross-clamped without distal malperfusions, it is divided at the level of the right PA and cold blood cardioplegia is given both by retrograde coronary sinus infusion as well as antegrade via direct coronary ostia cannulation. The right coronary is commonly involved in the dissection process and cannulation for cardioplegia gives the first opportunity to assess for the presence of false lumen-induced stenosis or occlusion or even for the presence of complete disruption. If cardioplegia extravasation is demonstrated in the very proximal portion of the right coronary, a segment of saphenous vein should be obtained by a second surgical team to bypass the right coronary arterial system in case a right button ostial repair proves impossible. Left main coronary problems are rare as they are usually fatal out-of-hospital. If they are noted at this point in the operation, additional vein grafts can be obtained to deal with them.

Next, the aortic root is exposed to assess whether it is reparable. If the intima within the aortic root is intact, and the aortic valve leaflets are normal, the root is usually reparable. Mild dilatation of the sinotubular junction up to about 34 mm in maximum diameter can be tolerated with an adequate repair. If the aortic root is severely aneurysmal, consideration of either a valve-sparing root replacement or a mechanical or biological root replacement should be planned at this time. With normal leaflets and intact root intima, the aortic root is mobilized circumferentially insuring adventitial root integrity. The aorta is then transected approximately 3 mm above the sinotubular junction, and three Teflon pledgeted monofilament sutures are placed approximately 2 mm above each of the three commissures. These resuspension sutures allow reestablishment of appropriate commissural geometry. Attention is directed next to the sinuses of Valsalva. The noncoronary sinus is nearly always dissected down to the annulus. Any debris or thrombus within the noncoronary sinus false lumen is cleared. Teflon felt is then fashioned to completely fit within the dissected space of the noncoronary sinus and is then inserted between the intima and the adventitia as a "neomedia." It is secured in place either by utilizing 5.0 or 4.0 monofilament sutures at the level of the sinotubular junction. Attention is next directed to the right coronary sinus of Valsalva. This is involved in over 50% of acute type A dissections and often requires very similar repair technique as used in the noncoronary sinus. Teflon felt is placed between the intima and the adventitia, fashioned in such a way to avoid any narrowing of the right coronary lumen as it courses through the dissected sinus. The left coronary sinus of Valsalva is then inspected. It is rarely involved in the dissection, but when it is, it is treated in a

similar manner as the right coronary sinus with a similar Teflon felt inserted between the intima and the adventitia and secured in the same manner.

If the aortic root is not reconstructable either because of pre-existing root disease or because of significant intimal root destruction and/or leaflets disease or tears, the coronary ostia are mobilized for root replacement. Valve-sparing root replacement is an excellent option in patient <50 years old, with collagen vascular disease and normal leaflets. A mechanical valved conduit also remains an excellent option in younger patients. A biologic root replacement, with a porcine root or handsewn pericardial composite root, should be considered in anyone over 60 years of age as the avoidance of coumadin greatly simplify postoperative management. Patients in this age group who have suffered an acute type A dissection are not likely to outlive their biologic prosthesis.

Usually, adequate cooling will be reached prior to completion of root replacement. To maximize the overall operative efficiency, all proximal work is stopped once flatline EEG, core temperature of 16°C to 18°C, and 50 minutes of cooling are reached, and attention is directed to the aortic arch. Antegrade cardiopulmonary bypass is interrupted, and either antegrade cerebral perfusion via the axillary artery or retrograde cerebral perfusion is begun via the snared SVC cannula. Antegrade perfusion is maintained at 10 cm^3/kg/min and mean pressures are 40 to 70 mmHg. For retrograde perfusion, flows of 150 to 250 cm^3/min are typically used to a target jugular venous pressure of 20 to 25 mmHg. Jugular venous pressure is measured via the right internal jugular sheath used to introduce the PA catheter. The ascending aortic cross-clamp is removed and the aorta is assessed. Now the goal is to excise the remainder of the ascending aorta and to prevent arch malperfusion once antegrade cardiopulmonary bypass is reestablished. The remainder of the ascending aorta is, therefore, debrided. The majority of the undersurface of the arch is excised. The details of the true and false lumen relationships are assessed carefully as there can be significant individual variation. The amount of aortic arch removed depends in large part on the relationship of the arch vessels to the arch false lumen. In general, the entire lesser curvature of the arch and the majority of the anterior and posterior aortic arch are excised leaving a lip of aortic tissue encompassing the left subclavian, left common carotid, and innominate artery. A thin Teflon-felt strip is placed within the false lumen areas around the arch vessels and as necessary into the dissected portions of the residual proximal descending thoracic aorta. Again, the primary goal here is to prevent arch malperfusion with the secondary goal to create a layer that will robustly hold sutures for aortic arch reconstruction. The strip of Teflon felt is secured between the intima and the adventitia around the arch vessels and the dissected portion of the proximal descending thoracic aorta with a few interrupted or short running of 5.0 monofilament nonabsorbable sutures. A Dacron graft is then appropriately beveled and sewn to the reinforced arch tissue utilizing running 4.0 monofilament sutures. Multiple interrupted pledgeted monofilament reinforcement sutures are placed wherever necessary to further buttress the suture line. Deairing maneuvers are meticulously performed.

Occasionally, the aortic arch disruption is so severe that a total aortic arch replacement is necessary. Selective antegrade cerebral perfusion can be carried out via manually inflated balloon tip cannulas inserted in the innominate and the left common carotid arteries. A Dacron graft is sewn to the neo-media-reinforced proximal descending thoracic aorta usually utilizing the elephant-trunk technique. The arch graft is then sewn either to a reinforced island containing the arch vessels or to the individual branches as necessary. Prior to resuming antegrade cardiopulmonary bypass, deairing maneuvers are then performed with the aid of retrograde cerebral perfusion. In our experience, <10% of acute type A dissections required total aortic arch replacement, with another 5% requiring a separate graft to the innominate artery only.

After either a "hemiarch" or a "total arch repair," the Dacron graft is directly cannulated and antegrade cardiopulmonary bypass is reestablished if central aortic or femoral cannulation were used as the initial bypass strategy. Antegrade reperfusion after arch repair minimizes potential malperfusion or arch suture line disruptions. If flow is reestablished via the right axillary artery in the setting of a distal innominate artery dissection, a right common carotid artery malperfusion could develop. Furthermore, the innominate artery false lumen may still have a sufficient distal entry point to sufficiently pressurize the arch suture line to cause disruption of the reconstructed aortic arch. Direct cannulation of the aortic arch graft should be performed in these situations to prevent such local perfusion anomalies, minimizing the risk of arch suture line disruption and maximize true lumen flow. A few minutes are taken early after full flow is established to assess the arch suture line and insure adequate hemostasis. This may be the only opportunity to treat significant bleeding points from the reconstructed aortic. Full rewarming is then allowed to begin while the proximal repair is completed.

The appropriate sized Dacron graft to restore normal sinotubular junction geometry is selected and it is then sewn to the reinforced sinotubular junction with 4-0 nonabsorbable monofilament suture. The proximal reconstruction is then tested by infusing cardioplegia solution into the repaired or replaced aortic root to assess for the presence of significant aortic valve insufficiency as well as suture line hemostasis. The proximal reconstruction is then sewn to the arch graft with running 4-0 nonabsorbable monofilament suture. Cardiac deairing maneuvers are performed and the heart is allowed to reperfuse. During reperfusion, the peripheral arterial cannulation site is appropriately repaired. After full rewarming, the patient is weaned off cardiopulmonary bypass. Early postpump, it is important to assess for significant aortic valve insufficiency. Any more than trace or 1+ AI will result in poor long-term valve durability. If the patient can tolerate going back on cardiopulmonary bypass, 2+ and higher AI should be fixed now, either by valvular replacement within the repaired root or by complete root replacement if necessary. Once the cardiac repair is noted to be satisfactory, the systemic vascular system is assessed to exclude new malperfusion syndromes, or to reassess preoperative ischemic regions. The vast majority of preoperative limb ischemia will resolve after adequate proximal aortic repair. If one limb remains pulseless with good perfusion in the contralateral limb, a femoral–femoral bypass can be performed. If both lower extremities continue to have poor pulses at this stage, an axillary–femoral or axillary–bifemoral reconstruction is favored over opening the abdomen and subjecting the patient to very morbid additional surgery. Stent grafting may provide an effective and durable treatment for abdominal and bilateral lower extremities malperfusions in this setting as well. In the future, a stent graft may be deployed routinely in the proximal descending thoracic aorta via the open aortic arch at the original surgery. This may significantly increase the likelihood of obliterating the distal false lumen, with minimal added operative risk, hopefully sparing many patients from late dissecting aneurysms.

ACUTE TYPE B DISSECTION

Acute type B aortic dissection is defined by the development of the primary tear distal to the origin of the left subclavian artery. Complicated type B dissection is defined as the presence of end-organ malperfusion or rupture. Overall, early risk of mortality is between 5% and 10% in the early period. The major early morbidity and mortality are due to malperfusion syndromes, which in the current era, are typically managed with endovascular intervention. Malperfusion of the viscera or lower extremities may occur as a result of true lumen compression at the level of the aorta itself (pseudocoarctation) or impingement of the true lumen at the level of the branches (Fig. 58.7). Branches may also be sheared off the true lumen and only be supplied by false lumen flow. Rupture is a less common variant occurring in <5% of cases.

The mainstay of early therapy for uncomplicated acute type B dissection remains medical management primarily consisting of strict blood pressure control. Beta-blocking agents provide excellent control of absolute systolic pressure and also minimize aortic wall stress by minimizing the blood pressure rate of rise (dP/dT). In the early phase, intravenous control is usually achieved with esmolol and labetalol. Additional IV vasodilators can be utilized including calcium blockers, such as nicardipine. We try to avoid nitroprusside, except for very short periods very early in the stabilization process. Once the patient has passed the acute phase, oral beta blockers and calcium channel blocking agents are titrated to maintain appropriate blood pressure and heart rate levels. Many of these patients may have presented with a very malignant pattern of hypertension and often require chronically three or four drugs including beta blockers, calcium blockers, clonidine, a central alpha-agonist, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, and diuretics.

It is very important to minimize aortic stress early in the process primarily to avoid rupture, control pain, stabilize treated or mild malperfusions, and allow stabilization of the weakened aorta. Once in the chronic state, tight blood pressure control is maintained to limit aneurysmal degeneration. Many of these patients have severe atherosclerotic distal vascular beds, including cerebrovascular intracranial disease, as well as renovascular and occlusive disease in their abdominal visceral and lower extremity beds. These patients may not respond favorably to "normalization" of their blood pressure. Often a balance has to be reached between minimizing aortic disruption and allowing adequate perfusion of very extensively diseased vascular beds.

Surgical interventions in type B dissection need to be tailored to the specific problems encountered by the individual patient. Unlike type A, most patients with type B dissection are more likely to have significant atherosclerotic disease, with the tear often within a ruptured plaque. In severely atherosclerotic aortas, dissection extension may occasionally be quite limited due to the fibrotic media resisting establishment of a false lumen. Endovascular intervention for complicated type B dissection is highly effective to treat malperfusion and rupture and is associated with <10% mortality versus >50% mortality for traditional open repair. Endovascular therapy allows for more expeditious reexpansion of the true lumen and avoids the massive bleeding and physiologic insult associated with open dissection repairs. Given these dramatic improvements, endovascular techniques have virtually completely supplanted open surgery when technically feasible. Current technique consists of establishing iliofemoral access and placement of a wire in the true lumen using intravascular ultrasound guidance. A covered aortic stent graft is placed across the intimal entry point at the level of the subclavian artery, which is usually at least partially covered to achieve a proximal landing zone of at least 2 cm proximal to the entry tear. Malperfusion syndromes are typically treated with initially stent grafting the proximal third to

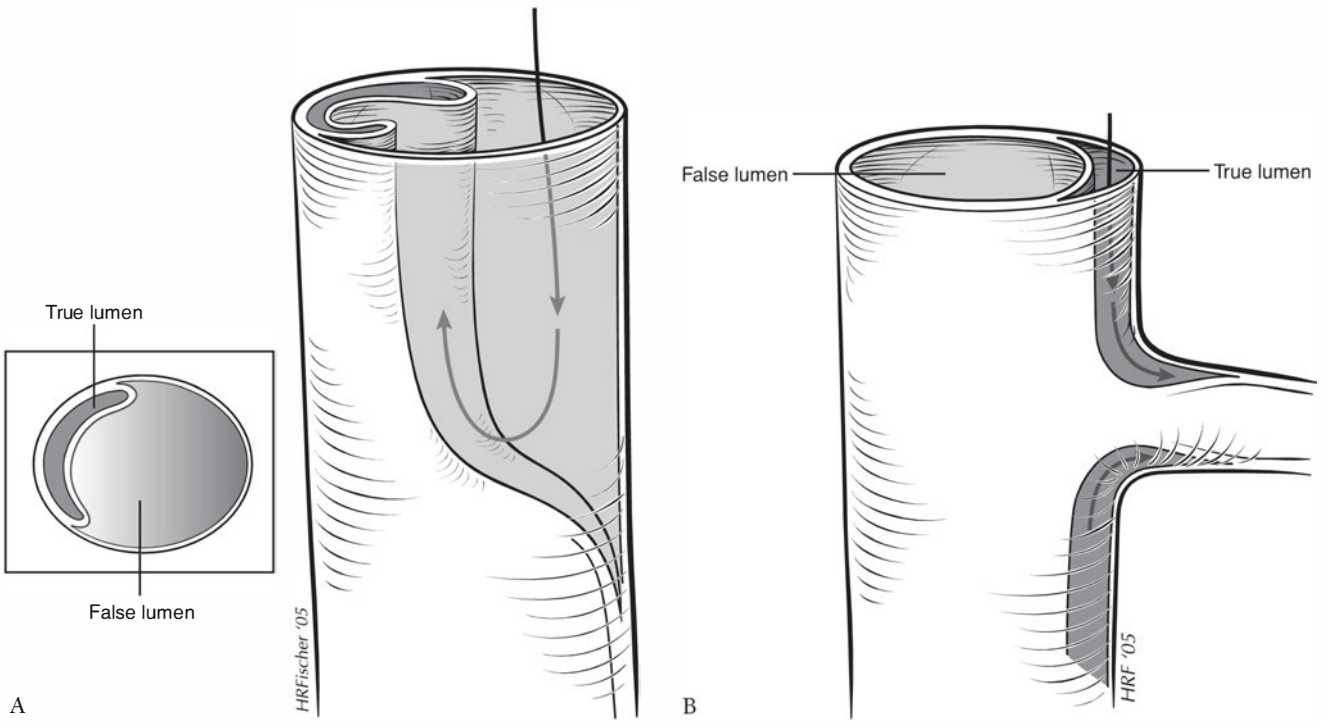


Fig. 58.7. (**A**) Pseudocoarctation. (**B**) Malperfusion syndrome.

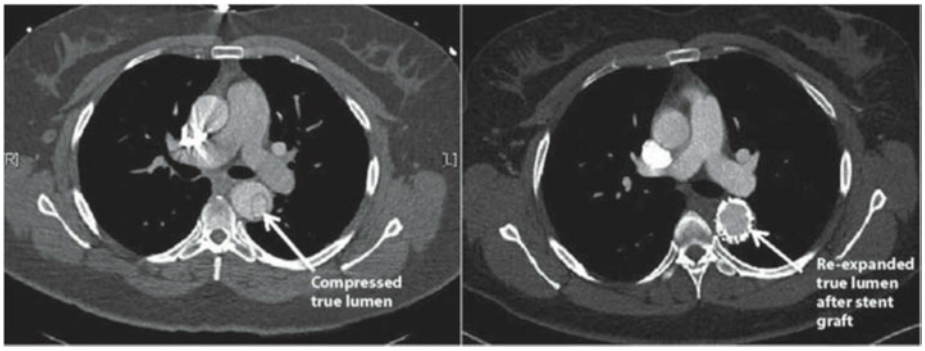


Fig. 58.8. TEVAR for acute Type B dissection.

half of the descending thoracic aortic true lumen (Fig. 58.8). In situations with pseudocoarctation, where the true lumen is totally collapsed, extension of the stent grafts to the level of the celiac artery may be required to achieve reexpansion. This may be extended even further distally with bare-metal stents across the visceral segment. Individual branch vessel malperfusions either in the iliofemoral system or the visceral segment may also be stented open with bare-metal stents as adjunctive procedures. Ruptures are treated with complete coverage of the thoracic aorta from left subclavian artery origin to the celiac artery origin.

Open surgical techniques for dealing with malperfusion include placement of a short segment graft in the descending aorta to reestablish the true lumen, extra-anatomic bypasses, and open surgical fenestration. If anatomy is not favorable for stent grafting, limb malperfusion is expeditiously managed with femoral–femoral bypass.

SPECIAL CASES: INTRAMURAL HEMATOMAS AND RETROGRADE EXTENSIONS OF TYPE B DISSECTIONS

Intramural hematoma is a variant of aortic dissection in which there is hematoma within the aortic wall in the absence of a classical intimal tear and no flow within a false channel. More common in older patients with severe atheroma of the aorta, intramural hematoma typically occurs as a result of bleeding into an atherosclerotic plaque or spontaneous bleeding in the vasovasorum of the aorta. They are best classified in the same manner as classic aortic dissections but may be managed somewhat differently. Type A aortic intramural hematoma is generally managed with urgent surgery as there is a significant risk of progression to frank aortic dissection or rupture. As these patients are frequently older and may have more comorbidities, medical management with strict blood pressure control and reversal of anticoagulants may be employed in patients with suitable anatomy who are poor surgical candidates. Favorable anatomy for medical management includes an absolute aortic diameter including hematoma of <4.5 cm with a hematoma thickness of <0.5 cm and absence of pericardial effusion. Serial CT angiogram should be performed within 48 hours of presentation for the determination of progression. Type B intramural hematoma is generally managed medically unless there is concern for rupture or impending rupture. In these cases, thoracic aortic stent grafting is the treatment modality of choice with full subclavian to celiac coverage. These cases are particularly high risk for spinal cord ischemia as the intercostal arteries are frequently sheared off by the hematoma and spinal perfusion is already compromised.

Occasionally, both type A and B dissections may present with either a partially or completely thrombosed false lumen, in which there is a true intimal tear but no flow within the false lumen. This may be indistinguishable from an intramural hematoma but typically has a more classic spiral dissection appearance with a compressed true lumen, as opposed to intramural hematoma, which has a more circumferential appearance without true lumen compression. Our approach has been to treat such patients with a thrombosed false lumen in the ascending aorta in the same manner as a standard type A dissection, thus proceeding to surgery without delay. Often these patients have a higher degree of atherosclerotic disease which may account for the lack of further progression of their dissection or for the early thrombosis due to sluggish false lumen flow secondary to minimal reentry sites. A type B dissection with thrombosed false lumen should be treated in the same manner as a standard type B dissection, with primary medical treatment. However, a partially thrombosed false lumen when pressurized is more likely to compromise true lumen flow leading to pseudocoarctation. Appropriate assessment and potential stenting or extra-anatomic bypass of the pseudocoarctation should be considered.

Aortic dissection may rarely arise in the distal arch and then progress both retrograde and antegrade. This process may on occasion be difficult to classify within the Stanford system and it may be difficult to decide whether the initial management should be surgical or medical. Most of these patients will have significantly atherosclerotic aortas with the false lumen becoming pressurized early in the descending thoracic aorta exerting significant retrograde forces to extend the dissection retrograde as the path of least resistance. If the retrograde extension ends at the level of the aortic arch and does not involve significant portions of the ascending aorta, the primary additional risk to a standard type B dissection is a cerebrovascular accident. The risk of rupture is relatively low. While our management remains individualized, we would initially observe patients with this type of retrograde extension and treat them as type B dissections. If, however, the ascending aorta becomes involved, either acutely or during follow-up, these patients should be treated as a standard type A dissection as they do suffer from the same risks of rupture, aortic valve insufficiency, and aortic root compromise leading to coronary malperfusion. We find, furthermore, that these patients have a higher surgical risk because the aortic arch is often completely destroyed and very difficult to piece back together. These are the classes of patients in whom an often very technically challenging total arch replacement may need to be performed, with an attendant higher mortality and a higher incidence of neurologic complications. Hybrid arch debranching procedures may by particularly useful in this setting as this limits the complexity of the arch repair. In this hybrid procedure, the ascending aorta is replaced as in a standard dissection, with an open distal anastomosis to the distal ascending, avoiding a deep arch anastomosis. The head vessels are rerouted using extra-anatomic bypasses from the ascending aortic graft. A stent graft is then deployed using fluoroscopic guidance with a proximal landing zone in the ascending aortic graft and distally in the descending aorta.

LONG-TERM FOLLOW-UP AND CHRONIC DISSECTIONS

Any patient with a valve-sparing repair or replacement of their aortic root needs lifelong surveillance to monitor the long-term durability of their aortic valve function. Patients with <1+ AI at the time of primary surgery have had an extremely durable repair utilizing the technique described. If the valve fails, regurgitation is typically the problem and an aortic valve replacement alone is often sufficient as the aortic root has been reinforced sufficiently to make the incidence of root dilatation very low.

All patients who have undergone proximal repair for an acute type A dissection will need lifelong surveillance of the residual aorta. Approximately, 90% will have a residual type B-like dissection beyond the completion of the arch repair. This residual dissection is at risk for aneurysm formation. If a dissecting aneurysm develops, they are candidates for repair based on standard aneurysm management criteria. We find 30% to 40% of patients will require downstream aneurysm surgery after proximal type A repair over 5 to 10 years of follow-up, and the risk will be higher the younger the patient, with a risk as high as 70% to 90% in patients with collagen vascular disease. Any patient who requires a distal aortic arch and descending thoracic aortic replacement must have a very careful evaluation for the presence of significant aortic valve insufficiency. Full cardiopulmonary bypass through the left chest with hypothermia leading to fibrillation can be very problematic in the presence of more than 1+ aortic valve insufficiency. It is difficult to control the ascending aorta from the left chest and significant left ventricular distension can occur, even with a well-positioned LV vent. This can lead to cardiac and pulmonary dysfunction at the completion of the distal arch and descending thoracic replacement. To prevent such problems, the aortic valve may need to be replaced ahead of the distal aortic repair.

Occasionally, a patient will survive an undiagnosed acute type A dissection and will present months to years later with a large ascending dissecting aortic aneurysm with variable amount of AI. These patients are treated surgically in an urgent manner because the risk of rupture is higher than a nondissected ascending aneurysm of equal size. These aneurysms may have grown quite rapidly and present occasionally with compression symptoms to surrounding structures. At surgery no attempt should be made to obliterate the distal false lumens as many distal vascular beds will have become dependent on false lumen flow. The dissection flap should be resected as far as reachable to maintain free flow in all distal lumens. Depending on the sinus involvement and the chronicity, valve resuspension techniques may not be effective. If the aortic leaflets appear relatively preserved, a valve-sparing root replacement may be a good option. In the chronic setting, total arch replacement with branched arch grafts is more often required, and it carries a significantly lower morbidity and mortality compared with the acute setting.

CONCLUSION

Aortic dissection remains a catastrophic diagnosis with a time-related, high early mortality rate. Improvements in mortality for the treatment of acute type A dissections has been achieved by improvements in diagnostic techniques, expeditious transfer to the operating room prior to cardiovascular collapse, better physiologic understanding of intraoperative malperfusion syndromes especially affecting the arch vessels, intraoperative neurologic monitoring leading to real-time surgical maneuvers to reverse abnormalities, precise repair or replacement of the aortic root, and precise and durable anastomoses at the level of the arch. Type B dissections have remained a relatively low-risk early process where medical management remains the mainstay. Thoracic aortic stent grafts have converted extremely high-risk complicated type B dissections into rapidly managed problems with generally good outcomes. Application of stent graft technology to uncomplicated type B dissection to promote distal remodeling, and eventually, novel catheter-based prostheses for type A dissection represent the future directions of innovation in this field.

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EDITOR'S COMMENTS

Dr. Bavaria and his team have set the standards for the surgical treatment of aortic dissection. I agree completely with their overall strategy. Certainly, the treatment of type A dissection is a surgical emergency. I very much like their approach of bringing patients urgently to the operating room and ruling them out there. Obviously, if they come in with a diagnosis and appropriate imaging that is one thing but certainly the most urgent case just need to be in the operating room.

I also agree entirely with their approach at doing their best to spare the aortic root unless there is evidence of annuloaortic ectasia. I agree with their approach of replacing sinuses if needed based on where the tear occurs. I also agree entirely with cannulation strategy. I think axillary approach may be a little more time-consuming in patients who are serious and femoral cannulation is appropriate. We almost always use a side arm graft to use central cannulation once the distal repair is done. We have a somewhat different approach to the anastomoses. I have used no glue and very little pledgeted material but very small needles to do the repair. I have found this in fact makes hemostasis easier because one can easily identify bleed points and fix them with a small suture.

Finally, I very much agree that distal dissections following the proximal repair are the norm rather than the exception. There are multiple reentry points and clearly there needs to be surgical follow-up of these patients for the rest of their lives.

ILK

59 Descending Thoracic and Thoracoabdominal Aortic Aneurysms

Matthew L. Stone and John A. Kern

INTRODUCTION

Aortic aneurysms are defined as a doubling of the normal aortic diameter for a particular body surface area, age, and gender. Advancements in open surgical technique and the advent of endovascular treatment have supported a significant improvement in outcomes and survival for many with descending thoracic and thoracoabdominal aortic aneurysms. Since the inception of successful thoracic and thoracoabdominal aortic reconstruction by Etheridge and DeBakey in 1955 and 1956, commitment to technical refinement and risk stratification has expanded the treatment potential for patients with thoracoabdominal aortic aneurysmal disease. The promise of current treatment has introduced preventative standards for reconstruction in the setting of chronic aortic aneurysm while advancing the success of treatment for acute aortic rupture and malperfusion. A principle achievement over the past decade of care has been the standardization of operative and critical care techniques for the maintenance of branch vessel and end-organ perfusion, which have minimized rates of postoperative organ failure and paraplegia. As we embark on a promising future for open and endovascular repair techniques, a continued commitment to evidence-based practice is imperative to further the curative potential for patients with descending thoracic and thoracoabdominal aortic disease.

The focus of this chapter is to review the current diagnostic standards and operative principles for the surgical treatment of thoracic and thoracoabdominal aortic aneurysmal disease. Open and endovascular techniques are presented as both independent and hybrid approaches to thoracic and thoracoabdominal aortic reconstruction.

ANATOMIC PRINCIPLES

Descending thoracic aortic aneurysms arise in the thoracic aorta distal to the origin of the left subclavian artery. Thoracoabdominal aortic aneurysms span the diaphragmatic hiatus at the level of T12 and introduce important considerations for pleural entry, peritoneal access, and diaphragmatic conservation for aortic repair. In addition, phrenic nerve localization and preservation are imperative within the pericardium as it crosses the left atrium and terminates distally on the abdominal surface of the diaphragm.

Thoracoabdominal aortic aneurysms may be classified according to the anatomic extent of disease as proposed by the Crawford classification system: type (or extent) I (24%) involving the proximal descending thoracic to proximal abdominal aorta, type II (26%) involving the proximal descending thoracic to infrarenal abdominal aorta, type III (26%) involving the distal descending thoracic and abdominal aorta, and type IV (24%) involving the abdominal aorta and including visceral vessel segments (Fig. 59.1). An understanding of the visceral and somatic branches along the thoracoabdominal aorta provides the foundation for operative techniques that maintain spinal cord and organ perfusion. Paired somatic branches arise from the third intercostal arteries and continue to the fourth lumbar arteries. The critical vascular zone of the spinal cord from T4 to L1 vertebral levels is characterized by the least prominent blood supply and is the zone at which interference with the circulation is most likely to result in paraplegia. Importantly, this anatomic guideline may vary based on the presence and extent of preexisting aortic reconstruction. The artery of Adamkiewicz is the largest anterior medullary feeder for the supply of the lumbar cord and arises from the lower intercostal or lumbar artery on left in 65% to 80% of cases between T6 and L4 levels. The lower intercostal and lumbar artery supplying the artery of Adamkiewicz should be preserved or reconstructed to maintain blood supply to the lumbar spinal cord to minimize the risk of spinal cord injury. Perfusion through medullary arteries maintains spinal cord blood supply proximal to the aortic cross-clamp and determines postoperative neurologic function. Visceral branches are located on the ventral aorta as the celiac axis, superior mesenteric, inferior mesenteric, and paired or multiple renal arteries.

Localization of somatic and visceral branches during each individual aortic reconstruction guides decisions regarding clamp placement, perfusion techniques, and the need for reimplantation or graft reconstruction. Multidetector row computed tomography (CT) provides characterization of the critical spinal cord blood supply for targeted reconstruction and may be considered for elective complex repairs. The anterior spinal artery is a principal component of the extensive intraspinal and paraspinous collateral blood supply to the anterior spinal cord, with 75% of all segmental arteries providing direct anterior spinal artery-supplying branches. This extensive collateral network imparts a responsibility to define dominant intercostal and lumbar arteries for each individual patient to guide operative planning for reconstruction and preservation of blood supply to the spinal cord.

EPIDEMIOLOGIC AND DIAGNOSTIC CONSIDERATIONS

The prevalence of thoracic aortic aneurysms has tripled over the past two decades to a current estimate of 10.4 cases per 100,000 person-years. Patients with thoracic aortic aneurysms are at a mean age of 59 to 69 years with a 2:1 to 4:1 male predominance. Unlike ascending aortic aneurysms, descending aneurysms are often associated with significant atherosclerosis, yet this may represent more of an association than causal relation. The natural history of aortic aneurysm formation is diverse and may

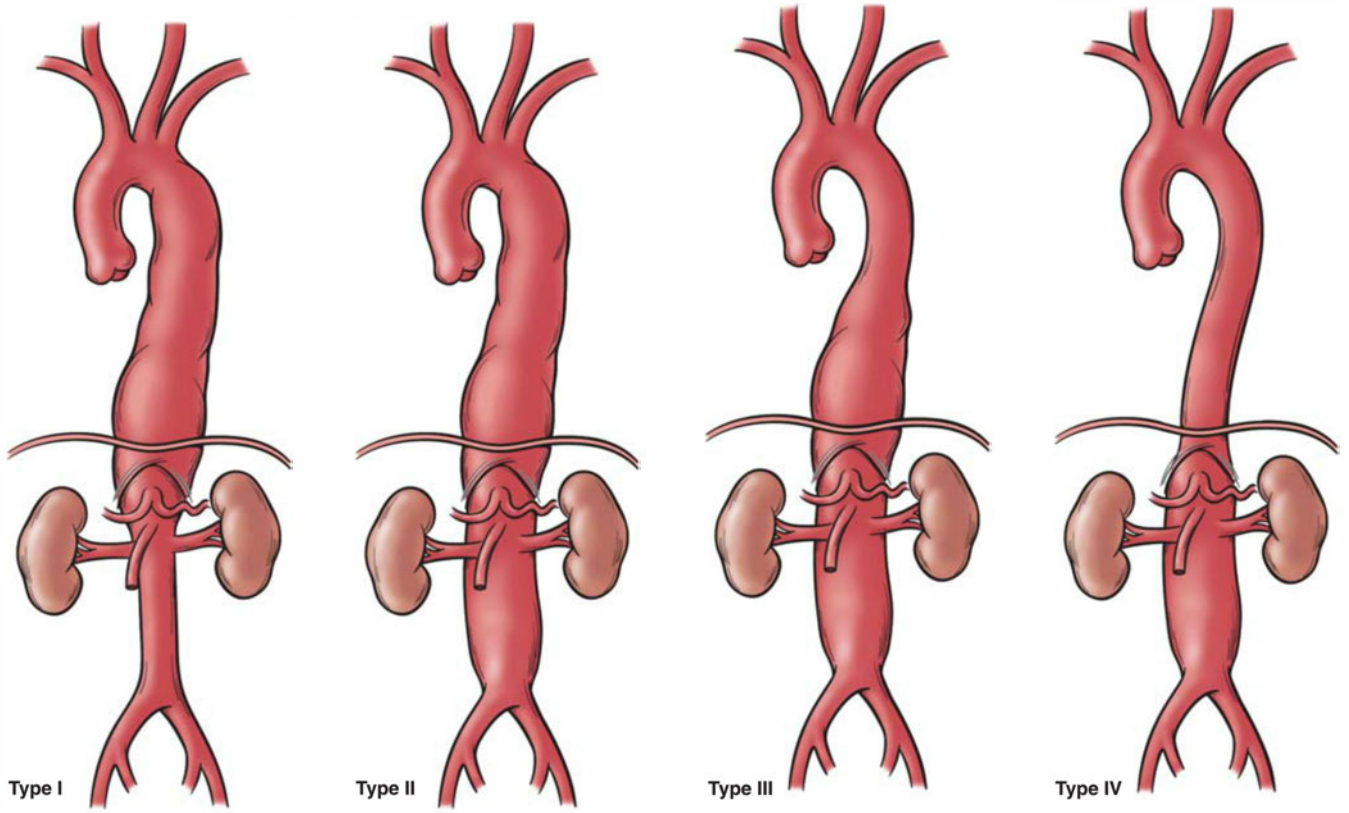


Fig. 59.1. Crawford classification of thoracoabdominal aortic aneurysms.

result from cystic medial necrosis, chronic dissections, aortitis, traumatic transaction, or conditions related to the degeneration of the aortic media such as Marfan syndrome, Ehlers–Danlos, annuloaortic ectasia, trauma, infections, mycotic conditions, syphilis, or idiopathic causes. Hypertension predisposes a patient to aortic dissection and subsequent potential aneurysm formation secondary to the increased thrombogenicity and incomplete decompression of the aortic false lumen. In retrospective review, the incidence of degenerative aneurysms without dissection was 73%, concomitant chronic dissection was 23%, and acute aortic dissection was 4%.

History and physical examination remain principal tenants to the management of a patient with presumptive descending thoracic and thoracoabdominal aortic aneurysm. The primary aim of physical examination is to exclude acute dissection, rupture, and malperfusion. Neurologic deficits, acute abdominal pain, hematuria, and lower extremity ischemia are indications of aneurysm-related malperfusion or distal embolism and prompt immediate intervention. Hypotension with hemorrhage into the left chest and pericardium represent rupture and support emergent resuscitation and repair. Acute or chronic enlargement of the thoracic aorta may manifest as chest and back pain or hoarseness secondary to recurrent laryngeal nerve compression. A nonproductive cough or hemoptysis may represent contained rupture or bronchial irritation, while gastrointestinal hemorrhage may occur in the setting of an aortoenteric fistula.

While an estimated 65% of patients with thoracic aortic aneurysms are symptomatic at the time of operative intervention, an increasing number of asymptomatic aneurysms are now detected on unrelated imaging. Primary risk factors for the development and size progression of thoracic aortic aneurysm include hypertension, smoking, and chronic obstructive pulmonary disease. Aortic wall tension and associated risk of rupture increase as the aneurysmal segment increases in diameter, as represented by Laplace's law. The descending thoracic aorta is estimated to grow on average 0.19 cm per year and can attain a growth rate as high as 0.28 to 0.48 cm per year in the presence of aortic dissection.

IMAGING

The choice of imaging technique for evaluation of the thoracic aorta is determined by patient-related factors (hemodynamic stability, renal function, contrast allergy) and institutional capabilities (technologic capability, expertise). CT-induced contrast nephropathy and magnetic resonance (MR)-associated gadolinium nephrogenic systemic fibrosis are principal considerations for patients with borderline kidney function (serum creatinine $>$ 1.8 to 2.0 mg/dl).

Chest X-ray has demonstrated specificity for acute aortic pathology of 86% in prospective study of patients undergoing evaluation for acute thoracic aortic disease. This imaging modality is, however, insufficient to definitely exclude thoracic aortic aneurysm in high-risk patients and lacks anatomic detail necessary for directed treatment. CT angiography (CTA) is accepted as the primary diagnostic imaging modality for the thoracic aorta with a demonstrated accuracy of 92% for all-inclusive abnormalities of the thoracic aorta and has an established efficacy in the prediction of the need for hypothermic circulatory arrest during surgical repair. Image acquisition should include thoracic branch vessels and femoral vessels to determine aneurysm extension and to guide potential endovascular access. Three-dimensional CTA reconstruction may provide additional advantages to operative planning.

MR angiography (MRA) provides similar advantages to CTA without the limitations of radiation exposure or iodinated contrast. Phase-contrast techniques and two-dimensional time-of-flight modalities have increased the application of MRA in the setting of thoracic aortic aneurysm and dissection, with beneficial applications in the determination of flow dynamics within the false channel. First-line adoption of this imaging modality in the setting of thoracic aortic aneurysm remains limited by institutional capabilities and the time required for acquisition. Current aortic sizing for intervention and endograft sizing are based on the external aortic diameter derived from CTA or MRA or the internal aortic diameter on echocardiography. In addition to providing insight regarding aortic size and anatomic aneurysm characteristics, both CTA and MRA guide the selection of safe sites for arterial cannulation and cross-clamp application.

The overall risk of rupture at 5 years following the initial diagnosis of descending thoracic or thoracoabdominal aortic aneurysm is estimated to be 20% and is dependent on the aortic size at diagnosis: 0% for aneurysms <4 cm diameter, 16% for those 3 to 5.9 cm, and 31% for aneurysms 6 cm or more. In addition, yearly composite adverse outcomes of rupture, dissection, and death are estimated to occur in 14.1% of patients with aneurysms 6 cm and greater in diameter in the absence of surgical intervention. These data support size-related treatment as represented by the expert American Heart Association consensus guidelines for intervention based on Class I evidence:

1. Open repair is recommended for chronic descending thoracic aortic dissections with an aortic diameter exceeding 5.5 cm.
2. Endovascular repair should be considered for degenerative aneurysms exceeding 5.5 cm, saccular aneurysms, and postoperative pseudoaneurysms.
3. Elective surgery is recommended for thoracoabdominal aneurysms exceeding 6.0 cm or less in the setting of connective tissue disease with limited endovascular options and elevated surgical morbidity.
4. Additional revascularization procedures are recommended for patients with end-organ ischemia or visceral artery atherosclerotic disease.
5. Patients with symptoms consistent with thoracic aneurysm enlargement warrant prompt surgical intervention unless candidacy is limited by comorbid disease or quality of life.

For endovascular techniques, a suitable landing zone of normal diameter with no circumferential thrombus 2 to 3 cm distal to the left subclavian or left common carotid artery and proximal to the celiac axis is required. For patients with anatomic limitations, endovascular techniques may also create landing zones for candidate aneurysms. In addition, a femoral vessel diameter > 7 mm and an aortic curvature <60 degrees optimize sheath advancement and stent deployment. Endograft sizing recommendations are to aim for 10% to 20% over sizing at the proximal landing zone. These guidelines represent current suggested thresholds within the rapidly advancing field of endovascular aortic aneurysm repair and should not be accepted as absolute criteria. With both endovascular and open repairs, the decision for surgical intervention and reconstruction is founded on a calculated heightened risk of nonoperative medical management that exceeds the risk of the selected operation. A preoperative understanding of potential ischemic heart disease, obstructive lung disease, and renal impairment is imperative prior to operative intervention. Medical management and optimization of end-organ function prior to endovascular or open repair is a critical consideration for all patients to minimize perioperative morbidity and mortality.

OPEN DESCENDING AND THORACOABDOMINAL RECONSTRUCTION

Anesthesia and Monitoring

Induction and maintenance of general anesthesia are achieved by double-lumen endotracheal intubation. A single-lumen endotracheal tube may be utilized for aneurysms isolated to the lower thoracic and upper abdominal aorta. Large-bore peripheral access, central venous pressure monitoring, continuous arterial pressure assessment, pulse oximetry, and foley catheter are imperative to guide intraoperative resuscitation and support. Transesophageal echocardiography, continuous electrocardiographic monitoring, and a pulmonary artery catheter provide additional adjuncts for intraoperative cardiac evaluation and are routine at our institution. Femoral artery access is needed with cardiopulmonary bypass to maintain balanced pressures. Temperature monitoring is achieved at two access sites to estimate cerebral (blood, esophageal, tympanic membrane, nasopharynx) and visceral (bladder, rectal) temperatures. Cerebrospinal fluid (CSF) drainage is the principal modality for spinal cord protection utilized at our institution due to the established benefit and low-risk of this protective strategy. Motor-evoked potential (MEP) and somatosensory-evoked potential (SSEP) monitoring may be applied in addition to CSF drainage for spinal cord protection and monitoring. Following anesthesia induction, the patient is rolled to expose the left flank and chest wall with the pelvis rolled half-anteriorly to enable access to the left femoral vessels (Fig. 59.2). The table is flexed to open the intercostal spaces, axillary rolls are applied, and pressure points are padded with care.

Incision and Exposure: Thoracic Aneurysms

An extended posterolateral thoracotomy is performed to expose the entire length of the thoracic aorta. The latissimus dorsi and a minimal portion of the serratus anterior muscles are divided. Left lung ventilation is terminated prior to entry into the pleural cavity. Aneurysms of the upper or middle thoracic aorta are accessed through a single intercostal space, which may be facilitated by division of one of the ribs posteriorly. We prefer two separate sites of entry through the fourth interspace and the seventh or eighth interspace for more extensive descending thoracoabdominal aneurysms. The fourth interspace is critical to proximal clamp application. Careful entry into the pleura allows anterior retraction of the lung under sponges to expose the aorta. The pleura over the aorta is divided. The intrathoracic course of the vagus nerve, recurrent laryngeal nerve, and proximal phrenic nerve at the aortic arch is identified. The aorta is then taped immediately proximal to or at the origin of the left subclavian artery where careful, sharp dissection allows for future application of the aortic cross-clamp. The thoracic duct and adjacent lymph vessels should be avoided. Intraoperative lymph leaks should be immediately repaired. The identification of the esophagus may be augmented by the direct palpation of the nasogastric tube or transesophageal echocardiography scope to avoid inadvertent injury.

Following achievement of the proximal aortic dissection, attention is directed to the aorta distal to the aneurysm. In the setting of extensive thoracic aortic involvement, a second pleural entry through the separate intercostal incision may be needed. Careful sharp dissection of the distal aorta is performed to achieve adequate control.

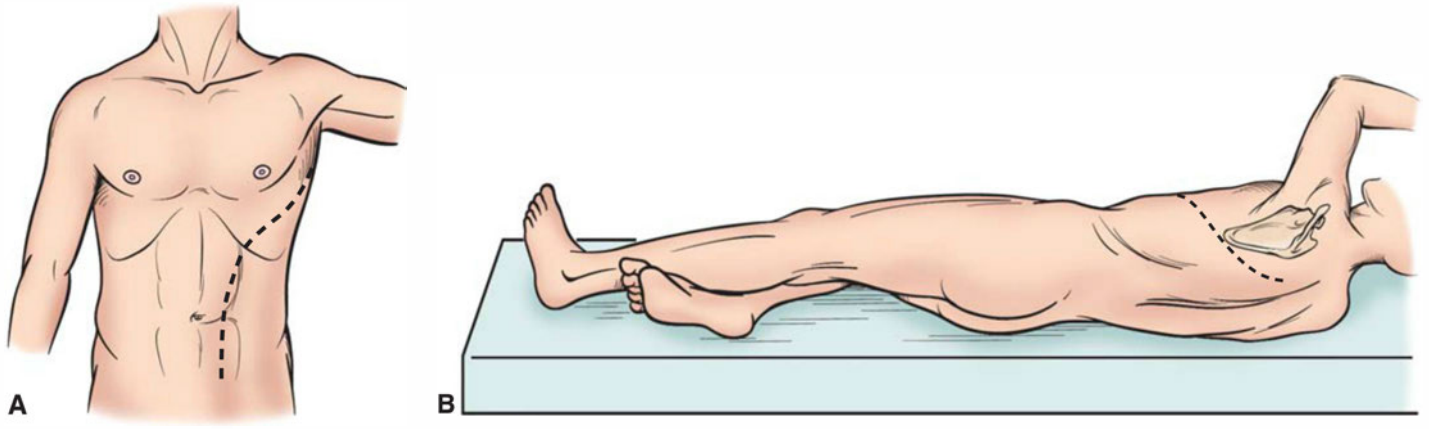


Fig. 59.2. (**A**) Thoracoabdominal incision as determined by extent of aortic aneurysm repair. (**B**) Posterior incision extension with patient positioned to expose the left flank while the pelvis is rolled half-anteriorly to enable access to the left femoral vessels.

Importantly, no attempts are made in the dissection of the aneurysm along its entire length to avoid potentially hazardous bleeding; however, posterior mobilization of the aneurysmal segment may be necessary to support clip application to the intercostals prior to opening of the aorta.

Incision and Exposure: Thoracoabdominal Aneurysms

Our standard incision is started posterolaterally over the ribs of the seventh, eighth, or ninth interspace dependent on the proximal extent of the aneurysm. The incision is then advanced across the ninth interspace at the costal margin to curve inferiorly to run parallel and left-lateral to the midline and rectus sheath. Single right lung ventilation is initiated and the left pleural space is entered. The abdominal muscles are divided and the peritoneum is preserved. Special care is needed at the lateral edge of the rectus abdominis muscle where the peritoneum and transversalis fascia are closely apposed to the abdominal wall. While the retroperitoneal approach is our preferred technique, we recognize that the intraperitoneal approach affords mobilization of the spleen and left colon, which may enable better access to the abdominal aorta in select patients. The diaphragm may be taken down with a curved incision along the costal margin with care to preserve a 3-cm rim along the posterior aspect of the rib. The tendinous center of the diaphragm is then conserved when anatomically feasible to improve postoperative respiratory recovery and weaning time. This limited phrenotomy technique allows passage of the graft through the natural hiatus of the diaphragm. The incision is continued down to the crura, and the left crus may be divided to expose the aorta beneath. A radial diaphragmatic incision may also be utilized and has become our preferred technique (Fig. 59.3).

Progressive retraction of the peritoneum and its contents will facilitate exposure of the retroperitoneum and a self-retaining retractor is necessary. The aorta is located medial to the iliopsoas muscle. Mobilization of the left kidney from the bed of the psoas muscle may provide additional exposure and is preferred at our institution. Importantly, this step is deferred in patients with a retroaortic left renal vein. Anterior visceral branches are identified and sharply dissected with attention to mobilization should reimplantation or bypass be required. Dissection is performed proximal and distal to the aneurysmal segment of aorta and each site is tapped in preparation for cross-clamping.

Hemodynamic Support during Aortic Cross-Clamping

Proximal aortic cross-clamp application induces a significant increase in cardiac afterload. Sudden afterload reduction following clamp release is associated with an acute relative hypovolemia and systemic hypotension. These periods

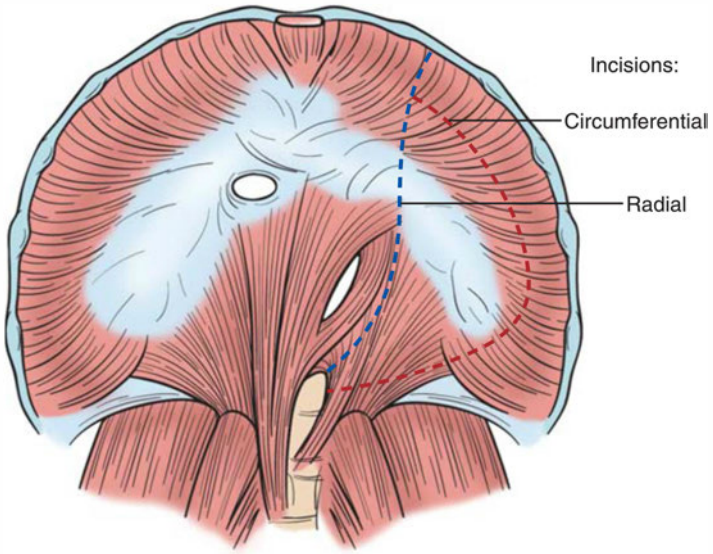


Fig. 59.3. Diaphragmatic incisions for thoracoabdominal aortic aneurysm repair.

of hemodynamic change are managed through the following intraoperative strategies.

Pharmacologic Manipulation and Clamp Application Technique

Direct and cooperative communication between surgeon and anesthesiologist is imperative during periods of cross-clamp application and release. Our approach to cross-clamp application and distal aortic perfusion involves active distal aortic perfusion with femoral-to-femoral bypass for all but type IV and V aortic aneurysms, for which we minimize supraceliac clamp times with no active distal perfusion. This perfusion strategy maintains distal aortic perfusion with warm, oxygenated blood and provides the time necessary to safely perform the most complex of repairs. With this strategy, maintenance of distal aortic blood pressure is imperative and is achieved through the titration of pharmacologic agents and pump flow rates to maintain optimal perfusion. Infusion of nitroglycerine, trimethaphan, or sodium nitroprusside prior to the application of the aortic cross-clamp may be performed to lower systolic blood pressure to 70 to 80 mmHg. The anticipated acute rise in blood pressure with aortic cross-clamp application should necessitate aggressive blood pressure control until planned clamp release. Sodium bicarbonate, calcium, and rapid volume infusion may be initiated prior to unclamping to prevent acute hypotension. Vasopressor agents may also be utilized to augment these resuscitative measures to maintain blood pressure upon clamp release. Importantly, our principal focus is to carefully titrate both volume and pump flow rate to maintain target pressure, often eliminating the need for these additional interventions throughout the critical periods of clamp application and release.

Progressive clamp application and release over a period of 2 to 4 minutes may blunt the imposed physiologic insult and hemodynamic response. In assessment of end-organ ischemia, cross-clamp time is measured from the first click of clamp application until its complete release. Despite these preventative and resuscitative measures, hemodynamic instability may occur and should be anticipated with appropriate preparation.

Extracorporeal Circulation

Extracorporeal circulation support provides after load reduction and continuous end-organ perfusion during the aortic cross-clamp period. Techniques for the maintenance of extracorporeal circulation include passive aorto-aortic shunt, left atriofemoral bypass, and femorofemoral cardiopulmonary bypass.

Left Atriofemoral Bypass

Atriofemoral bypass is achieved through an inflow circuit cannula that is introduced into the left atrial appendage or inferior pulmonary vein. The left atrial inflow blood is powered through a centrifugal or roller pump and returned to a nondiseased region of femoral artery or aorta below the level of the distal clamp. Femoral-to-femoral bypass has mostly replaced this technique at our institution due to the potential for air in the circuit and the absence of a heater–cooler pump mechanism. This technique may, however, be beneficial in patients with poor hemodynamics, impaired cardiac function, or during prolonged cross-clamp times. Improvement in long-term survival with distal aortic perfusion in addition to measures for cerebral protection supports the efficacy of both techniques in the mitigation of ischemic end-organ injury.

The technique for initiation of circuit support involves exposure and taping of the left femoral artery prior to systemic heparinization. The pericardium is opened to expose the left atrial appendage and a purse string is placed. We prefer the use of the inferior pulmonary vein to avoid opening of the pericardium and potential injury to the fragile left atrial appendage. Prior to aortic cross-clamp application, heparin (100 U/kg) is administered to achieve an activated clotting time of 200 seconds. A heparin-bonded circuit provides a measure for the reduction of systemic heparin. The pump inflow cannula is placed in the left atrial appendage or inferior pulmonary vein and the pump outflow cannula is inserted in the left femoral artery to close the circuit. The pump is activated prior to clamping to achieve a high flow rate for reduction of preload prior to the significant increase in afterload imposed by clamp application. Pump flows are maintained and adjusted to achieve target pressure above and below the aortic cross-clamp of 80 to 100 mmHg. Following reconstruction, pump flow can be terminated to increase preload prior to clamp release and the associated afterload reduction.

Femorofemoral Cardiopulmonary Bypass

Femorofemoral bypass is our preferred technique for distal aortic perfusion and is especially beneficial in the setting of a heavily calcified proximal aorta. The principal benefit to this technique is the maintenance of warm, oxygenated blood perfusion to the distal aorta and branch vessels with precise control of perfusion pressure and temperature. The inclusion of cooling capabilities within the femorofemoral bypass circuit allows the achievement of circulatory arrest when necessary. Additionally, pump sucker return to the pump reservoir minimizes transfusion requirement.

The technique for circuit achievement involves femoral vein and artery exposure preferentially on the left side for cannulation following systemic heparinization. Longer activated clotting times are required secondary to the circuit components. An extended 21-F multistage femoral vein cannula is advanced from the left femoral vein toward right atrium to achieve adequate venous drainage of 1.5 to 3 L. Arterial access is achieved at the left femoral artery, left internal iliac artery, or by direct cannulation of the distal aorta with a 15- to 17-F arterial cannula. Cardiopulmonary bypass is initiated immediately prior to aortic cross-clamp application. In patients requiring circulatory arrest, the patient is cooled to 15 to 18 degrees Celsius and circulatory arrest is initiated to allow creation of the proximal anastomosis. The left ventricle should be vented upon fibrillation in the setting of aortic insufficiency. Prior to clamp release, additional volume may be needed in the reservoir to allow rapid transfusion and high pump flows upon clamp release and anticipated hypotension.

Spinal Cord Protection

The inherent threat of unavoidable temporary cord ischemia during repair and the potentially preventable lasting ischemia resulting from inadequate perioperative cord perfusion in patients with descending thoracic and thoracoabdominal aneurysms have supported significant technical advancements in spinal cord protection. Central to methods of cord protection is an understanding of the axial network and the supplying segmental, subclavian, and hypogastric arteries and the protective effect of permissive hypothermia on neural tissue. This collateral network may increase flow through alternative routes when another is reduced; however, this network may also result in steal that will result in decreased nutrient flow to the cord. Steal may occur secondary to the absence of visceral and iliac artery perfusion, during the cross-clamp period, or as a result of pharmacologically induced arteriovenous

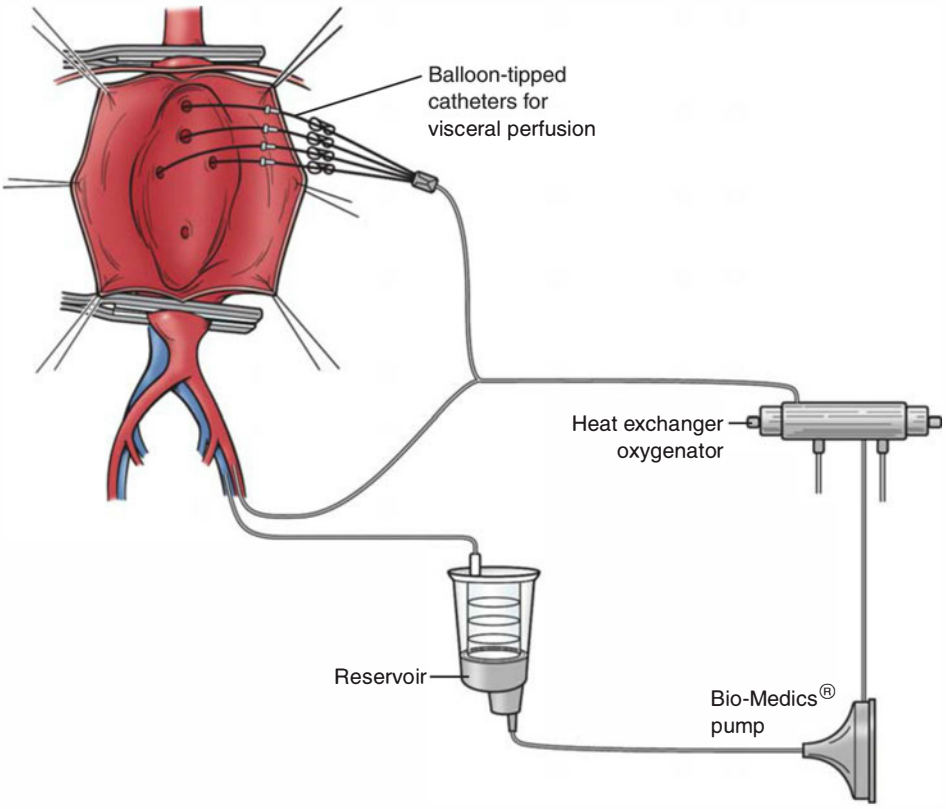


Fig. 59.4. Individual visceral vessel cannulation strategy for cardiopulmonary bypass and maintenance of visceral perfusion during thoracoabdominal aortic aneurysm reconstruction.

shunting when bleeding intercostals into an excluded aortic segment. With these considerations, we have adopted the liberal use of multihead perfusion cannulae and pediatric coronary sinus catheters for the maintenance of perfusion to the visceral and subclavian aortic branches and dominant intercostal arteries, respectively (Fig. 59.4). The detection of potentially reversible delayed paraplegia may be achieved by MEP and SSEP monitoring with immediate assessment of function postoperatively. It is not our routine practice to perform MEP or SSEP monitoring. The adoption of the following provocative techniques for reduction of spinal cord ischemic-induced injury has resulted in a significant decrease in the incidence of paralysis following open repair that is dependent on the extent of repair: 15% for type I, 30% for type II, 7% for type III, and 4% for type IV. Additional predictors of delayed neurologic deficit for thoracoabdominal aneurysms include emergent operative status, prolonged aortic cross-clamp time, level of cross-clamp, hypogastric artery exclusion, aortic rupture, preoperative renal dysfunction, prior abdominal aortic aneurysm repair, acute dissection, and extent II involvement. These techniques and the avoidance of hemodynamic instability and significant blood volume loss have offset the influence of cross-clamp time on induced spinal cord injury.

1. **Distal aortic perfusion.** The presented techniques of atriofemoral and femorofemoral cardiopulmonary bypass have proven efficacious in the reduction of postreconstruction neurologic deficit, particularly in extent II aneurysms. Lower body pressure monitoring is imperative to maintain appropriate cord perfusion which may necessitate flow rates as high as 3 to 3.5 L/min.
2. **Intrathecal vasodilators and topical cooling.** Experimental porcine studies have established the protective effect of intrathecal administration of vasodilators during the aortic cross-clamp period. This technique dilates spinal arteries and prevents spasm and may be combined with local cooling of the cord with 4°C saline to extend the potential aortic cross-clamp time. Topical cooling of the spinal cord is associated with increases in CSF pressure; however, this technique has proven to be a protective adjunct in the reduction of postoperative paralysis and paraplegia. Additionally, prospective clinical study has demonstrated the efficacy of combined CSF drainage and intrathecal papaverine administration in the reduction of spinal cord injury for high-risk thoracoabdominal aortic operations.
3. **Cerebrospinal fluid drainage.** Randomized and nonrandomized clinical trials in addition to cohort studies have demonstrated the efficacy of CSF drainage for the prevention of postoperative paraplegia. In randomized prospective study, the incidence of paraplegia or paraparesis was reduced with CSF drainage from 13.0% to 2.6%, $P = 0.03$. A lumbar catheter is introduced into the subarachnoid space at the L3 or L4 intervertebral space and connected to a pressure transducer and drainage set to achieve CSF pressure monitoring and drainage. CSF pressure is maintained $<$10 mmHg with mean perfusion pressures of 85 to 90 mmHg to enhance spinal cord perfusion. Lumbar drains are generally removed on postoperative day 2 in patients without neurologic deficit. Our preference in high-risk patients is to leave the lumbar drain in-place for 72 hours, while in select low-risk patients we consider removal at 24 hours.
4. **Reattachment of intercostal and lumbar arteries.** Retrospective study has demonstrated acceptable rates of paralysis and paraplegia following sacrifice without reimplantation of as many as 15 intercostal and lumbar arteries during thoracoabdominal aneurysm reconstruction. In addition, porcine studies suggest that all segmental arteries may be ligated with preservation of spinal cord function. Prompt ligation of nonimplanted intercostal arteries is recommended to avoid steal from the cord circulation. While multimodality approaches have implemented complete intercostal reimplantation, the demonstrated efficacy remains undefined and the prolonged ischemia imposed by this technique may increase the risk of postreconstruction paralysis and paraplegia.
5. **Hypothermia.** Moderate (29 to 32°C) to profound ($<$20°C) hypothermia is associated with improved outcomes following thoracoabdominal and descending aortic operations. The proposed mechanism of hypothermia-induced protection involves the reduction of excitatory neurotransmitter release, decreased free oxygen radical production, decreased postischemic edema,

and stabilized central nervous system blood flow. Hypothermia is achieved through passive cooling, fluid resuscitation, and titration of the extracorporeal circuit heat exchanger.

6. **Pharmacologic agents.** Mannitol (0.25 to 1.0 g/kg), high-dose barbiturates, methylprednisolone (30 mg/kg), calcium-channel blockers, adenosine 2A agonist, naloxone, and local anesthetic agents have a demonstrated efficacy in experimental models of spinal cord ischemia and multimodality clinical protocols. Methylprednisolone, naloxone, and mannitol have achieved variable clinical adoption. The proposed mechanism of spinal cord protection involves decreased spinal cord edema and improved free oxygen radical scavenging.
7. **Minimizing aortic cross-clamp times.** Debate persists regarding the effect of aortic cross-clamp time on rates of paralysis and paraplegia following thoracic and thoracoabdominal aortic aneurysm repair after the near-universal adoption of distal aortic perfusion. Despite these conflicting reports, the clamp-and-sew technique remains a foundational technical principle in the reconstruction of thoracoabdominal and descending thoracic aortic aneurysms.

Our institutional approach to spinal cord protection incorporates minimal clamp times, femorofemoral bypass, and CSF drainage for all extent I and II aneurysms and select extent III aneurysms or complex reconstructions. Special care is imperative in clamp application to avoid placement in segments with significant atheroma or mural thrombus to minimize the risk of embolization to the spinal cord and viscera. Distal aortic perfusion is maintained by femorofemoral bypass. Beveling of the anastomosis or revascularization with a branched aortic graft is performed at the T11 to L1 level when feasible and without a significant extension of total clamp time.

In patients with proximal extension or aortic calcification limiting proximal clamp application, femorofemoral bypass and hypothermic circulatory arrest provide support measures to allow completion of the proximal anastomosis. The graft is then cannulated to initiate warming to the great vessels, lower extremities, and viscera while the distal anastomosis is completed. Alternative cannulation strategies with the axillary artery may be utilized to minimize embolic stroke potential in patients with extensive atheroma.

AORTIC GRAFT SELECTION

Direct proximal and distal measurements or a sizing device should guide graft size selection. Zero-porosity polyethylene terephthalate (Dacron) grafts have eliminated the need for preclotting or bleeding through the graft. Sidearm grafts may also be selected to achieve complex reconstruction or revascularization of visceral and intercostal vessels. Suture material should be selected and loaded prior to cross-clamp application. Our preferred suture is a long, double-armed 3-0 or 4-0 polypropylene suture for each aortic anastomosis with felt reinforcement of the aortic suture line.

TECHNIQUE FOR AORTIC ANASTOMOSES

Following application of the proximal and distal cross-clamps, the aneurysm is incised longitudinally with a large blade close to the site of the anastomosis. The aortotomy is extended proximally and distally with scissors to expose the lumen of the aneurysm and to allow evacuation of laminated thrombus. In extended aneurysms, the clamps should be placed in close proximity to minimize the potential for exclusion of dominant segmental arteries to the spinal cord. The distal clamp is transferred to the site of the distal anastomosis at the completion of the proximal anastomosis. Avoidance of distal clamp application with cell-saving suction may be utilized with the clamp-and-sew technique.

TUBE GRAFT PLACEMENT

The proximal aortotomy is fashioned to accommodate a transverse anastomosis. In chronic dissection, complete transection is recommended to identify each aortic layer for subsequent incorporation into the anastomosis. Complete transection may also be of benefit when feasible in the setting of chronic atherosclerotic aneurysm to facilitate the anastomosis.

Side towels cover retractors and clamps and the graft is placed onto the field. The proximal anastomosis is initiated on the graft and the aorta is picked-up for the first four sutures as a parachute stitch. The graft is approximated to the aorta and the anastomosis is completed expediently with large aortic bites to achieve hemostasis (Fig. 59.5). The suture line is completed sewing toward oneself beginning with the back wall until reaching the anterior suture line. The other arm is then brought from below. The aorta may be taken outside-in or inside-out as long as bites are both rapid and sufficient.

Following completion of the proximal anastomosis, the graft is clamped at 4 to 5 cm below the suture line and the proximal clamp is released to allow inspection and immediate repair of any sites of inadequate hemostasis (Fig. 59.5B). Repair is achieved with a standard-length 3-0 polypropylene suture as a mattress stitch, pledgeted with polytetrafluoroethylene. Branched visceral vessel and intercostal reconstructions are then performed prior to the distal aortic anastomosis.

Attention is directed to the distal suture line and retraction may be moved from the upper to lower incision when necessary. The aortotomy is extended down the aorta to the distal aneurysm neck and squared horizontally at the site of the proposed anastomosis. Complete transection of the aorta at the distal margin is favored only in the setting of a chronic dissection flap (Fig. 59.5C). In subacute dissection, a long tongue is fashioned away from the flap to allow a graft anastomosis to the outer aortic layer to maintain perfusion to both the true and false lumen distally. The anastomosis is then created in a similar manner. Notification to the anesthesia team upon nearing completion will allow preparation for cross-clamp release. As the distal clamp is removed, bypass flows are titrated and weaned to maintain optimal perfusion pressures. With this technique, minimal hemodynamic compromise should occur.

REIMPLANTATION OF INTERCOSTAL ARTERIES

Inspection for dominant, large intercostals is performed during the initial dissection, marking each for potential revascularization. Our preferred approach is to incorporate a 6-mm sidearm graft with 4-0 or 5-0 polypropylene suture to critical intercostal arteries rather than the island patch technique to minimize the potential for aneurysmal degeneration (Fig. 59.5C).

In the setting of extensive thoracoabdominal disease with visceral artery involvement, our preferred technique involves mobilization of the celiac axis, superior mesenteric artery (SMA), and right renal artery for reconstruction with a branched aortic graft (Fig. 59.6). During such complex reconstructions, distal aortic perfusion and progressive clamp advancement are performed to minimize visceral ischemic time. The proximal anastomosis is completed first and the graft clamped just below the anastomosis. The proximal

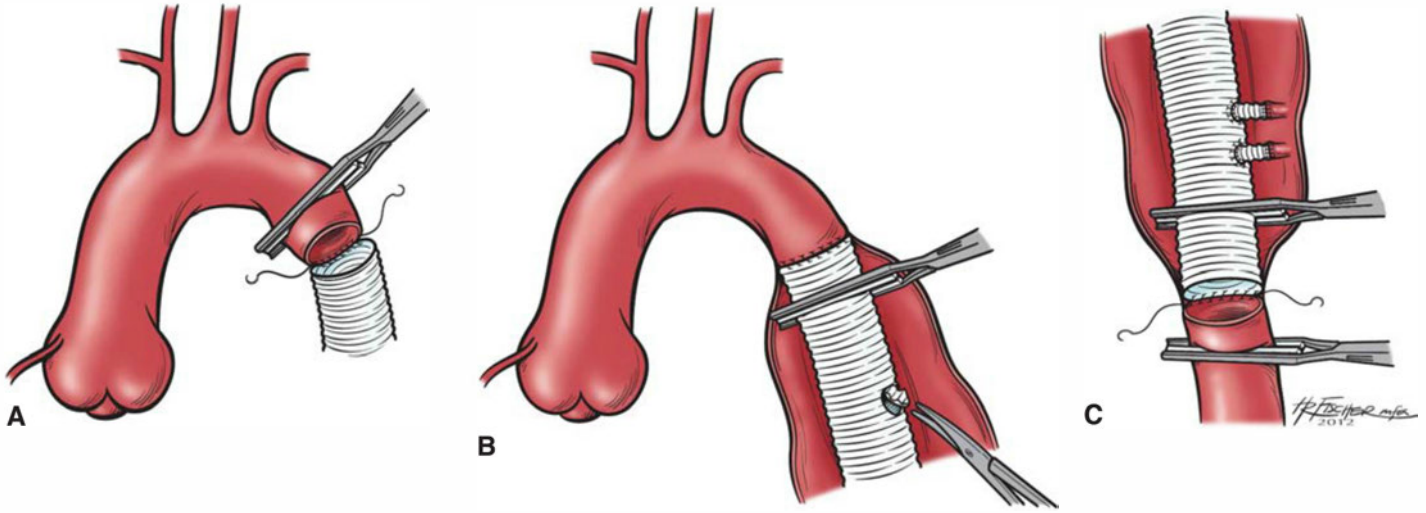


Fig. 59.5. Transection of the aorta in cases of aortic dissection to ensure that all aortic layers are incorporated into the anastomosis. (**A**) Completion of the proximal aortic anastomosis. (**B**) Sequential clamping strategy with removal of an ellipse from the graft for reimplantation of dominant intercostal arteries. (**C**) Intercostal artery revascularization is completed prior to the distal anastomosis with 6 mm sidearm graft. The distal is then performed with complete transection of the aorta in the setting of concomitant dissection.

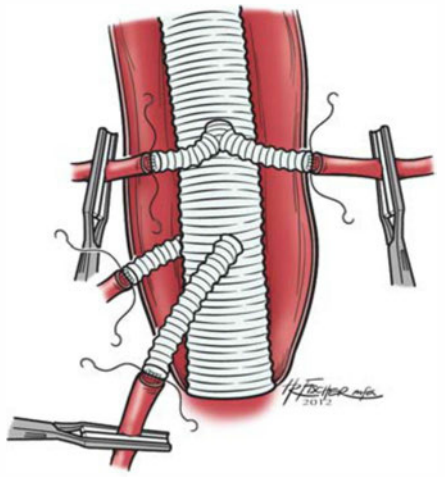


Fig. 59.6. Branched graft reconstruction of the celiac, superior mesenteric, and right renal arteries during the period of distal aortic perfusion for the restoration of visceral inflow. Note: left renal artery not depicted within aneurysmal segment.

clamp is removed and the distal clamp moved below the site of the arteries to be reconstructed. Anastomoses are performed with a 3-0 or 4-0 polypropylene suture. Following proximal aortic anastomosis completion, branched vessels are sequentially clamped and branched graft anastomoses are performed in the following order: right renal artery, celiac artery, SMA, and left renal artery. The clamp is then moved to below the reimplanted intercostal and visceral vessels and the distal anastomosis is completed.

In many individuals, visceral vessels may neighbor the aneurysmal segment, yet their origins remain uninvolved. For these select patients, we favor an oblique distal anastomosis that preserves an anterior tongue of aorta from which the visceral vessels arise. This aortic tongue is subsequently incorporated in the aortic anastomosis, effectively maintaining visceral perfusion without the requirement for additional anastomoses.

Multiside-branched grafts have emerged as our preferred technique for branch vessel revascularization, with particular benefit for patients in which the origin of visceral vessels is splayed, occlusive, or aneurysmal. Multihead perfusion catheters from the bypass arterial line are recommended with this technique to individually perfuse visceral arteries through coronary sinus balloon catheters during sequential revascularization as described.

COMPLETION OF THE OPERATION

Hemostasis at all suture lines and adequate distal perfusion are confirmed following the achievement of all anastomoses. Extracorporeal support is weaned and the patient is adequately warmed. Adequate blood pressure and a satisfactory rhythm are confirmed. Protamine sulfate may be administered and cannulation sites are repaired. The graft is covered with the remnant aneurysm sac to prevent later fistula formation to the lung or intestine. The retroperitoneal tissue is approximated over the abdominal portion of the graft. If deficient, greater omentum may be used. 19- to 24-F small-diameter channel drains are placed into the pleural cavity at the apex and base. In addition, we routinely place a 19- to 24-F drain into the retroperitoneum. Topical hemostatic agents and anti-fibrinolytics may be used in the setting of persistent and generalized lack of adequate hemostasis, although the efficacy of antifibrinolytic therapy has not been demonstrated in retrospective analysis. The collapsed lung is reinflated and the operative field is irrigated. The ribs are approximated with multiple interrupted, heavy, absorbable sutures, taking care to avoid the neurovascular bundles. Intracostal sutures placed through holes that are drilled in the lower rib have been demonstrated to decrease postoperative neuralgia and are recommended. The soft tissues are approximated in layers. Postoperative mechanical ventilation is necessitated in all patients until acidosis is reversed and hemodynamic stability at normothermia is achieved.

ENDOVASCULAR REPAIR TECHNIQUE

Advancements in thoracic endovascular aneurysm repair (TEVAR) have inspired the clinical adoption of this treatment for many with distinct aortic pathologies or limited candidacy for open surgical

reconstruction. The potential avoidance of thoracotomy, extracorporeal circulatory support, aortic cross-clamp time, and prolonged hospital length of stay support consideration of endovascular descending thoracic and thoracoabdominal aneurysm repair in patients with significant comorbid conditions (advanced age, cardiac, pulmonary, and renal dysfunction). Importantly, patients who are not candidates for open surgical repair have significantly poorer outcomes following TEVAR than TEVAR-treated patients that would have been open repair candidates. These data support further research advancement to define the potential expansion of treatment criteria for patients with asymptomatic descending and thoracoabdominal aneurysms.

Absolute contraindications to TEVAR include insufficient proximal and distal landing zones or an aortic diameter <22 mm. In the clinical translation of this guideline, off-labeled uses of non-FDA approved devices have been described. Extreme tortuosity of the thoracic aorta should be considered in operative planning as this may impair the ability to safely advance guide wires into the thoracic aorta. In the presence of laminated thrombus or significant atheromatous plaque, we carefully consider the application of endovascular therapy secondary to the significant risk of distal embolization. Relative and debated contraindications include a young age as long-term outcomes remain undefined, unstable patients requiring preoperative imaging, and patients with chronic renal insufficiency, although intravascular ultrasound (IVUS) may be performed with minimal contrast material.

At this time, three thoracic aortic stent platforms are approved by the U.S. Food and Drug Administration for the treatment of thoracic aortic aneurysms: the W.L. Gore TAG endograft platform (Flagstaff, AZ), the Cook Zenith TX2 (Bloomington, IN), and the Medtronic Talent (Santa Rosa, CA) thoracic endograft systems. Common design features of thoracic endograft systems include an elongated shaft for delivery to the proximal aorta from the groin, larger diameters and lengths, increased flexibility, and a tubular, nonbifurcated design. The selection of the appropriate endograft is determined by the individual patient anatomy. Endograft devices and approaches tailored to distinct pathologies are under current clinical investigation and development. The Talent (Medtronic) is available in larger diameters to 42 mm and incorporates a proximal bare metal stent for proximal fixation. The Zenith (Cook) device provides tapered graft options and an exposed distal stent for improved fixation. In addition to endograft system components, a stiff wire such as the Lunderquist (Cook) wire for tracking, a marker pigtail for length projection, and balloons for graft effacement provide intraprocedural adaptations to achieve satisfactory graft positioning within the thoracic aorta. All wires are to be at least 260 cm for access from the groin.

Thoracic endografting should be performed in an appropriately equipped specialized operating room with intraprocedural angiography capability. The patient is positioned supine on the operative table with exposure of the predetermined access vessels (femoral, iliac, terminal aorta) and upper extremity vascular access sites as necessary. General anesthesia is achieved with a single-lumen endotracheal tube; however, regional anesthesia may be utilized in select patients. Arterial pressure monitoring and pulse oximetry are established on the upper extremity. Large-bore peripheral intravenous access, Foley catheter placement, and temperature monitoring are achieved. CSF pressure monitoring and drainage and SSEP assessment are techniques to minimize the risk of paraplegia in patients requiring extensive coverage of the descending aorta or in patients with prior history of abdominal aortic aneurysm repair. Our general practice is to always place a lumbar drain in patients undergoing thoracoabdominal aneurysm repair. While recognizing the potential benefit of SSEP monitoring, we have not found that SSEP monitoring significantly alters intraoperative treatment. Thoracoabdominal aortic aneurysms may involve the ostia of the celiac axis or the SMA and inferior mesenteric artery (IMA), necessitating coverage upon endovascular repair. In these select patients, a first-stage debranching operation to achieve SMA-to-celiac artery, aorta-to-SMA, or aorta-to-celiac artery bypass grafting is performed. Branched, fenestrated endografts provide a promising innovation for future study and development. Additionally, snorkel or stove-pipe techniques are continuing to evolve and we have utilized each with some success in extent III aneurysms. When selecting an approach to bypass and reconstruction, it is important to consider that the ostia of the celiac artery axis may be covered without sequelae in the setting of documented and appropriate collateral flow through the SMA.

The common femoral or common iliac arteries are the principal sites for device access. Angiographic access may be obtained through the contralateral groin or the left brachial artery. Access through the upper extremity provides localization of the left subclavian artery and may facilitate true lumen cannulation in the setting of aortic dissection. In patients with insufficient femoral arteries, a retroperitoneal approach may be necessitated for infra-aortic or iliac artery access. The device is introduced and positioned prior to angiographic imaging at an oblique angle to splay the thoracic aorta and identify visceral vessels. Hemodynamic monitoring and control are imperative during device deployment, as balloon molding and pulse pressure may shift the graft. Overlap of at least 5 cm is recommended to avoid a junctional endoleak. Intussusception of the graft components may be necessary in the setting of aortic tapering. Compliance with device-specific guidelines is recommended for access diameter and deployment. Following deployment of the endograft, postreconstruction angiography is performed to confirm satisfactory localization and exclusion of the aneurysmal segment. Sheaths are removed and hemostasis is confirmed following either percutaneous device closure or direct surgical repair of access vessels. Immediate postoperative care and resuscitation parameters follow the presented guidelines for open repair.

Common iliac artery conduits or direct common iliac artery puncture provide alternative techniques for access in patients with size-limiting femoral arteries. While the most widely accepted approach to iliac artery access is to sew an 8 to 10 mm conduit onto the artery, direct access and primary closure are often sufficient. If the proximal landing zone is insufficient, extra-anatomic bypass may be performed to maintain perfusion to branch vessels. The most common indication for extra-anatomic bypass is the need to cover the ostia of the left common carotid or innominate arteries. The left subclavian artery is intentionally covered in approximately 50% of cases. In our experience, TEVAR exclusion of the origin of the left subclavian artery necessitated pre- or postoperative revascularization in 40.7% of patients. Exclusion of the left subclavian artery without revascularization may also increase the risk for perioperative stroke or spinal cord ischemia. In addition to selective revascularization, subclavian to carotid artery transposition is recommended when the aortic lesion is within 15 mm of the subclavian orifice. This technique is aimed at preventing type II endoleaks and perfusion of a dissected false lumen and is also recommended when the ipsilateral vertebral

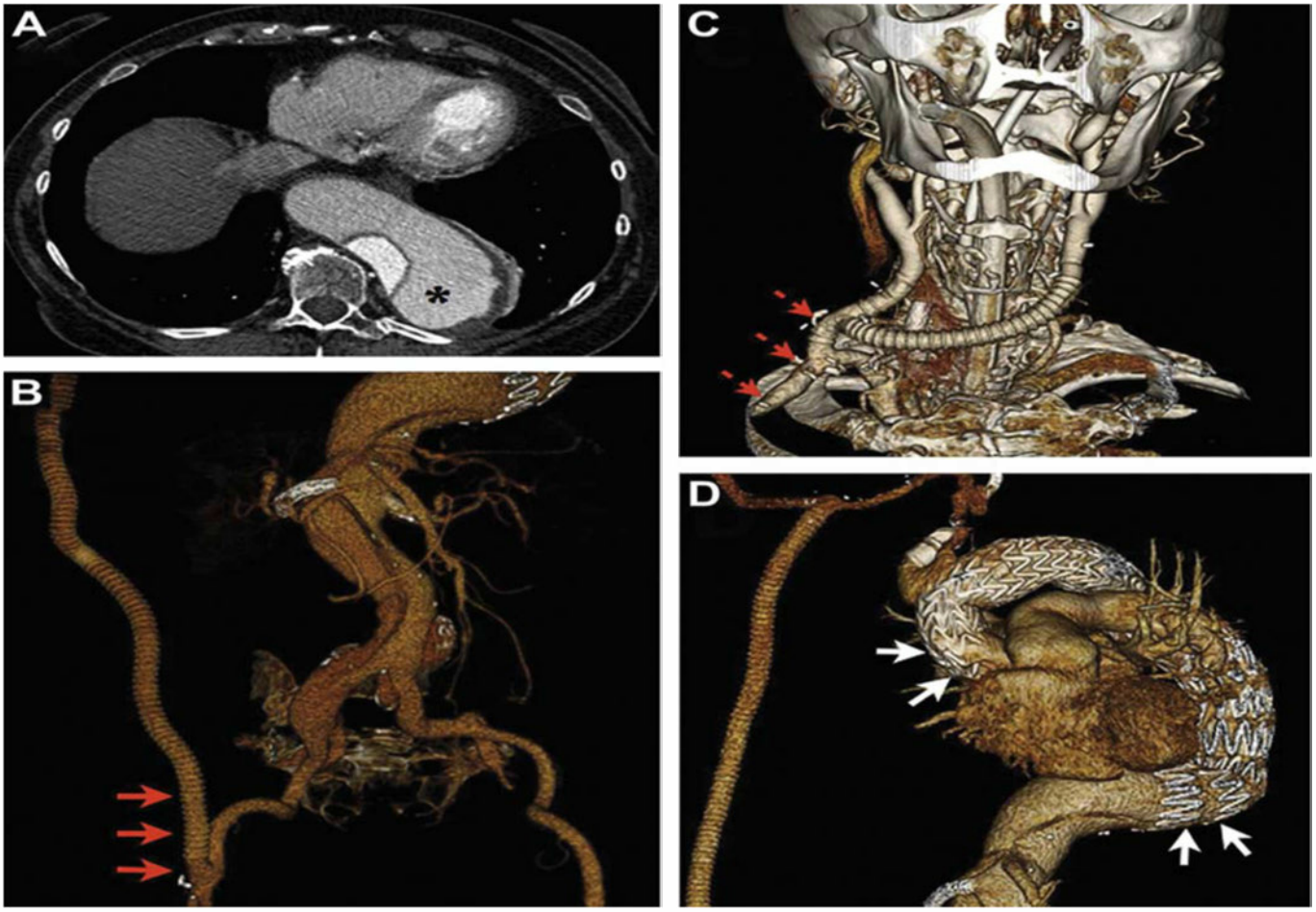


Fig. 59.7. (**A**) Axial image demonstrating the true lumen of the mid-distal aortic arch dissection (*asterisks*). (**B**) Extra-anatomic right common femoral to axillary artery bypass (*red solid arrows*) to establish in-flow. (**C**) Right subclavian–carotid–carotid bypass with cerebral flow conduits (*dashed red arrows*) and aortic arch debranching. (**D**) Thoracic endografting from sinotubular junction to the diaphragm with expanded proximal and distal landing zones. (Images used with permission, Annals of Vascular Surgery 2012;26(3):420.)

artery is patent and dominant, when the ipsilateral internal mammary artery may be needed for future coronary revascularization, and when extensive coverage of intercostals contributing to spinal cord perfusion is anticipated. In patients who have absolute contraindications limiting their candidacy for open repair, extreme measures are possible to enable endovascular repair (Fig. 59.7). In each patient with the potential need for revascularization, transcranial Doppler angiography or CTA is recommended to confirm communication of the vertebral arteries at the basilar artery. Our preferred approach is to obtain a preoperative CTA of the head and neck with the performance of bypass in the setting of left vertebral artery dependence and anticipated stent graft coverage of the origin vessel from the aorta. Carotid to subclavian artery bypass is implemented in patients with a patent internal mammary artery conduit that is perfusing a coronary vessel and is combined with proximal subclavian artery ligation. With these considerations, endovascular therapy is emerging as an increasingly viable option for repair. However, further study and device design advancements are needed to define and address acceptable aortic diameters, landing zones, and treatment advantages for not only aneurysmal disease but also aortic dissection and transection.

THE HYBRID APPROACH

Hybrid techniques that incorporate open and endovascular repair techniques provide a promising approach to treatment in patients with complex thoracic aortic aneurysms with or without concomitant dissection. In patients with descending thoracic aortic aneurysms with proximal extension into the aortic arch, we have successfully performed open arch debranching and revascularization of branch vessels followed by TEVAR of the arch and descending thoracic aorta (Fig. 59.8). Such a technique has been demonstrated through a right anterior minithoracotomy at the third to fourth intercostal space, supporting further clinical advancement to provide patients with complex aortic pathologies the potential benefit of a minimally invasive approach to reconstruction. Additionally, our emerging treatment strategy for extent I and II aneurysms is to perform endovascular treatment of the proximal aneurysm extension, converting extent I or II disease into an extent III or IV aneurysm that is amenable to open surgical repair (Fig. 59.9). This approach provides the patient with a lower predicted mortality and paraplegia risk than that of an open extent I or II aneurysm repair. In patients with combined aortic arch and descending aorta involvement, endovascular treatment of the descending segment up to the subclavian artery enables open arch reconstruction through a median sternotomy, and reconstruction completion upon sewing the proximal graft to the descending

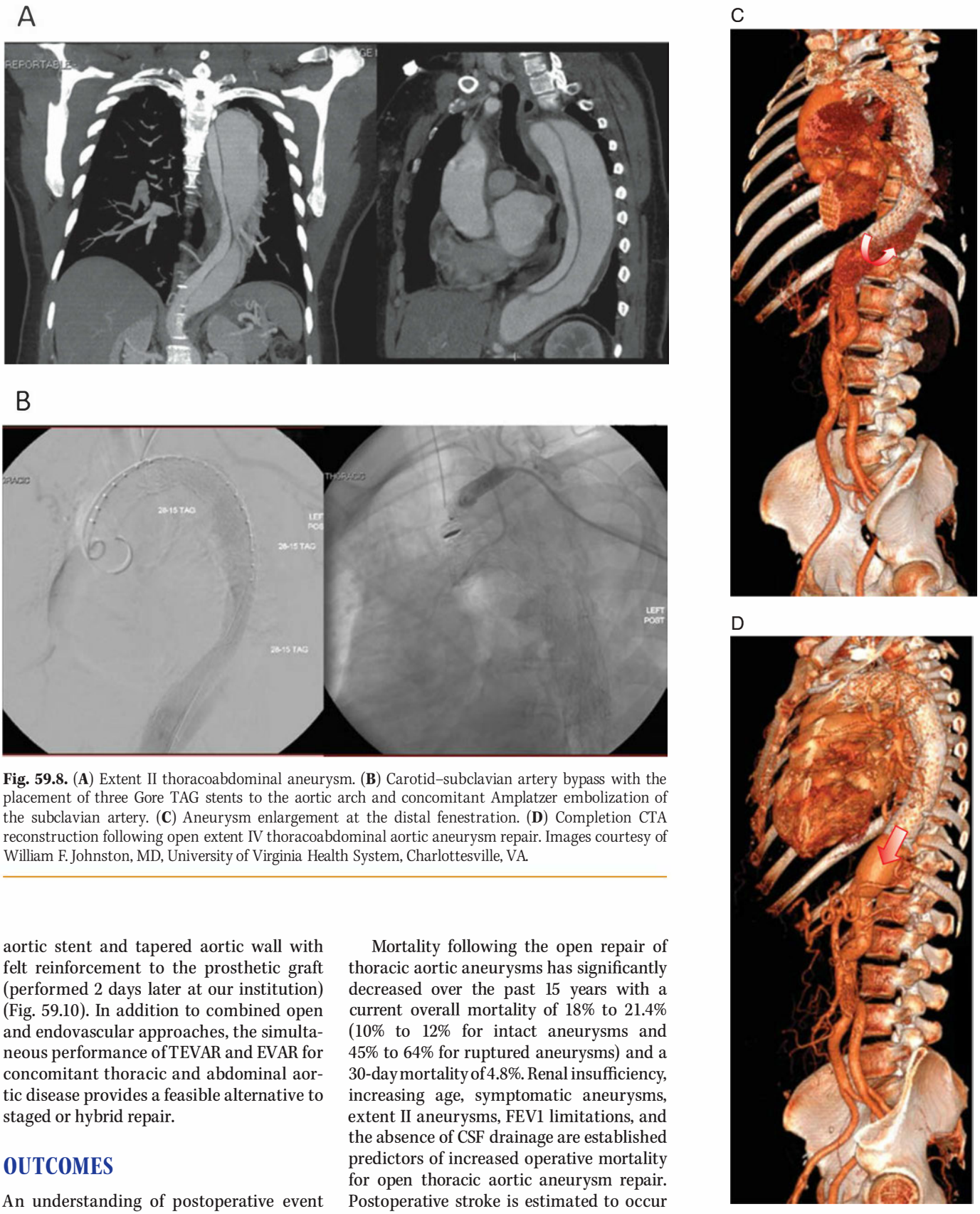


Fig. 59.8. (**A**) Extent II thoracoabdominal aneurysm. (**B**) Carotid–subclavian artery bypass with the placement of three Gore TAG stents to the aortic arch and concomitant Amplatzer embolization of the subclavian artery. (**C**) Aneurysm enlargement at the distal fenestration. (**D**) Completion CTA reconstruction following open extent IV thoracoabdominal aortic aneurysm repair. Images courtesy of William F. Johnston, MD, University of Virginia Health System, Charlottesville, VA.

aortic stent and tapered aortic wall with felt reinforcement to the prosthetic graft (performed 2 days later at our institution) (Fig. 59.10). In addition to combined open and endovascular approaches, the simultaneous performance of TEVAR and EVAR for concomitant thoracic and abdominal aortic disease provides a feasible alternative to staged or hybrid repair.

OUTCOMES

An understanding of postoperative event chronology and preoperative risk stratification guides the selection of medical, open, or endovascular therapies for the treatment of descending thoracic and thoracoabdominal aortic aneurysms for patients across all risk stratifications.

Mortality following the open repair of thoracic aortic aneurysms has significantly decreased over the past 15 years with a current overall mortality of 18% to 21.4% (10% to 12% for intact aneurysms and 45% to 64% for ruptured aneurysms) and a 30-day mortality of 4.8%. Renal insufficiency, increasing age, symptomatic aneurysms, extent II aneurysms, FEV1 limitations, and the absence of CSF drainage are established predictors of increased operative mortality for open thoracic aortic aneurysm repair. Postoperative stroke is estimated to occur in 2% to 21% of patients following open repair and is dependent on the extent of the descending thoracic aorta replacement. In addition to stroke, paralysis and paraplegia are dependent on the extent of repair and are estimated to occur in 3% to 5% of patients following open repair. Additional predictors of paraplegia or paresis include total aortic clamp time, rupture, advanced age, proximal aortic aneurysm, renal dysfunction, and the application of distal

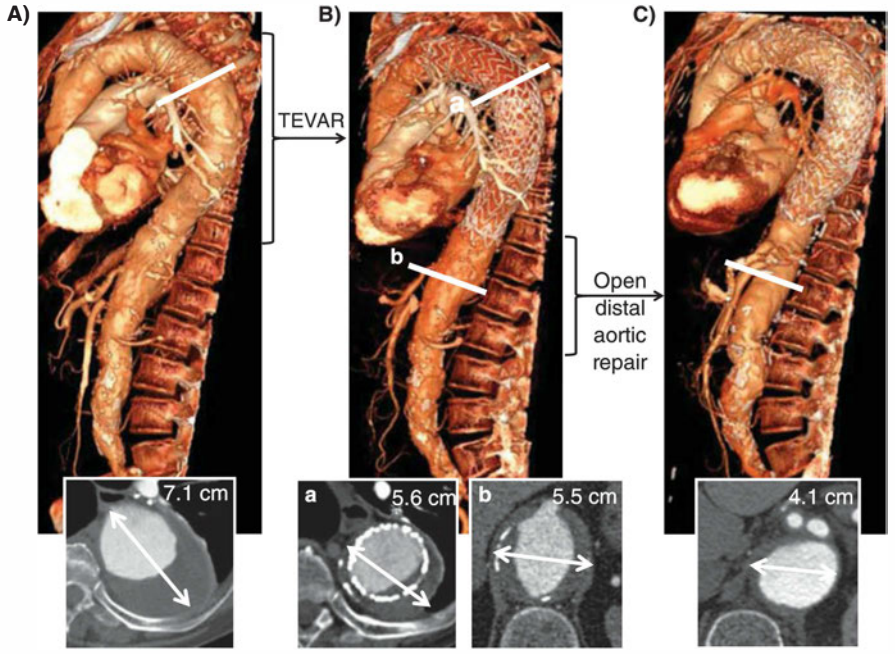


Fig. 59.9. Hybrid repair of an extent II thoracoabdominal aortic aneurysm. (**A**) Initial presentation of a three-dimensional computed tomography reconstruction. (**B**) Proximal aortic aneurysm extension treatment with thoracic endovascular aneurysm repair. (**C**) Open distal aortic aneurysm repair at site of distal fenestration and aneurysmal enlargement. (Images used with permission. Journal of Vascular Surgery 2012;56(6):1495-1502.)

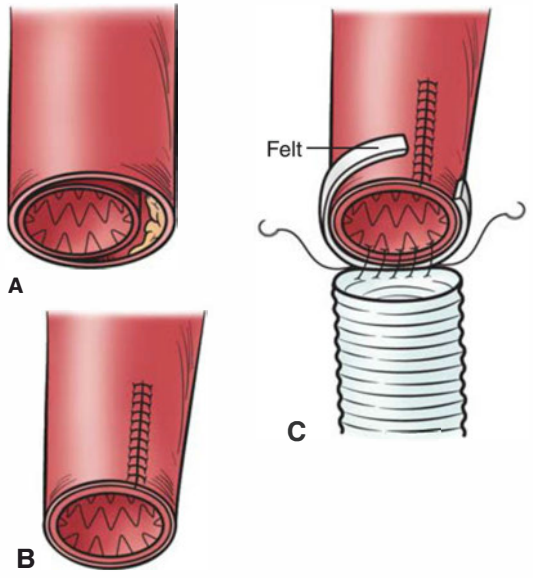


Fig. 59.10. Staged hybrid approach. (**A**) Distal thoracic aorta with endograft in the true lumen and thrombus in the false lumen. (**B**) Thrombus is removed, the dissection septum is divided, and the aortic wall is cut along the *dashed line* to taper the aortic wall. (**C**) The anastomosis is crafted with an end-to-end attachment of the endograft and tapered aortic wall with felt reinforcement of the prosthetic graft. (Images adapted from Journal of Vascular Surgery 2012;56(6):1495-1502.)

aortic perfusion and CSF drainage. Distal aortic perfusion and advanced techniques for revascularization target the prevention of postoperative renal dysfunction, with 13% of patients requiring temporary postoperative dialysis and 2% necessitating chronic renal replacement therapy. These primary postoperative outcomes remain the principal focus of surgical technique advancement and preoperative risk stratification for the open repair of descending thoracic and thoracoabdominal aortic aneurysms.

Adverse device-related postoperative events following TEVAR occur in 10% to 12% of patients in the initial 30-day postoperative period. The most common immediate intraoperative injury involves avulsion or tear of the external iliac and common femoral arteries upon sheath removal. Immediate arterial exposure and repair are performed with bypass from the common iliac to common femoral artery. Postoperative stroke is estimated to occur 2.5% to 5.8% of patients following TEVAR. Atheroemboli from the transverse aortic arch that are dislodged upon guide wire and device manipulation represent the principal etiology for stroke following TEVAR. Higher stroke rates are demonstrated in patients with a preoperative history of stroke undergoing proximal descending thoracic or complete descending thoracic aortic replacement. In addition to cerebral embolization, distal embolization following TEVAR results in acute mesenteric ischemia in 3% of patients.

Long-term complications include graft endoleak, continued aneurysm growth, metal fatigue, stent fracture and kinking, detachment, migration, perforation, and infection of the implanted device. Endoleaks are estimated to occur in 10% to 20% of patients and most commonly result from proximal attachment site failure, type IA. Endoleaks often warrant reintervention to prevent aneurysm enlargement and the associated increased risk of aortic rupture. For patients presenting with rupture post-TEVAR, complete endovascular relining is performed to provide proximal and distal landing zone extension.

Nationwide inpatient data from the United States demonstrate equivalent mortality, complication rates, and failure to rescue for TEVAR and open aortic repair with more frequent routine discharge for TEVAR. These data also suggested that TEVAR is an ideal alternative to open repair for ruptured thoracic aortic aneurysm in smaller hospitals where open expertise may be lacking. While clinical trials have suggested a reduction in 30-day cardiovascular morbidity and aneurysm-related mortality, superiority in neurologic and long-term outcome remains undefined. As our understanding of endovascular technology progresses, multidisciplinary collaborative decision-making is imperative to select the appropriate therapy for an individual patient based on underlying pathology, concomitant risk factors, and projected natural history of aneurysmal disease if medically treated.

FOLLOW-UP RECOMMENDATIONS

The potential for mid-term and long-term complication following descending thoracic and thoracoabdominal aneurysm

repair necessitates a commitment to dedicated surgical follow-up. The current standard for postoperative endovascular and open descending thoracic and thoracoabdominal aortic aneurysm repair involves physical examination and repeat imaging with CTA chest and chest radiography at 1, 6, and 12 months. The initial interval to postoperative evaluation may be modified as determined by the immediate in-hospital postoperative course.

CONCLUSIONS

The past two decades of open surgical technique refinement and the advent of endovascular technology have significantly advanced our capability to treat complex descending thoracic and thoracoabdominal aortic aneurysms in high-risk patients. We consider the factors most important in the determination of a safe operation to be careful assessment of the patient's overall condition, especially noting chronic lung disease or coronary artery disease; planning of an operation that replaces in one setting the shortest possible segment of aorta; assessment before aortic cross-clamping of the likely complexity of the repair and planning for distal aortic perfusion if appropriate as well as CSF drainage; and rapid and precise anastomotic techniques that minimize cross-clamp times and allow precise management of hemodynamic stability.

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EDITOR'S COMMENTS

This chapter on descending thoracic and thoracoabdominal aortic aneurysms by Drs. Stone and Kern has provided the comprehensive approach to descending thoracic aortic surgery at The University of Virginia. I would like to highlight a couple of comments. There is not a specialty that has changed as rapidly as this one has. The use of TEVAR for the majority of purely descending aortic aneurysms has revolutionized the field. One could argue about the issues with TEVAR but avoiding major thoracotomies certainly is worthwhile in the majority of patients. I think Dr. Kern has highlighted that the combination of TEVAR in addition to more limited repairs of the visceral vessels is an interesting one and probably will become a standard of practice. I also totally agree with the use of branch grafts for type 3 and 4 thoracoabdominal aneurysms where the visceral vessels are splayed to prevent recurrent aneurysms particularly in patients with Marfans Syndrome. I think that this field will continue to evolve. More improved branch grafts will allow percutaneous approaches to perhaps replace open surgery for even more complex situations.

ILK

60 Ascending Aortic Aneurysms

Matthew L. Stone and Irving L. Kron

There is no disease more conducive to clinical humility than aneurysm of the aorta.
–Sir William Osler, c 1900

INTRODUCTION

Since the first reported ascending aortic replacement by Cooley and DeBakey in 1956, operative technique refinement and critical care advancement have led to a significant improvement in survival for patients with ascending aortic aneurysmal disease. An improved understanding of distinct aortic pathologies and predisposing factors for aortic rupture and dissection has inspired a commitment to risk stratification and guidelines for elective surgical intervention. Improvements in diagnostic imaging and an increase in the incidental identification of aortic pathology on unrelated imaging have supported a significant increase in the incidence of thoracic aortic aneurysms to a projected rate of 10.4 cases per 100,000 person-years.

Isolated aneurysms of the ascending tubular aorta that taper to a normal diameter in the distal ascending aorta with a normal diameter at the sinuses of Valsalva are relatively uncommon and are estimated to occur in 13.5% of cases, with the majority of cases involving either the aortic annulus and the sinotubular junction or the aortic arch and branch vessels. Therefore, techniques for aortic valve conservation or replacement and branch vessel reconstruction are often needed in the operative reconstruction of the ascending aorta. The two surgical approaches to ascending aortic aneurysms involving the aortic root include aortic valve-sparing operations and composite replacement of the aortic valve and ascending aorta with a heart valve conduit. Aneurysms isolated to the ascending aorta without aortic root involvement are selectively approached by replacement of the aneurysmal segment with a composite ascending aortic graft.

In addition to anatomic principles for reconstruction, an understanding of distinct aortic pathologies and concomitant aortic valve disease is needed in the operative repair of ascending aortic aneurysms. Annuloaortic ectasia comprises a spectrum of aortic pathologies that involve dilation of the aortic annulus. This condition is predominately associated with an aneurysm of the aortic root; however, may occur independent of aneurysm formation and manifest as bicuspid or tricuspid aortic valve insufficiency or occur in patients with a subaortic ventricular septal defect. Importantly, ascending aortic aneurysms may also occur in the absence of annuloaortic ectasia. Dilation of the aortic sinuses results in aortic insufficiency when the aortic annulus and sinotubular junction dilate, resulting in poor leaflet apposition and central insufficiency. As the aortic root dilates, the aortic cusps become thinner, overstretched, and may develop stress fenestration in the commissural areas. This is an important consideration for the aortic valve repair at the time of ascending aortic aneurysm reconstruction.

The focus of this chapter is to review diagnostic standards and surgical principles for reconstruction in the treatment of aortic root and ascending aortic aneurysm with or without annuloaortic ectasia.

IMAGING

Aortic dimensions remain a principle consideration for intervention and reconstruction of the aortic valve, root, and proximal ascending aorta. Chest radiography may impart the initial diagnosis of ascending aortic aneurysm, with the findings of a convex contour to the right superior mediastinum or a loss of the retrosternal air space on lateral examination. Computed tomography (CT) and magnetic resonance imaging (MRI) provide detailed aortic wall assessment in addition to the capability for three-dimensional reconstruction, branch vessel evaluation, and aortic valve characterization. The reliability and reproducibility provided by these novel imaging techniques in coordination with transesophageal echocardiography (TEE) have largely replaced traditional aortography for the evaluation of the proximal ascending aorta, aortic valve, and aortic branch vessels, as aortography is limited in the determination of the presence of intraluminal hematoma. Aortography may be used selectively to localize the coronary ostia and to detect concomitant aortic regurgitation or coronary atherosclerotic disease. Measurements obtained with CT or MRI should be obtained perpendicular to the axis of blood flow and represent the external diameter of the aorta. Echocardiographic measurements are obtained in a similar plane; however, should be calculated for the internal aortic diameter. The dimension of the aortic root should be obtained at the segment of widest diameter, typically the mid-sinus level. In addition to dedicated aortic imaging, duplex carotid ultrasonography is indicated in patients older than 65 years of age or in younger patients with symptomatic presentation, physical exam findings, or additional atherosclerotic vascular disease consistent with an increased risk for cerebrovascular disease, as undetected carotid disease is a significant risk factor for stroke following ascending aortic replacement.

CT provides an estimated diagnostic accuracy of 92% for abnormalities of the thoracic aorta, with correct prediction of a requirement for circulatory arrest in 94% of patients. Electrocardiogram gating is required for ascending aortic disease to eliminate motion artifact that can resemble aortic dissection. In addition, CT provides a reliable method for the localization of calcification and evaluation of aortic valve morphology, function, and coronary artery anatomy. Axial CT introduces the potential for misinterpretation of the proximal aortic diameter, as ascending aortic elongation results in a C-shape of the aorta and resultant vertical plane orientation of the aortic valve.

MRI provides a comparable sensitivity and specificity for thoracic aortic pathology

that may exceed that afforded by CT. The principle advantages to MRI include the identification of anatomic dissection variants and the enhanced diagnosis of branch vessel disease, aortic valve pathology, and left ventricular dysfunction in the absence of radiation or iodinated contrast. Clinical adoption has been limited by the time required for image acquisition, an inability to utilize gadolinium in patients with renal insufficiency, and a contraindication for use in patients with metallic implants including pacemakers.

Transthoracic echocardiography (TTE) provides an understanding of biventricular function while enabling visualization of the aortic valve and the proximal several centimeters of the ascending aorta to above the sinotubular junction. This capability supports the application of TTE in the evaluation of patients with proximal aortic aneurysms to determine the potential presence of concomitant valvular pathology or ventricular dysfunction. Importantly, TTE lacks reliability in mid-ascending aortic assessment. Thus, TTE is not recommended as the primary imaging modality for ascending aortic aneurysm surveillance. TTE may, however, be used to follow patients with aortic disease limited to the aortic root. TEE allows the assessment of biventricular function, segmental wall motion, and the potential for coronary artery ostia involvement in the setting of concomitant proximal aortic dissection. Intraoperative TEE provides a principle technique for the confirmation of preoperative diagnoses and the detection of pericardial or pleural effusions and aortic regurgitation. In addition, TEE enables the determination of the extent of aortic dissection and the identification of the intimal tear location, while providing characterization of aortic aneurysms and the confirmation of appropriate true lumen flow upon commencement of cardiopulmonary bypass.

ETIOLOGY

Aortic aneurysm formation is most commonly the result of medial degeneration, which is characterized by noninflammatory smooth cell loss, fragmentation of elastic fibers, and basophilic ground substance accumulation in the medial layer of the vessel wall. Debate persists regarding the pathogenesis of aortic medial degeneration and the resultant ascending aortic aneurysmal dilation; specifically, whether medial lesions are the result of primary connective tissue defects or arise secondary to hemodynamic forces, or both. Functional and structural asymmetry and decreased elastic performance of the aorta are proposed mechanisms for aneurysm formation, implicating advanced age, coronary artery disease, chronic smoking, and 17β-estradiol deficiency as primary predisposing factors to aneurysm formation. An increased incidence of ascending aortic aneurysm is also demonstrated in the setting of congenital or acquired aortic valve malfunction, an aortic bicuspid or unicuspid valve, and in hereditary connective tissue disorders including Marfan syndrome (fibrillin-1 gene mutation), Ehlers-Danlos syndrome type IV (deficiency in type III collagen), Loeys-Dietz syndrome, and familial thoracic aortic aneurysm and dissection. Medial degeneration may, therefore, be the result of damage and repair events in the aging aorta, abnormal postvalvular hemodynamics (excessive injury), and connective tissue disorders (impaired repair).

In addition to medial degeneration, established etiologies for thoracic aortic aneurysm formation include atherosclerosis, infection, inflammation, trauma, cardiopulmonary resuscitation, and chronic dissection. While atherosclerosis is considered a principal factor in the development of descending aortic aneurysms, dedicated study has demonstrated only a minor role for atherosclerosis in the pathogenesis of aneurysms of the ascending aorta. Inflammatory conditions include bacterial or fungal aortitis, Takayasu arteritis, and giant cell arteritis. Prior to improved diagnosis and the development of effective antibiotic therapies, syphilitic aortitis was the most common cause of ascending aortic aneurysmal enlargement, characterized by an obliterative end arteritis of the vasa vasorum. Risk factors associated with the development of thoracic aortic aneurysms include hypertension, smoking, and chronic obstructive pulmonary disease. Dedicated preoperative risk stratification and determination of underlying cardiopulmonary function, potential bleeding diatheses, and underlying connective tissue defects are critical in the operative planning for reconstruction of the ascending aorta.

EPIDEMIOLOGY AND PATHOPHYSIOLOGY OF BICUSPID AORTIC VALVE DISEASE

Bicuspid aortic valves (BAVs) represent the most common congenital heart anomaly, occurring in 0.9% to 2% of the population. Patients with a BAV demonstrate significantly increased wall stress, accounting for the increased propensity for aortic dilation, rupture, and dissection in this patient population. In addition to increased wall stress, patients with BAV disease have larger dimensions at the aortic annulus, sinus of Valsalva, the sinotubular junction, and the ascending aorta irrespective of the valve hemodynamic function. Microscopic studies and gene expression profiles for patients with BAV and concomitant ascending aortic aneurysm have demonstrated that the media of the aorta above the BAV is consistently abnormal and independent of both inflammatory response and the hemodynamic function of the valve. These data demonstrate that aortic root dilation is a morphological correlate of intrinsic structural aortic abnormalities in patients with BAV disease.

In retrospective study of patients with BAV disease, the aortic valve morphology was found not to be predictive of the pathologic anatomy of the thoracic aorta; however, patients with predominant aortic insufficiency demonstrated the highest incidence of aortic root dilation. In patients with a BAV and an absence of baseline aortic aneurysm, a 16-year mean follow-up (6,530 patient-years) has demonstrated an estimated age-adjusted relative risk of 86.2 for aneurysm formation in comparison to the general population. In patients with BAVs, aortic dissection incidences for patients 50 years and older at baseline was 17.4 per 10,000 patient-years and increased to 44.9 per 10,000 patient-years in individuals with concomitant aortic aneurysm. These epidemiologic data support a research commitment to risk-stratification modeling and selective preventative reconstruction in patients with BAV disease.

ANATOMY OF THE AORTIC ROOT

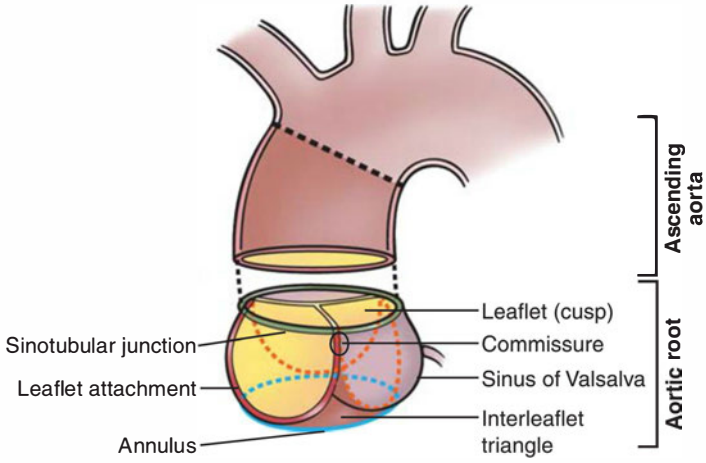
The aortic valve is described as the aortic root, given the anatomic and functional relationships of the aortic cusps and surrounding structures. The aortic root is defined by four distinct anatomic components: the aortic annulus, the aortic cusps, the aortic sinuses or sinuses of Valsalva, and the sinotubular junction. The aortic annulus is the attachment between the aortic root and the left ventricle. The aortic root is attached to the ventricular myocardium throughout 45% of its circumference and to the anterior leaflet of the mitral valve and the membranous septum in the remaining 55%. The aortic annulus forms a superior border to the three triangular structures of the left ventricular outflow

tract. The triangle below the right and left cusps consists of ventricular muscle and is rarely affected by connective tissue disorders of the aortic root. The additional two triangles are fibrous and flatten to acquire a broader base in patients with a dilated aortic annulus. The semilunar aortic cusps have a base and free margin extending from commissure to commissure. The sinotubular junction refers to the ridge above the commissures and is functionally important for the aortic root as it suspends the aortic cusps. Dilation at the sinotubular junction moves the free margins of the aortic cusps away from one another to result in central aortic insufficiency with ineffective coaptation. The aortic sinuses are anatomically described as the arterial walls contained within the aortic annulus and sinotubular junction (Fig. 60.1). Isolated dilation of the sinuses has no effect on valvular competence. The aortic sinuses maintain coronary artery blood flow during the cardiac cycle while maintaining eddies and currents to facilitate closure of the aortic cusps during diastole.

The aortic root contains a proportionally high content of elastin with a resultant compliance that diminishes with anatomic progression into the aortic arch and descending aorta. Systolic expansion of the ascending aorta represents the transfer of a portion of left ventricular kinetic energy into potential energy within the aortic wall. Diastolic recoil transfers this potential energy into kinetic energy for resultant forward flow. The transverse diameter of the aortic annulus is approximately 15% to 20% larger than the sinotubular junction diameter in young patients. The aging process results in a loss of elastic fibers within the arterial wall and the aortic root becomes less compliant, an effect that is associated with the presence of diastolic left ventricular dysfunction. An understanding of these dynamic physiologic and disease-specific changes in the ascending aorta will guide the approach to reconstruction.

PRESENTATION

Acute rupture of the aneurysmal ascending aorta is manifested as severe anterior chest pain with sudden-onset of congestive symptoms resulting from associated cardiac tamponade. Patients with acute rupture or dissection necessitate emergent resuscitation and operative intervention. In the absence of rupture or dissection, ascending aortic aneurysms are primarily identified on unrelated imaging or during surveillance of associated anomalies. The predominant symptoms of aneurysmal enlargement result from distal arch involvement and impingement on nearby structures and may manifest as chest pain, back pain, dyspnea, hoarseness, or transient neurologic deficit. Ascending aortic enlargement may result in airway compression or superior vena cava obstruction. Heart failure symptoms may arise in the setting of aortic valve regurgitation secondary to aortic root or ascending aortic dilation. On physical examination, a widened pulse pressure or diastolic murmur signifying aortic insufficiency may be present in the setting of sinotubular ridge or aortic root enlargement. BAV disease may manifest as asymptomatic stenosis or regurgitation of the valve on physical examination.



Aortic valve: Three leaflets only

Aortic root: All components (*Sinuses of Valsalva, interleaflet triangles, sinotubular junction, leaflet attachments, leaflets, annulus*)

Fig. 60.1. Defined terminology for the aortic valve and aortic root components. (Adapted with permissions: Sievers et al. Eur J Cardiothoracic Surg 2012, 41(3):478-482.)

INDICATIONS FOR INTERVENTION

A significant research commitment to define intervention criteria for the ascending aorta and aortic valve has provided disease-specific thresholds for operative reconstruction to prevent the life-threatening risks of rupture and acute dissection. The following recommendations are provided as evidence-based guidelines supported by the American Heart Association consensus statement:

- Surgical repair is recommended for asymptomatic patients with an acceptable physiologic status and a degenerative ascending aortic aneurysm, chronic aortic dissection, intramural hematoma, penetrating atherosclerotic ulcer, mycotic aneurysm, or pseudoaneurysm in the setting of an ascending aortic or aortic sinus diameter of 5.5 cm or greater.
- Elective reconstruction is recommended for patients with disorders of the ascending aorta (Marfan syndrome, Ehlers–Danlos syndrome, Loeys–Dietz syndrome, Turner syndrome, BAV, or thoracic aortic aneurysm and dissection) at 4 to 5 cm to avoid acute dissection or rupture.
- Operation should be considered for patients with a growth rate of 0.5 cm per year or greater in an aorta that is $<$5.5 cm in diameter.
- Concomitant repair of the aortic root or replacement of the ascending aorta is recommended for patients undergoing aortic valve repair or replacement with an ascending aortic or aortic root diameter of $>$4.5 cm.

Ascending aortic replacement alone is performed in patients with normal sinuses of Valsalva and a nonaneurysmal sinotubular junction. Individuals with concomitant aortic valve disease should undergo separate valve and aortic replacements. Patients with Loeys–Dietz syndrome and an internal aortic diameter of 4.2 cm by echocardiography or a 4.6-cm external aortic diameter by CT or MRI are considered candidates for operative repair. When contemplating pregnancy, women with

Marfan syndrome should be considered for aortic replacement when the aortic diameter exceeds 4.0 cm in diameter. In addition to this guideline, patients with Marfan syndrome or a BAV are recommended to undergo ascending aortic replacement when the maximal cross-sectional area (cm^2) of the ascending aorta or root divided by height (m) is >10. This guideline is applied to patients with Marfan syndrome to account for the 15% risk of aortic dissection at a diameter <5 cm. In addition, sinus conservation is not recommended in Marfan patients secondary to the requirement for reoperation.

Patients without significant aortic root dilation, elderly patients, or young patients with minimal dilation should be considered for separate valve and ascending aortic replacement in the presence of aortic valve disease. In the setting of concomitant aortic root and sinuses of Valsalva dilation, excision of the sinuses in combination with a modified David reimplantation operation should be performed. Aortic root reconstruction with reimplantation of the native valve provides an alternative to alloprosthetic composite replacement. With this technique, a level of coaptation within the tube graft is essential to achieve valve competence. If this operation is not considered technically feasible, a Bentall procedure is performed and involves a composite valve graft with mechanical or biologic components. This technique provides the capability to replace both the aortic root and ascending aorta in selected patients. In the setting of aortic regurgitation secondary to BAV disease, repair of the aortic valve is recommended in the absence of severe fibrosis or calcification and may necessitate root remodeling or tailoring of the sinotubular junction. Aortic allografts with coronary reimplantation may be considered in patients with endocarditis, women anticipating pregnancy, and young adults with active lifestyles or contraindications to anticoagulation. Pulmonary autografts are considered a congenital operation due to the proposed growth potential of the autograft.

While these guidelines represent accepted criteria for intervention, recent research efforts have established that aortic valve replacement alone may be sufficient in patients without connective tissue disorders with moderate poststenotic ascending aorta dilatation (50 to 59 mm), reserving aortic replacement for patients with a long-life expectancy. These data demonstrate that further long-term outcome research is needed in the evaluation of concomitant aortic valve and ascending aortic aneurysmal disease.

The selection of an appropriate valve substitute in patients with aortic valve stenosis requiring valve replacement should be determined by comorbid disease, risk of complications associated with anticoagulation and reoperation, and the projected life expectancy. In addition to aortic root replacement for valvular regurgitation, composite valve graft conduits provide the capability for the treatment of the stenotic aortic valve and ascending aorta.

Coronary artery bypass grafting may be performed at the time of ascending aortic reconstruction and is indicated in 25% of patients. In addition, patients with moderate-to-severe mitral regurgitation should undergo mitral valve replacement or repair concurrent with aortic replacement.

OPERATIVE PREPARATION AND INITIAL EXPOSURE

Central venous access is obtained through the internal jugular vein and hemodynamic pressure monitoring is placed into the radial artery. Pulmonary artery catheter monitoring is recommended to follow intraoperative filling pressures and cardiac output. Large-bore peripheral intravenous access is achieved for intraoperative resuscitation. In preparation for potential circulatory arrest, continuous pressure monitoring through the femoral artery is recommended as the radial artery waveform may be dampened. General endotracheal anesthesia is initiated and maintained through a single-lumen endotracheal tube. A nasopharyngeal temperature probe is recommended to monitor cooling and rewarming.

Neurologic Protection and Cardiopulmonary Bypass

A collaborative commitment to efficient and effective communication is essential between the anesthesiology and operative teams to minimize the duration of circulatory arrest and the potential for neurologic injury. Cardiopulmonary bypass and selective hypothermia are required for operations that involve replacement of the aortic arch. Cerebral protection may be achieved by profound hypothermia alone, direct antegrade perfusion of one or more brachiocephalic arteries, or retrograde perfusion with cold oxygenated blood infused into the superior vena cava during the arrest period. Following median sternotomy, pericardiotomy, and heparinization, the ascending aorta is cannulated in the region to be excised. Epiaortic ultrasound may aid cannulation in the setting of aortic calcification. Alternative to the ascending aorta, the axillary and femoral arteries provide potential sites for the arterial inflow of the cardiopulmonary bypass circuit. If the arch is involved and circulatory arrest is indicated, the right axillary artery, femoral artery, or aneurysmal aorta is cannulated. The right axillary artery enables a single arterial cannulation strategy for the entire operation, permitting selective antegrade cerebral perfusion.

Venous cannulation is achieved by a single two-stage cannula or bicaval cannulation strategy, if retrograde cerebral perfusion is to be performed (Fig. 60.2). A temporary period of retrograde perfusion is preferred when sustained cerebral perfusion is not planned to flush gaseous and embolic debris prior to the initiation of cardiopulmonary bypass. To support a period of retrograde cerebral perfusion, a connection is established between the arterial and superior vena cava cannulae. This connection is clamped during cardiopulmonary bypass and opened with alternative clamping of the arterial inflow and caval cannulae to provide retrograde cerebral perfusion.

Cardiopulmonary bypass is initiated upon achievement of an activated clotting time >480 seconds. A retrograde cardioplegia cannula is directed into the coronary sinus and a sump drain is placed into the left ventricle through the right superior pulmonary vein across the mitral valve. Cardiopulmonary bypass flows are increased and moderate systemic hypothermia (28°C) and hemodilution (Hct: 15% to 25%) are achieved. Cold blood hyperkalemic cardioplegia (4°C) is administered antegrade into the aortic root (300 ml/min) for 2 minutes followed by 2 minutes of retrograde flow (200 ml/min). Following the initiation of cooling, the ventricle is examined for distention to confirm appropriate sump placement. Cooling is advanced until the achievement of electroencephalogram (EEG) silence and the head is packed in ice. Proximal aortic reconstruction may begin during cooling following the administration of cardioplegia solution into the aorta proximal to a preplaced aortic cross-clamp. The patient is cooled for an additional 5 minutes following EEG silence to a core temperature that is typically below 18°C. Whole-body cardiopulmonary bypass is then discontinued and retrograde cerebral perfusion is initiated with the placement of a superior vena cava tourniquet. Retrograde cerebral perfusion should be maintained at a perfusion pressure of 20 to 40 mmHg at mild or profound hypothermic temperatures. Alternatively, antegrade perfusion techniques should maintain a perfusion pressure of 50 to 80 mmHg instituted through direct

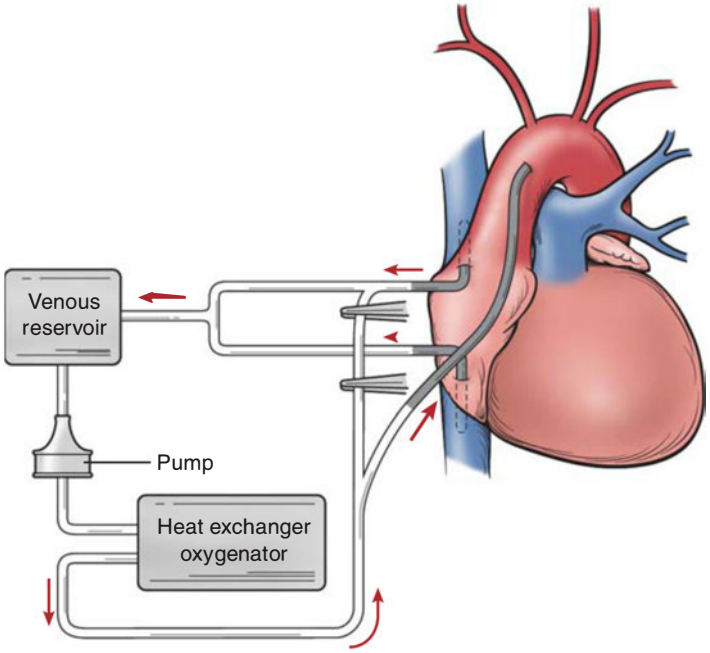


Fig. 60.2. Bicaval cannulation technique for cardiopulmonary bypass with alternative clamping strategy for the performance of retrograde cerebral perfusion.

cannulation of the brachiocephalic arteries, side-graft anastomosis to the axillary artery, or direct cannulation of the aortic graft.

The patient is transitioned to a Trendelenburg position. The aortotomy is then performed and cardioplegia solution is administered into the coronary ostia, and is to be repeated every 10 minutes into the ostia, coronary sinus, or both. The aneurysmal aorta is then excised through an incision down the lesser curvature with the preservation of all uninvolved vessels. When retrograde cerebral perfusion is utilized, a transition to whole body cardiopulmonary bypass is performed following completion of the distal anastomosis. If utilizing the ascending aorta as the site of initial arterial access, a sidearm graft may provide antegrade cerebral perfusion following completion of the distal anastomosis. Pump suckers aid in this transition when placed into the proximal unclamped graft as flow is slowly resumed. Following completion of the proximal anastomosis, deairing is achieved through a catheter placed into the graft. The patient is then placed in deep Trendelenburg position, flows from the bypass circuit are reduced, and the cross-clamp is removed. Sump suction allows decompression of the heart and flow from the bypass circuit is resumed. Rewarming is initiated and directed to achieve a difference in perfusate and core body temperature $<10°C$ until a core body temperature of 27°C is achieved, upon which a perfusate temperature of 37°C is maintained. TEE is performed to confirm the absence of air bubbles prior to removal of the deairing needle. The retrograde cardioplegia cannula and sump drain are removed and the patient is weaned from cardiopulmonary bypass.

Independent risk factors for stroke in ascending aortic operations requiring hypothermic circulatory arrest include age >60 years, emergency operation, new preoperative neurologic symptoms, presence of clot or atheroma, concomitant cardiac procedures, and total cerebral protection time. Permanent neurologic injury is most commonly the result of thromboembolic event and is independent of the method of intraoperative cerebral protection. In retrospective review of ascending aorta-hemiarch replacement, comparable rates of permanent neurologic dysfunction were demonstrated between patients receiving deep hypothermic circulatory arrest (12.5%) or antegrade selective cerebral perfusion (7.6%; $P = 0.075$). The incidence of transient neurologic dysfunction increases linearly in relation to patient age and the duration of hypothermic circulatory arrest, with adequate cerebral protection afforded if the arrest period is kept <60 minutes.

AORTIC VALVE-SPARING OPERATIONS

Aortic Root Remodeling

The David technique for aortic root remodeling and aortic valve-sparing ascending aortic reconstruction is described in a dedicated chapter of this text. In brief, aortic root remodeling is achieved through excision of the three aortic sinuses, preserving a remnant of arterial wall cusp attached to the aortic annulus and coronary artery ostia. A Dacron fabric strip is applied to the fibrous components of the left ventricular outflow tract and the commissures of the aortic valve are resuspended into the tailored graft. The neoaortic sinuses are sutured to the aortic annulus with a continuous 4-0 polypropylene suture and the coronary arteries are then re-implanted. The Cabrol technique involves the end-to-end anastomoses of a single 6- to 8-mm Dacron graft to the coronary arteries with subsequent side-to-side anastomosis of the graft to the reconstructed aorta in the setting of low-lying coronary ostia or prior scar. The distal aorta is clamped and cardioplegia solution is injected into the reconstructed aortic root to confirm aortic valve competence. The distal aortic graft is sutured to the ascending aorta to complete aortic continuity. In patients with a connective tissue disorder, the aortic annulus may dilate postreconstruction. It is, therefore, preferred to perform aortic valve reimplantation in patients with aortic root aneurysm with or without annuloaortic ectasia.

Reimplantation of the Aortic Valve

Reimplantation of the aortic valve into the reconstructed aortic root is similarly achieved through the excision of the three aortic sinuses. A triangular segment of the graft is excised that corresponds to the commissure between the right and left cusps. Horizontal mattress 4-0 polyester sutures are passed inside-to-out on the left ventricular outflow tract below the aortic annulus. These sutures are placed in a single horizontal plane. These preplaced sutures are then passed inside–out through the aortic graft. The aortic valve is placed within the graft and the left ventricular outflow tract sutures are tied on the outside. The distal end of the graft is cut to a length of 5 to 6 cm and the three commissures of the graft are pulled gently upward. The commissures are secured to the graft with transfixing 4-0 polypropylene sutures from the inside-to-out and left untied. The levels of coaptation and commissure alignment are evaluated with gentle traction applied to each of the sutures to ensure appropriate geometry and optimal function of the reconstructed aortic annulus.

The three commissural sutures are tied on the outside of the graft, and the remaining aortic sinuses and aortic

annulus are secured against the graft. The left and right coronary arteries are reimplanted into their corresponding neoaortic sinuses and hemostasis is confirmed. The distal end of the graft is sutured to the ascending aorta with a running 4-0 polypropylene suture. Neoaortic sinuses, as described in the modified David technique, provide improved accommodation to the resultant mechanical stress imposed on the aortic valve leaflets within a straight tubular graft.

Repair of Aortic Cusp Prolapse

Aortic cusp elongation at the free margin results in valvular prolapse. This condition is corrected by central plication of the cusp. The nodule of Aranti is folded to the aortic side and plicated with a 5-0 polypropylene suture. Additional sutures may be applied separate from the initial site of plication and toward the commissures of the prolapsing cusp to further shorten the free margin. Plication elevates the level of cusp coaptation and corrects excessive motion.

In addition to plication, the free margin may also be reinforced by weaving a 6-0 expanded polytetrafluoroethylene suture from commissure to commissure and anchoring the suture on the outside of the aorta or graft to correct fenestration in a commissural area or excessive free margin elongation.

Aortic Root Replacement

Aortic root aneurysms with damaged aortic cusps necessitate replacement of the ascending aorta with a conduit containing a valve. Options for ascending aorta and aortic valve replacement include mechanical valve containing Dacron grafts, biologic or bioprosthetic aortic root prostheses, and pulmonary autografts or aortic valve homografts. The Bentall procedure incorporates a valved-graft conduit for reconstruction and replacement of the aortic valve and is our preferred approach to the treatment of both an irrepairable aortic valve or the reconstruction of the aortic root in a patient with concomitant acute aortic dissection and annuloaortic ectasia.

Following intraoperative evaluation of the aortic valve and confirmation of irrepairable cusp damage, the aortic valve is excised and the annulus debrided of all calcium, scar, and any abnormality. Each sinus is excised with a remnant arterial cusp attached to the aortic annulus, and a 5-mm rim is conserved around each coronary artery. Mobilization of the coronary arteries should be performed to provide reimplantation without tension.

The selected replacement valve is secured to the aortic annulus with inverted horizontal mattress sutures of 2-0 polyester sutures with pledgets. Simple interrupted sutures may be applied in patients with a small aortic annulus size to body surface area to allow implantation of a larger valve. Round openings are fashioned in the aortic graft to allow reimplantation of the respective coronary arteries without tension or kinking. Particular attention is directed to the positioning of the right coronary artery, as right ventricular motion may induce kinking. The size of each coronary ostia should not exceed twice the value of the coronary artery diameter to reduce the risk of coronary button aneurysm formation, particularly in patients with Marfan syndrome. A continuous 5-0 polypropylene suture is utilized to secure the coronary buttons to the respective coronary ostia. Teflon felt is not recommended. 4-0 polypropylene suture is applied to fashion the distal graft-aorta anastomosis. The length of the graft should not exceed 6 to 7 cm.

Application of a stented bioprosthetic valve is achieved with concomitant graft and bioprosthesis fixation to the aortic annulus with multiple interrupted 2-0 polyester sutures on the ventricular side of the annulus. If concern is present for bioprosthetic valve failure, the valve may be sutured to the graft prior to fixation to the aortic annulus. This is our preference.

Isolated Ascending Aorta Replacement

Following the commencement of cardiopulmonary bypass, the aorta is cross-clamped proximal to the innominate artery. The heart is arrested with cold blood cardioplegia and the aorta is transected below the clamp, preserving a sufficient aortic cuff for the subsequent distal anastomosis. The proximal aorta is then transected superior to commissures. An appropriately sized Dacron graft is chosen and distally sewn to the aortic cuff with a 3-0 or 4-0 polypropylene suture. An aortic valve replacement may be performed at this time in the setting of a normal diameter aortic root. The graft is then fashioned to the desired length and the proximal anastomosis is completed with a continuous 3-0 or 4-0 polypropylene suture (Fig. 60.3). A catheter is inserted into the graft following completion of the proximal anastomosis to deair the graft. The patient is subsequently weaned from cardiopulmonary bypass.

Open Distal Anastomosis Technique

In the setting of significant atherosclerotic burden at the distal ascending aorta or concomitant aneurysmal aortic arch disease, an open distal anastomosis under circulatory arrest may be necessitated. Arterial inflow for cardiopulmonary bypass is maintained through the right axillary artery, femoral artery, or ascending aneurysmal aorta. The presented setup for retrograde cerebral perfusion and cardiopulmonary bypass is assumed. The patient is cooled to achieve EEG silence and a nasopharyngeal temperature of 15°C. Trendelenburg positioning is assumed and approximately one-fourth of the patient's blood volume is drained into the venous reservoir. The aorta is transected distal to the furthest extent of aneurysmal disease. Beveling of the graft may be necessitated in the setting of hemiarch involvement to preserve native branch vessels. Involvement of the brachiocephalic vessels supports aortic transection distal to the left subclavian artery. The distal anastomosis is completed as previously described. An ellipse of the graft may be removed to accommodate reimplantation of the brachiocephalic vessels, completed with a felt-enforced 3-0 polypropylene running suture. Selective antegrade cerebral perfusion or slow retrograde cerebral perfusion through the superior vena cava is maintained during the brachiocephalic

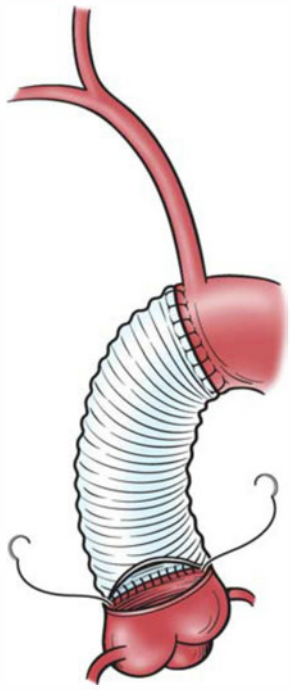


Fig. 60.3. Completion of the proximal ascending aortic anastomosis following transection of the aorta superior to the commissures.

anastomosis to avoid embolization of air or debris. If selective antegrade perfusion is utilized throughout systemic circulatory arrest, a period of retrograde perfusion is recommended to similarly protect against induced embolic injury. Upon completion of the distal and potential brachiocephalic anastomoses, cardiopulmonary bypass is initiated through a preplaced 8-mm side-arm graft, direct cannulation of the ascending aortic graft, or initial axillary artery cannula utilized for continuous antegrade cerebral perfusion. The avoidance of femoral artery cannulation for resumption of bypass limits that potential for embolic injury. Additional cardiac procedures may now be performed as indicated for coronary revascularization or concomitant mitral valve disease. The proximal anastomosis is completed to the remnant proximal aorta above the sinotubular ridge. Deairing of the graft is performed through the direct insertion of a catheter into the graft and the patient is subsequently weaned from cardiopulmonary bypass.

OUTCOMES

Significant advancements in surgical technique have supported a reduction in operative mortality to approximately 1.2% to 3.7% for elective ascending aortic aneurysm resection, independent of etiology. Malperfusion resulting from acute aortic dissection is the primary determinant of early mortality for proximal aortic operations, supporting preventative reconstruction in the setting of at-risk aneurysmal disease. Predictors of mortality following elective ascending aortic reconstruction include: New York Heart Association class (III or IV), advanced age, preoperative dissection, reoperation status, prolonged cardiopulmonary bypass, and the requirement for concomitant coronary artery bypass grafting or arch reconstruction. Patients with connective tissue disorders such as Marfan syndrome demonstrate a low operative risk with an associated 30-day morality of 1.5% with prophylactic surgical replacement of the ascending aorta, supporting elective intervention to prevent aortic catastrophe. Aortic root replacement and distal aortic reconstruction may be approached with low operative mortality independent of the type of operation. As our understanding of valvular and aortic pathologies evolves, a commitment to patient-specific treatment promises to improve outcomes for patients with diverse pathologies and across all risk stratifications with ascending aortic aneurysmal disease.

Bleeding complications in the early postoperative period present the most common threat to immediate survival with indication for re-exploration in 4.5% to 17% of patients, particularly in the setting of prolonged cardiopulmonary bypass times and concomitant cardiac or aortic operations. As described, neurologic dysfunction represents a principle complication following aortic root and ascending aortic reconstruction. In retrospective review, transient neurologic dysfunction is noted in 9.8% of patients following ascending aortic and proximal aortic arch reconstruction, with a demonstrated increased incidence of permanent neurologic deficit, hospital stay, and a significantly impaired quality of life. Debate persists regarding the optimal strategy for cerebral protection. Deep hypothermic circulatory arrest provides a principle method for cerebral preservation, with a demonstrated effectiveness as the sole protection strategy. Retrograde cerebral perfusion has an established efficacy in the reduction of postoperative stroke that is most pronounced in patients receiving prolonged hypothermic circulatory arrest. The duration of deep hypothermic circulatory arrest is the primary predictor of transient neurologic dysfunction, regardless of the application of retrograde cerebral perfusion. These data support further research commitment to patient-specific strategies for preventative cerebral protection during aortic root and ascending aortic reconstruction.

The described David procedure affords a reliable technique for the reconstruction of the aortic root, with an estimated 4.8% of patients demonstrating relevant valve dysfunction. A 3-year survival of 95% and 89% is achieved with the David procedure for aortic root and ascending aortic aneurysmal disease, respectively. In patients with ascending aortic aneurysm and concomitant dilatation at the sinotubular junction, reduction in the sinotubular junction may be an alternative technique for the treatment of aortic insufficiency in the setting of normal aortic cusps with a 5-year freedom from aortic insufficiency of 91%. Long-term follow-up data at 10-years following the David reimplantation technique have demonstrated a 92% survival with a freedom from moderate or severe aortic insufficiency of 94%. From this study, 90% of patients achieved a New York Heart Association function class I with the remaining 10% demonstrating class II presentation at 10 years.

In review of the presented Bentall procedure, mechanical and biologic valve conduits demonstrate comparable long-term survival supporting patient-specific selection. The presence of clot or atheroma is an independent predictor of both short- and long-term outcome following the Bentall procedure, with an associated eightfold increase in in-hospital mortality. Patients under the age of 50 years may select a mechanical aortic valve for the Bentall procedure, with consideration for the low incidence of long-term valve-related complication. Alternatively, patients above the age of 50 years may benefit from a biologic aortic valve with comparable long-term outcomes and the requirement for only a valve replacement rather than replacement of the composite root should postoperative valve dysfunction arise. Review of experience with a totally biological composite stentless aortic valved conduit implanted utilizing the Bentall technique has demonstrated a freedom from valve-conduit-related mortality of 88% over a 7-year follow-up period. In this study, >50% of patients achieved a New York Heart classification of I or II, with no reported valved conduit failures.

Reoperation following ascending aorta reconstruction is associated with an estimated mortality of 3% following elective intervention, with an incidence of 38% for emergent reoperations. In addition to indications for reoperation, distal thromboembolism, prosthetic valve endocarditis, and aortic graft infection represent principle considerations in the long-term follow-up of patients following ascending aortic reconstruction.

CONCLUSIONS

The presented techniques for aortic root and ascending aortic reconstruction for aneurysmal disease provide a promising hope for cure for many with diverse aortic pathologies. Outcomes following ascending aortic and aortic root reconstruction provide reliable long-term survival and quality of life with the achievement of acute aortic event prevention. As we continue to advance our understanding of aortic disease, operative technique refinement will be founded on patient-specific reconstruction and an enhanced understanding of aortic fluid dynamics and disease-specific aneurysmal change.

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EDITOR'S COMMENTS

This chapter is a complete and comprehensive summary of the issue of surgery for ascending aortic aneurysms. As noted by the authors, various types of aneurysms affect components of the aortic root and each type should be addressed based on its underlying pathologic and physiologic characteristics.

As greater information has been obtained about the relative risk of rupture or dissection for various sizes of aortic aneurysm, guidelines have been developed to suggest when surgical therapy should be undertaken. It is important to note that the guidelines are not absolute, but that the rate of progression of dilation may also be considered a primary indicator for operative therapy independent of aortic aneurysm size. It is particularly of concern that patients with Loeys-Dietz Syndrome have a high risk of aortic aneurysm rupture even at relatively smaller sizes. Therefore, the guidelines would suggest that operative intervention is appropriate in patients with Loeys-Dietz syndrome when the aortic root is only 3.5 to 4 cm in diameter. In children when the aortic root exceeds 3 cm in diameter, in a patient with Loeys-Dietz Syndrome, operative intervention should be undertaken. If these patients attain adulthood, as aortic root diameter >4 cm is generally considered an indication for operation. In addition, extensive searching for aneurysms in the rest of the vascular tree is recommended in this particular genetic abnormality.

The issue of root dilation in patients with conotruncal defects is a significant one. Most patients with tetralogy of Fallot will have some evidence of aortic root dilation over time, especially of the aorta above the sinotubular junction. In addition, patients who have had repair of hypoplastic left heart syndrome with patch augmentation of the aortic arch and patients with transposition of the great arteries who have had an arterial switch operation commonly have root dilation and aortic arch dilation. While it has been shown that the aortic roots dilate in patients with transposition after the arterial switch, at the present time there is a relatively low incidence of aortic insufficiency and dissection and rupture have not been reported. Nevertheless, these patients will need to be followed carefully as they age since progressive dilation may require intervention in some patients, preferably with preservation of aortic valve function with a David-type valve sparing root replacement.

The surgical techniques for aortic aneurysm resection and root replacement have been well described by the authors of this chapter. It is important in the David operation of valve sparing root replacement when the aortic leaflets are reimplanted into the Dacron graft to place interrupted sutures at the base of the sinuses and commissures of the aortic root through the graft, but not circumferentially. Placing mattress sutures circumferentially around the annulus and tying them to the graft can cause narrowing of the proximal graft anastomosis due to a purse string effect and can create left ventricular outflow tract obstruction. The majority of the hemostasis in the David operation involves suturing the aortic wall containing the aortic commissures and leaflets inside the graft. Thus, the proximal suture line can be simply tacked with a few interrupted sutures to prevent migrations, but hemostasis is not critical at this site since the graft is actually outside the aortic root.

TLS

61 Arch Aneurysms

Joseph S. Coselli, Susan M. Trocciola, Kim I. de la Cruz, and Scott A. LeMaire

INTRODUCTION

The aorta is divided into two major segments: the proximal aorta and the distal aorta. The proximal aorta includes the aortic root, ascending aorta, and transverse aortic arch. The transverse aortic arch is anatomically defined as the segment of aorta that includes the origins of the brachiocephalic vessels: the innominate artery, left common carotid artery (LCCA), and left subclavian artery (LSCA) (Fig. 61.1). The descending thoracic aorta starts distal to the origin of the LSCA and continues to the diaphragmatic hiatus, where it joins the abdominal aorta. Criado and colleagues, as well as Ishimaru and associates, have each separately mapped the arch into five anatomic zones for endovascular procedures (Fig. 61.1). This schema has mostly been used to describe proximal landing zones for disease that involves the descending thoracic aorta, but it is becoming increasingly valuable in describing combined open and endovascular (hybrid) approaches to aortic arch repair.

Aortic aneurysm can occur anywhere along the aorta and is defined as a permanent, localized dilation that is at least 50% greater than the normal diameter at that anatomic level. Most arch aneurysms are contiguous with an aneurysm involving the adjacent ascending or descending thoracic aorta. Isolated arch aneurysms are less common and generally appear as localized, saccular outpouchings.

Aneurysms of the aortic arch can be caused by a variety of diseases but most commonly result from medial degenerative disease and chronic aortic dissection. Additional causative factors include genetically triggered disorders (such as Marfan, Ehlers–Danlos, and Loeys–Dietz syndromes), infection, trauma, and aortopathy related to having a bicuspid aortic valve.

Surgical repair of the aortic arch remains one of the most difficult challenges in cardiothoracic surgery. This is largely because one must interrupt the natural flow of blood to both the brain and downstream organs. The first successful graft repair of a transverse aortic arch aneurysm was reported in 1957 by DeBakey, Crawford, Cooley, and Morris, who used an early form of antegrade cerebral perfusion (ACP).

For many years, aortic arch surgery was associated with high mortality and neurologic morbidity. Recently, dramatic advances in aortic arch surgery have improved outcomes. These advances include new or refined strategies for cerebral protection and arterial perfusion, revised reattachment strategies for the brachiocephalic vessels, and new options for treating extensive aortic disease. In addition, endovascular techniques that have recently revolutionized distal aortic repair are increasingly being applied to the aortic arch.

DIAGNOSIS AND PREOPERATIVE ASSESSMENT

Although most aneurysms are asymptomatic and are incidentally discovered through imaging studies performed for other reasons, some patients with aortic arch aneurysm present with symptoms, which are often related to the aneurysm's proximity to nearby structures. For example, patients may present with hoarseness because of stretching of the left recurrent laryngeal nerve. Patients may have dysphagia from esophageal compression, respiratory symptoms due to airway compression, or edema of the upper body from superior vena caval compression. Other problems that can arise from aneurysms involving the aortic arch and the adjacent segments include thromboembolic events such as stroke, and aortic valve regurgitation from associated root enlargement. Chest wall compression can cause chronic dull or aching retrosternal or mid-scapular pain. New severe pain usually indicates aneurysm rupture or acute dissection. Arch aneurysms can rupture into the pleural cavity (usually on the left), mediastinum, esophagus, or tracheobronchial tree. Dissection involving the arch can cause signs and symptoms of cerebral malperfusion, including syncope and neurologic deficits.

Whereas a chest radiogram may reveal a dilated aorta, definitive evaluation of an aortic arch aneurysm generally requires computed tomography or magnetic resonance imaging. These modalities can image the entire aorta and elucidate its relationship to surrounding structures and brachiocephalic vessels. Echocardiography is generally not useful in evaluating the aortic arch because of acoustic shadowing from the trachea. However, many patients with arch aneurysms have associated ascending aortic, valvular, or cardiac disease; therefore, echocardiography is useful for both planning the operation and assessing risk. Similarly, angiography is generally not used on a routine basis, but it is usually performed in patients who are already undergoing left heart catheterization to evaluate coronary artery disease.

Because there is a well-known association between aneurysmal aortic disease and carotid artery stenosis, it is useful to perform a screening ultrasound of the carotid arteries on patients with arch aneurysms. The risk of stroke is significant during aortic arch repair, and cannulation strategies may need to be revised in an individual patient on the basis of preoperative findings. Additional investigation may include pulmonary function testing, cardiac stress testing, nuclear imaging, and coronary catheterization.

INDICATIONS

Aortic aneurysms are repaired in order to prevent dissection, rupture, and death. Examining the natural history of aortic aneurysms has helped clarify the risk of these catastrophic complications. Elefteriades and his group at Yale found that aortic diameter is a strong predictor of rupture or dissection, especially when a "hinge point" is reached—a diameter of 6.0 cm in the

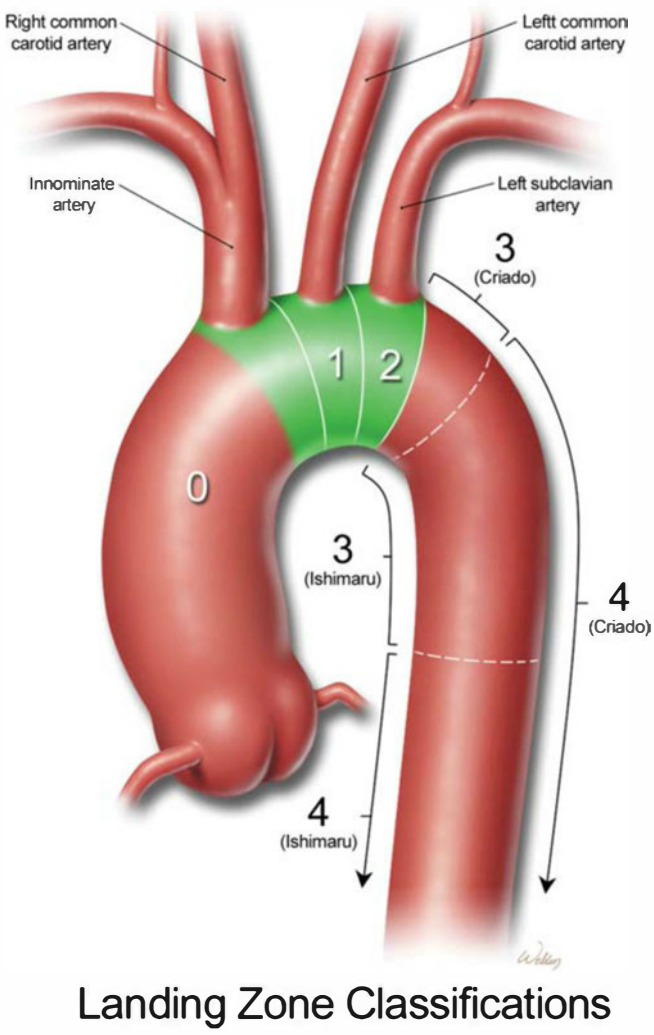


Fig. 61.1. Illustration of the proximal aorta. The arch is the short segment that includes the origins of the three brachiocephalic arteries—the innominate artery, the left common carotid artery, and the left subclavian artery. The Criado and Ishimaru landing zones are used to describe aortic anatomy during endovascular repair. The landmarks for defining the proximal aorta are the same in both systems. Zone 0 includes the ascending aorta and the origin of the innominate artery. Zone 1 includes the origin of the left common carotid artery. Zone 2 includes the left subclavian artery origin. The systems differ in their description of the distal aorta. In the Criado system, Zone 3 is a short section of the aorta that comprises the 2 cm immediately distal to the origin of the left subclavian artery, and Zone 4 begins where Zone 3 ends. In the Ishimaru system, Zone 3 is longer, extending to an imaginary border at the end of the arch curvature, whereupon Zone 4 begins. (Used with permission from Baylor College of Medicine, Houston, TX.)

ascending aorta or 7.0 cm in the descending aorta. Thus, the investigators recommended replacing the ascending aorta at a diameter of 5.5 cm and the descending aorta at 6.5 cm (5.0 and 6.0 cm, respectively, for patients with Marfan syndrome) or if an aneurysm of any size becomes symptomatic.

Current guidelines make several scenario-based Class IIa recommendations for the repair of aortic arch aneurysms, with the caveat that the approach to repair is generally dictated by adjacent disease of the ascending or descending aorta. For isolated arch aneurysms in low-risk, asymptomatic patients, it is reasonable to replace the arch when the aortic diameter exceeds 5.5 cm or is growing more than 0.5 cm per year. Saccular aneurysms tend to grow more rapidly than fusiform aneurysms and often warrant earlier intervention. For aneurysms limited to the ascending aorta and proximal arch, it is appropriate to perform a partial arch replacement along with the ascending aortic repair. Replacing the entire arch is reasonable in cases of chronic dissection and aneurysms that extend into the proximal descending thoracic aorta. Again, repair is indicated for symptomatic patients regardless of aortic diameter unless the patient has a limited life expectancy. Although the scenario of asymptomatic aortic arch aneurysm in patients with triggered disorders is not specifically addressed in these guidelines, patients with asymptomatic ascending aortic aneurysm in this circumstance may undergo elective repair at an aortic diameter of 4.0 to 5.0 cm, depending on the particular disorder.

PERFUSION AND CEREBRAL PROTECTION

There are several different surgical options for managing aneurysms that involve the transverse aortic arch. The surgical approach depends on the extent of involvement and the need for cardiac and cerebral protection. Repairs of the ascending aorta and arch are performed through a sternotomy with cardiopulmonary bypass (CPB), whereas aneurysms that involve the distal arch and descending/thoracoabdominal aorta are performed through a thoracotomy or thoracoabdominal incision.

Most aortic arch repairs necessitate a temporary period of systemic circulatory arrest combined with varying degrees of hypothermia to protect the brain and other vital organs. Over the last several years, advances in cerebral protection strategies and surgical techniques have been shown to improve outcomes in open arch repair; therefore, we have gradually incorporated these changes into our practice. Changes include the increasing use of ACP rather than retrograde cerebral perfusion (RCP), the use of the axillary or innominate artery instead of the femoral artery as a cannulation site for inflow, and refined hypothermic targets—mild 28.1 to 34°C) or moderate hypothermia (20.1 to 28°C)—rather than deep (14.1 to 20°C) or profound hypothermia (≤14°C).

The well-established role for hypothermia in cerebral protection stems from the fact that the cellular metabolic activity of the brain slows with cooling, reducing cerebral oxygen demand. However, the use of profound hypothermic circulatory arrest (HCA) during arch repair has important limitations; increasing rates of stroke and death have been associated with longer HCA times, especially those longer than 30 minutes. Nevertheless, for selected limited repairs of the aortic arch (i.e., those with an anticipated circulatory arrest time of <30 minutes), the use of HCA alone is considered a reasonable strategy for providing adequate brain

protection. Because many aortic arch repairs are quite complex and require longer periods of HCA, two cerebral perfusion strategies—RCP and ACP—were developed to supplement HCA by delivering cold, oxygenated blood to the brain for the purpose of reducing the risk associated with repair. RCP involves directing blood from the CPB circuit into the head through the snared superior vena cava cannula. Although this approach still has some proponents, recent evidence suggests that RCP is less beneficial than ACP, and today, the use of RCP in aortic arch surgery is generally in decline.

Unlike RCP, ACP provides cerebral flow via the brachiocephalic arteries. Early on, the brachiocephalic vessels were cannulated directly, but this approach was soon abandoned because of the high rate of stroke associated with it. Renewed interest in ACP developed after Japanese surgeons showed improved outcomes with the use of flexible balloon-tipped catheters to deliver ACP during arch repair. Contemporary ACP techniques commonly involve cannulating either the right axillary artery (Fig. 61.2) or the innominate artery and subsequent connection to the arterial limb of the bypass circuit. This procedure carries an inherent risk, although small, of brachial plexus and vascular injury. Once systemic circulatory arrest is induced, the innominate artery is occluded proximally with either a Rumel tourniquet or a vascular clamp, and bypass flow is reduced to approximately 10 ml/kg/min. This allows either the axillary or the innominate artery cannula to deliver oxygenated blood to the brain via the right common carotid artery.

When the circle of Willis is intact, unilateral ACP via the right common carotid artery can deliver blood flow to the left side of the brain. Methods to help determine the adequacy of cerebral cross-circulation include preoperative imaging and intraoperative monitoring. Our preferred method of intraoperative monitoring is brain near-infrared spectroscopy (NIRS), which measures cerebral oxygenation; an additional perfusion catheter can be inserted into the LCCA if NIRS monitoring indicates inadequate perfusion. Antegrade cerebral protection via the right axillary artery or innominate artery has become our standard adjunct for cerebral perfusion during HCA and avoids the problems that can develop from cannulating the femoral artery. These problems include plaque embolization, which can result in stroke and other complications; also, in cases of aortic arch dissection, the retrograde pressurization of the false lumen can result in cerebral malperfusion.

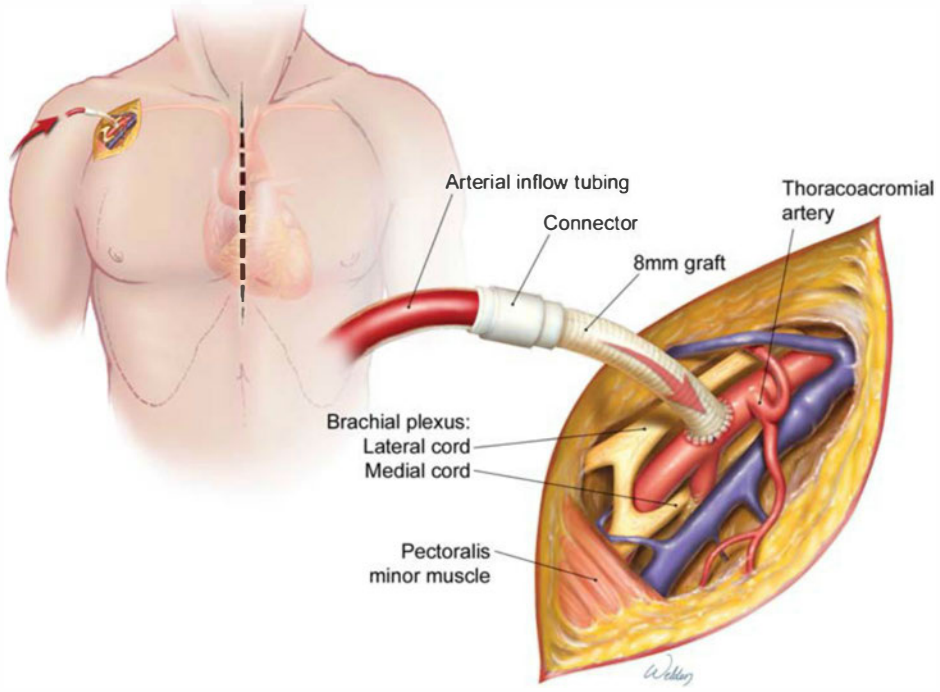


Fig. 61.2. Cannulation of the axillary artery via 8-mm graft for use in delivering cardiopulmonary bypass inflow and antegrade cerebral perfusion. (Image adapted from Gravlee GP, Davis RF, Stammers AH, et al., eds. Cardiopulmonary Bypass: Principles and Practice. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2008, Chap. 32, Fig. 1A.)

Recently, there has been a trend toward using warmer temperatures during HCA with ACP. The primary advantage of mild (28.1 to 34°C) and moderate (20.1 to 28°C) HCA is a reduced risk of the severe and life-threatening coagulopathy that occasionally results from profound hypothermia. Potential drawbacks of warmer temperatures include the increased risk of neurologic deficit, including spinal cord ischemia. Because of the importance of temperature management in patients subjected to HCA, the decision regarding where to measure body temperature is important. The jugular venous bulb provides the best approximation of brain temperature, but few centers use this approach. Nasopharyngeal and esophageal temperatures lag behind the jugular venous bulb temperature but serve as reasonable, practical options. Bladder and rectal temperatures are poor indicators of brain temperature and are generally not reliable during HCA cases.

OPTIONS FOR REPAIR

There are many different options available for repairing aneurysms that extend into the transverse aortic arch; a brief overview of several approaches is presented below. It is important that the surgeon develop a consistent approach that allows the anesthesiologist, perfusionist, and nurses to function as a team with the goal of limiting circulatory arrest and cardiac ischemic times. The guiding principle should be economy of steps (i.e., avoiding unnecessary activities), especially during circulatory arrest. Aneurysms involving only a portion of the transverse aortic arch are usually treated with partial arch repair. For more extensive aneurysms, the entire arch is typically replaced. If the aneurysm extends into the descending thoracic aorta, the aneurysm can be repaired in either one or two stages. Additionally, the arch can be repaired by using open surgical techniques, experimental endovascular techniques, or a combination of both open surgical and endovascular approaches (i.e., hybrid arch procedures).

Partial Arch Repairs

Partial arch repairs, such as patch graft aortoplasty and hemiarch approaches, are less complex than other aortic arch repairs because repair does not directly involve the brachiocephalic vessels. Patch graft aortoplasty can be used to repair saccular aneurysms that originate from the lesser arch curvature and involve <50% of the aortic

circumference. A fusiform ascending aortic aneurysm that extends into only the proximal portion of the arch (in a patient with a normal-sized distal arch) can be repaired with a hemiarch approach by using a beveled graft to replace a portion of the lower curvature of the arch (Fig. 61.3). Of note, for acute dissections that involve the arch (DeBakey I or II), we often use the hemiarch technique.

Total Arch Repairs

More extensive arch aneurysms often involve total replacement of the arch, with reattachment of the brachiocephalic vessels and a distal anastomosis at the proximal descending thoracic aorta. In the past, we reattached the brachiocephalic vessels to one or more openings made in the graft ("island patch technique") (Fig. 61.4) or by using smaller, individual grafts to replace each vessel at its origin. There are two disadvantages to the island patch technique. First, this technique can result in difficult-to-control bleeding from the posterolateral aspect of the anastomosis. Second, the aortic tissue left behind can become aneurysmal, necessitating a second operation; this problem is most common in patients with connective tissue disorders.

Y-Graft Techniques

In 2002, Spielvogel and colleagues described the trifurcated graft technique (also called the double Y-graft technique) that they devised to simplify arch reconstruction by essentially "debranching" the aortic arch without strictly following its original anatomic structure. This technique appears to improve outcomes by reducing embolization and minimizing cerebral ischemia. In this technique, single or double Y-branched grafts are created by suturing one or two branch grafts (usually 8 mm in diameter) to a main trunk (usually 12 mm in diameter) at a 45-degree angle; commercially available, prefabricated Y-grafts can also be used. Each brachiocephalic branch vessel is typically sutured end-to-end to separate distal ends of the Y-graft (Fig. 61.5). To expedite repair, debranching can be performed during the cooling process before HCA is achieved. The double Y-graft can then be used to provide ACP while the aortic arch is replaced with a graft. The Y-graft approach is commonly used with the collared elephant trunk graft, and because the brachiocephalic vessels have been debranched, it is possible to move the distal anastomosis forward from its traditional location just distal to the LSCA to a position in line with the origins of the LCCA or innominate artery. These adjustments in technique are thought to promote a hemostatic distal anastomosis.

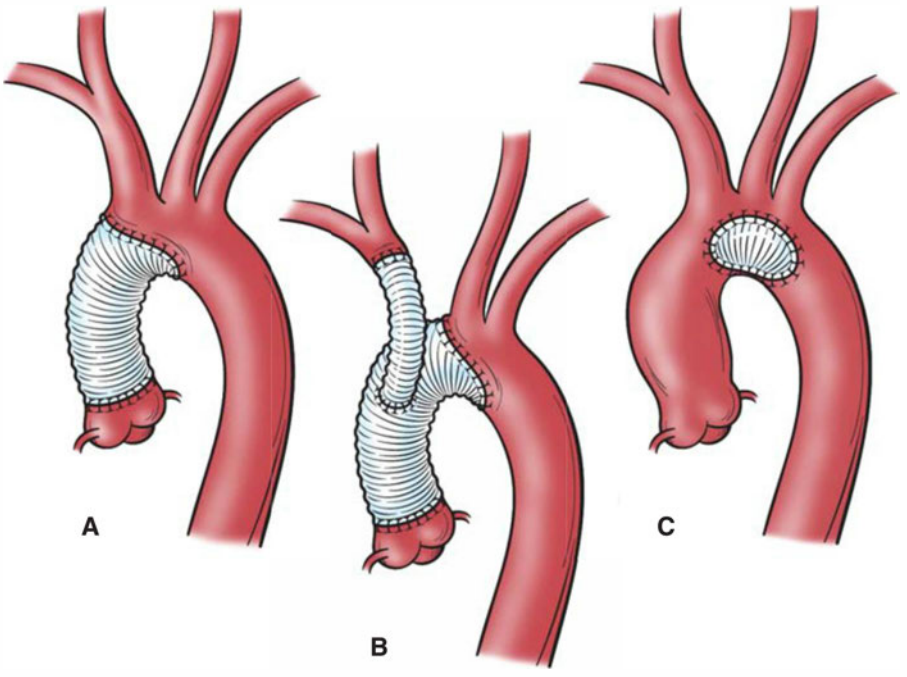


Fig. 61.3. Traditional partial arch repairs. These include (**A**) the hemiarch repair, (**B**) modifications of the hemiarch, such as a beveled arch with graft replacement of the innominate artery, and (**C**) patch repair of the aortic arch. In contemporary arch repair, patch repairs are infrequent.

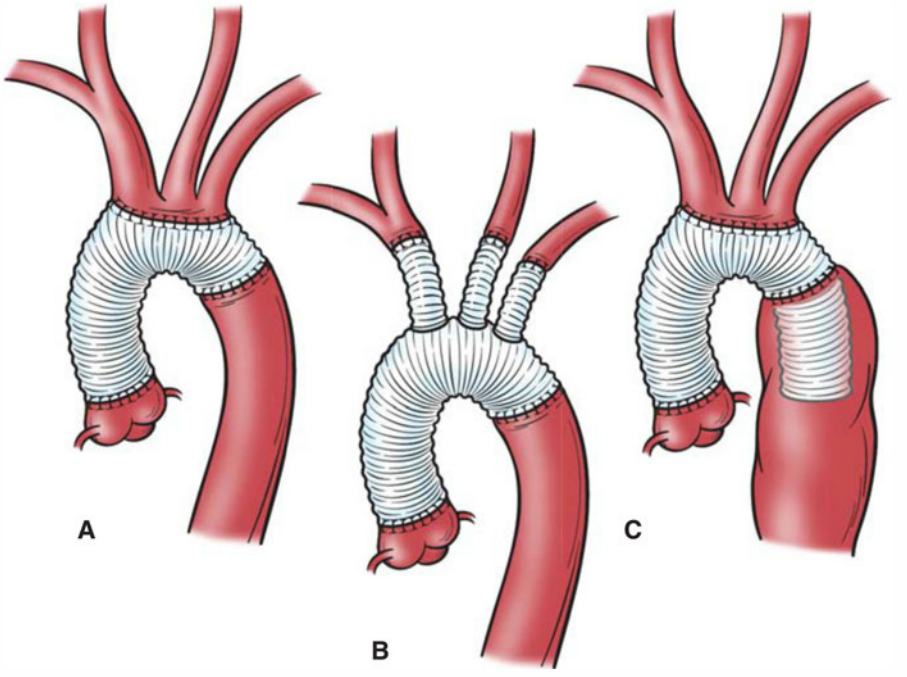


Fig. 61.4. Traditional total arch repairs. These include (**A**) the island approach, (**B**) the branched graft approach, and (**C**) the traditional elephant trunk approach (shown with a concomitant island brachiocephalic artery reattachment).

Elephant Trunk

For extensive aneurysms involving the entire thoracic aorta, repairs are generally performed in two stages, because the

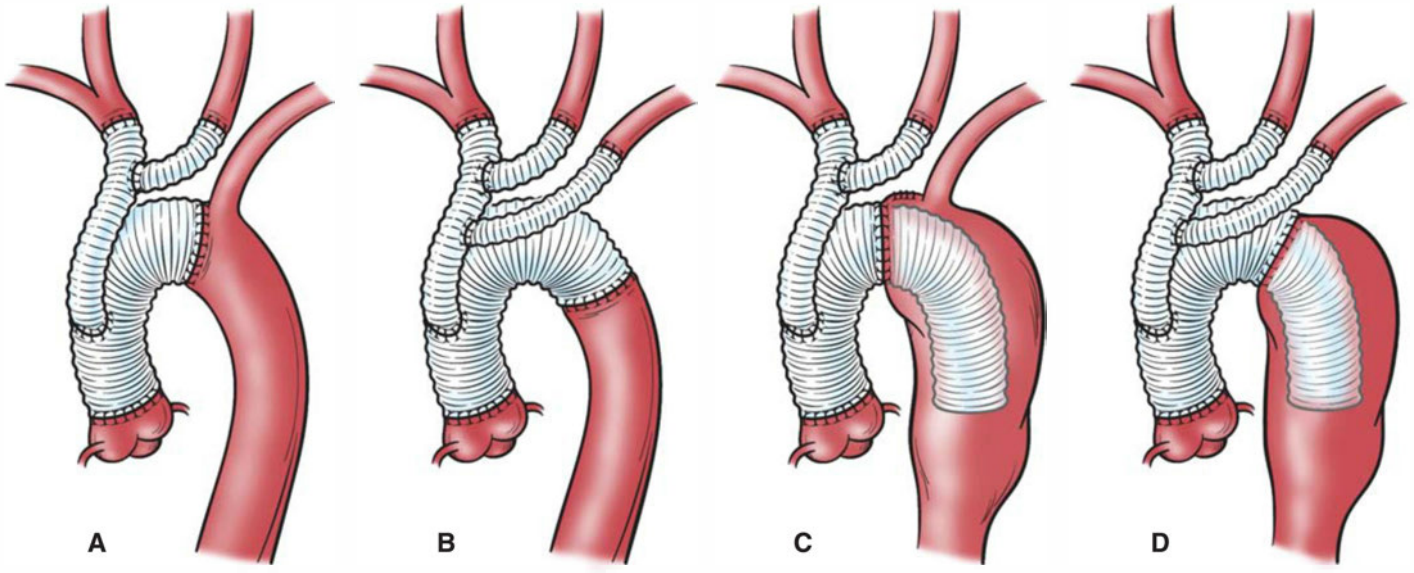


Fig. 61.5. Contemporary Y-graft arch repairs. These include (**A**) the single Y-graft approach, (**B**) the double Y-graft approach, (**C**) the elephant trunk approach with a single Y-graft, and (**D**) the elephant trunk approach with a double Y-graft.

surgical exposure needed to repair the proximal and distal aorta differs. The decision whether to address the proximal or distal aorta first is based on symptoms, aneurysm size, and cardiac disease; the symptomatic segment is addressed first. Borst's elephant trunk procedure involves repairing the proximal aorta first, then performing a completion procedure to repair the distal portion of the aneurysm. During the first stage of the elephant trunk procedure, the distal anastomosis is constructed and a portion of the graft is left suspended within the proximal descending thoracic aorta (Fig. 61.4C). This "trunk" is then used during subsequent distal aortic reconstruction (Fig. 61.6) and offers many advantages, including easier dissection, shorter durations of aortic clamping, and safer clamping. Avoiding dissection around the distal arch reduces the chance of injury to the pulmonary artery and esophagus, which may be obscured by adhesions. Also, clamp times are usually shorter during the second stage because the proximal anastomosis is graft-to-graft, which makes hemostasis easier to achieve. Finally, because the aorta does not need to be clamped proximal to the LSCA, this technique reduces the risk of stroke from potential embolization of atherosclerotic debris.

An important change that improves the elephant trunk approach is the recent introduction of the collared or "skirted" aortic arch graft. The distal anastomosis of the aortic arch for the first stage of the elephant trunk procedure was traditionally done by invaginating the graft within itself,

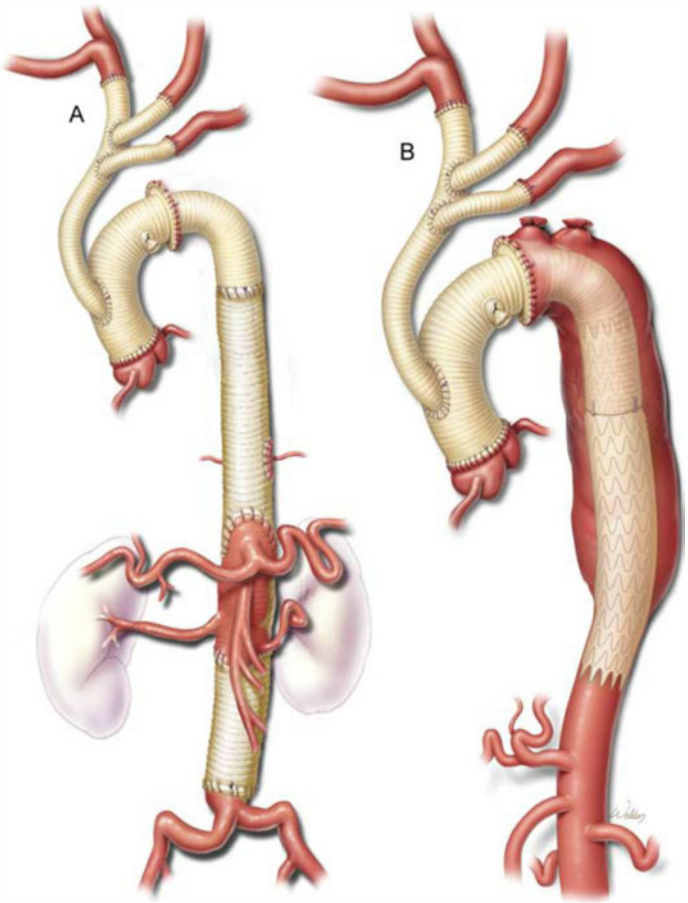


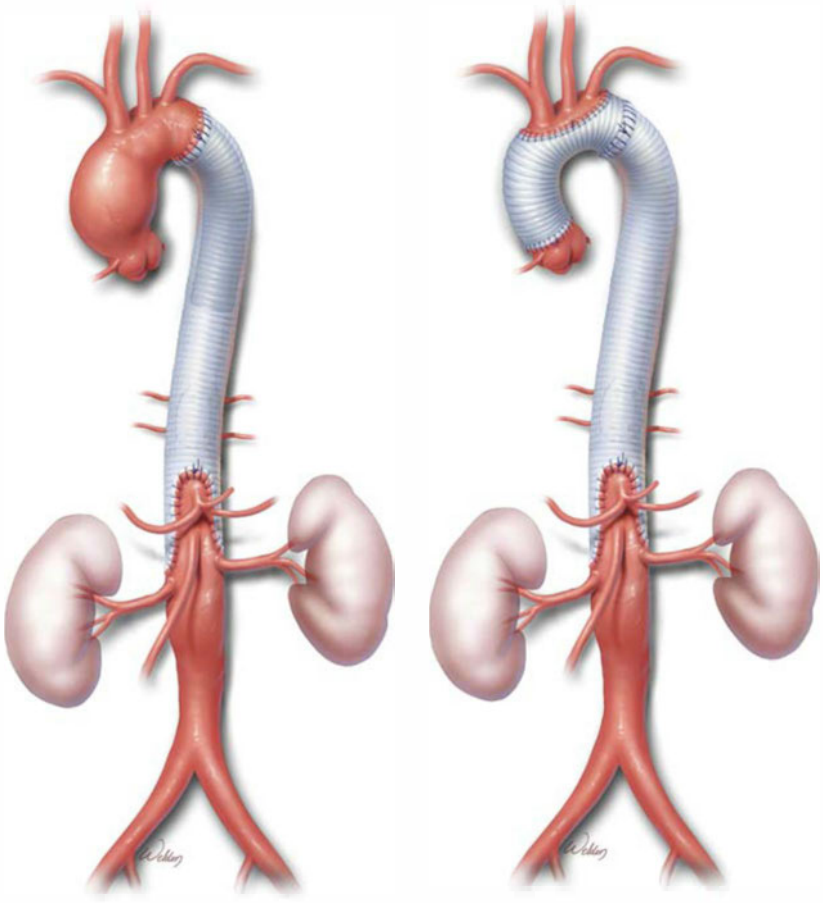
Fig. 61.6. Illustration of second-stage elephant trunk repairs. Completed (**A**) open and (**B**) endovascular completion repair in patients who underwent first-stage elephant trunk repair of the aortic arch by the Y-graft technique. (Used with permission from LeMaire SA, Price MD, Parenti JL, et al. Early outcomes after aortic arch replacement by using the Y-graft technique. Ann Thorac Surg 2011;91:700-707; discussion 707-708, figure 4. Copyright of The Society of Thoracic Surgeons.)

mimicking an intussusception. One of the problems of this technique was the substantial size discrepancy between the graft and the remaining diseased distal aorta. This size discrepancy could create undue tension on the anastomosis and result in bleeding or pseudoaneurysm formation at a later time. The collared elephant trunk graft facilitates a tension-free distal anastomosis by providing a graft collar that can be easily cut to match the distal aortic diameter; this helps to simplify the repair.

In patients with extensive aneurysmal disease in which the thoracic or thoracoabdominal aorta is symptomatic or substantially larger than the proximal aorta, the distal repair is often performed first with the "reverse elephant trunk" and followed by the second-stage replacement of the proximal aorta (Fig. 61.7). The advantage of the reverse elephant trunk technique is that it eliminates the distal anastomosis during the second stage of the repair. A drawback of open elephant trunk approaches is the inherent between-stage recovery period, during which the unrepaired portion of the aorta may rupture. In general, there is usually a 3- to 6-week interval between stages, although if the patient is symptomatic, this period may be shortened.

Extensive Single-Stage Repair

Alternatively, it is possible to replace the entire thoracic aorta during a single procedure. Kouchoukos and colleagues describe a single-stage repair for extensive disease involving the ascending aorta, the arch, and variable portions of the descending thoracic aorta; this repair involves performing a bilateral anterior thoracotomy with a transverse sternotomy (clamshell) incision. In this procedure, the arch anastomosis is done first during a short period of HCA in order to minimize the duration of cerebral ischemia.



A B

Fig. 61.7. Postoperative drawing illustrating (**A**) the completed thoracoabdominal repair with the invaginated reversed elephant trunk graft suspended in the proximal portion of the graft. The completion repair may be done in the traditional manner (**B**) or by using the Y-graft technique. (Used with permission from Coselli JS, LeMaire SA, Carter SA, et al. The reversed elephant trunk technique used for treatment of complex aneurysms of the entire thoracic aorta. Ann Thorac Surg 2005;80:2166-2172; discussion 2172, figures 4 and 8. Copyright of The Society of Thoracic Surgeons.)

Hybrid Arch Repair

The increasing interest in endovascular approaches to aortic arch repair has largely been driven by the need to reduce the high morbidity and mortality traditionally associated with open arch surgery and with the use of HCA. Such approaches are particularly attractive in high-risk patients with a poor likelihood of surviving a traditional open repair, either because of existing comorbidities (such as poor cardiac or pulmonary function) or emergent scenarios (such as rupture). Purely endovascular approaches to aortic arch repair are largely experimental and have limited applicability in day-to-day practice. Hybrid repairs, which are more accessible and increasingly widely used, involve "debranching" some or all of the brachiocephalic arteries, essentially lengthening the branchless aorta, and using an off-label application of standard "tube" stent grafts to exclude the diseased portion of the aortic arch.

The previously mentioned anatomic zones (Fig. 61.1) help to indicate the extent of hybrid arch repair. Zone 0 designates a repair that involves the entire aortic arch, necessitating the rerouting of all the brachiocephalic vessels because the stent graft will cover the origins of the innominate artery, LCCA, and LSCA (Fig. 61.8). When Zone 1 is planned as a landing zone, both the LCCA and the LSCA are rerouted, because their origins will be excluded by the stent graft.

These arch repairs are generally performed in specially designed hybrid operative suites that combine standard cardiothoracic operative equipment with specialty imaging devices. Hybrid arch repair typically involves performing a median sternotomy to provide sufficient exposure to reroute brachiocephalic vessels and to restore inflow from an anterolateral aspect of the ascending aorta, similar to the Y-graft techniques described previously. The debranching procedure may be done off-pump and without HCA, although in practice, a period of CPB and even HCA is not uncommon. For saccular aneurysms, hybrid arch repair is particularly attractive because the disease is localized, and healthy aortic tissue is usually readily available for adjacent proximal and distal landing zones.

Hybrid Elephant Trunk Repair

Hybrid elephant trunk repairs combine a standard open arch repair with an extended, distal endovascular repair (Fig. 61.6B). These repairs can be performed simultaneously with the open arch repair or at a later time. The primary benefits of hybrid elephant trunk repairs are that they are less

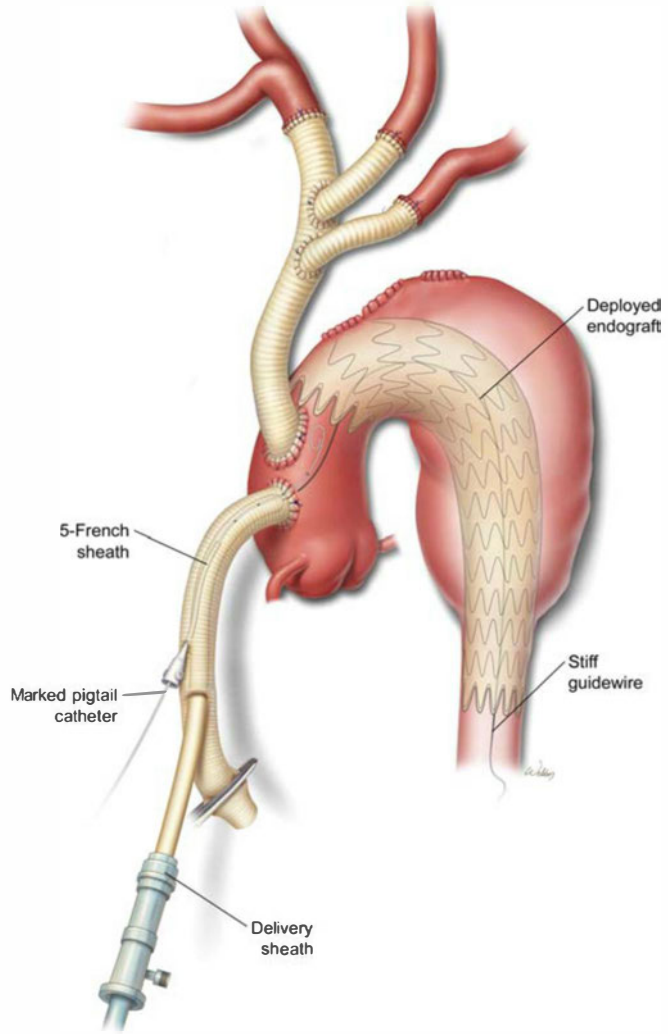


Fig. 61.8. A Zone 0 hybrid arch repair. This procedure is an option in select patients with limited arch disease. The brachiocephalic vessels are debranched onto a double Y-graft, and a separate graft is used as a conduit for antegrade endovascular deployment of the stent graft. (Used with permission from Baylor College of Medicine, Houston, TX.)

invasive than the traditional open elephant trunk completion procedure and that they can be performed sooner; by reducing or eliminating the interval between operations, the chance of between-stage death from rupture or other causes is reduced. Additionally, many patients are unwilling or unable to withstand a second open procedure, and a combined strategy allows for extended descending thoracic aortic coverage without requiring a second open procedure that involves a thoracotomy. Hybrid elephant trunk repairs are not suitable for patients with extensive distal aortic disease because endovascular approaches require secure landing zones, and it may not be possible to identify a suitable distal landing zone for the stent graft.

A modification of this approach is the frozen elephant trunk, in which the stent graft is deployed antegrade during the initial arch repair procedure. In the United States, this approach generally involves direct suturing to affix the stent graft to both the aortic wall and the replacement graft; this approach should minimize the risk of device migration by securing the endograft. Outside the United States, the frozen elephant trunk is a specialty device that has a Dacron-only section sutured to a stent graft. Spinal cord injury, as a result of ischemia, has been reported as an occasional complication of the frozen elephant trunk approach.

SURGICAL TECHNIQUES

Contemporary Approach to Aortic Arch Repair

Preparation for open repair of the transverse aortic arch includes placing a central line, a Swan–Ganz catheter, a left radial arterial line, and NIRS optodes for cerebral oxygenation monitoring. A standard median sternotomy is performed, and arterial inflow is established through the axillary or innominate artery. If the axillary artery is to be used, an incision is made in the deltopectoral groove. The pectoralis major muscle is spread along its fibers to expose the pectoralis minor muscle, which is then divided to expose the axillary artery. Care is taken to avoid the lateral and medial cords of the brachial plexus. A 3- to 4-cm segment of artery is freed, and a vessel loop is placed around it. A partial occluding clamp is placed on the artery after 5,000 units of heparin are given intravenously. An 8-mm Dacron graft is then anastomosed to the axillary artery with a running 6-0 polypropylene suture. The graft is clamped, the partially occluding clamp is removed, and the graft is attached to the arterial arm of the bypass circuit. Alternatively, the arterial cannulation can be achieved through an 8-mm graft anastomosed to the innominate artery by a similar process. Full-dose heparin is given before the cannula is attached to the CPB circuit. Venous drainage is achieved by placing a dual-stage venous cannula in the right atrium with its tip in the inferior vena cava. We routinely place a retrograde cardioplegia cannula in the coronary sinus and a sump drain in the left ventricle via the right superior pulmonary vein.

CPB is initiated and ventilation is discontinued once an adequate activated clotting time (>480 seconds) is achieved. The innominate artery is encircled with a tourniquet in preparation for snaring when CPB is discontinued. Once CPB has been initiated and the heart is fully decompressed, cooling can safely begin; the target temperature is 24°C (as measured with a nasopharyngeal probe). The head is packed in ice to enhance cerebral protection and prevent topical rewarming. During cooling, we give mannitol for cerebral protection, and we administer magnesium, lidocaine, and amiodarone to help maintain a sinus rhythm for as long as possible. Immediately after fibrillation (which usually occurs at approximately 30 to 32°C), retrograde cardioplegia is delivered into the coronary sinus. This induces diastolic arrest that is maintained by hypothermic conditions. Alternatively, if the aorta can be safely cross-clamped and the aortic valve is competent, cardioplegia can be given antegrade into the aortic root. Cardioplegia can also be administered directly into the coronary ostia once the aorta is opened under circulatory arrest. Cardioplegia is given about every 6 minutes thereafter.

If a hemiarch procedure is going to be performed, the patient is cooled to the target temperature before HCA is initiated. We now cool to a moderate temperature of 24°C; subsequently, CPB pump flows are brought down to about 10 ml/kg/min. Simultaneously, the Rumel tourniquet around the innominate artery is cinched down to allow perfusion of the brain through the right common carotid artery and to the right arm via the distal innominate or right axillary artery. The aorta is opened, and the arch vessels are visualized from within the opened arch. During this procedure, if NIRS monitoring indicates a significant drop in cerebral oxygenation from baseline, a perfusion catheter can be inserted directly into the LCCA. To incorporate the lesser curve of the arch, a beveled anastomosis is made with a running 3-0 or 4-0 polypropylene suture. The anastomosis may be reinforced with a second layer of running polypropylene suture or interrupted, pledgeted mattress sutures. After the distal anastomosis is completed, the innominate snare is released, CPB flows are increased, and the graft is deaired and then clamped. The proximal aspect of the repair is then performed as the patient is slowly rewarmed while receiving full CPB flows.

If the entire arch is to be repaired, we now generally use a Y-graft approach (Fig. 61.9). While the patient is being cooled

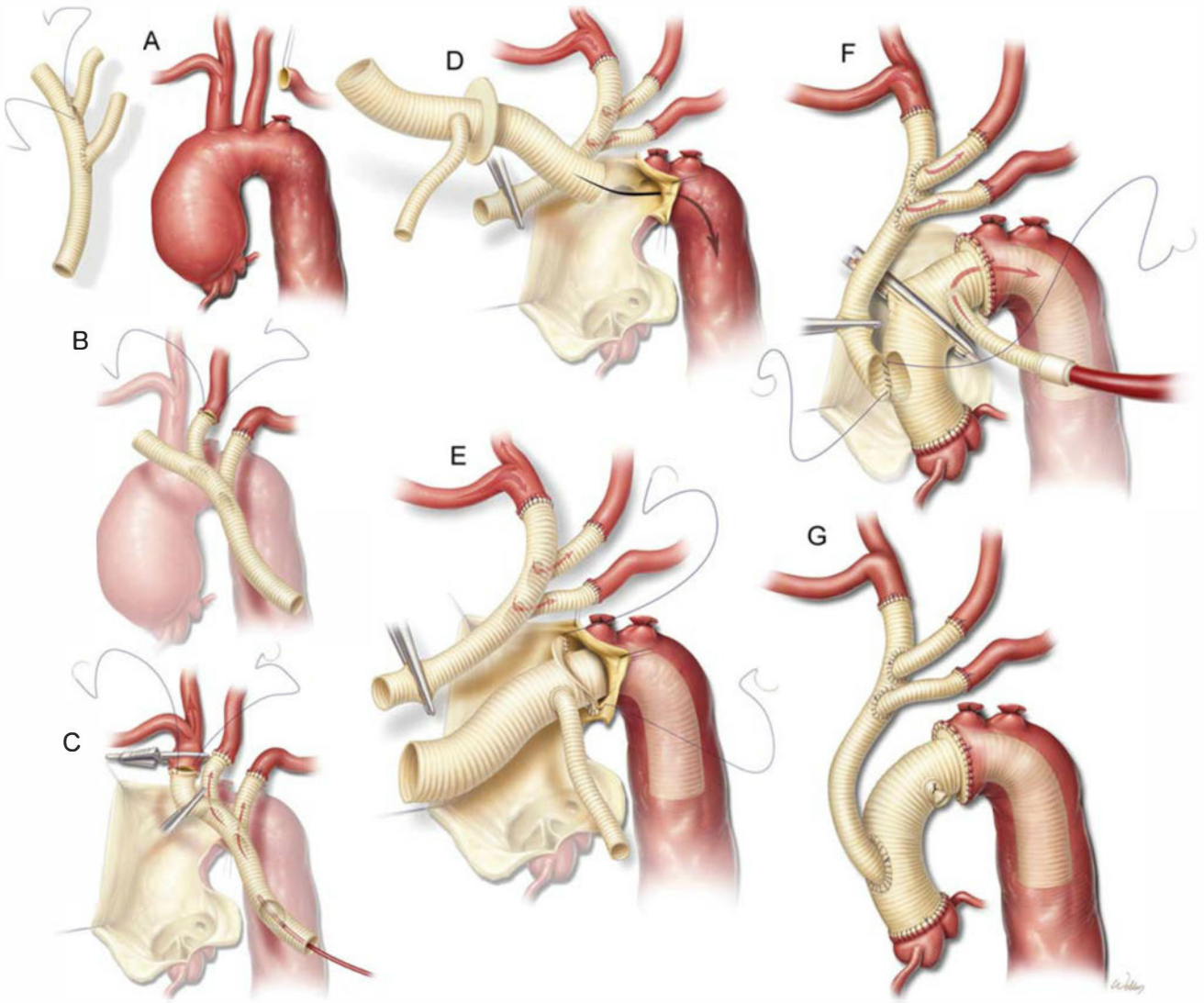


Fig. 61.9. Drawings illustrating the Y-graft technique. (**A**) The proximal portions of the brachiocephalic arteries are exposed, and a double Y-graft is created by suturing two side grafts to a main graft at 45-degree angles. (**B**) The first two branches of the graft are sewn end-to-end to the transected left subclavian and left common carotid arteries. The proximal ends of the transected brachiocephalic arteries are ligated. (**C**) A balloon-tipped perfusion cannula is placed inside the double Y-graft and used to deliver antegrade cerebral perfusion. After systemic circulatory arrest is initiated, the innominate artery is clamped, transected, and sewn to the distal end of the main graft. (**D**) A clamp is placed on the proximal aspect of the Y-graft, thereby directing flow from the axillary artery to all three brachiocephalic arteries. The arch is then replaced with a collared elephant trunk graft. (**E**) The distal anastomosis between the elephant trunk graft and the aorta is created between the innominate and left common carotid arteries. (**F**) The aortic graft is clamped, and a second limb from the arterial inflow tubing of the cardiopulmonary bypass circuit is used to deliver systemic perfusion through a side branch of the arch graft while the proximal portion of the ascending aorta is replaced. Once the proximal aortic anastomosis is finished, the main trunk of the double Y-graft is cut to an appropriate length, and the beveled end is then sewn to an oval opening created in the right anterolateral aspect of the ascending aortic graft, which completes the repair (**G**). (Used with permission from LeMaire SA, Price MD, Parenti JL, et al. Early outcomes after aortic arch replacement by using the Y-graft technique. Ann Thorac Surg 2011;91:700-707; discussion 707-708, figure 2. Copyright of The Society of Thoracic Surgeons.)

to a moderate target of 24°C, the Y-graft debranching process is begun. The proximal aspects of the brachiocephalic vessels are exposed. Next, the LSCA is ligated near its origin with a heavy silk tie and transected. One of the branches of the Y-graft is then sewn end-to-end to the LSCA with a 5-0 or 6-0 polypropylene suture. In the same manner, the LCCA is ligated and transected, and the distal end is sewn end-to-end to another side branch of the Y-graft. A single Y-graft approach may be used if the distal portion of the arch is not aneurysmal; in this case, only the LCCA is anastomosed to the Y-graft. The graft is then deaired and clamped. Once HCA is achieved, the arch is opened, the proximal portion of the innominate artery is ligated, and the distal end of the innominate artery is anastomosed end-to-end to the main trunk of the Y-graft. During the innominate artery anastomosis, if NIRS monitoring shows a significant drop in cerebral oxygenation from baseline, a perfusion catheter can be inserted directly into the Y-graft to perfuse the LCCA. Once the innominate anastomosis has been performed, the Rumel tourniquet around the innominate artery is removed. The Y-graft is deaired and clamped proximally to maintain flow from the axillary artery to the two or three previously anastomosed brachiocephalic arteries while the aortic graft is placed.

Next, the distal aortic anastomosis is performed. When the descending thoracic aorta is not aneurysmal, a straight or curved graft may be used to complete an end-to-end distal anastomosis. More commonly, the aneurysmal disease extends into the descending thoracic aorta, and a collared elephant trunk graft is used to perform the distal anastomosis. Because the aortic arch has already been debranched, this distal aortic anastomosis can be performed more proximally within the boundaries of the transverse aortic arch (proximal to the original anatomic location of the LSCA or LCCA), rather than at the traditional location just beyond the LSCA. This anastomosis is performed with either a 3-0 or a 4-0 polypropylene suture in a continuous manner. Often, this distal anastomosis is then reinforced with either another layer of running suture or pledgeted mattress sutures. If it appears that the residual descending thoracic aortic aneurysm is amenable to subsequent endovascular repair, metallic clips are placed on the distal edge of the elephant trunk graft to facilitate later stent-graft delivery by aiding fluoroscopic visualization. Once the distal anastomosis is complete, the graft is deaired, a clamp is applied to the proximal portion of the graft, and distal systemic perfusion is reestablished via a side branch (usually 8 mm) of the collared elephant trunk graft. Full CPB is reestablished via the axillary artery perfusion cannula and the side branch of the elephant trunk graft.

Rewarming to 36.5°C can be initiated immediately after CPB is restarted, but it is occasionally prudent to delay rewarming if extensive reconstruction is required proximally, such as repair involving the aortic valve or root. The difference between perfusate and core body temperature is kept to $<$10°C until a core body temperature of 27°C is achieved, after which the perfusate is kept at 37°C for the remainder of rewarming.

During rewarming, the proximal aortic anastomosis is completed, often near the level of the sinotubular junction. After the main trunk of the Y-graft is cut to an appropriate length, the proximal end is anastomosed to a small opening made on the right anterolateral aspect of the ascending aortic graft (proximal to the aortic cross-clamp) in an end-to-side manner with a 4-0 or 5-0 polypropylene suture. The length and orientation of the Y-graft are carefully planned to prevent any kinking of the graft when it is pressurized and compressed by the closed sternum.

The graft is deaired and the aortic cross-clamp is released. The retrograde cardioplegia catheter is removed. Temporary pacing wires are placed. The patient is separated from bypass as soon as the physiologic temperature is reached and appropriate criteria are met. When it is time to remove the arterial cannula, the side arm of the graft is clamped and divided. A heavy silk suture is used to tie off the side arm graft flush with the aortic graft, and a suture ligature is used to definitively obliterate the side arm lumen. If the side arm is not tied flush with the graft, contrast may enter the residual side arm during future computed tomography scans, and this leakage might be interpreted as an anastomotic leak or rupture.

Elephant Trunk Completion

The second portion of the elephant trunk procedure may be performed with open or endovascular techniques (see section Hybrid Elephant Trunk) (Fig. 61.6). Our technique for distal aortic repair is more fully described elsewhere. In brief, our approach is based on the distal extent of repair, and additional surgical adjuncts are used for more extensive repairs (such as Crawford extent I and II thoracoabdominal aortic repairs). We begin by placing a double-lumen endobronchial tube and monitoring arterial pressure with right radial and femoral arterial lines. For extensive distal aortic repairs, a cerebrospinal drain is placed in the intervertebral space at L3–L4 or L4–L5. The patient is then positioned on the right side with the thorax at a 75-degree angle to the bed and the hips at a 45-degree angle to allow the exposure of the vessels in the groin in the event that they are needed to establish CPB.

A posterolateral thoracotomy incision is made in the fifth or sixth interspace. If the aneurysm extends below the diaphragm, the incision is extended toward the umbilicus in a curvilinear manner. We do not remove a rib, and we only occasionally need to divide the posterior aspect of a rib to complete an elephant trunk procedure. The costal margin and rectus abdominis muscle are divided, connecting the thoracotomy to the abdominal incisions. The retroperitoneal reflection is dissected with electrocautery; this mobilizes the spleen, descending colon, and left kidney, thereby exposing the abdominal aorta. An arcuate incision is made in the diaphragm, staying approximately 1 to 2 cm from the tendinous portion, where the neurovascular bundle runs, and leaving a 3- to 4-cm margin attached to the chest wall to facilitate later closure with No. 1 polypropylene suture. Self-retaining retractors on the upper and lower ribs, and gentle reflection of the abdominal contents to the right by the surgical assistant, provide stable exposure of the thoracoabdominal aorta.

If the aneurysm is less extensive and a short cross-clamp time is anticipated, an expeditious clamp-and-sew technique can be performed. Otherwise, left heart bypass is used with outflow from the left pulmonary vein and inflow to the distal descending thoracic aorta. A heparin dose of 1 mg/kg is given, and the pericardium overlying the left inferior pulmonary artery is opened. A pledgeted 4-0 polypropylene purse-string suture is placed in the pulmonary vein, which is then cannulated. The aorta is cannulated distally in the descending thoracic portion, or in the abdominal portion in the case of thoracoabdominal aneurysms. Flow is initially run at 500 ml/min until the cross-clamp is applied, after which flows are adjusted (usually to 1.5 to 2.0 L/min) to maintain a right radial mean arterial pressure of 60 to 70 mmHg. The aorta is clamped in a place that includes the free-floating arm of the elephant trunk graft. Because the graft should extend some distance into the descending aorta, the clamp does not have to be applied flush against the arch.

A second clamp is placed on the aorta proximal to the arterial cannula. The isolated segment of aneurysmal aorta is opened, and the free-floating portion of the graft is retrieved and clamped. The first clamp is removed, which allows the graft to be stretched inferiorly because its corrugated structure gives it a tendency to shorten.

An appropriately sized graft is anastomosed to the free-floating arm of the elephant trunk graft with 2-0 or 3-0 polypropylene suture. It is not necessary to routinely reinforce this anastomosis because the graft-to-graft connection is quite hemostatic. Left heart bypass is discontinued, and the arterial cannula is removed. The distal clamp is also removed, and back-bleeding from the aorta is dealt with by suctioning blood to a cell-saver system. The remainder of the aneurysmal aortic segment is opened. Intercostal vessels with vigorous back-bleeding are oversewn. Depending on the extent of the aneurysm, one may need to reattach the intercostal arteries, visceral branches, or both to the graft in addition to performing the distal anastomosis.

The graft is then cut to size, and the distal anastomosis is performed with running 3-0 or 4-0 polypropylene suture. During this time, the anesthesiologist is alerted a few minutes before the clamp is removed so that appropriate adjustments can be made to preload, afterload, contractility, and acid–base status. The patient is placed head-down, and the aortic clamp is slowly removed. The cannula in the pulmonary vein is removed, protamine is given, and meticulous care is taken to achieve a hemostatic field, after which the wound is closed.

Reverse Elephant Trunk

In the first stage of the repair, the descending thoracic or thoracoabdominal aorta is replaced by using our standard techniques, with one important difference: the proximal anastomosis is constructed with approximately 10 cm of graft invaginated within itself (Fig. 61.7). This folded edge of the invaginated graft is sewn end-to-end to the aorta with 3-0 or 4-0 polypropylene.

The second portion of the repair is performed after an appropriate convalescence (ideally about 4 weeks) in a manner similar to that described above for aortic arch repair. This is done via a median sternotomy after HCA and ACP are established. The Y-graft technique is used to anastomose the brachiocephalic vessels. Forceps are then used to reach into the aorta beyond the arch to pull the invaginated portion of the previously placed graft from within itself. The proximal portion of the graft is then anastomosed to the ascending aorta. Finally, the main trunk of the Y-graft is sutured to an oval opening in the ascending aortic graft.

Hybrid Arch Repair—Zone 0

This repair can be performed without CPB. The chest is entered through a median sternotomy, and heparin (1 mg/kg) is administered. With a side-biting, partially occluding clamp, a small anterolateral portion of the ascending aorta is isolated; care is taken to ensure that this site is proximal to the proposed endovascular landing zone. The main branch of a Y-graft is anastomosed end-to-side to a small opening in the isolated portion of the ascending aorta with continuous 4-0 polypropylene suture. The side-biting clamp is then removed, and a straight clamp is placed on a proximal section of the Y-graft trunk. The innominate artery is clamped and divided, and its proximal aspect is then oversewn near its origin with 3-0 polypropylene suture; the artery is then anastomosed to the distal aspect of the Y-graft main trunk with continuous 5-0 polypropylene suture in an end-to-end fashion. The LSCA is then divided and oversewn at its origin with 3-0 polypropylene suture; subsequently, its distal transected end is anastomosed end-to-end to one of the side branches of the Y-graft with continuous 5-0 polypropylene suture. Similarly, the LCCA is divided and oversewn at its origin with 3-0 polypropylene suture, and its distal transected end is anastomosed end-to-end to the remaining side branch of the Y-graft with continuous 5-0 polypropylene suture. As each brachiocephalic artery is rerouted, clamps are repositioned to restore circulation expeditiously.

After the aortic arch has been debranched, a 10-mm graft is anastomosed with continuous 4-0 polypropylene suture in an end-to-side manner to an anterolateral portion of the ascending aorta isolated by a side-biting, partially occluding clamp. This graft will serve as a conduit for antegrade deployment of one or more stent grafts, depending on the extent of the aneurysm. Through this conduit, a pigtail catheter is inserted through a guide wire so that the adequacy of the arch debranching process can be assessed aortographically. Then, a 24-F sheath is inserted through the conduit over a stiff guide wire, and one or more stent grafts (the number depends on the extent of aortic disease) are advanced into the aortic arch and descending thoracic aorta. Care is taken to ensure that the Y-graft inflow from the proximal ascending aorta is not occluded by the proximal stent (Fig. 61.8). The stent grafts are often ballooned to ensure an adequate seal at the proximal and distal landing zones, as well as at any overlapping sections; ballooning is not performed if there is an aortic dissection. A completion arteriogram is then performed to ensure that the aneurysm has been excluded, that there are no endoleaks, and that no suture lines have been disrupted during this process. The 10-mm graft is oversewn at its base with 3-0 polypropylene suture, because leaving any stump could confound later imaging studies or cause thrombus formation with the potential for distal embolization. Protamine sulfate is then given to reverse the effects of heparin, tubes are placed for drainage, and the chest is closed.

Hybrid Elephant Trunk

The first part of the procedure is similar to our method of total aortic arch replacement with the Y-graft technique and the collared elephant trunk graft approach (see sections Options for Repair and Y-Graft Techniques). During the cooling phase of CPB (to 24°C), the brachiocephalic arteries are debranched in an end-to-end manner onto a Y-graft (a 10- or 12-mm main trunk and two 8- or 10-mm side branches), which is then used to provide ACP. The aortic arch is transected at the level of the LCCA, and a collared elephant trunk graft is distally anastomosed with 2-0 polypropylene suture in a running, continuous manner. A second suture of 3-0 polypropylene is run around the anastomosis for reinforcement. Under direct vision, the stent graft is deployed antegrade through this graft over a stiff Amplatz wire to cover the proximal aspect of the descending thoracic aorta. One or more stent grafts may be deployed at this time, and if necessary, these stent grafts are overlapped by at least 2 cm. Often, the endograft is then ballooned to create an adequate seal (this step may be omitted in cases of aortic dissection) and may be secured at the proximal landing zone to both the collared elephant trunk graft and the aortic wall by placing pledgeted 3-0 polypropylene sutures in a mattress manner. The collared elephant trunk graft is then deaired, and the cross-clamp is applied to its proximal portion. Full CPB is reestablished via the 8-mm perfusion graft in the right axillary or innominate artery and the 8-mm side branch of the collared elephant trunk graft. The proximal portion of the aortic repair is then carried out while the patient is rewarmed back to physiologic temperature. The main trunk of the Y-graft is anastomosed to an opening in the right anterolateral section of the ascending aortic graft in an end-to-side manner with continuous 4-0 polypropylene suture (Fig. 61.6B). Deairing is done through the anastomosis and subsequently via a 19-gauge needle placed in the ascending aortic graft before the aortic cross-clamp is released. The retrograde cardioplegia cannula is removed, a temporary right ventricular pacing wire is

placed, and once the appropriate temperature is reached (36.5°C) and deairing is completed, the patient is weaned from CPB.

Alternative two-stage approaches involve advancing the stent graft in an antegrade manner from a brachial artery or advancing the wire in a traditional retrograde manner from the femoral artery into the existing elephant trunk. Often, metallic clips are placed on the distal edge of the elephant trunk graft to facilitate radiographic visualization during the second-stage repair.

OUTCOMES

Aortic arch repair carries substantial risks—primarily the risks of operative death or stroke. The prolonged use of CPB and HCA has been long associated with increased mortality, neurologic morbidity, and other complications. Furthermore, certain patient-specific characteristics (such as acute aortic dissection, previous median sternotomy, urgency of repair, an incomplete circle of Willis, and a history of stroke) may enhance operative risk. Although it is difficult to interpret outcomes across these various approaches, results appear to be generally improving, particularly for elective repair of the aortic arch in patients without aortic dissection. In general, less extensive open aortic arch repair (such as the hemiarch approach) tends to carry less risk than extended arch repair (such as elephant trunk approaches). In very high-risk patients, a hybrid approach has been advocated as an effective alternative to open repair, because this approach is believed to cause fewer cardiac, pulmonary, and other complications, as well as less short-term mortality and morbidity. The primary drawbacks of hybrid approaches appear to be the increased risk of stroke (due to endovascular manipulation within the aortic arch) and the increased risk of spinal cord ischemia (when repairs extend into the descending thoracic aorta), as well as uncertain durability.

Our contemporary experience regarding aortic arch repair in 243 patients is presented in Table 61.1 and includes outcomes for traditional partial-arch approaches (such as the hemiarch approach), traditional full-arch approaches, and our initial use of the Y-graft approach. Many of the patients had conditions that tend to enhance operative risk: 83 patients (34%) had a prior median sternotomy and 91 (37%) had acute dissection of the ascending aorta. Many of these arch repairs involved concomitant repair of the aortic valve (148 patients, 61%). We have been gradually shifting toward more moderate HCA temperatures, and in this series, the median lowest nasopharyngeal temperature was 22.0°C for patients who underwent the arch repair with the Y-graft approach, 19.4°C for the traditional partial-arch approach, and 16.3°C for the traditional full-arch approach.

Table 61.1 Outcomes in Contemporary Aortic Arch Repair by Operative Approach

Operative approach	Patients, *n*	Operative survival, *n* (%)	Stroke, *n* (%)	Renal failure, *n* (%)	Reoperation for bleeding, *n* (%)
Traditional partial arch replacement	165	155 (94%)	7 (4%)	4 (2%)	9 (5%)
Traditional total arch replacement	23	22 (96%)	3 (13%)	1 (4%)	3 (13%)
Y-graft technique	55	54 (98%)	3 (5%)	3 (5%)	4 (7%)
Total	243	231 (95%)	13 (5%)	8 (3%)	16 (7%)

Renal failure is defined as the need for dialysis at time of hospital discharge.

FINAL COMMENTS

Approaches to transverse aortic arch repair are becoming increasingly diverse, and over the past several years, our approach to these operations has gradually changed, largely through the introduction of various techniques aimed at reducing the risk of neurologic complications. The selection of one approach over another should be based on the extent of aortic disease, as well as patient-specific comorbidities such as advanced age, severe atherosclerosis, chronic renal dysfunction, and preexisting cardiac dysfunction. Regardless of the approach used for transverse aortic arch repair, postoperatively, all patients should be routinely assessed for late complications, including the development of pseudoaneurysms, patch aneurysms, and new aneurysms in other sections of the aorta. This is particularly important in hybrid arch repairs, because these are off-label applications of endovascular devices, and the durability of these repairs is unknown.

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EDITOR'S COMMENTS

Dr. Coselli and colleagues probably have the largest experience in the United States with arch aneurysms. They present an erudite approach to this complex disease process. I believe these are some of the most difficult procedures in the adult cardiac surgery but I have to say that a hybrid approach using TEVAR, which has simplified some of the procedures.

Many patients can be treated with a hemiarch procedure and I agree entirely with Dr. Coselli that hypothermia to the 24°C to 25°C level is all that is needed in these situations. More complex arch aneurysm repairs can be done with traditional open techniques and direct anastomosis to the branch vessels as Dr. Coselli suggested.

The combination of hybrid approaches, including direct revascularization of the arch vessels through a sternotomy followed by TEVAR, is a very attractive approach in much more complex or difficult situations. I believe that this technology is continuing to change, and a procedure that used to be greatly feared now can be done electively with relatively low risk of mortality or stroke.

ILK

62 Acute Traumatic Aortic Transection

Benjamin J. Pearce, Jay D. Pal, and John H. Calhoon

Acute traumatic aortic transection (TAT) remains a devastating complication of blunt trauma with a mortality of 80% at the scene and is the second leading cause of death from blunt injuries. Further, up to half of the patients surviving to hospital admission perish within the first 4 hours. Due to the mechanism of injury, TAT is rarely an isolated finding and confounding extrathoracic injuries complicate both the diagnosis and management strategy. A multidisciplinary approach including trauma services, critical care management, radiology, and the cardiovascular surgeon is critical to assessing the spectrum of injuries and decisively executing a therapeutic plan.

Increasing sophistication of trauma systems and the introduction of multidetector computed tomographic angiography (CTA) have led to the increase in both the initial survival of TAT patients to hospital admission and diagnosis of aortic pathology. The ability to obtain quick, high-resolution imaging with multiplane reconstruction through CTA has replaced traditional angiography in the diagnosis of TAT. In addition, the liberal application of computed tomography scanning has allowed for better assessment of concurrent injuries and allowed for appropriate planning of either an open or endovascular surgical approach. Further, documented decrease in rupture rates with the administration of anti-impulse therapy has allowed for more patients to reach the operating room and a small subset of patients to be treated without surgery.

Despite these advances, TAT continues to be a highly lethal entity. Operative mortality approaches 25% in recent American Association for the Surgery of Trauma (AAST) prospective trials on management of TAT. Data are promising for the ability of thoracic endovascular aortic repair (TEVAR) to improve outcomes in this population; however, the applicability of this technology to all patients is controversial and still limited by anatomic and device-specific constraints. Continued experience in open surgical repair (OSR) for TAT is critical in approaching these complex patients. The most efficacious approach to management will be appropriate selection of both the timing and technique of intervention.

NATURAL HISTORY AND PATHOLOGIC CONSIDERATIONS

Eighty percent of acute TAT occur as a result of motor vehicle collisions. Other causes include kicks, falls from heights, and crush injuries. Most series report that 20% of motor vehicular accident fatalities are a result of TAT. The incidence is higher in the young adult population and has an increased male-to-female ratio. More than one half of the transections occur at the aortic isthmus, 25% can occur in the arch and great vessels, and the remaining throughout the descending and abdominal aorta. Recent pooled data of OSR only trials for meta-analysis reported the need for open arch reconstruction in TAT at 4.1%. This was associated with significantly elevated mortality. Similarly, concomitant repair of injury at the isthmus and abdominal aortic branches has increased mortality and morbidity, likely due to the greater force and associated injuries needed to create this spectrum of disease.

Multiple mechanisms of injury have been reported by autopsy study and ex vivo mechanical testing of the tensile strength of the aorta. The most commonly accepted theory is the sudden movement of the heart and arch against the fixed position of the descending thoracic aorta. In this deceleration model, the greatest tension point is the aortic isthmus, resulting in the spectrum of injuries in TAT. Other proposed mechanisms include a sudden rise in blood pressure, high-pressure reflected wave from sudden occlusion of the aorta at the diaphragmatic hiatus, and stretching or "pinching" of the aorta by the osseous structures in the thorax. Regardless of mechanism, the natural history of TAT is such that immediate death was historically reported in 80% to 90% of cases. In a contemporary autopsy study of 242 cases of fatal TAT, Burkhart et al. (*J Trauma* 2001) published immediate mortality in 57%, 37% within 4 hours of admission, and only 6% who died >4 hours after hospital arrival.

A basic understanding of the underlying pathology can also greatly aid in diagnosis and decision making regarding surgical timing. TAT is a continuum of disease spanning from subintimal hemorrhage to complete transection, and extent of injury likely plays the ultimate role in survival. Parmley et al. (*Circulation* 1958) classified the injuries as intimal hemorrhage only, intimal hemorrhage with laceration, medial laceration, complete laceration of all layers, pseudoaneurysm formation, and free aortic hemorrhage. The fine detail provided by CTA allows for classification of these injuries along this spectrum at diagnosis.

DIAGNOSIS

With a history of any deceleration injury, there should be a suspicion for possible acute aortic transection. Symptoms on arrival at the emergency room in a conscious patient range from nonspecific complaints to those of chest pain, intrascapular pain, hoarseness, dyspnea, dysphasia, and frank paralysis. Physical examination findings include rib fractures, sternal fractures, and even imprints of the steering wheel on the chest. The cervical and thoracic spines are also vulnerable, and these should be carefully examined. Hypertension in the arms, so-called coarctation syndrome, has been reported. It is important to carefully examine the pulses in the upper and lower extremities and check for any differences. A baseline neurologic examination is extremely important in these patients because paraplegia may be present preoperatively and may be due to other causes such as spinal fractures and dislocations.

In the era of liberal CT scanning, chest roentogram has a less important role, but diligent examination is still warranted,

especially in patients with otherwise underwhelming presentation. An initial chest radiograph should be obtained as soon as the patient's cervical spine is cleared. An upright posteroanterior (PA) film is desirable; however, the most upright anterior–posterior (AP) film possible is the best alternative. Supine chest radiographs usually reveal a widened mediastinum. Any abnormality seen in the mediastinum on an upright PA chest film should raise the suspicion for aortic transection and prompt further investigation. Various specific radiographic findings associated with aortic transection include mediastinal widening, obscurity of the aortic knob, obliteration of the AP window, tracheal deviation, depression of the left mainstem bronchus, widening of the paravertebral stripe, deviation of the esophagus (seen most commonly as deviation of the nasogastric tube), and a left hemothorax. Fractures of the first rib and/or scapula should raise the suspicion of an aortic injury due to the extreme force required to fracture these bones. A history of a deceleration injury in conjunction with an abnormal mediastinum seen on the chest radiograph should prompt further investigation.

CTA has replaced arch angiography as the modality of choice in investigating blunt aortic injury (Fig. 62.1). Magnetic resonance imaging (MRI) has also been advocated, especially considering the physiologic information which can be gained from functional MRI; however, it is often too cumbersome to obtain in the acute trauma situation. Transesophageal echocardiography (TEE) has been shown to be a useful diagnostic tool, but this test may not be readily available in all institutions, and its accuracy is operator anatomy dependent. With TEE, there is also a risk of free aortic rupture due to patient gagging or Valsalva from inadequate sedation causing increase in intrathoracic pressure. In our experience, TEE has been most useful when the patient was sent to the operating room emergently for other life-threatening injuries and there remains a question regarding the diagnosis of acute TAT. Intraoperatively, TEE is useful in helping to rule in or out aortic injury. Operative exploration is now justified without the use of arteriography in the acute setting with a suggestive clinical picture along with other diagnostic radiologic findings such as that seen on plain radiographs, CT scans, and intraoperative TEE findings. Associated injuries are common, and acute TAT is rarely seen as an isolated entity. Injuries to other organ systems (intracranial, intra-abdominal), including major fractures (spine, pelvis), must be considered and treated if a successful outcome is to be obtained. When associated injuries are present, the cardiovascular surgeon should be involved with the treatment and prioritization of these injuries.



A



B

Fig. 62.1. (**A**) Preoperative CTA demonstrating traumatic PSA of aorta at the level of LSCA. (**B**) Intraoperative angiogram demonstrating patent subclavian and vertebral transposition and stent-graft placement in the same patient. CTA, computed tomographic angiography; PSA, pseudoaneurysm; LSCA, left subclavian artery.

Three recent papers have attempted to grade CT findings of TAT along the lines of pathologic injury. The Simplest of these is the Vancouver simplified grading systems described by Lamarche et al. (*J Thorac Cardiovasc Surg* 2011). In order from least to most severe: Grade I is defined as an intimal flap/thrombus/hematoma <1 cm in length; Grade II as intimal flap/thrombus/hematoma >1 cm; Grade III pseudoaneurysm formation; and Grade IV free contrast extravasation. Furthermore, the authors correlated these findings to outcomes in a small subset of patients, advocating observation for all Grade I and some Grade II injuries with no mortality. Grade III injuries were treated with intervention and hospital survival in 90% and Grade IV with only 33% survival despite intervention. These favorable data can be used to guide operative decision making.

CTA can be both diagnostic and used for planning of operative intervention. Sizing of landing zones for TEVAR, presence of anomalous great vessel anatomy, patency of vertebral circulation, evidence of prior coronary grafting from internal mammary, angulation of aortic arch, and presence of other injuries can all be assessed with appropriate CTA. When CTA is being performed for suspected TAT, it is advantageous to scan the entire vascular tree from neck to femoral vessels. This allows for assessment of access vessels

in the event of possible TEVAR without adding to the contrast administration. One must consider that in younger patients with compliant vessels, intake CT scans may underestimate the actual size of the aorta and iliac system. Several millimeters of variance have been described in the under-resuscitated state. Furthermore, the more dynamic pulsation of the aorta in a young patient can be misrepresented on a static scan. This can lead to inappropriate oversizing of an endograft as will be described later. In patients with a documented anaphylactic reaction to iodine, or concern for renal failure, a noncontrasted thin cut CT scan can still be a useful adjunct in evaluating TAT and for operative planning and can be augmented by other modalities such as TEE and intravascular ultrasound (IVUS).

MEDICAL THERAPY

The use of anti-impulse therapy was first applied in the management of descending thoracic dissection and has expanded to use in the management of acute traumatic aortic injuries. This therapy decreases the wall stress tension in the aorta by means of beta-blockade and, if necessary, vasodilating agents. Prospective studies have demonstrated a decrease in the incidence of spontaneous rupture with the institution of appropriate anti-impulse therapy. This is an important aspect in the management of multiply injured patients where other life-threatening injuries need to be addressed first, prior to aortic repair.

The goals of therapy are to reduce systolic blood pressure to $<$100 mmHg. This is best achieved with a combination of pain control, inotropic/chronotropic blockade, and afterload reduction. Often overlooked is the importance of adequate analgesia. The contribution of acute pain to the cascade of catecholamine release is significant and appropriate treatment with intravenous opiates is warranted. One of the most feared complications of TAT is paraplegia from spinal cord infarction either spontaneously or from treatment. As such, the use of intrathecal analgesia is contraindicated.

Initiation of therapy with beta-blockade is the standard of care. The goal heart rate is $<$80 bpm. By initiating beta-blockade early, reflex tachycardia and catecholamine release are blunted with the initiation of afterload reducing agents. Choice of a beta-blocker, for example labetalol, with α and β adrenergic effects may achieve appropriate cardiac and peripheral effects with a single agent. However, this may precipitate pulmonary complications in at-risk patients and thus initiation of therapy with a short acting agent, for example esmolol, may be more prudent in selected cases. Initiation of sodium nitroprusside infusion after beta-blockade has been an effective agent. However, recent data have shown similar efficacy of nicardipine infusion in treatment of acute type B dissection without the risk of cyanide toxicity and may be applied to TAT.

TIMING OF REPAIR

While expectant management of a potentially fatal injury pattern can be anxiety provoking, the scope of injuries in the TAT patient often makes this management advantageous. Perceived advantages of delayed repair include allowing for appropriate resuscitation, time for full evaluation and treatment of other life-threatening injuries, stabilization of intracranial or solid-organ injuries, which may potentially hemorrhage with heparinization, improvement in pulmonary mechanics in patients requiring significant ventilator support, correction of hypothermia and coagulopathy, and allow for proper sizing and acquisition of endografts if TEVAR is planned.

An inherent bias exists in the literature regarding this issue. Intuitively, the patients with the most severe grade of TAT will necessarily require a more immediate operation and have expected worse outcomes. The AAST evaluated this issue in a prospective manner. Two major groups were examined—those with and without major extrathoracic injuries. In the group with major extrathoracic injuries, the mortality in the early repair group was 21.6% versus 3.2% in the delayed repair group. Even when adjusting for severity of injuries and modality of repair in a regression analysis, this trend toward improved overall survival in delayed repair was evident. A similar trend was described in patients with no other associated injuries. The authors in the AAST advocate delayed repair when feasible in all comers, although it is more difficult to justify observation of a potentially fatal injury in an otherwise stable patient based on these data alone. Conversely, a recent consensus statement from the Society of Vascular Surgery states that most patients should be repaired within 24 hours when feasible. Careful consideration of the entire clinical picture combined with appropriate grading of the injury on CTA will lead to the most judicious timing of intervention.

CHOICE OF MODALITY

TEVAR for TAT has rapidly gained acceptance as an alternative to OSR. In fact, in a recent meta-analysis of OSR versus TEVAR for TAT, TEVAR was performed in the majority of cases (60%). There are currently several device-specific trials attempting to gain Federal Drug Administration (FDA) indication for TAT; however, it must be mentioned that this remains an off-label therapy for commercially available grafts in the United States at this time.

The perceived advantages of TEVAR in other aortic pathologies also make it an attractive option in TAT as well. The devices can be delivered from remote sites without the need for a major cavity operation in patients who may have associate injuries in these areas. In addition, heparin administration is significantly less than that needed for cardiopulmonary bypass, and TEVAR can be performed under regional anesthesia in many cases. No aortic cross-clamping is needed attenuating the hemodynamic effects on both the heart and distal vasculature. Reported rates of spinal cord infarction are significantly less with TEVAR than OSR, and paraplegia can often be managed expectantly without the need of prophylactic spinal drainage.

However, as with any new technology, TEVAR has developed a new set of complications. Rupture of access vessels due to the large-bore devices was common in early experiences and still remains an important consideration in operative planning. Careful attention to evidence of anomalous arch anatomy or prior coronary bypass is crucial to prevent coverage of vital branch vessels. Proper oversizing of the endograft is difficult in the young trauma patient, and angulation of the distal arch can make delivery and seal of the proximal aspect of the stent difficult, both of which may lead to proximal type I endoleak or collapse of the endograft. Type I and Type II endoleaks are present in up to 20% of cases necessitating repeat intervention.

Similar to evaluating delayed versus immediate intervention, bias exists in evaluating the literature regarding OSR versus TEVAR. In five of six meta-analyses comparing the two modalities, operative mortality was in favor of TEVAR, but in those studies reporting timing or severity of injury, OSR was more often the modality chosen. However, in more contemporary series, the perioperative mortality and paraplegia rate have favored TEVAR. In the same cohort of patients from the prior study, the AAST investigators evaluated OSR versus TEVAR in a prospective manner. In patients

with and without other associated injuries, the mortality was significantly lower with TEVAR as was paraplegia (2.9% OSR. 0.8% TEVAR). There was no associated difference in ICU stay, systemic complications, or days of ventilator dependence.

The pooled data on TEVAR are limited by a mean follow up of only 18 months. Similar to evaluation OSR versus TEVAR for aneurysm disease, it appears that the survival benefit of TEVAR is lost after 1 year, likely due to the not insignificant morbidity and mortality of subsequent interventions required in patients treated with TEVAR. Perhaps, the most important factor limiting application of TEVAR to all cases of TAT is the lack of long-term data regarding performance of the grafts in young patients. Concern exists regarding potential for migration as the aorta dilates with age. Further, it is unknown whether these devices will suffer material fatigue when implanted for extended periods of time. Several reports describe successful salvage of failed endografts with open repair.

No clear data exist to definitively favor one modality over the other. A strong trend toward decreased perioperative mortality and paraplegia in patients treated with TEVAR for TAT. This technique can expand therapy to patients who are not candidates for open repair. Evaluation of anatomy, associated injuries, urgency of repair, and patient age will lead to proper operative decision making.

OPEN SURGICAL MANAGEMENT

The surgical preparation of a patient with an acute TAT should be done in an orderly manner. Every attempt to approach this problem as elective surgery should be made if the patient is stable. It is of paramount importance that the patient and the family be thoroughly counseled about the procedure and specifically about the perioperative risks of paraplegia. This is a lethal injury, and the possibility of death from other injuries or from the transected aorta should be thoroughly discussed with the family. The other major risks of the operation should be discussed. These include the possibility of infection, renal failure, respiratory distress syndrome, vocal cord paralysis, and complications from other associated injuries.

If at all possible, a double-lumen endotracheal tube should be placed for the procedure because this will greatly facilitate exposure of the injury. If there is a question of a cervical spine injury, it should be ruled out before the patient is taken to the operating room. A cervical collar makes anesthetic intubation difficult and limits access to the neck, should the placement of additional intravenous lines be necessary. Appropriate monitoring and resuscitative lines should be placed. A cardiopulmonary perfusionist should be available. The blood pressure in the upper extremities should be monitored through a right radial artery catheter, because during the repair the left subclavian artery (LSCA) will need to be occluded and inaccurate readings will be obtained from a left radial artery catheter.

The left groin is left free in case access is needed for bypass procedures or for other lines. Our preference is to place a Swan sheath for its size and to use a true Swan–Ganz catheter only when hemodynamic questions arise. We do not routinely monitor cerebral spinal fluid (CSF) pressure, because this adds time and complexity to the procedure with little known benefit in the acute trauma setting. All patients receive prophylactic antibiotic therapy mainly aimed at prevention of *Staphylococcus aureus* infection in anticipation of potentially placing a prosthetic graft in the aortic position. Several sizes of aortic grafts should be in the room and made available. Our routine is to size the aorta intraoperatively, either visibly or with sizers, which generally gives us an accurate idea of what size of graft to use. Our graft preference is a pretreated woven graft, which does not require preclotting.

After all the preoperative plans have been made, the patient is taken to the operating room, placed in the supine position, and anesthetized. The induction of anesthesia and intubation is a very critical point in the procedure. Every attempt should be made to avoid severe blood pressure fluctuations, because any hypertension can precipitate a rupture. The patient is then placed in the right lateral decubitus position. Positioning and padding of the extremities are to be performed at this juncture. In addition, we like to position the patient so the left groin is available in case the femoral artery or femoral vein needs to be exposed during the procedure. The patient is prepared and draped. Our preference is to use an antiseptic-impregnated plastic drape to keep as much skin covered as possible considering the probability of a prosthetic graft being used during the procedure.

A full posterolateral thoracotomy incision is made from just below the left nipple. Both the latissimus dorsi and serratus anterior are divided, and the fourth intercostal space should be chosen for entering the pleural cavity as a fifth interspace incision can make the exposure difficult. It is usually not necessary to excise a rib. However, if exposure is inadequate on opening of the chest cavity, our procedure has been to resect 1 cm of rib (posteriorly, behind the paraspinous ligament) below our initial incision to allow additional exposure.

Once the pleura is entered, the left lung is deflated and adequate ventilation ensured. After the lung is deflated, often a formidable, large pulsating hematoma will be seen in the mediastinum. The surgeon should proceed deliberately to obtain proximal and distal control. Distal control is usually obtained first, because this is easiest to perform and avoids precipitating free rupture. It is important when dissecting around the distal aorta to avoid the intercostal artery branches, which are paired and come off each side. It is preferable not to sacrifice any intercostals but to go between the intercostals and encircle the aorta with an umbilical tape

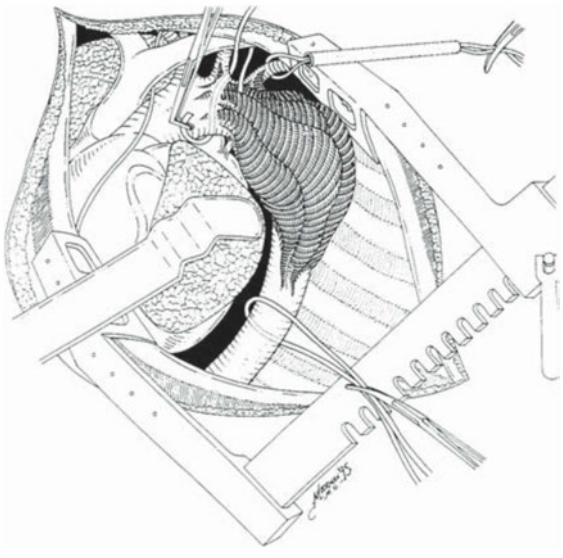


Fig. 62.2. The lung is retracted superiorly and anteriorly, exposing the descending thoracic aorta, which is encircled away from the hematoma. The intercostal vessels should be avoided. With blunt dissection of the plane complete, the transverse aorta is encircled proximal to the ligamentum arteriosum between the subclavian and the carotid. A tourniquet has been placed on the left subclavian and secured.

(Fig. 62.2). The umbilical tapes are placed to provide a handle in case of sudden rupture and to facilitate the placement of vascular clamps. It is much easier to pull up on these tapes and place a clamp on the aorta than to try to get both a hand and a vascular clamp in the wound. The other advantage of a tape is to ensure that when the clamp is placed, it is completely around the aorta. Attention is then turned superiorly. The subclavian artery can be palpated easily and often is involved in the periaortic hematoma. Control of the subclavian artery is important and should be obtained even when the angiogram suggests that the injury is distal to its takeoff. We prefer a Rommel tourniquet on the subclavian artery, because it takes up very little space in the surgical field and is more than adequate to provide hemostasis during the repair. Placement of the proximal aortic clamp is one of the most important technical parts of the operation. It is preferable to place this clamp distal to the left carotid, but well proximal to the takeoff of the LSCA. Although aortic transections are usually distal to the takeoff of the LSCA, this extra portion of aorta is often necessary to obtain a good repair. In addition, intimal tears may extend further proximally than can be appreciated by preoperative imaging. Attempts to obtain control distal to the takeoff of the LSCA are often met with rupture of the pseudoaneurysm, because, invariably, the hematoma is entered or friable aortic tissue is encountered in this region. When a clamp is placed distal to the left subclavian, the tear will often extend into the clamp, and it may be necessary to obtain more proximal control (which may not be possible without exsanguination occurring first). The vagus nerve runs in this area, with the recurrent branch as well. The recurrent branch is almost never seen because of the hematoma, but the vagus can usually be seen stretched by the hematoma. By staying anterior to this nerve, one should easily be able to obtain proximal control of the aorta. A finger can usually be placed gingerly between the left carotid and the left subclavian around the aorta in this area to create a proximal clamp site. At this point, an umbilical tape is passed to provide a handle to place the clamp at a later time.

One should be aware that multiple aortic tears are possible. The tear at the isthmus is sometimes associated with a tear in the arch, the ascending aorta, or the great vessels, and this should be anticipated as well and preferably ascertained on preoperative imaging. On rare occasions, the clamp will have to be placed between the innominate and left carotid to gain adequate proximal control.

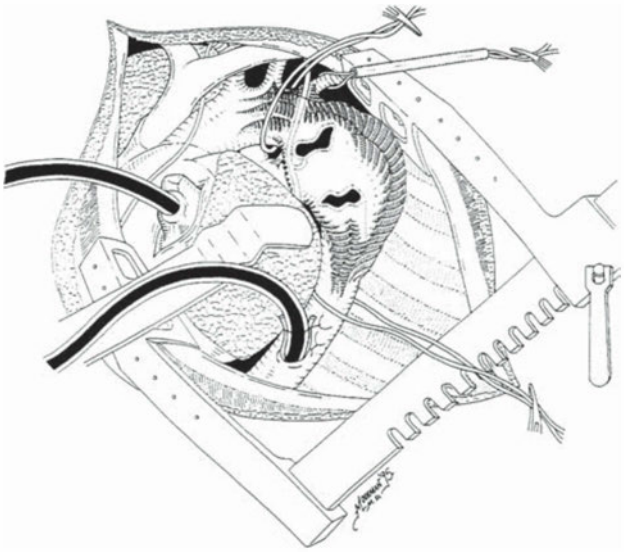


Fig. 62.3. Arterial access is achieved in the descending aorta well away from the hematoma below the tape placed for distal control through purse-string sutures. Alternatively, the femoral artery may be cannulated.

When proceeding with partial left heart bypass, it is necessary to establish a drainage line and a return line to the patient (Fig. 62.3). Many sites are available for this. Our preference is to use the juncture of the left atrium with the superior pulmonary vein (or, alternatively, via the left atrial appendage) for drainage and the descending aorta distal to our distal clamp for return. The lung should be retracted laterally and the pericardium opened with care to avoid injury to the phrenic nerve. We usually open the pericardium posterior to the phrenic nerve immediately anterior to the pulmonary veins, but occasionally have opened it anterior to the phrenic nerve. The surgeon should inspect the pericardium before deciding on which side of the phrenic nerve to incise the pericardium. Usually, the left atrial appendage can be identified easily because it beats within the pericardium. In the decubitus position, there is almost an up-and-down motion of the appendage. When the pericardium is opened, the left atrial appendage almost "pops" up into the wound. On occasion, the pericardium can be ruptured from the blunt trauma, and access can be obtained through the rupture. The tip of the appendage may appear friable, it is for this reason that we have occasionally obtained venous drainage at the confluence of the left superior pulmonary vein and the left atrium. This area takes sutures well. To avoid tears in the left atrial appendage, we place a purse string of 2-0 braided polyester suture in the best appearing tissue. The tip of the appendage is amputated, and a venous drainage catheter is placed into the left atrium. Depending on the size of the patient, a 26-, 28-, or 30-F wire-reinforced venous cannula is selected. With a 28-F drainage catheter, flows can be obtained in the 3- to 4-liter range without difficulty. During the placement of this catheter in the atrium, communication between the anesthesiologist and the surgeon is important as the clamp is removed from the appendage. A Valsalva maneuver is performed to prevent entry of air into the left side of the heart as the cannula is placed, and a Rommel tourniquet is secured and tied to the cannula. The connection to the inlet tubing of the pump is freed of air.

Distal perfusion can be obtained in several areas. If it appears reasonable, the descending aorta well away from the injury is cannulated through the thoracotomy wound. An 18- to 20-F flexible cannula is usually sufficient for adequate flow. Alternatively, if descending aortic cannulation is not feasible, an incision is made in the left groin. The femoral artery is dissected and a femoral artery cannula inserted. In patients with multiple injuries or fractures with possible delayed or remote bleeding, this circuit can be used without heparin or a very minimal amount of heparin, such as 5,000 units.

At this point, a graft is chosen. We have used both knitted and preclotted woven grafts. The knitted grafts are easier to sew, but must be preclotted before heparinization.

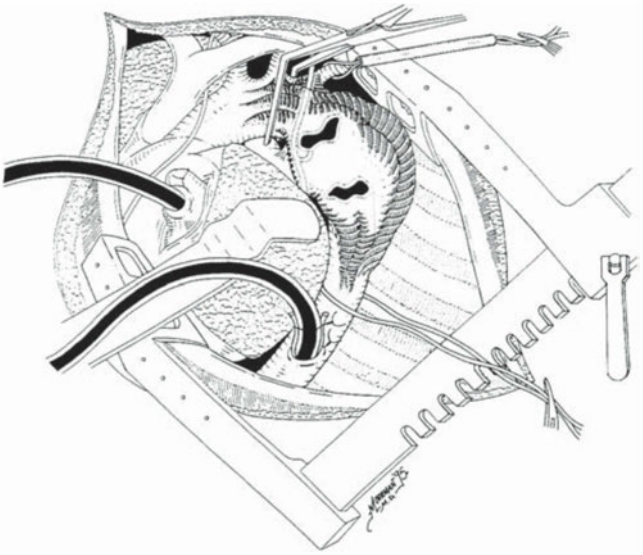


Fig. 62.4. The proximal aortic clamp is placed on the transverse arch as the left heart bypass circuit is initiated. Pump flows are adjusted as the clamp is placed to ensure adequate proximal and distal aortic perfusion pressures.

Appropriate vascular clamps are chosen to be used for the proximal and distal control. Before the clamps are placed for repair of the transection, mannitol (12.5 to 25 g [0.25 to 0.5 mg/kg]) is administered to maximize renal perfusion.

Once all the above steps are accomplished, the proximal aortic clamp is applied (Fig. 62.4). Proximal hypertension is alleviated by increasing left heart assist to unload the left heart as the clamp is secured. The distal clamp is applied, and then the snare on the subclavian artery is secured. The operation should proceed expeditiously but in an orderly manner. The hematoma is opened, usually met with a disconcerting amount of blood that should be anticipated. With the clamps properly placed, active bleeding should not be seen from any of the clamp sites. Bleeding from intercostal collaterals is expected; however, this should not be a cumbersome amount. If the clamps are not placed all the way across the aorta, time will be wasted in obtaining proximal or distal control, and a large amount of blood will be lost. Once the hematoma is entered, the site of the tear will be obvious. In the case of complete transection, the stumps may be retracted and separated by several centimeters. Once the free ends are identified, the edges of both sides should be freshened, excising to good tissue (Fig. 62.5). Sometimes, a primary repair may be possible. However, in the majority of cases a graft will be needed to bridge the gap without undue tension. Sometimes, bleeding from intercostals can be problematic, and these can be controlled with bulldog clamps, hemoclips, or silk ligatures as needed. We try not to sacrifice any intercostals unless they are involved directly in the tear and cannot be salvaged.

We prefer to address the proximal aorta first because this is typically the more difficult anastomosis as a result of relatively limited exposure around the ligamentum and the friable and unpredictable nature of the tissue in this area. Our preference is to use a double-arm 4-0 polypropylene suture without pledgets and to perform the anastomosis in a simple running manner. Care should be taken not to place the sutures blindly and deep in this area because the esophagus and recurrent laryngeal nerve are immediately adjacent and posteromedial to this area. After the proximal anastomosis is completed, the graft is stretched and cut to length. An important point is to cut the graft appropriately. Too long a graft will lead to kinking; too short a graft will create tension and tearing of the aorta as sutures are placed. The distal suture line is then done with a running 4-0 Prolene suture. Just before the anastomosis is completed, the distal clamp is removed to check for hemostasis and to deair the aorta (Fig. 62.6). The anastomosis is completed and the proximal clamp removed after deairing. Weaning from left heart assist is done simultaneously with the removal of the proximal clamp. Sometimes, hypotension will develop, which requires reapplication of a clamp either partially or totally to restore the proximal blood pressure; then, as volume is added, an adequate pressure proximally and distally can be maintained.

At this point, the proximal and distal suture lines can be further examined. Sometimes bleeding is seen and can be controlled with simple sutures. However, bleeding from needle holes or just oozing

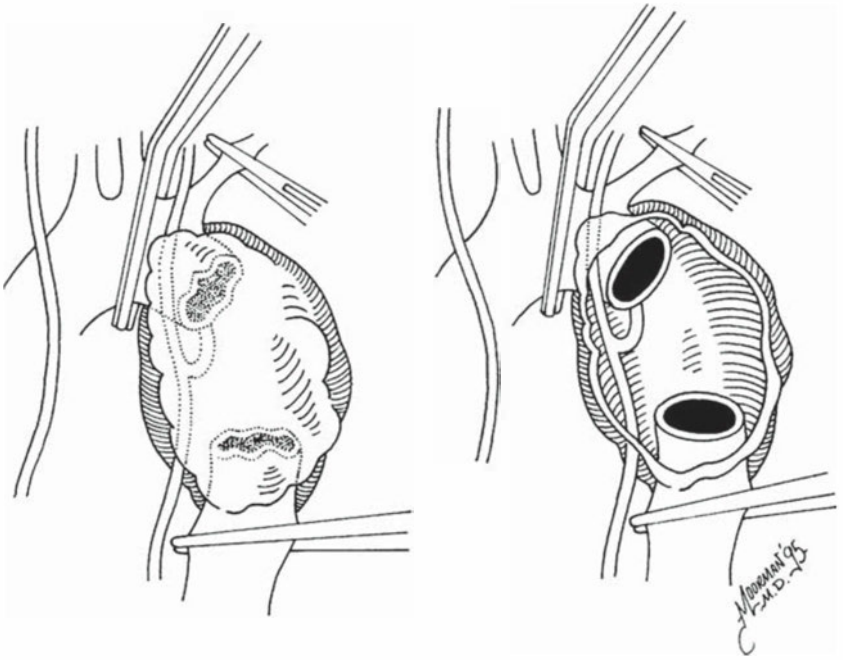


Fig. 62.5. The hematoma is opened after left heart assist is achieved. The friable aortic edges are carefully debrided to good tissue, any intercostal bleeders are controlled, and the recurrent nerve is avoided.

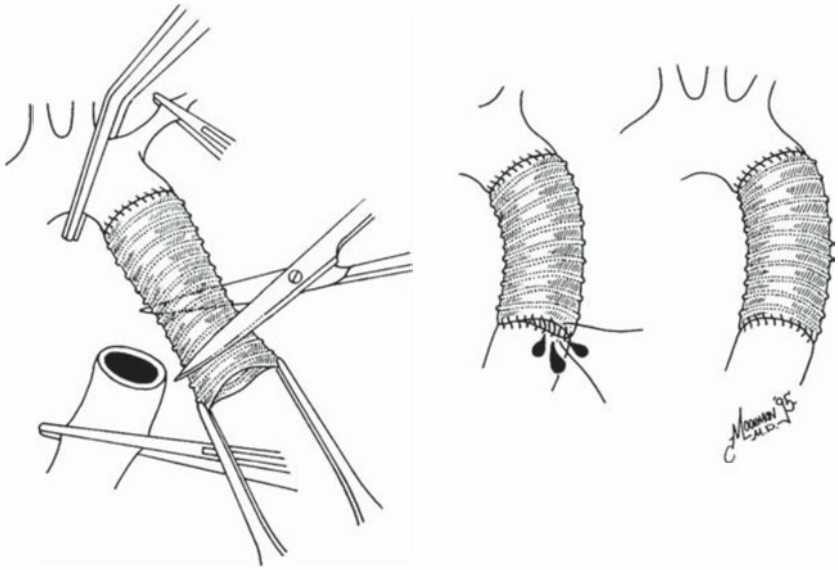


Fig. 62.6. With the proximal anastomosis completed, the graft is trimmed to appropriate length, and torsion is avoided. Before the clamps are removed, deairing should be performed by releasing the inferior clamp and backbleeding the graft.

from distal suture lines should be watched; if heparin has been administered, it should be reversed with protamine to allow these sites to clot. Usually, packing the area and waiting 5 to 10 minutes will reveal complete hemostasis. At this point, the bypass circuit, if used, can be removed and the previously placed purse strings tied to ensure hemostasis. Before the thoracotomy is closed, we cover the graft, especially the suture lines, with mediastinal pleura using absorbable sutures to exclude the graft site from the lung.

Over the years, our technique for repair has evolved from one in which all cases were done in a clamp-and-sew manner to one in which we nearly routinely use left heart bypass from the left atrium to the descending aorta or to the left femoral artery using a centrifugal pump with very little heparinization. However, when this is not technically feasible, or in cases of free rupture, we do not hesitate to perform the clamp-and-sew technique. We do not use passive shunts because flow cannot be regulated. The other advantage of using a centrifugal type of left heart assist is the ability to warm or cool the blood as a heat exchanger can be easily added to the circuit.

We do not believe that any one technique has been conclusively proven to prevent paraplegia. It is our opinion that expeditious and timely repair of this lesion is the most important aspect in preventing this complication. Much has been made of the 30-minute ischemia period as a contributing factor to paraplegia. However, there have been many cases reported of clamp times much longer than this with no evidence of paraplegia. The complication of paraplegia is probably multifactorial, and time represents only one factor. However, partial left heart assist may benefit in other ways, because it does maintain distal perfusion pressure and allows us to unload the left heart and avoid hypertension proximal to the clamp. It, therefore, may help to avoid the use of vasodilating agents such as nitroprusside, which have been known to cause decreased blood flow to the spinal cord. Most importantly, it maintains perfusion to the lower body and kidneys during the period of cross-clamping. It also allows the opportunity to either cool or warm the patient in an expeditious manner. Although it does add complexity to the operation, we believe that its benefits are well worth the effort.

ENDOVASCULAR MANAGEMENT

Operative preparation, monitoring, and blood availability for TEVAR should proceed similar to that for open repair. Anesthetic technique can be either general or sedation with local anesthetic if femoral access is assured. Due to the potential for paraplegia and need for postoperative monitoring of neuromotor function, we do not recommend spinal or epidural anesthetic. The patient is placed supine and prepping/draping of the patient should allow access to both groins as well as the abdomen and lower chest. This will facilitate the ability to change to an iliac, or aortic, exposure if difficulty is encountered during femoral access. We also find preparation of the right brachium/axilla helpful in case the need arises for percutaneous, or open, brachial/axillary artery access to aid in the repair. Notification of the perfusion team is warranted in the event that conversion to open repair is needed, but a circuit need not be prepared in the majority of cases.

TEVAR may be performed in standard operating rooms with a mobile C-arm capable of digital subtraction angiography. However, if available, fixed imaging equipment will significantly improve imaging in the thoracic region, decrease parallax, and allow for a larger field of view and less need to move the image intensifier (II) during diagnostic imaging and deployment of the endograft. Another critical adjunctive imaging technique is IVUS. An 8.2-F Visions catheter (Volcano Corp., San Diego, CA) allows for diameter imaging up to 60 mm. IVUS can precisely image the relationship of the intimal injury to the arch anatomy and often can detect discreet injuries not picked up by CTA. In addition, IVUS allows for measurements to be taken in systole and diastole of the proposed landing zones resulting in more precise sizing of the endograft. Pull back measurements can be made in the center line of flow to ensure proper length of device as well.

The newest generation of endografts has a broader range of sizes and smaller diameters to allow treatment in most patients. In addition, these devices have lower profile delivery systems allowing most to be deployed from femoral access. Historically, abdominal cuffs have been utilized to treat smaller diameter aortas; however, these devices have several limitations. They are shorter and may require multiple components to treat the injured area. In addition, they come mounted on shorter shafts and may not reach the injured area through femoral access, especially as most TAT occur near the isthmus. Lastly, the thoracic endografts are engineered to withstand the unique pressures and hemodynamics present in the distal arch and have improved delivery systems for this area. When at all possible, a thoracic device should be used for TEVAR in TAT (Fig. 62.7).

In choosing an appropriate access site, the diameter and nature of the iliac vessels is evaluated on CT. Most new generation thoracic endografts can be delivered through 20- to 22-F outer diameter sheaths. An access vessel of 7 mm should be more than adequate. In the younger patient and those without atherosclerosis on CTA, the external iliac arteries are often compliant

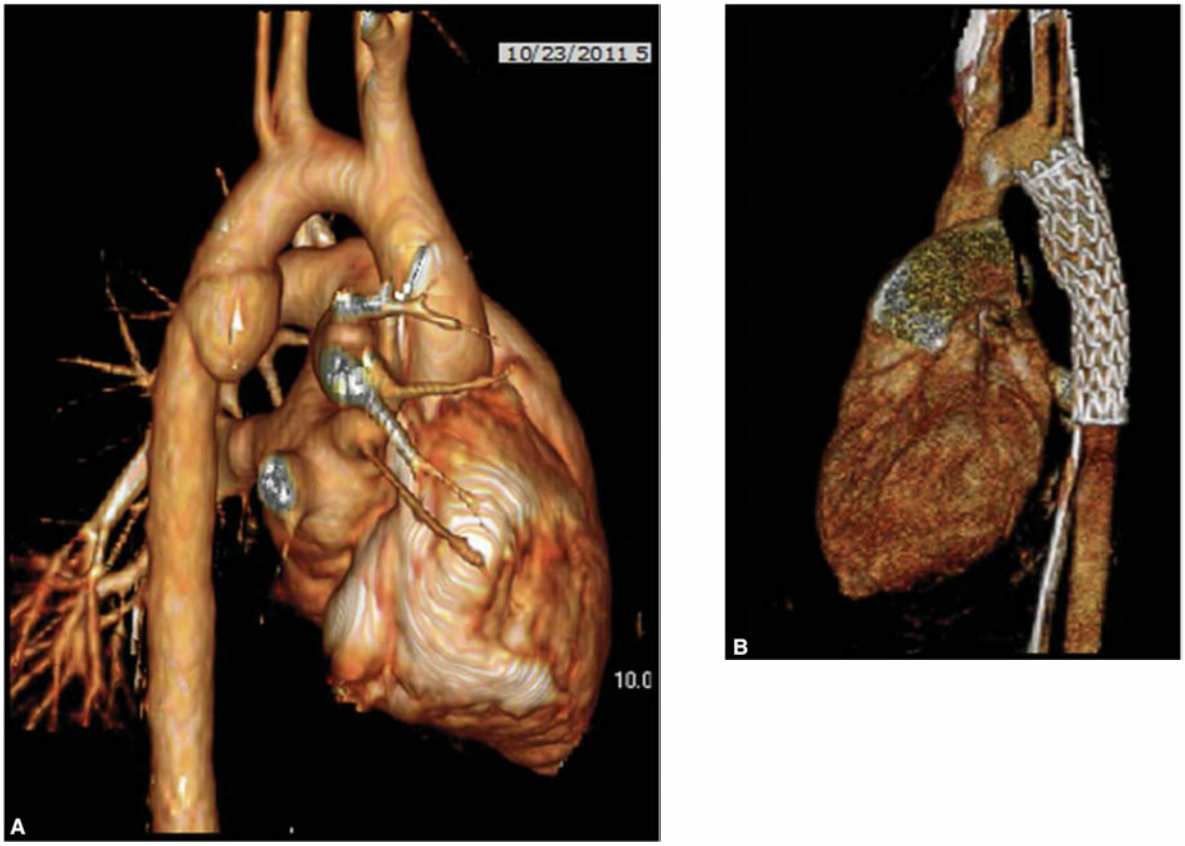


Fig. 62.7. (**A**) CTA demonstrating traumatic PSA of aorta in an 18-year-old male with anomalous vertebral anatomy. (**B**) Postoperative CTA demonstrating complete exclusion of PSA with new generation conformable, tapered stent graft in same patient. CTA, computed tomographic angiography; PSA, pseudoaneurysm.

and can still receive the device. Again, IVUS can be critical in evaluating the true size and presence of atherosclerosis when performing TEVAR. In pooled analyses of over 25 experiences with TEVAR in TAT, the need for iliac or aortic conduit was only 8.7%.

Once the patient is properly prepared, an oblique incision is made over the femoral artery and an appropriate length of common femoral artery is dissected and controlled with a Potts vessel loop. Unless bulky atherosclerosis is encountered in the femoral, dissection of the bifurcation is not warranted. Access is gained to the exposed artery and a 9-F sheath is placed. Diagnostic access is then gained percutaneously in the contralateral femoral or right brachial artery (the left brachial is avoided due to the involvement, and possible need to cover, the left subclavian). A guidewire is then placed from the 9-F sheath into the ascending aorta. Over this, the IVUS catheter is introduced and evaluation of the iliac and aorta commences. If any question exists regarding access vessels, this is the appropriate time to change to a conduit. If necessary, the common iliac artery or distal aorta can be exposed through a retroperitoneal exposure in either lower quadrant. Identification of the ureter is critical and control of the iliac is facilitated by circumferential control with vessel loops. Appropriate heparin administration is given for an ACT >250. A smaller dose of heparin may be used if bleeding from other injuries is a concern, although frequent flushing of sheaths and attention to flushing at the conclusion of the case is warranted. A 10-mm Dacron conduit is then sewn end to side on the common iliac or distal aorta and then accessed with a 9-F sheath.

Sizing of the aorta in systole allows for precise oversizing of the endograft. In TAT, we recommend oversizing of only 10% to 15% to decrease the incidence of endocollapse and decrease the "stretch" of the graft on the aorta, which may precipitate further propagation of dissection either proximally or distally. Newer devices now have tapered configurations to aid in appropriate sizing as the aorta is often significantly larger at the distal arch than mid-thoracic region. Pull back measurements are made to ensure 15 mm of seal on either side of the injury and the shortest component chosen that covers the injured site to limit the number of intercostal arteries sacrificed. At this point, the endograft can be opened and prepared on the back table, along with an appropriate compliant aortic occlusion balloon.

A marking flush catheter is then placed in the ascending aorta and diagnostic aortogram performed. A left oblique position of the type II should be utilized to open up the arch, usually around 40 degrees. A 30-cm^3 injection is usually adequate. Confirmation of arch anatomy, vertebral anatomy, site of injury, and presence of internal mammary grafts is made at this juncture. Selection of the vertebral arteries is not routine unless definitive coverage of the left subclavian is planned, and any question exists regarding vertebral anatomy or fetal posterior circulation from preoperative imaging. The proposed landing sites are marked on the imaging screen and length measurements confirmed. At this point, the patient should be heparinized as above.

Working over the stiff guidewire, the device sheath (or device itself depending on manufacturer) and then the device are advanced to the area of intended delivery. The flush catheter is positioned just proximal to the proximal landing zone and a magnified aortogram is performed. The

critical anatomy is marked and the device is then advanced past the landing zone to remove any energy built up in the delivery shaft and then slowly pulled back to the intended mark. This is often the most critical step in deployment. The angulation of the arch at the isthmus can be severe, especially in young patients. A major limitation is the ability to track the device over the curve of the distal arch. Significant force is placed on the outer curve of the aorta and this can precipitate rupture in a tenuous injury. The newer generation devices have improved tips for trackability; however, this can still be a challenge. Occasionally, right brachial access is needed. A snare system is used to create a "floss" of the patient where the wire is tethered both outside the right brachial sheath and in the access sheath. The "floss" should be performed using a floppy glidewire and then exchanged for a stiff wire through a catheter. This technique will greatly improve the trackability of the device over the arch and decrease the stress on the outer curve of the aorta. Conversely, we have had to convert to a sternotomy and antegrade access in this situation when the device cannot pass.

Once in proper position, the graft is deployed via the specific manufacturer's recommendation. We routinely leave the flush catheter behind the device to preserve access behind the graft in case of inadvertent coverage of a branch vessel. It can then be retracted over a wire and replaced through the graft after deployment. The device is removed and gentle molding with a compliant balloon is recommended for any areas that do not fully conform to the aortic wall. This is not mandatory, however, is often helpful in eliminating the "bird-beak" effect in the proximal landing zone. A completion angiography is performed, and repeat IVUS can confirm apposition of the stent to the aortic wall.

The catheters and balloon are removed. Final removal of the large-bore access sheath is the last critical component of the case. It is during this step that the external iliac artery can be sheared from the iliac bifurcation (the so-called "iliac on a stick"). This will precipitate exsanguinating hemorrhage. We, therefore, advocate removal of the sheath over a guidewire left in the aorta. This facilitates rapid placement of an occlusion balloon into the aorta for proximal control and then expeditious open or endovascular repair of the iliac injury.

Once the device is removed and pulsatile inflow confirmed, the femoral arteriotomy is closed, or conduit is oversewn. Routine angiography of the iliac is not necessary, but if any question exists regarding iliac injury, the diagnostic catheter from either the contralateral femoral or brachium can be used for diagnostic purposes. Pulses in the feet and wrist are confirmed to be the same as preoperative and protamine is administered. After removal of all devices, the patient can be transferred back to the ICU for further management of his injuries. If necessary, blood pressure can now be liberalized and we recommend maintaining a systolic pressure of at least 110 mmHg to aid in spinal cord perfusion. Neuromotor checks are performed frequently and expectant management of paraplegia is appropriate. A follow-up CTA is recommended before discharge. Any signs of lower extremity malperfusion, sudden drop in urine output, and severe right arm hypertension should alert the intensive care team to the potential for endograft collapse which is a life-threatening emergency. Emergent CTA angiography is warranted, although if the necessary personnel are in the hospital, proceeding directly to the angiography suite is appropriate as well.

The management of the LSCA warrants special consideration. Due to the relatively low risk of paraplegia with coverage of subclavian collaterals in patients with TEVAR for TAT, the Society for Vascular Surgery recently released guidelines recommending elective revascularization of the LSCA. In the majority of cases, coverage is well tolerated and upper arm arterial insufficiency or vertebrobasilar symptoms can be managed with delayed revascularization. With the exception of hypoplastic right vertebral artery and patent left internal mammary artery (LIMA) grafts, traumatic TEVAR can proceed without this step as delay in aortic repair and additional operation with heparin administration is not warranted in most cases.

CONCLUSION

Improvements in trauma systems, critical care, and imaging of the injured patients have greatly altered the landscape of this lethal injury. Managing the spectrum of aortic transection and other associated injuries in these complex patients remains a challenge. Advances in both the medical management of the TAT patient and a careful selection of patients appropriate for delayed repair have improved overall outcomes. Application of both precise surgical technique and mastery of endovascular skills allows for appropriate selection of modality to best treat the individual patient.

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EDITOR'S COMMENTS

The authors have demonstrated the changes in management for this frequent lethal injury. When I was a resident, diagnosis was by angiography, emergent surgery was required, and procedure was done open. Frequently, the procedures were done with the so-called clamp and run technique and the push was to keep the clamp times below 30 minutes.

Things have completely changed and I believe for the better. Diagnosis is now CT angiography, then there is suspicion of injury. The patients are often treated electively particularly if multiple other injuries are present. There have been a very few cases of a rupture occurring during the waiting period. Repair has pretty much all switched over to TEVAR. It is very rare to do an open repair for this injury anymore. This has been done without any significant comparative data of open repair versus TEVAR.

There are several issues that need to be discussed. I am in full agreement with elective subclavian artery revascularization following TEVAR repair. I think this is an appropriate technique and can easily be done with a carotid subclavian bypass. I am in full agreement with TEVAR but the long-term results are not clear. These are young patients with normal-sized aortas, and what will happen to repairs in the long run is not certain.

Having said all this, we now can avoid rushing patients having multiple organ injuries to the operating room. I believe this has been a huge advance and believe we salvage more patients than we used to.

ILK

63 Heart Transplantation

Christopher T. Salerno and Edward D. Verrier

INTRODUCTION

The techniques developed by Norman Shumway and Richard Lower at Stanford University set the stage for heart transplantation to become the therapy of choice for patients with end-stage heart failure. The introduction of transvenous endomyocardial biopsy and the advent cyclosporine immunosuppression dramatically increased patient survival and marked the beginning of the modern era of successful cardiac transplantation. Heart transplantation is now a widely accepted therapeutic option for end-stage cardiac failure, with more than 3,300 procedures performed annually.

RECIPIENT SELECTION

Patients with end-stage heart disease who are being considered as potential candidates for cardiac transplantation should be evaluated by a multidisciplinary committee to ensure an equitable, objective, and medically justified allocation of the limited donor organs. This process should select patients with the greatest chance of postoperative survival and rehabilitation. The primary objective of the recipient selection process is to identify patients with irreversible cardiac disease not amenable to other therapy (optimization of medical therapy, revascularization, ventricular remodeling, valve repair/replacement, biventricular pacing). In our center, the evaluation for a ventricular device is part of the transplant selection process. It is desirable to identify patients who will most likely resume a normal active life and be compliant with the rigorous postoperative medical regimen. Recent successes and the introduction of improved immunosuppression have significantly expanded the eligibility criteria. Patients with New York Heart Association class III or IV symptoms despite optimal medical therapy should be considered for cardiac transplantation. Most patients present with end-stage heart failure due to ischemic heart disease or idiopathic dilated cardiomyopathy. However, the spectrum of known causes of end-stage cardiomyopathy includes infectious (viral), inflammatory, toxic, metabolic, and familial etiologies. Patients selected for cardiac transplantation should have a predicted 2-year survival of $<$50%. Other less common indications for cardiac transplant include refractory angina, life-threatening arrhythmias, and chronic cardiac allograft rejection. Contraindications to cardiac transplant are primarily based on comorbid illnesses. Examples of widely accepted contraindications include active infection, irreversible hepatic dysfunction, and fixed pulmonary hypertension (pulmonary artery systolic pressure $>$60 mmHg, transpulmonary gradient $>$15 mmHg, pulmonary vascular resistance $>$6 Wood units). Many centers have demonstrated that a left ventricular assist device (LVAD) can be used to reverse fixed pulmonary hypertension and allow patients to become transplant patients. Age in no longer considered to be a contraindication to cardiac transplant.

Preoperative Recipient Evaluation

Evaluation for cardiac transplantation begins with a comprehensive history and physical examination, chest roentgenogram, and lab work including complete blood count, coagulation screen, erythrocyte sedimentation rate, uric acid level, liver function tests, fasting lipid panel, and infectious disease serologies (hepatitis A, B, and C, herpes simplex virus, human immunodeficiency virus, rapid plasma regain, rubella, measles, *Toxoplasma*). All patients should undergo an exercise test with peak exercise oxygen consumption (peak VO_2) measurement. Available studies suggest that patients with peak VO_2 $<$14 ml/min/kg have improved survival and significant functional benefit with transplantation. A right heart cardiac catheterization study should be performed to rule out irreversible pulmonary hypertension. For patients with ischemic cardiomyopathy, coronary angiography should be reviewed or repeated to confirm the inoperability of coronary artery disease. For patients with nonischemic cardiomyopathies and prolonged or atypical symptoms, endomyocardial biopsy should be performed to rule out the possibility of a medically treatable illness.

Most centers also include nutritional labs, thyroid function studies, fasting and postprandial blood sugar, creatinine clearance, 12-lead electrocardiogram, echocardiogram, pulmonary function tests, panel reactive antibody screen, HLA typing, vascular screening exams (abdominal ultrasound, carotid and lower extremity Doppler flow studies), esophagogastroduodenoscopy, psychosocial evaluation, dental evaluation, financial analysis, and screening studies for malignancy (stool guaiac, prostate-specific antigen, mammogram, Papanicolaou smear).

Patients listed for transplantation should be examined routinely for re-evaluation of recipient status. Repeat right heart catheterization is indicated when follow-up echo studies suggest worsening or persistent pulmonary hypertension. Actively listed patients should undergo a repeat right heart catheterization at least twice annually.

DONOR AVAILABILITY AND ALLOCATION

Donor organ availability is the primary factor limiting the application of heart

transplantation. As a result, 20% to 40% of patients on the waiting list may die before transplantation. Organ allocation is based on recipient priority status, time on the waiting list, and proximity. Highest priority is given to local status 1A patients with the longest accrued waiting time. The allocation system is designed to provide the most critically ill patient with a heart while minimizing allograft ischemic time. Although only 25% of patients are classified as status 1 at the time of listing, 48% progress to status 1 by the time of transplantation. The mean waiting period for a status 2 candidate is currently >1 year, whereas status 1 patients wait a mean of 60 days. There is significant variation in waiting times from region to region. Due to this variability, many more patients require an LVAD as a bridge to transplant, in some regions as many as 85% of blood type O patients require an LVAD bridge.

Donor Selection

Potential cardiac donors undergo a rigorous screening evaluation. The local organ procurement agency should provide the implanting program with patient's age, height, weight, gender, blood type, hospital course, cause of death, routine laboratory data, and viral serologies. Additional required data include electrocardiogram, chest roentgenogram, arterial blood gas, and echocardiogram. Coronary angiogram is indicated in a selective manner. The presence of advance donor age (male donors >45 years of age, female donors >50 years of age), risk factors for atherosclerotic coronary artery disease (tobacco abuse, diabetes, significant family history), and occasionally the mechanism of death should be evaluated to determine the need for coronary angiogram. When a member of the transplant team arrives for organ procurement, a secondary screening is performed. This secondary screening allows the recovering surgeon to confirm and review the data presented by the procurement agency. The most important donor screening occurs in the operating room at the time of organ procurement. The heart is examined to identify ventricular or valvular dysfunction previous infarction, coronary atherosclerosis, or myocardial contusion. If direct examination of the heart is unremarkable, the procurement surgeons proceed with donor cardiectomy.

Matching potential recipients with the appropriate donor is based primarily on blood group compatibility and patient size. As a rule, ABO barriers should not be crossed in heart transplantation because incompatibility frequently results in fatal hyperacute rejection. Donor weight should ideally be within 30% of recipient weight except in pediatric patients, where closer size matching is required. If the recipient has elevated pulmonary vascular resistance (>4 Wood units), a larger donor is preferred to reduce the risk of right ventricular failure in the early postoperative period. When the recipient has an LVAD, special consideration should be given to the location of the donor; in these cases, the variable time required to explant the device mandates that the implanting surgeon be able to control the cross-clamp time and organ arrival times. When the percentage (or the panel) reactive antibody (PRA) is ≥10% to 15%, a prospective negative T-cell cross-match between recipient and donor sera is recommended before transplantation. Some centers also insist on a negative B-cell cross-match before transplantation. A positive cross-match is an absolute contraindication to transplantation. A cross-match is always performed retrospectively, even if the PRA is absent or low. Retrospective studies have also demonstrated that better matching at the HLA-DR locus results in fewer episodes of rejection and infection and an overall improved survival. Because of current allocation criteria and limits on ischemic time of the cardiac allograft, prospective HLA matching is not always logistically possible. Many centers have begun to selectively use virtual cross-matching in cases where recipient serum is not available.

There have been recent reports that "extended" donors can be safely used to obtain similar short-term results observed with standard donors. The term "extended" refers to patients with advanced age, high-risk social behavior, limited coronary artery disease, or positive viral serologies. When used, the extended donors are matched to high-risk recipients in which increased risks are acceptable.

DONOR HEART PROCUREMENT

During the secondary donor screening, the location of central and arterial lines should be noted. The location of these lines may affect the conduct of the organ harvest. It is important to continually monitor the donor's volume status and urine output during the harvest to maintain organ function. The donor is positioned in the supine position and widely prepped from chin to knees. To facilitate both thoracic and abdominal organ recovery, the harvest is begun with a midline incision from sternal notch to pubis, including a median sternotomy. The pericardium is incised and a pericardial well is created. The heart is examined to assure no previously unidentified anomalies exist. After this inspection, the harvesting surgeon should notify the implanting surgeon regarding the suitability of the heart for transplantation.

The heart is then mobilized for cardiectomy. It is important for the harvesting surgeon to know whether any extended lengths of vessels will be necessary to facilitate implantation. For our preferred method of implantation, the bicaval technique, an extended length of superior vena cava (SVC) is required. The SVC is mobilized from the right atrium to the innominate vein. SVC mobilization usually includes dissection of the right pulmonary artery and ligation of the azygous vein. If additional caval length is required, the innominate vein can be harvested en bloc. The inferior vena cava (IVC) is dissected and mobilized circumferentially. Encircling the SVC and IVC with umbilical tapes or heavy suture may assist with retraction during cardiectomy. The aorta is dissected from the pulmonary artery and then isolated with umbilical tape. If hemodynamic instability is encountered during the harvest, clamping of the abdominal aorta at the iliac bifurcation may be helpful. Once the mobilization of the abdominal organs is complete, the donor is given 30,000 units of heparin intravenously. A purse-string suture is placed in the ascending aorta, through which an antegrade cardioplegia catheter is placed. The central venous pressure line is pulled back to beyond the caval–innominate junction. After assurance is given that the abdominal team is ready to proceed, the SVC is clamped or ligated distal to the azygous vein (this avoids sinoatrial nodal injury) (Fig. 63.1).

If the abdominal IVC is vented, the IVC is clamped at the level of the diaphragm. The heart is vented by transecting the left inferior pulmonary vein (or left atrial appendage if the lungs are being concomitantly harvested) and incising the anterior IVC proximal to the clamp. The aortic cross-clamp is applied, and the heart is arrested with cold cardioplegic solution. Adequate perfusion pressure is assessed by palpating the aortic root. The left ventricle should also be monitored to assure that it does not become dilated. Rapid cooling of the heart is achieved with 10 L of topical cold saline (4°C). After a successful arrest, cardiectomy is initiated by completing the transaction of the IVC (Fig. 63.2). The heart is wrapped in an ice-soaked lap pad, and the apex of the heart is elevated. Proceeding from inferior to superior, first on one side and then the other, one divides the remaining intact pulmonary veins and branch pulmonary arteries. When the lungs are being procured for transplantation, the procedure is modified to retain adequate left atrial cuffs and pulmonary arteries for both the heart and

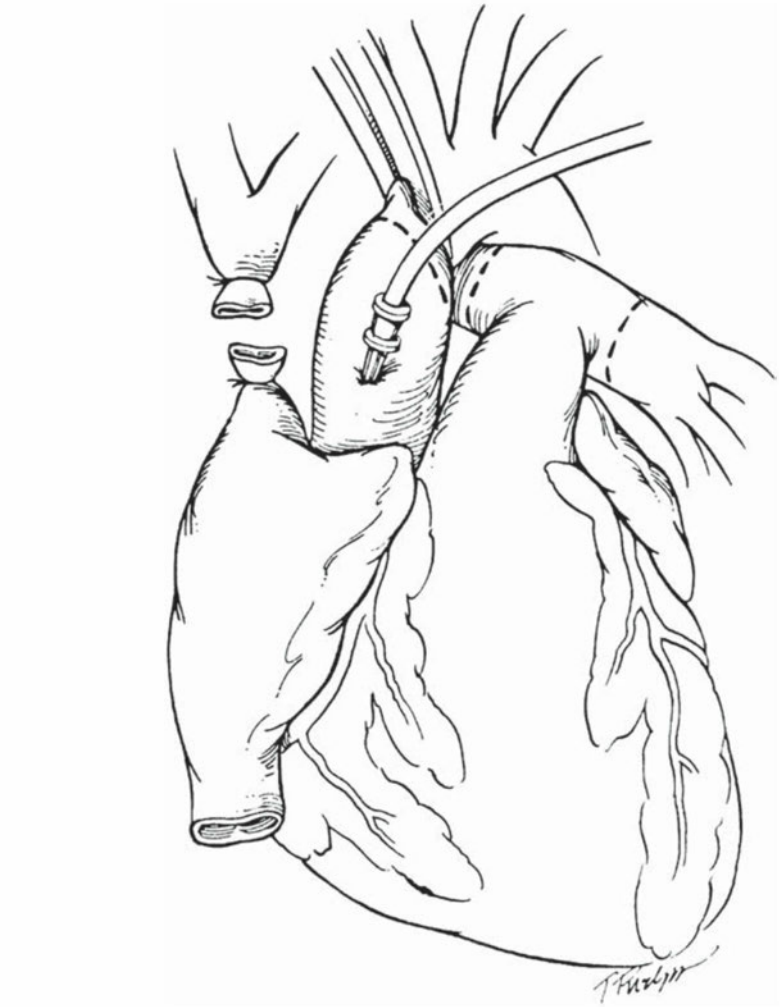


Fig. 63.1. Standard donor cardiectomy. *Dotted lines* identify aortic and pulmonary artery sites of division.



Fig. 63.2. Standard donor cardiectomy. After transaction of the inferior vena cava the heart is retracted cephalad and the pulmonary veins are transected.

lung implantations. The ascending aorta and SVC are transected last. The underlying disease of the recipient should be taken into account when determining the required length of aorta and SVC.

After explanation, the allograft is carefully taken to the back table and placed in a basin of cold saline for inspection and final preparation. The heart is examined for evidence of patent foramen ovale, vascular injuries (or inadequate length), and valvular abnormalities. Any positive findings should be disclosed to the implanting surgeon. The donor heart is then sequentially placed in two sterile bowel bags, each filled with cold saline, a sterile saline-filled air-tight container, and finally a standard cooler of ice for transport.

Several common pitfalls have been identified. Avoiding the following mistakes is vital to successful organ harvest:

1. Failure to monitor the heart closely during multiorgan dissection.
2. Failure to heparinize.
3. Right or left ventricular distension.
4. Failure to adequately cool the heart during harvest and transport.

ORGAN PRESERVATION

Most preservation techniques of the cardiac allografts permit a "safe" ischemic period of 4 to 6 hours, because ischemia times >4 hours have traditionally been associated with poorer outcomes. Multiple factors contribute to postoperative myocardial dysfunction including insults associated with suboptimal donor management, hypothermia, ischemia–reperfusion, and depletion of energy stores. The preservation method used by >90% of transplant centers is a single flush of a cold crystalloid cardioplegic solution followed by hypothermic storage. No single preservation regimen has demonstrated consistent clinically significant superior myocardial protection. Hypothermia is the cornerstone of organ preservation, and some experimental evidence suggests that 4°C provides the best protection. Perfusion of the donor heart with cardioplegic solution to achieve electromechanical arrest is an invaluable adjunct to topical hypothermia. Crystalloid solutions are classified as "intracellular" or "extracellular." Intracellular solutions, characterized my moderate-to-high concentrations of potassium and low concentrations of sodium, purportedly reduce hypothermia-induced cellular edema by mimicking the intracellular milieu. Examples of commonly used intracellular solutions include University of Wisconsin, Euro-Collins and Bretschneider (HTK). Extracellular

solutions, which are characterized by low-to-moderate potassium and high sodium concentrations, avoid the theoretical potential for cellular damage and increased vascular resistance associated with hyperkalemic solutions. Celsior, Hopkins, and St. Thomas Hospital solutions are representative extracellular cardioplegic solutions. Some centers choose to augment their solution with highly oncotic additives (mannitol, lactobionate, raffinose, and histidine), which theoretically counteract intracellular osmotic pressure to reduce hypothermia-induced cellular edema in the allograft. Several other groups have reported using other additives, including Krebs-cycle substrates and free radical scavengers. Recent developments include the investigation of continuous hypothermic perfusion using portable machines. In animal models, this technology appears to safely extend cold ischemia time up to 24 hours. During graft implantation, myocardial preservation can be augmented with continued use of topical cold saline and retrograde blood cardioplegia.

ORTHOTOPIC HEART TRANSPLANTATION

Nearly all heart transplants are performed in an orthotopic manner. The technique has changed little what was originally described by Shumway and Lower in the 1960s. The transplant surgeon must be well organized to avoid any technical or nontechnical mishaps. Special attention should be paid to assuring that an ABO mismatch does not occur. At our center, the operative surgeon must confirm the donor and recipient blood type prior to incision and again upon arrival of the donor organ. Patients who have been fully anticoagulated preoperatively should be corrected with a combination of vitamin K and fresh-frozen plasma. After the organ procurement team has confirmed that the donor allograft is acceptable, the recipient is taken to the operating room and placed under general anesthesia. Preferentially, central line placement should avoid the right internal jugular to minimize difficulties with future myocardial biopsies. High-dose narcotics are usually the primary agents for induction and maintenance of anesthesia. Inotropic and vasoactive agents should be readily accessible for the rapid management of induction-induced hypotension. Inhaled agents may be added, but their potential myocardial depressant effects limit widespread use in this patient population. Antibiotics should be given at least 30 minutes before skin incision. At our center, we routinely use aminocaproic acid therapy to minimize perioperative blood loss.

Operative Preparation of the Recipient

The heart is exposed via a medium sternotomy and vertical pericardiotomy. If the patient has had prior cardiac surgery, a femoral arterial line may be placed in case cardiac decompensation requires the emergent placement of an intra-aortic balloon pump or the initiation of peripheral cardiopulmonary bypass. In a patient with an LVAD, peripheral arterial cannulation may be preferential, because this often facilitates the aortic anastomosis. The patient is heparinized and prepared for cardiopulmonary bypass. In most cases, the aortic cannula is inserted just proximal to the origin of the innominate artery. Bicaval cannulation is performed with venous cannulae placed at the IVC–atrial junction and directly into the SVC. Umbilical tape snares are passed around the SVC and IVC. Bypass is initiated, the caval snares are tightened, and any additional cardiac mobilization is performed. After the donor heart has arrived, the ascending aorta is cross-clamped and the cardiectomy is performed. The aorta and main pulmonary artery are transected above the semilunar valves. Most centers now perform bicaval atrial anastomosis, and hence the SVC and IVC are transected at the caval–atrial junction. The left atrium is entered anterior to the right pulmonary veins and then incised along the atrioventricular groove to leave an adequate cuff for allograft implantation. Great care should be taken to avoid injury to the left superior pulmonary vein when excising the left atrial appendage. If a biatrial anastomosis is required, a right atrial cuff can be preserved.

To facilitate implantation, the proximal 1 to 2 cm of aorta and pulmonary artery are separated from one another with electrocautery. A weighted sucker placed in the left atrial remnant will keep the field free of blood and augments donor heart myocardial protection. Some centers routinely flood the operative field with CO_2 to assist with graft deairing.

Timing of donor and recipient cardiectomies is critical to minimize allograft ischemic time and recipient bypass time. Frequent communication between the procurement and transplant teams permits optimal coordination of the procedures. Ideally, the recipient cardiectomy is completed at the time of cardiac allograft arrival. At our institution, we have a protocol that mandates five telephone calls: first, when the recovery team arrives at the harvest site and assesses the donor, next after visualization of the organ in the operating room, then before cross-clamp at the harvest site, next on leaving the remote site, and finally when arriving locally. At any time, implantation team should be allowed to slow down the harvest process as needed to minimize organ ischemia time. In an era when many recipients have an LVAD at the time of transplant, this is especially crucial, preferably the harvesting surgeon should not cross-clamp the donor heart until given approval by the transplanting surgeon.

Implantation

The donor heart is removed from the transport cooler and placed in a basin of cold saline. The aorta and pulmonary artery are separated using electrocautery or sharp dissection. The left atrial cuff is prepared by connecting the pulmonary vein orifices and excising excess atrial tissue (Fig. 63.3). The cuff is tailored to the size of the recipient's left atrium. The tricuspid apparatus and intra-atrial septum are inspected. There have been several recent reports suggesting that there should be a low threshold for performing a donor tricuspid annuloplasty at the time of transplantation. Recipients are predisposed to increased right-sided heart pressures in the early postoperative period owing to pre-existing pulmonary hypertension and volume overload. Both the conditions are poorly tolerated by the recovering right ventricle. To avoid refractory arterial desaturation associated with right-to left shunting, if present, patent foramen ovale is over sewn.

Implantation begins with an end-to-end left atrial anastomosis (Fig. 63.4). A 54-inch, double-armed 4-0 polypropylene suture is passed through the recipient left atrial cuff at the level of the left superior pulmonary vein and then through the donor left atrial cuff near the base of the atrial appendage. The allograft is lowered into the recipient mediastinum, and the suture is continued in a running manner caudally and medially to the inferior aspect of the intra-atrial septum. The second arm of the suture is run along the roof of the left atrium and down the intra-atrial septum. Any size discrepancy between donor and recipient atria should be continually assessed and corrected. At our center, we often leave a weighted sucker in the left atrium (as a vent) at the completion of the suture line. The suture line is temporarily controlled with a Rummel tourniquet. After completion of all of the anastomosis, the heart is deaired, the vent is removed, and the suture line is secured. Some centers introduce a bubble-free cold line into the left atrial appendage for continuous hypothermic

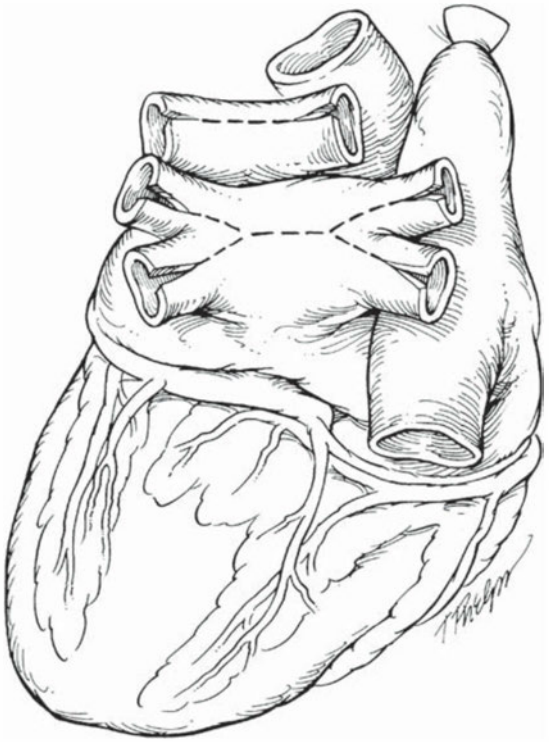


Fig. 63.3. Allograft preparation for orthotopic transplant. The pulmonary vein orifices are joined to form a left atrial cuff.

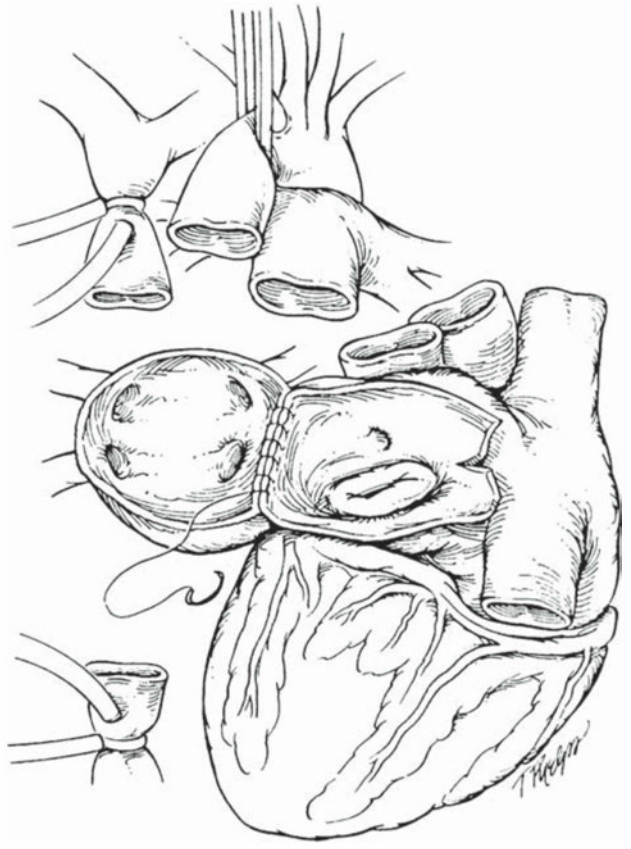


Fig. 63.4. Bicaval orthotopic heart transplant starts with the left atrial anastomosis.

saline lavage (50 to 75 ml/min) and evacuation of intracardiac air.

After completing the left atrial anastomosis, attention is turned to the IVC anastomosis. An end-to-end donor-to-recipient anastomosis should be performed using a 4-0 polypropylene suture. Again, great care should be taken to correctly manage any size discrepancy. This is often the most difficult of the five anastomoses and often is more easily performed from the right side of the table. Next, an end-to-end SVC-to-SVC anastomosis is constructed using a 5-0 polypropylene suture. We prefer to lock the posterior row of the anastomosis to minimize the risk of purse-stringing. When prolonged ischemia is expected after the completion of the two caval anastomoses, a dose of cardioplegia can be administered in either an antegrade or retrograde manner.

Next, an end-to-end pulmonary artery anastomosis is performed. It is crucial that the pulmonary artery ends be trimmed to eliminate any redundancy that might cause kinking and lead to right heart dysfunction. Using a 4-0 polypropylene, one starts the anastomosis at approximately 3 o'clock on the recipient pulmonary artery from outside to inside. The posterior wall of the anastomosis is then performed from the inside of the vessel, whereas the anterior wall is completed from the outside. If adjunctive hypothermia was used, the patient is rewarmed at this time. Finally, an end-to-end aortic anastomosis is performed in a similar manner. If a significant donor–recipient size mismatch is encountered when performing the aortic anastomosis, this may be handled by either beveling the aorta or creating a vertical aortotomy. In cases where prolonged ischemia times are encountered, the aortic anastomosis can be performed directly after the left atrial cuff anastomosis while the pulmonary artery and caval anastomosis can be performed with the cross-clamp off. The caval tapes are then released and the heart is filled with blood. Before the aortic anastomosis is secured, the lungs are ventilated and the heart is manipulated to facilitate deairing through the anastomosis. An aortic root vent/cardioplegia needle is then placed. If desired, a dose of warm cardioplegia can be administered before removal of the cross-clamp. Lidocaine is given, and the patient is placed in a steep Trendelenburg position. The aortic cross-clamp is removed, and deairing is continued via the root vent until no air is visible on transesophageal echocardiography.

Several maneuvers are performed during the resuscitation phase. Atrial and ventricular pacing wires are placed, pleural and mediastinal chest tubes are placed, the pulmonary artery catheter is advanced into proper position, and a thorough exploration for bleeding is undertaken. In patients with an implanted AICD or pacemaker, the device is removed at the time of transplant. The leads are transected during the

recipient cardiectomy and the generator is removed while the heart is being resuscitated. The patient is then weaned from cardiopulmonary bypass with the assistance of inotropes and nitric oxide as indicated. After removal of the cannulae and assurance of hemostasis, the wound is closed in the standard manner.

To assure excellent results, it is crucial to be hypervigilant during the early resuscitation period. This is especially important in cases with recipient pulmonary hypertension, prolonged organ ischemia, or when using a slightly undersized donor heart. In these cases, right ventricular failure may be potentiated by subtle changes in cardiac volume, acid–base status, or pulmonary artery pressure. The cardiac surgeon should work closely with the anesthesiologist to assure that all metabolic derangements are promptly corrected (e.g., hyperglycemia, hyper- or hypokalemia, respiratory or metabolic acidosis, hypocalcemia). Volume resuscitation should be closely monitored, especially when blood products are required to correct a coagulopathy. Pulmonary hypertension can be mitigated with rational use of inotropes, nitric oxide, and ventilator management. Finally, per the local protocol, immunosuppressive therapy should be started in the operating room.

ALTERNATIVE TECHNIQUES FOR ORTHOTOPIC HEART TRANSPLANTATION

Although most centers now perform the bicaval anastomosis, the original biatrial technique described by Shumway and Lower and revised by Barnard can often prove useful. We prefer the bicaval technique due to the associated increase in 5-year survival (81% vs. 62%), decreased risk of tricuspid regurgitation, reduced postoperative dependence on diuretics and inotropes, and lower incidences of atrial dysrhythmias, conduction disturbances, mitral and tricuspid valve incompetence, and right ventricular failure. To perform a right atrial cuff anastomosis, a curvilinear incision is made from the inferior vena caval orifice toward the right atrial appendage of the allograft (Fig. 63.5). The right atrial anastomosis is performed in a running manner similar to the left with initial suture placed at the most superior aspect of the intra-atrial septum.

Some centers have advocated total-heart transplantation, which involves complete excision of the recipient heart with bicaval end-to-end anastomosis and bilateral pulmonary venous anastomosis.

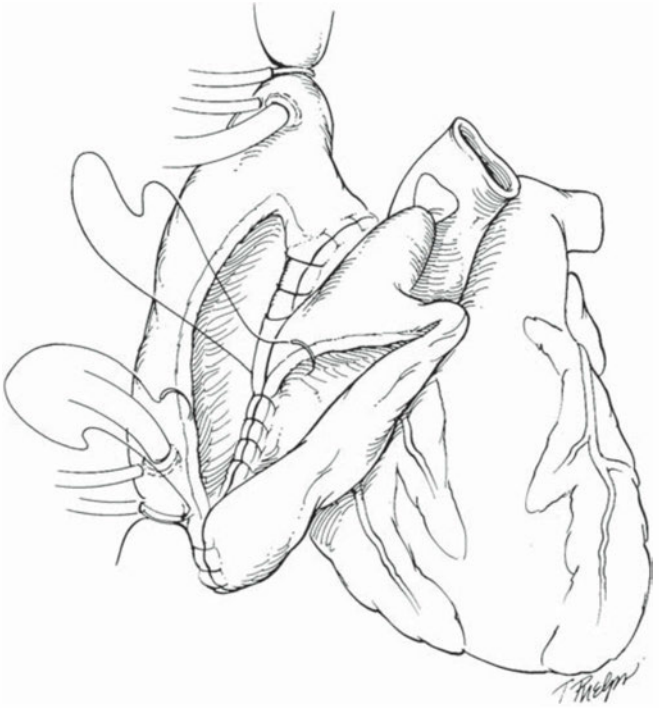


Fig. 63.5. Biatrial orthotopic heart transplant. After completion of the left atrial anastomosis, full-thickness bites are taken through the interatrial septum. Note the curvilinear incision made in the right atrium.

RECIPIENTS WITH CONGENITAL ANOMALIES

Transplantation in adults with previous palliative procedures for congenital anomalies is uncommon; however, it is critical that a generous donor cardiectomy be performed so that sufficient tissue is available for optimal reconstruction. Several authors have described techniques detailing heart transplantation in recipients with "L" transposition, hypoplastic left heart, previous Fontan procedure, and situs inversus. In most situations, an allograft cardiectomy that includes the branch pulmonary arteries, aortic arch, and innominate vein is required. The details of these specific reconstructions are well described in the literature and beyond the scope of this text.

REDO STERNOTOMY

At least 30% of heart transplant recipients will have had a previous median sternotomy. The use of preoperative chest X-rays or computed tomographic imaging may help to determine whether a potential space exists posterior to the sternum. External defibrillation pads should be placed before making an incision. In patients with previous coronary artery bypass surgery, an angiogram should be reviewed to determine which, if any, grafts are still open. If the proximal aorta must be retained, the stumps of any existing bypass grafts must be oversewn to minimize the risk of pseudoaneurysm formation. In patients who have had multiple previous median sternotomies or are redo transplants, it may help to expose the femoral vessels. If emergent bypass is required, it may help to start with a single venous cannula and transition to bicaval cannulation after the control of the situation has been achieved.

After completing a substernal dissection, the sternum is divided with oscillating saw (anterior table) and Mayo scissors (posterior table). The right atrium, SVC and IVC, and aorta are then mobilized to facilitate cannulation. The reminder of the heart is then mobilized and a low threshold for early use of cardiopulmonary bypass is observed. The implantation is performed in the standard manner.

TRANSPLANT AFTER VENTRICULAR ASSIST DEVICE

A growing number of patients will have a ventricular assist device (VAD) in place at the time of transplantation. These patients can provide many additional challenges. Femoral arterial cannulation is often preferred in these patients due to the risk of re-entry injury and to facilitate the

aorta–aorta anastomosis. If one chooses to primarily cannulate centrally, the outflow graft from the VAD often provides a flexible site for temporary arterial cannulation. After the initiation of cardiopulmonary bypass, the VAD is turned off, the aorta is cross-clamped, and the native cardiectomy is performed. It is often advantageous to amputate the left ventricular apex to facilitate cardiectomy and remove the VAD after reperfusion of the allograft. The extensive dissection required to remove the device predisposes these patients to bleeding. In addition, the current generation of LVADs requires that the patients be on anticoagulation; despite efforts to normalize the coagulation profile preoperatively, the patients often remain coagulopathic in the early postoperative period. Aggressive use of targeted diagnostics (thromoboelastogram, platelet functional assays, fibrinogen, PT, PTT) followed by directed transfusion is the basis of managing this potential problem. Some centers have advocated delayed primary closure in patients with uncontrollable posttransplant bleeding.

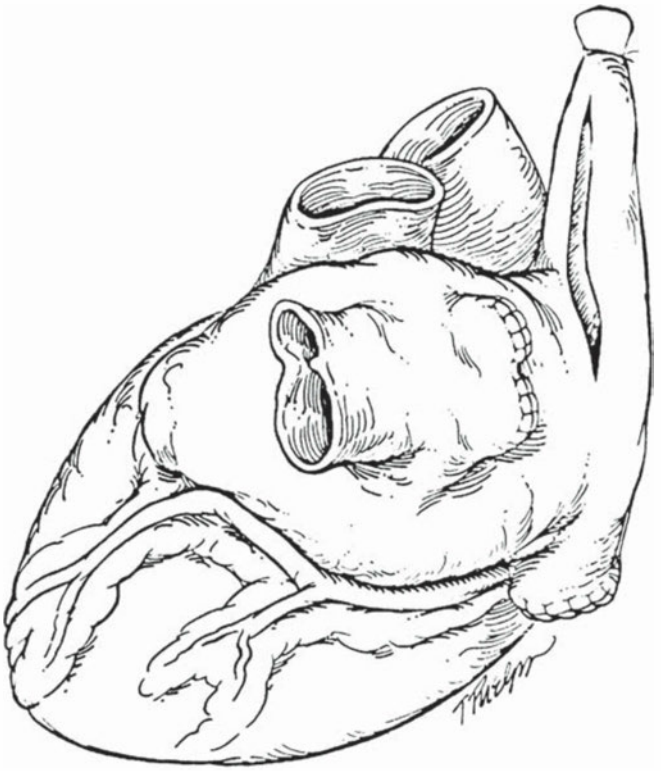


Fig. 63.6. Preparation of heart for heterotopic heart transplant.

HETEROTOPIC HEART TRANSPLANTATION

Heterotopic cardiac transplantation, the intrathoracic placement of an allograft in series with patient's heart, is rarely performed today. The procedure was first used clinically by Barnard in 1974. Today, the procedure may be indicated in cases with marked elevated pulmonary vascular resistance (pulmonary artery systolic pressure >60 mmHg and no pulmonary vasoreactivity) or when a donor heart is too small to sustain the recipient. Even in these selected cases, results have not been equivalent to orthotopic heart transplantation, with reported 1- and 5-year survival rates of 83% and 66%, respectively.

Similar to the cardiectomy performed for patients with congenital disease, the maximal lengths of aorta, SVC, and pulmonary arteries are procured. The IVC and the right pulmonary veins are oversewn, and a common left pulmonary vein orifice is created (Fig. 63.6). A linear incision is made along the long axis of the posterior right atrium extending 3 to 4 cm into the SVC. The recipient is cannulated in a bicaval manner as described, and the pericardium and right pleura are incised to permit placement of the allograft in the right chest. The sequence of anastomosis is as follows: donor to recipient left atrium, donor SVC to recipient right atrium, end-to-side aortic–aortic anastomosis, finally, end-to-side anastomosis joining the pulmonary arteries of donor and recipient (Fig. 63.7). The pulmonary artery anastomosis often requires the use of an aortic homograft of synthetic graft conduit permitting the donor heart to reside within the right pleural space.

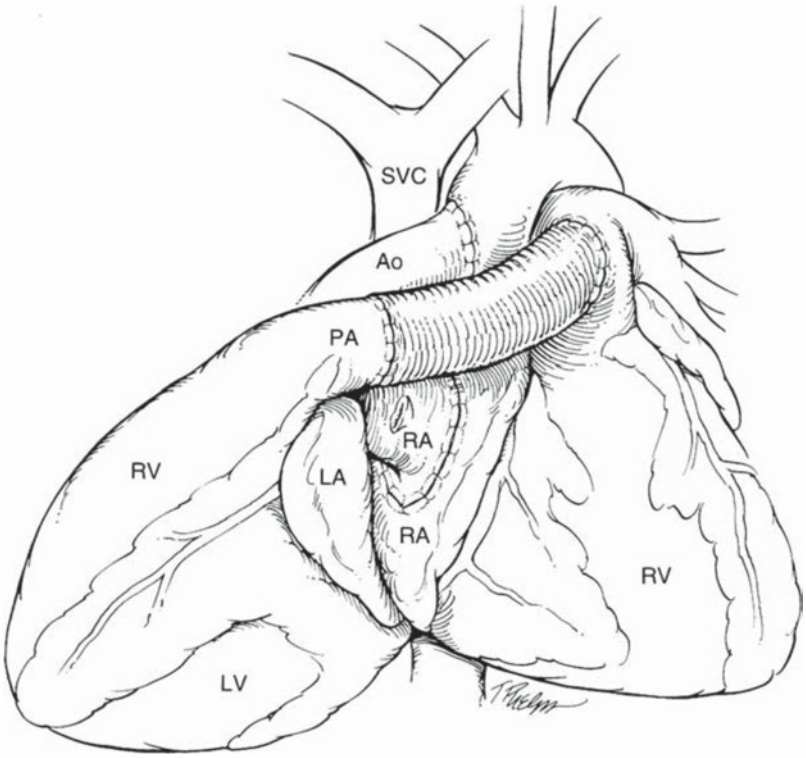


Fig. 63.7. Heterotopic heart transplant. An interposition graft has been used for the pulmonary artery anastomosis. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

POSTOPERATIVE MANAGEMENT

Postoperatively, the transplant recipient is usually admitted to a private room in the intensive care unit. Traditional "protective isolation" with elaborate positive-pressure air filtration systems has been abandoned by most centers because recent studies demonstrate no benefit over mask and hand-washing regimens. Contact with individuals with communicable diseases should be avoided.

Intensive care monitoring is analogous to that of any patient who has undergone cardiac surgery. A radial artery catheter, left internal jugular venous catheter, continuous telemetry, pulse dosimeter, oceanography, and Foley catheter are essential components for optimal postoperative care. Occasionally, a Swan–Ganz catheter is indicated in patients with increased pulmonary vascular resistance or if donor allograft dysfunction is anticipated. If the patient remains hemodynamically stable, all invasive monitoring lines and Foley catheter should be removed by 48 to 72 hours to minimize the risk of nosocomial infection. Chest tubes are removed on the first postoperative day if drainage is <25 ml/h.

Donor myocardial performance is transiently depressed in the immediate postoperative period. Allograft injury associated with donor hemodynamic instability and the hypothermic, ischemic insult of preservation contributes to the reduced ventricular compliance and contractility characteristic of the newly transplanted heart. With a biatrial anastomosis, abnormal atrial dynamic owing to the mid-atrial dynamics exacerbates the reduction in ventricular diastolic loading. An infusion of isoproterenol or dobutamine is initiated routinely in the operating room to provide temporary inotropic support. The positive chronotropic effect of isoproterenol is also therapeutic for the bradyarrhythmias that frequently complicate the early posttransplant period.

In patients with high pulmonary artery pressures or right heart failure, inhaled pressures nitric oxide is a therapeutic alternative. Intra-aortic or pulmonary artery balloon counterpulsation and right VADs have been utilized in patients unresponsive to medical therapy. Restoration of normal myocardial function usually permits the cautious weaning of inotropic support within 2 to 4 days.

Sinus or junctional bradycardia occurs in more than half of transplant recipients. The primary risk factor for sinus node dysfunction is prolonged organ ischemia. Adequate heart rate is achieved with isoproterenol infusion and/or temporary dual chamber epicardial pacing. Most bradyarrhythmias resolve over 1 to 2 weeks, although placement of a permanent pacemaker is necessary in 2% to 25% of patients; few are still pacer dependent at 6 months.

Most patients undergo an endomyocardial biopsy on approximately day 7 and are discharged shortly thereafter. All patients have close outpatient follow-up with intensive monitoring of their immunosuppression, antibiotic prophylaxis, and endomyocardial biopsies.

RESULTS

Cardiac transplantation continues to have operative mortality of approximately 5% due to primary graft nonfunction, acute rejection, sepsis, and pre-existing recipient comorbidities. Most patients will have an acute rejection episode during the first posttransplant, which is most often treated with corticosteroids. Patients with recurrent or refractory rejection are often treated with polyclonal antibodies, alternate immunosuppression, or antimetabolites. Chronic rejection is manifested by allograft coronary artery disease, for which the only available therapy is re-transplant. The use of calcineurin inhibitors (cyclosporine, FK506) is associated with a 3% to 10% incidence of chronic renal failure. The overall 1-year patient survival is now approximately 90%, with a 3-year survival of $>75\%$. Similar outcomes should be expected in patients that are bridged to transplant with an LVAD. There is every reason to believe that, with continued improvements in immunosuppression and treatment of transplant-related illnesses, these data will improve.

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EDITOR'S COMMENTS

Drs. Salerno and Verrier have done a complete analysis of heart transplantation. This has been a transformative procedure and is now practiced at many major cardiac surgical centers. The basic techniques have stood the test of time. The authors have done a very good job discussing about patient selection followed by the technical aspects.

I would like to focus on two issues that the authors have analyzed very thoughtfully. The most important is transplantation following a VAD. The majority of our patients now have VADs and these are different kinds of patients. We have seen issues with vasoplegia and have had to treat this systemic hypotension. In addition, the bleeding issues are real. I think there has to be excellent preparation and quick use of appropriate clotting factors including in some cases factor 7. The other important consideration is managing the right ventricle postoperatively. In the majority of patients, the left ventricle is working but often there is some preexisting pulmonary hypertension that would affect an unconditioned right ventricle. One must be very careful with administration of clotting factors to avoid RV distention and failure. It is critically important to get outstanding results. Programs are being very carefully monitored by CMS and I think the stakes are high for both the patient and the program.

ILK

64 Heart–Lung Transplantation

Ahmad Y. Sheikh, Hari R. Mallidi, and Robert C. Robbins

INTRODUCTION

Heart–lung transplantation represents the last hope for a small group of patients with severe combined end-stage disease. It was performed successfully at Stanford University in 1981, and these patients were the first to be completely supported by transplanted lung grafts. The Registry of the International Society for Heart and Lung Transplantation reports that >3,000 heart–lung transplants have been performed worldwide. In the earlier years, this included patients with primarily lung disease, such as emphysema or cystic fibrosis; more recently, it has been restricted to end-stage disease of both organs, such as patients with congenital heart disease and pulmonary vascular disease (*Eisenmenger syndrome*), or severe primary pulmonary vascular disease with end-stage right heart failure (*Cor pulmonale*). In addition, at our center, we have successfully transplanted patients with primary lung dysfunction in the setting of end-stage ischemic heart disease. About 20 to 30 heart–lung transplant procedures are performed yearly in the United States, with an additional 30 to 50 performed in the rest of the world.

RECIPIENT SELECTION

The primary objective in recipient selection is to identify individuals with progressively disabling cardiopulmonary or pulmonary disease who still possess the capacity for full rehabilitation after transplantation. Heart–lung transplantation has a greater potential for technical complications than either heart transplant or lung transplant alone, so that it is particularly important to have stricter recipient selection criteria to assure a reasonable chance of a successful outcome. To this end, older recipients (>60 years of age) have not usually been selected for heart–lung transplantation, whereas they are frequently candidates for heart transplant. Significant multisystem disease is a contraindication, although combined heart–lung and liver, or combined heart–lung and kidney, transplantation has been successfully performed. Contraindications include renal dysfunction, active malignancy, infection with HIV, hepatitis B or hepatitis C, severe liver disease, cachexia or obesity, drug or alcohol abuse, and inability to cooperate with treatment programs. Patients who have had previous thoracic surgery are evaluated on a case-by-case basis. Multiple previous thoracotomies or pleurodesis are not absolute contraindications, although they might represent higher risk for surgical teams without significant experience in performing the operation. A patient who is acutely ill in the intensive care unit and specifically on mechanical ventilation is generally considered to be too ill to undergo heart–lung transplantation.

Patients accepted for transplantation are listed on the National Transplant Registry on the basis of diagnosis, time on the waiting list, and ABO blood group. Because heart–lung transplant recipients are in competition with critically ill heart recipients who may have a more urgent status, the number of potential donors offered is low. The additional requirement for lung volumes of approximately equal or slightly smaller size further limits the potential donor pool. Like lung transplantation alone, patient height is a reasonable predictor for lung volume, and donors of significantly greater height should be avoided. The presence of preformed reactive antibodies (PRA) at a level of >25% requires a prospective specific crossmatch between the donor and recipient. The presence of a significantly elevated PRA will require specific measures, either pretransplant or posttransplant, to reduce the likelihood of hyperacute rejection of the donor organs, such as plasmapheresis and alternative immunosuppression. Diligent surveillance is also mandated in these patients to avoid the sequelae of late humoral rejection.

ORGAN PROCUREMENT AND PRESERVATION

Currently, all heart–lung block donors have sustained irreversible brain death but retain near-normal heart and lung function. Standard donor evaluation is performed, including physical examination, chest X-ray, 12-lead electrocardiogram, arterial blood gases, and serologic screening. A donor age of <50 years is preferred. Coronary angiography may be indicated in donors >40 years of age or with cardiac risk factors. Meticulous fluid management prevents excessive pulmonary edema and improves pulmonary and myocardial function. The last decade has seen significant progress toward better donor management, such that initially poor lung and heart function can be improved with aggressive treatment. Because of the already greater technical complexity of the heart–lung transplant operation, good donor organ function is extremely important. The criteria for donor selection are listed in Table 64.1.

The donor operation is performed via a median sternotomy, as shown in Figure 64.1. Some harvests may also be done with a bilateral transverse thoracotomy as shown, depending on the requirements of the abdominal organ teams. Both pleural spaces are immediately opened with inspection of both lungs carried out. The heart is inspected, to rule out significant contusions and anatomic anomalies. This is followed by palpation of the coronary arteries to rule out significant coronary artery calcifications. The lungs are briefly deflated, and the pulmonary ligaments are divided using electrocautery. After completely dissecting and removing the thymic remnant, the pericardium is removed to within about 2 cm of the phrenic nerves bilaterally. Umbilical tapes are placed around the ascending aorta and both venae cavae. The pericardium overlying the trachea is incised vertically, and the trachea is encircled in the superior part of the mediastinum at least five or six

Table 64.1 Heart–Lung Donor Selection Criteria

Age <50 years
Smoking history <20 pack-years
Arterial oxygen pressure of 140 mmHg on a fraction of inspired oxygen of 40% or 300 mmHg on a fraction of inspired oxygen of 100%
Normal chest X-ray
Sputum free of bacteria, fungus, or significant numbers of white blood cells on Gram and fungal staining
Bronchoscopy showing absence of purulent secretions or signs of aspiration
Absence of significant thoracic trauma
HIV-negative

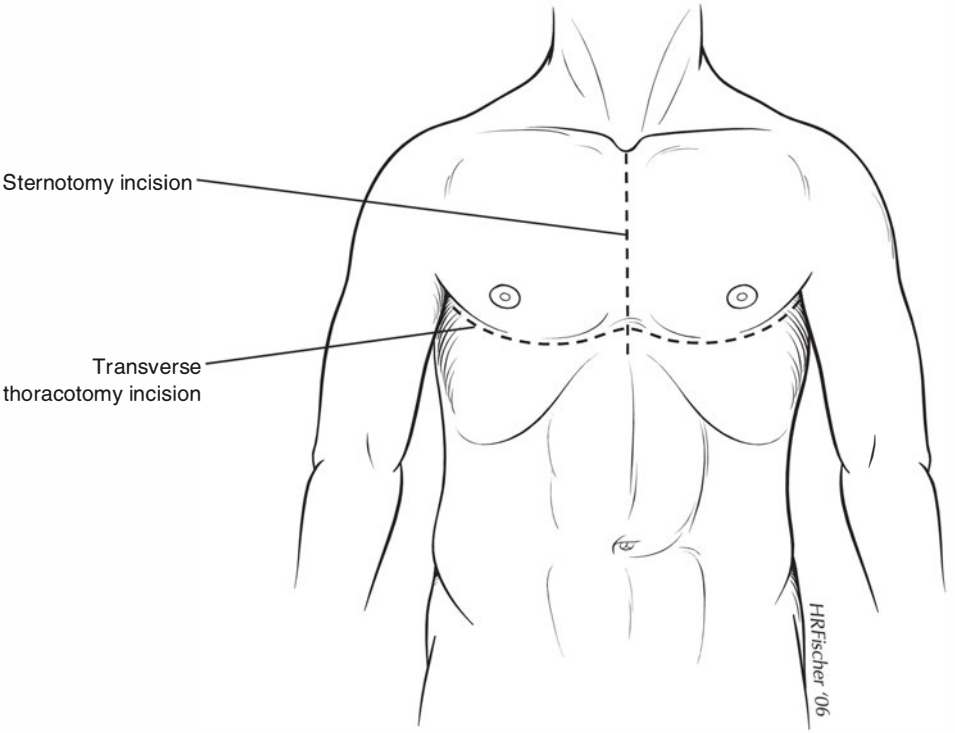


Fig. 64.1. Both the donor and recipient operations can be approached either through a median sternotomy or a bilateral transverse thoracotomy in the fourth intercostal space. Sternotomy is preferred, but bilateral transverse thoracotomy does provide better exposure of the posterior mediastinum and the apical spaces of both lungs.

complete tracheal rings above the carina. This maneuver can be facilitated by ligating and dividing the innominate vein.

An infusion of prostaglandin E_1 is started approximately 15 minutes before applying the aortic cross-clamp, beginning at a rate of 20 ng/kg/min, followed by incremental increases, to a target rate of 100 ng/kg/min. Careful monitoring of the mean arterial blood pressure should assure that it remains above 50 mmHg. Ventilation continues with a fraction of inspired oxygen (FiO_2) of 40% and a small amount of positive end-expiratory pressure (PEEP) (3 to 5 cm of water). Infusion lines are placed in the ascending aorta and the main pulmonary artery, as shown in Figure 64.2. The donor is then heparinized, the superior vena cava is ligated, and a straight Potts clamp is placed across the inferior vena cava. After the heart empties, the aortic cross-clamp is applied, and 10 ml/kg of cold crystalloid cardioplegia is rapidly infused into the aortic root. We prefer the Stanford formulation, but several other cardioplegia solutions are available. The inferior vena cava is then incised, and the tip of the left atrial appendage is amputated (Fig. 64.2), to avoid cardiac and lung distention from the return of the pulmonary perfusion. While antegrade cardioplegia is being delivered, a separate pulmonoplegia is flushed through the main pulmonary artery at a rate of 15 ml/kg/min for a period of 4 to 5 minutes. Ice-cold saline or Physiosol solution (Abbott Laboratories, North Chicago, IL) is immediately poured over the heart and lungs. During the cardioplegia and pulmonoplegia infusions, ventilation

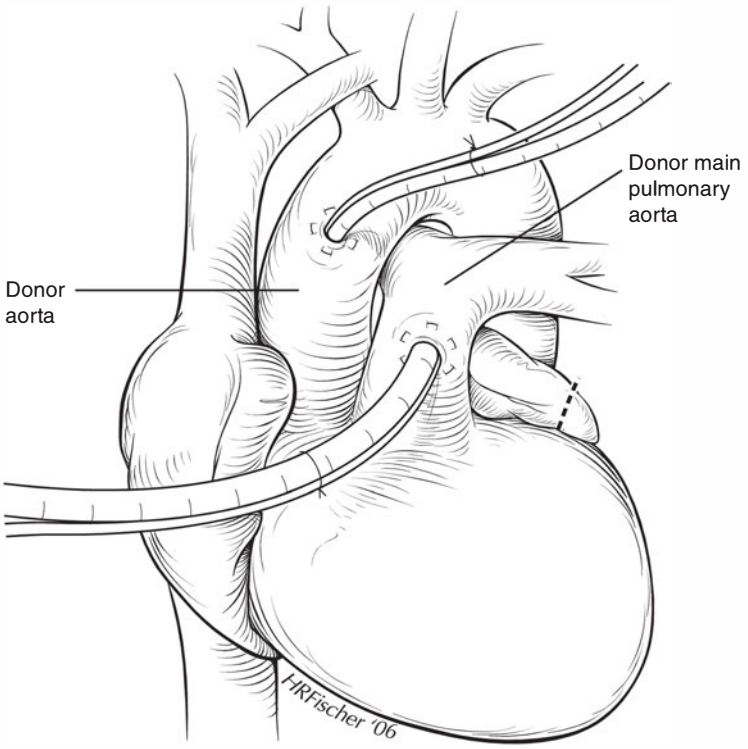


Fig. 64.2. After dissection and inspection of the donor organs, an infusion catheter is placed in the mid-ascending aorta and the mid-portion of the main pulmonary artery for the administration of cardioplegia solution and pulmonoplegia solution. The *dashed line* on the tip of the left atrial appendage shows the place that the heart is opened to allow decompression of the solution perfusing the lungs.

is maintained with half-normal tidal volumes of room air. On completion of these infusions and topical cold application, all solutions are aspirated from the thoracic cavity, and the lungs are fully deflated.

The heart–lung block is dissected free from the esophagus, commencing at the level of the diaphragm and continuing cephalad to the level of the carina. Dissection is kept close to the esophagus, and care is taken to avoid injury to the trachea, lung, and great vessels. The posterior hilar attachments are divided and the lungs inflated to a half-normal tidal volume, and the trachea is stapled at the highest point possible with a TA-55 stapler (United States Surgical, Norwalk, CT) at least four rings above the carina. The trachea is then divided above the staple line, and the entire heart–lung block is removed from the chest. After the heart–lung block is removed from the donor, it is wrapped in sterile gauze pads and immersed in ice-cold saline between 2°C and 4°C, placed in several sterile plastic bags, and then placed in a sterile plastic container. This is placed in an ice-filled insulated box and transported to the transplant center. Because nonpressurized aircraft might be employed, we believe it is important to avoid overdistention of the lung before stapling of the trachea, to prevent overexpansion during transport, especially if air transport is required.

With current preservation techniques and careful and atraumatic excision of the heart–lung block, procurements from as far as 1,000 miles from the transplant center and ischemic times up to 6 hours are reasonably well tolerated. The additional use of corticosteroids in the donor and white cell filtration before lung reperfusion in the recipient are also measures that contribute to improved preservation.

RECIPIENT OPERATION

The heart–lung transplant operation is composed of two phases of equal importance. The first is safe excision of the native organs with excellent hemostasis, followed by implantation of the donor organs and continued attention to excellent hemostasis.

Anesthetic monitoring includes arterial pressure, pulse oximetry, continuous electrocardiography, temperature, and urine output. A standard endotracheal tube is used, and transesophageal echocardiography may be helpful during the early stages of the operation, but should be removed during the time of recipient organ excision. This helps to prevent injury to the esophagus while dissecting in the posterior mediastinum. A median sternotomy is employed for the majority of recipients, with transverse thoracotomy (clam-shell) helpful for patients who have had extensive previous thoracotomies or pleurodesis (Fig. 64.1). In that situation, the bilateral thoracotomies offer better access to the posterior mediastinum and the apical areas of the thoracic cavity for a more meticulous dissection and better hemostasis. Otherwise, the median sternotomy is preferable for better chest wall mechanics postoperatively.

After sternotomy, both pleural spaces are opened widely just below the sternal edges. Anterior mediastinal thymic remnants and the anterior pericardium are carefully dissected and removed. The ascending aorta is dissected free and encircled with umbilical tape, followed by the superior vena cava and the inferior vena cava, as shown in Figure 64.3. Cannulation for cardiopulmonary bypass consists of a cannula in the high ascending aorta and separate vena caval cannulas, as seen in Figure 64.4. Once cardiopulmonary bypass has been initiated, the aorta is cross-clamped and tapes are placed down around the vena cava. The heart is then excised by dividing the great vessels just above the aortic and pulmonary valves in a manner quite similar to cardiectomy for heart transplantation (Fig. 64.5). When bicaval anastomoses will be used, we do like to leave a portion of posterior right atrium that connects the superior and inferior vena cavae to prevent them from retracting once the cardiectomy has been performed. The left atrium is divided across its mid-portion, leaving the pulmonary veins intact. Mobilization of the lungs is begun with separation of the pulmonary ligaments bilaterally. The pleural reflections over the anterior mediastinum are carefully opened, and dissection of the pulmonary veins is begun. As much as possible of the dissection of the hila can be performed before cardiopulmonary bypass, but cardiopulmonary bypass should be started if hemodynamic instability develops. Any pleural adhesions are divided using electrocautery with meticulous hemostasis. A useful adjunct helpful in maintaining hemostasis is the Argon Beam Coagulator (ConMed Corp., Utica, NY) for the control of diffuse bleeding from chest wall adhesions.

After cardiectomy, bilateral pneumonectomies are performed. The pulmonary veins and pulmonary artery are divided by stapling, as shown in Figure 64.6. In some particularly scarred hila, the electrocautery can also be used to divide the pulmonary veins and arteries.

In some patients, there may be significant bronchial artery collaterals, as well as other mediastinal vessels in the lymphatic and peribronchial tissue. All of these have to be carefully controlled with either the electrocautery or ligaclips. The right and left mainstem bronchi are stapled using the TA-30 (United States Surgical, Norwalk,

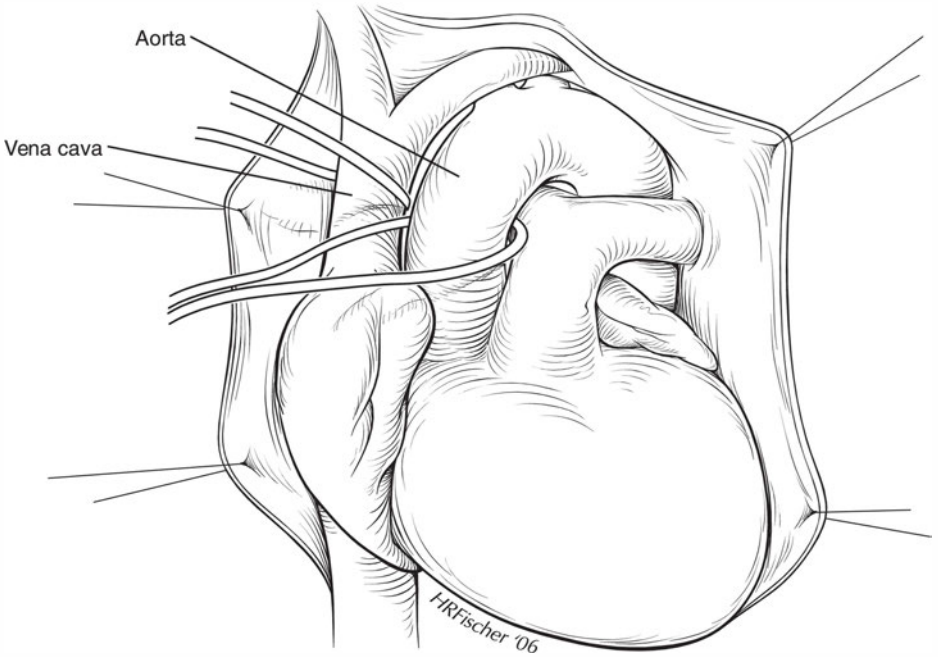


Fig. 64.3. After sternotomy, both pleural spaces are opened widely, just below the sternal edges. The aorta and vena cava are dissected and encircled with umbilical tapes.

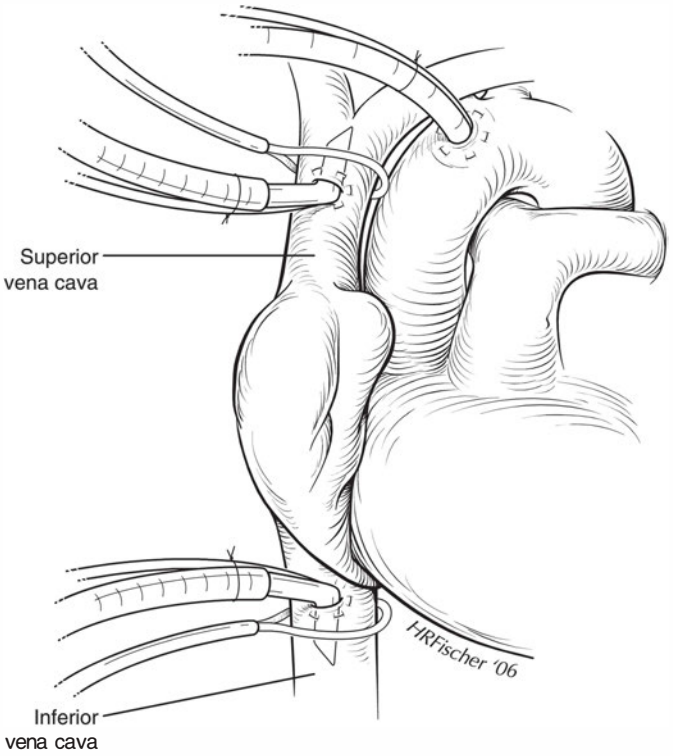


Fig. 64.4. After the median sternotomy, both pleural spaces are opened widely just below the sternal table. The ascending aorta and both vena cavae are dissected free and encircled with umbilical tapes. Cannulation for cardiopulmonary bypass includes a typical arterial return line in the high ascending aorta and a right-angled venous cannula in the superior vena cava and the inferior vena cava.

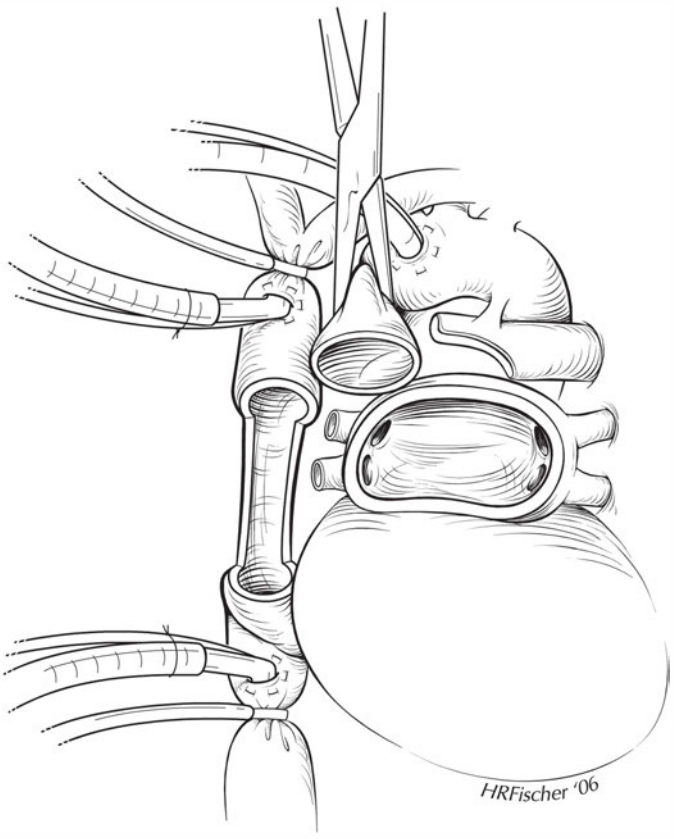


Fig. 64.5. Cardiectomy is performed in a manner quite similar to that for isolated heart transplantation. We prefer to leave an intact portion of posterior and medial right atrial wall that will connect the superior and inferior vena cavae and prevent their retraction, facilitating the anastomosis.

CT), dividing distally and then removing the lungs. At this point, very careful control of any bleeding in the posterior mediastinum is essential.

The next steps in preparing the recipient for implantation consist in creating openings for both the right and left hila in the lateral pericardial walls and dissecting the main bronchi back to the distal trachea, to divide the trachea and prepare it for the implantation.

Anterior to the pulmonary veins bilaterally, the left and right pericardium are opened and enlarged superiorly and inferiorly to provide enough room for the donor lungs. Careful identification of the right and left phrenic nerves should provide adequate protection against injury. By grasping the stapled ends of the right and left bronchus and using the electrocautery, careful dissection right against the bronchus can separate it from the posterior mediastinum and dissect it back into the central posterior mediastinum, where they meet at the carina (Fig. 64.7). Dividing the right pulmonary artery remnant just anterior to the trachea will help facilitate this exposure. We believe it is important to leave a portion of the left pulmonary artery intact adjacent to the underside of the aorta near the ligamentum arteriosum to preserve the left recurrent laryngeal nerve. It is also important to stay right on the bronchus and not to injure the tissue immediately posterior to the carina, where the right and left vagus nerves will be passing anterior to the esophagus. A common error is to divide or traumatize the vagii at this location, causing a high vagotomy with gastric paresis post transplant. Again, multiple bronchial vessels must be identified and carefully ligated using ligaclips or controlled with the electrocautery. Patients with Eisenmenger syndrome have fairly large bronchial collaterals, which are identified and ligated. Once perfect hemostasis has been obtained, the trachea is divided at the carina with a No. 15 blade. The surgeon should keep in mind that the membranous trachea should not be pulled down excessively and then divided, because this may remove a fair amount of the recipient posterior wall unnecessarily. Additional tracheal collaterals should be controlled at this point. The chest is now prepared to receive the heart–lung graft.

The donor heart–lung block is removed from its transport container and prepared by opening the trachea just below the staple line and carefully obtaining a swab culture, irrigating, and then aspirating the retained mucus. The suction used for this purpose should be kept separate from the

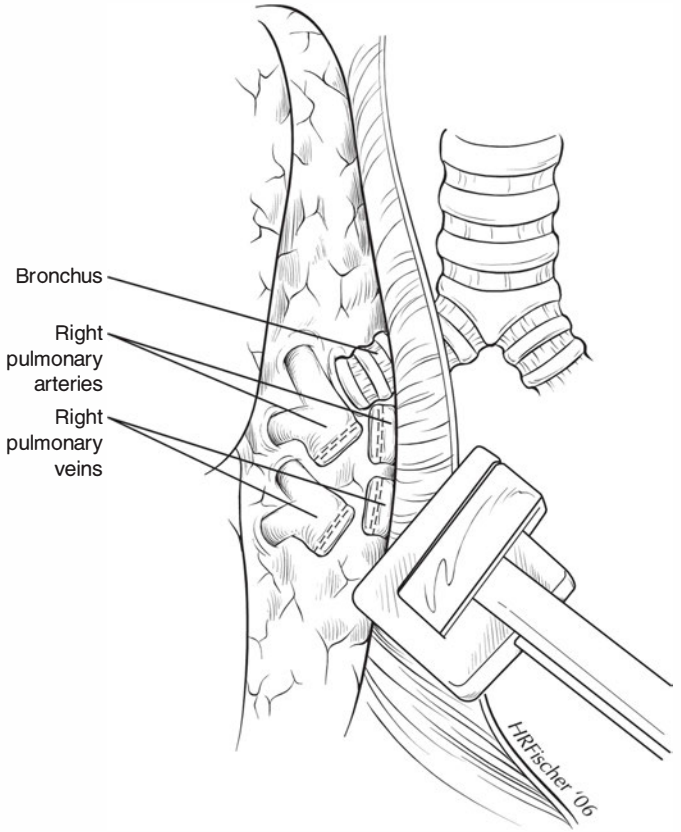


Fig. 64.6. Dissection of the lung hilum includes identification of the pulmonary veins and pulmonary artery, and stapling them on the recipient side, as a way to help control some of the mediastinal collateral adventitial vessels. Finally, the bronchus is also stapled to help maintain sterility of the thoracic cavity. The trachea will be divided and opened just before the tracheal anastomosis is performed.

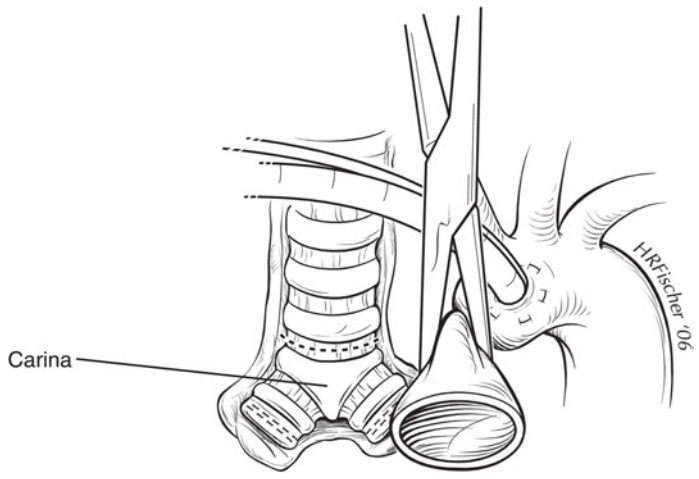


Fig. 64.7. The stapled ends of both the right and left bronchus can be grasped with a clamp and gently retracted to facilitate the retrograde dissection back to the carina and to the point of the incision in the trachea.

suctions used for the recipient operation. The cultures obtained are sent for fungal and bacterial examinations. The donor trachea is then trimmed down to leave just one cartilaginous ring intact above the carina. All of the peritracheal tissue should be retained as much as possible to facilitate post transplant vascularization of the anastomosis. Excessive dissection of this tissue will lead to possible necrosis or stricture, especially if a long left bronchial remnant remains with extensive skeletonization. Care should be taken to leave adequate tracheal tissue in the membranous portion posteriorly when transecting the donor trachea.

The heart–lung graft is then lowered into the chest, passing the right lung beneath the right phrenic nerve pedicle. The left lung is gently manipulated under the left phrenic nerve pedicle. An alternative technique that leaves one or both lungs anterior to the phrenic nerves has been described by Copeland's group. This maneuver facilitates rotation of the heart–lung block for better exposure during the post-bypass phase of the operation to inspect the posterior mediastinum for bleeding points.

Positioning of the donor organs before beginning the anastomosis is shown in Figure 64.8. The implantation begins with anastomosis of the trachea. In the adult patient, this is performed using continuous 3-0 polypropylene suture, as shown in Figure 64.9A. For children or neonates, appropriately smaller polypropylene suture is used. The posterior membranous portion is sutured first from the inside and from the left to the right, followed by completion of the anastomosis anteriorly, as shown in Figure 64.9B. The heart is wrapped with a sterile gauze pad, and topical myocardial hypothermia is initiated for the heart, with secondary topical cooling of both lungs.

Next, the bicaval venous anastomosis is performed beginning with the inferior vena cava, as seen in Figure 64.10. The donor interatrial septum should be looked at through the opening of the inferior vena cava to see whether a patent foramen ovale is present and needs closure. The inferior vena cava–right atrial junction is joined with a continuous 4-0 polypropylene suture. This is performed along the back wall of the right atrium, where the posterior atrial wall is intact, between the superior vena cava and inferior vena cava of the recipient. It is then continued along the free edge of the recipient inferior vena cava anteriorly. When this is completed, we usually proceed with anastomosis of the ascending aorta in a standard manner using 4-0 polypropylene suture. Finally, the superior vena cava is appropriately trimmed and sutured to the recipient vena cava using 5-0 polypropylene, with care to prevent purse-stringing of this anastomosis by leaving it intentionally looser. Excessive tension in this suture line will cause some stricture and possible later thrombosis.

Before release of the aortic cross-clamp, the amputated left atrial appendage is oversewn, and the pulmonoplegia site in the pulmonary artery is also repaired. A bubble vent in the anterior aorta is put to coronary suction as the tapes are removed from the vena cava, and blood is allowed to return into the heart and lung block. Just before this maneuver, we initiate white cell

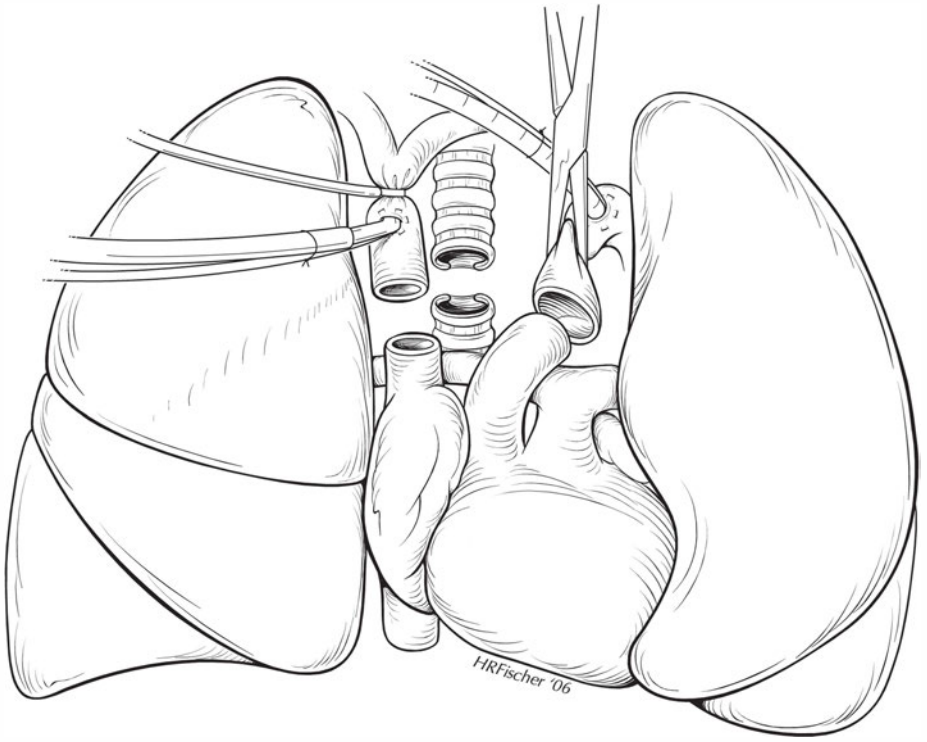


Fig. 64.8. The donor heart–lung block has been lowered into the thoracic cavity, positioning the right and left lung either anterior or posterior to the phrenic pedicle. The initial anastomosis performed will be the trachea.

filtration of the cardiopulmonary bypass circuit to reduce the exposure of recipient white blood cells to the donor lungs. The aortic cross-clamp is then removed so the heart will be reperfused.

The reperfusion and resuscitation of the heart–lung may require 30 to 45 minutes. Deairing and complete dilution of the pulmonoplegia solution are essential. Gentle ventilation with room air is performed prior to coming off of cardiopulmonary bypass. Weaning is initiated with an FiO_2 of 50% and adjusted based on the oxygen saturation measured peripherally. Some low-dose dopamine or isoproterenol may be helpful to maintain heart rate and improve renal perfusion. Methylprednisolone (500 mg intravenously) is given after protamine is administered. The appearance of the completed transplant is shown in Figure 64.11. Drains are left in both chest cavities and the mediastinum, and closure is routine.

CLINICAL MANAGEMENT IN THE EARLY POSTOPERATIVE PERIOD

The early postoperative management for heart–lung transplant recipients includes careful fluid and ventilatory management. The primary objective is to maintain adequate perfusion and gas exchange while minimizing intravenous fluid, cardiac work, and barotrauma. Careful and gentle pulmonary and endotracheal suctioning should be used to remove mucus and prevent atelectasis. Standard ventilatory weaning is initiated after the patient is deemed to be stable, awake, and alert. Typically, extubation is feasible within the first 24 hours after transplantation. As with all heart transplant recipients, some degree of transient sinus node dysfunction may be present in about 10% to 20% of patients. The use of isoproterenol or temporary cardiac pacing will usually lead to resolution within 1 week.

If pulmonary dysfunction develops, as manifested by an increasing requirement for FiO_2 with the development of a diffuse interstitial infiltrate on chest roentgenogram,

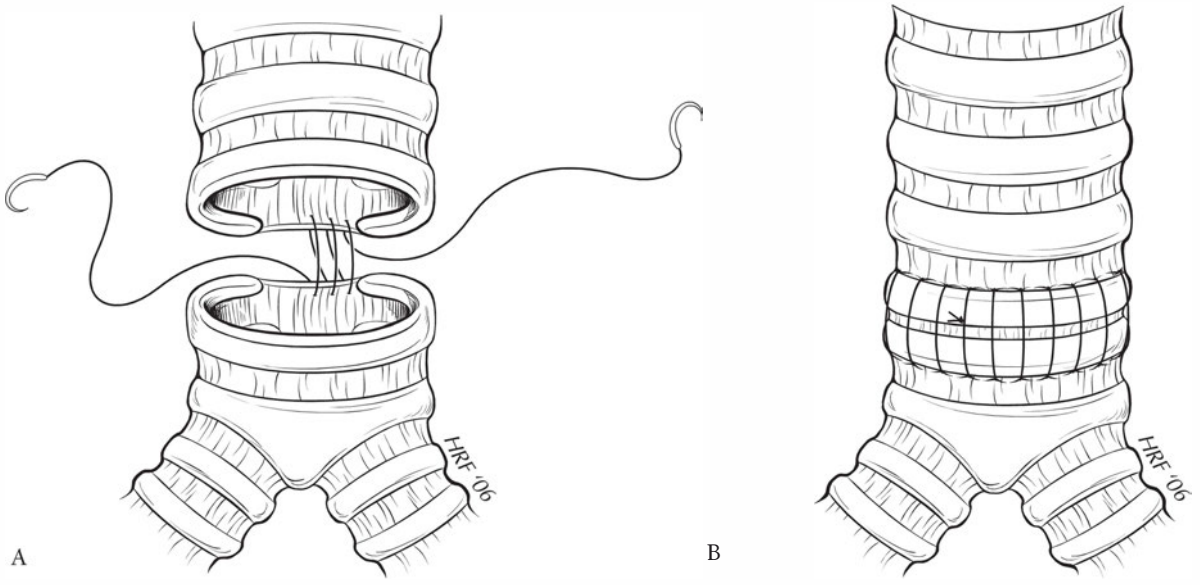


Fig. 64.9. (**A**) The tracheal anastomosis is started along the posterior wall at the junction between the cartilaginous portion and the membranous portion. A 3-0 polypropylene suture is used in a continuous running anastomosis from left to right and inside the trachea, being converted to an external suture line anteriorly. (**B**) The completed tracheal anastomosis is shown, with just one complete ring above the carina.

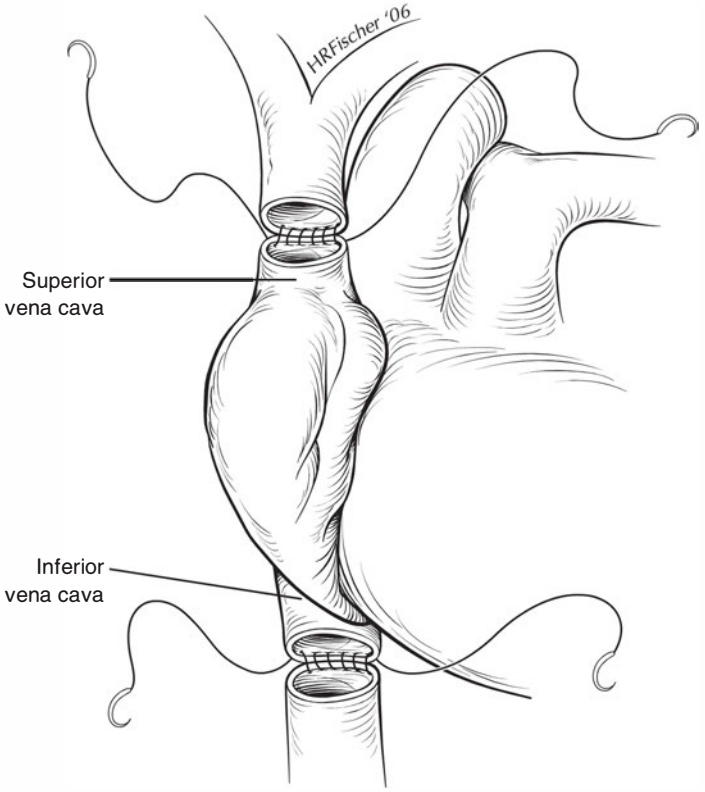


Fig. 64.10. The inferior vena cava anastomosis is started along the posterior wall and sutured from left to right. It is continued anteriorly, with care to avoid severe purse-stringing. Usually, this opening is large enough that this is not as much of a concern as with the superior vena cava.

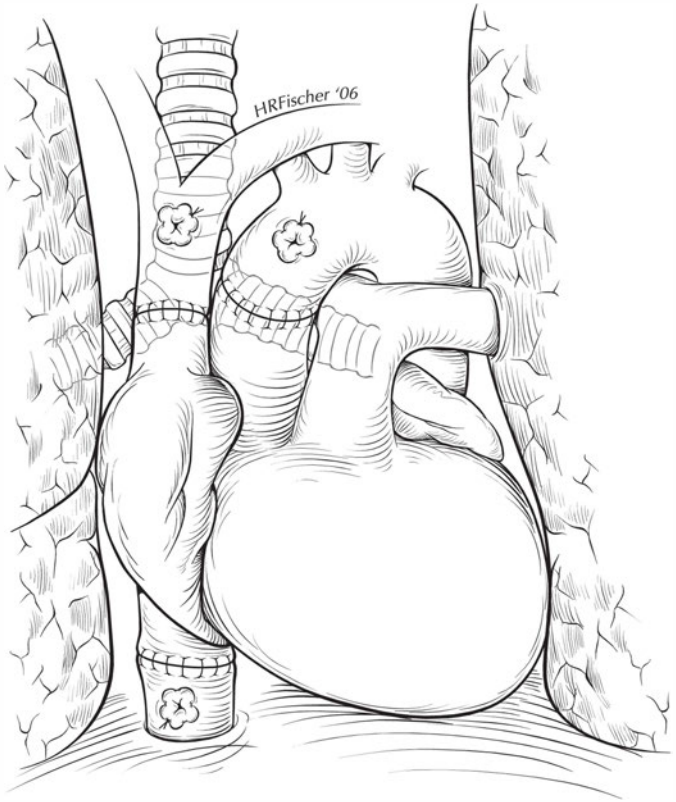


Fig. 64.11. The completed implantation of the heart–lung transplant. The tracheal anastomosis lies in the space between the superior vena cava and posterior to the ascending aorta. Angled chest drains are placed bilaterally and a straight drain in the anterior mediastinum. The chest closure is routine.

then the presence of a reperfusion injury should be considered. In addition, some element of obstruction of a pulmonary vein should be considered, because it has been reported after heart–lung transplantation. This is a particular risk when the heart–lung block is placed anterior to the phrenic nerves. This can be detected by transesophageal echocardiography. If primary graft dysfunction continues, the use of inhaled nitric oxide may be indicated. Temporary extracorporeal membrane oxygenation may be considered if iNO is not effective. Postoperative hemorrhage and the need for excessive blood product replacement will magnify any pulmonary dysfunction, again pointing out the importance of careful intraoperative hemostasis.

IMMUNOSUPPRESSIVE MANAGEMENT

The immunosuppression for heart–lung transplant recipients begins intraoperatively and continues for the patient's lifetime. The drug protocols used are similar to those for other lung transplant recipients. We have seen benefits from the use of induction therapy using rabbit anti-thymocyte globulin, with delay in the use of maintenance steroids for 2 weeks post transplant. However, for most patients we do not currently use induction therapy. During the early post transplant period, cyclosporine is initiated and switched to oral administration when extubation has occurred. Methylprednisolone is administered intraoperatively at graft reperfusion and continued for the first 24 hours at 125 mg intravenously every 8 hours. After 2 weeks, prednisone is started at a daily oral dose of 0.6 mg/kg and gradually tapered over the next month to 0.2 mg/kg/day.

INFECTION PROPHYLAXIS

In heart–lung transplantation, as in lung transplantation, very meticulous antiviral and antifungal prophylaxis are important components of postoperative management. Cytomegalovirus (CMV) prophylaxis with ganciclovir is used by most centers in any CMV-negative recipient receiving an allograft from a CMV-positive donor. Fungal prophylaxis against mucosal *Candida* infection includes the use of daily Nystatin mouthwash. *Pneumocystis carinii* prophylaxis consists of trimethoprim–sulfamethoxazole or aerosolized pentamidine. *Aspergillus* colonization is inhibited by the use of aerosolized amphotericin B early postoperatively. Recipients who are toxoplasma-negative and receive grafts from

toxoplasma-positive donors receive pyrimethamine prophylaxis for at least the first 6 months after transplantation.

GRAFT SURVEILLANCE

Routine clinical follow-up to monitor and modify the need for immunosuppressive drugs requires regular surveillance. This consists of serial pulmonary function tests, arterial blood gases, and bronchoscopic evaluation beginning at 2 weeks post transplant. Further surveillance bronchoscopies and biopsies are obtained on a regular schedule.

POSTOPERATIVE COMPLICATIONS

The importance of a meticulous and atraumatic operative procedure is highlighted by the fact that most of the early morbidity and mortality can be directly related to the quality of the donor organs and the need for excessive blood product administration due to postoperative hemorrhage. The most common cause of early death is due to primary graft failure or infection, together with multisystem organ failure due to poor primary graft function or excessive bleeding. This leads to a relatively high early postoperative mortality of 16% in our series of more than 200 heart–lung transplants.

If present, pulmonary dysfunction and multisystem organ failure are managed in a conventional manner, as for all critically ill postoperative patients. Careful attention to preservation of the graft, judicious fluid administration, and minimizing the need for excessive blood products postoperatively will reduce the incidence of this syndrome. Fortunately, as in heart or lung transplant-only recipients, acute rejection episodes and graft failure due to rejection are extremely unusual. The lung is more likely to experience rejection than is the heart, with about 67% of heart–lung patients having pulmonary rejection within the first year and only approximately 15% experiencing heart graft rejection.

CHRONIC REJECTION OR LATE COMPLICATIONS

The major long-term limitation of heart–lung transplantation, similar to isolated lung transplants, is the presence of chronic lung rejection presenting as obliterative bronchiolitis (OB). The diagnosis is confirmed with lung histology obtained by bronchoscopy, characterized by a dense eosinophilic submucosal scar, which partially or totally obliterates the lumen of small airways, accompanied by a decrease in arterial oxygen pressure, a decrease in the forced expiratory volume in 1 second, and a decrease in the forced expiratory rate between 25% and 75% of forced vital capacity. The only therapy is an augmentation of immunosuppression, which may be helpful in reducing the ongoing progression but usually does not reverse the disease.

RETRANSPLANTATION

When terminal respiratory failure develops secondary to OB or when chronic coronary artery disease develops (rarely) in long-term recipients, retransplantation of the heart–lung block, the lung alone, or of the heart alone has been performed. These operations represent technical challenges but have been reported to be successful in recipients who are otherwise in reasonably good physiologic condition. The same principles of meticulous donor procurement and preservation, attention to hemostasis, and careful perioperative management are essential. The major cause of death after retransplantation procedures remains multisystem organ failure.

AIRWAY COMPLICATIONS

Fortunately, the heart–lung graft facilitates revascularization of the tracheal anastomosis through the early development of coronary-to-bronchial collaterals. This has resulted in a very low rate of primary tracheal dehiscence, as well as a low rate of tracheal stenosis, which is usually due to ischemia. In fact, tracheal complications have tended to develop only in patients who required extended postoperative ventilation and increased PEEP. When complications develop, the diagnosis is made by bronchoscopy and can be treated by reoperation or bronchoscopy with dilation or airway stenting.

DOMINO-DONOR PROCEDURES

In patients who require a double-lung transplant, the strategy of performing a heart–lung transplant with donation of the recipient heart has been described as the "domino-donor transplant," with good results for both the heart–lung recipient and the heart recipient. These operations must be done with complete bicaval anastomosis so that an adequate donor heart is procured. Otherwise, the procedures are as already described.

HEART–SINGLE-LUNG TRANSPLANT

A small number of patients with congenital heart disease have been reported who have unilateral pulmonary vascular disease or severe developmental changes in one lung, which have been successfully transplanted with a heart–single-lung block. This strategy has also been used when severe scarring of one thoracic cavity has made it impossible to safely remove one of the recipient's lungs. A single bronchial anastomosis with appropriate pulmonary vein and pulmonary artery anastomosis are then performed. Good long-term results can be obtained.

LONG-TERM RESULTS

The long-term survival for heart–lung patients has gradually improved with better operative and postoperative management. Newer immunosuppressive drugs and better antimicrobial prophylaxis, particularly for CMV, have been major factors. In the last 10 years at Stanford, the 1-, 5-, 10-, and 15-year survival rates were 75%, 55%, 40%, and 30%, respectively, in 217 patients. The currently longest-living heart–lung transplant recipient is 23 years post transplant, with normal heart and lung function.

CONCLUSION

The evolution of heart–lung transplantation for end-stage cardiopulmonary disease has depended on careful operative technique. It can be a very straightforward operation in some patients and a very demanding and difficult procedure in others. The importance of appropriate donor organ procurement and preservation cannot be overemphasized. Meticulous hemostasis and the use of all possible measures to reduce postoperative hemorrhage are essential to good outcomes. Patients who experience successful procedures can look forward to long-term survival equivalent to any recipient of a bilateral lung graft, with very satisfactory and rewarding return to normal activities. Future improvements in immunosuppression and the ability to induce immunologic tolerance will continue the evolutionary improvement of heart–lung transplantation therapy.

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EDITOR'S COMMENTS

Heart–lung transplantation was developed at Stanford University under the leadership of Dr. Bruce Reitz. It has been a very exciting procedure and we have had the opportunity to do several of these procedures here at The University of Virginia. The difficult is getting heart–lung blocks. Double-lung transplantation has now been used for Eisenmenger's syndrome and certainly single-lung transplantation is available for other kinds of primary lung disease. We have also performed intercardiac repairs in addition to lung transplantation in patients with ventricular septal defects that resulted in Eisenmenger's syndrome. At present, the only major indication for heart–lung transplantation is cardiomyopathy in addition to end-stage lung disease. I think we will become much more clever as time goes on in terms of other application of lung transplantation. I believe heart–lung transplantation should be in every major transplant center's skill set but will be infrequently used.

ILK

65 Surgical Treatment of Atrial Fibrillation

Sameh M. Said and Hartzell V. Schaff

BACKGROUND

Atrial fibrillation (AF) is by far the most common cardiac arrhythmia, and the prevalence increases with age, from 0.5% for patients in their 1950s to almost 9% among patients 80 years of age and older. It is estimated that approximately 2.3 million people in the United States have AF, and the incidence will continue to increase with increasing age in the population.

AF has three main serious sequelae: (1) patient anxiety due to rapid heart rate, (2) deterioration of cardiac hemodynamics related to loss of synchronous atrioventricular contraction, and (3) increased risk of thromboembolic complications and stroke. The basis of AF management is directed toward these complications and includes control of heart rate, prevention of thromboembolism, and, when possible, reestablishing and maintaining sinus rhythm. There is continued controversy regarding the efficacy of rhythm control versus rate control strategies. Unfortunately, long-term success of medical treatment of AF is poor. Five years after entry into the AFFIRM trial, only 63% of patients assigned to the rhythm control group were in sinus rhythm. Furthermore, chronic medical therapy has a number of unwanted side effects.

For these and other reasons, there has been considerable interest in novel strategies for control and treatment of AF over the last three decades. One of the earliest approaches to control AF was "ablate and pace strategy" with atrioventricular nodal ablation and permanent pacemaker implantation. Although atrioventricular node ablation controls the heart rate, the patient remains in AF, is pacemaker dependent, and still incurs the risks of thromboembolic stroke. Compared with patients with AF managed medically, patients who undergo atrioventricular node ablation and permanent transvenous pacing exhibit similar exercise performance and ventricular function. In patients with diminished left ventricular function, atrioventricular nodal ablation and permanent pacing result in a small but significant improvement in left ventricular function compared with medical therapy alone.

Through the work of Cox and associates, surgical and catheter techniques for ablating AF have been developed and are highly refined. Patients with AF who come to surgical attention fall into two general groups. First, are patients with isolated (lone) AF who are symptomatic despite medical treatment and/or attempts at catheter ablation. In the second, larger group, are patients with AF and associated structural heart disease that requires operative treatment. Selection of a surgical ablative method differs depending on the indication for operation, the disability caused by the arrhythmia, and the anticipated morbidity of the procedure for AF and any additional necessary surgery. For example, the standard "cut-and-sew" maze procedure may be the most effective and appropriate treatment for a highly symptomatic patient with isolated AF, but the technique would not be appropriate for an elderly patient undergoing double-valve replacement who has few symptoms from AF and whose heart rate is easily controlled medically.

SURGICAL ABLATION

The Cox-Maze Procedure

The prototype for all current surgical procedures for AF is the maze procedure developed by Cox and coworkers, and we consider the standard "cut-and-sew" maze procedure to be the most reliable and effective method in creating complete transmural atrial lesions for AF ablation. We continue to use the "cut-and-sew" maze (Figs. 65.1–65.6) procedure in selected patients with only two modifications from the original description of the procedure. First, we have replaced the incision on the medial aspect of the right atrium from the cut edge of the right atrial appendage to the tricuspid valve annulus with a cryolesion; this avoids the division of the frequently seen branch of the right coronary artery that supplies the sinoatrial node and reduces the risk of postoperative sinus node dysfunction (Fig. 65.2A and 65.2B). The second modification involves the left atrium where we prefer to extend the incision that encircles the pulmonary veins to the orifice of the left atrial appendage, and then, after excising the appendage, close the orifice transversely as part of the encircling incision. Alternatively, cryolesions can be utilized as part of the lesion encircling the pulmonary veins when the exposure of this area is poor, and incisions may be difficult to close.

Many other surgical procedures have been proposed for the treatment of AF, and these operations utilize a variety of lesion sets and energy sources. Each of the modified procedures has potential advantages regarding ease of application and, in some cases, minimally invasive approaches. However, there is no conclusive evidence that any of the modified procedures yields better or even similar results to the original operation described by Cox.

Alternate Surgical Approaches for Atrial Fibrillation Ablation

Bilateral Pulmonary Vein Isolation

Pulmonary vein isolation (PVI) can be performed epicardially or endocardially

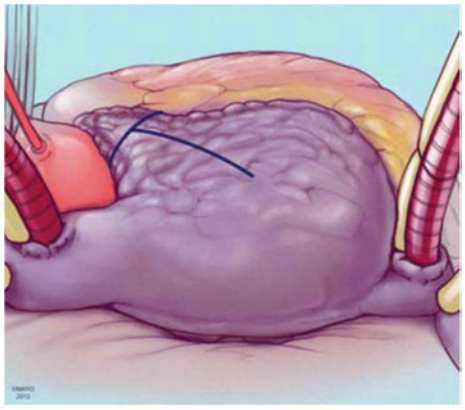


Fig. 65.1. Modified "cut-and-sew" Cox-maze III procedure. The first incision includes amputation of the right atrial appendage and extension on the right atrial free wall for approximately 5 cm. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

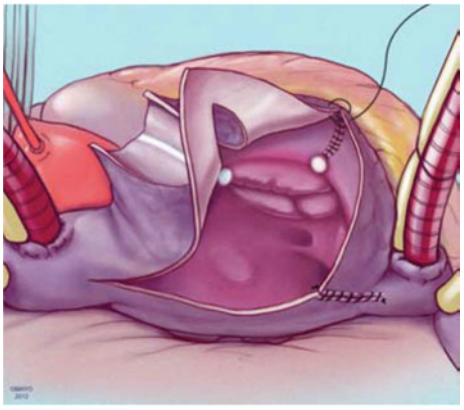


Fig. 65.3. Modified "cut-and-sew" Cox-maze III procedure. The vertical atrial incision is extended toward the tricuspid valve annulus, but it is stopped short of the right coronary artery. A cryolesion is then created at the remaining portion of the atrial muscle to the tricuspid valve annulus. Alternatively, this incision on the inside of the right atrium can be replaced with a cryolesion. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

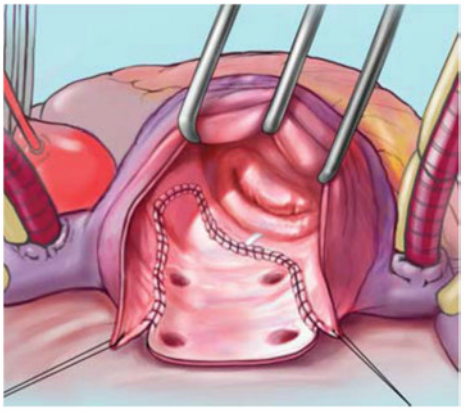
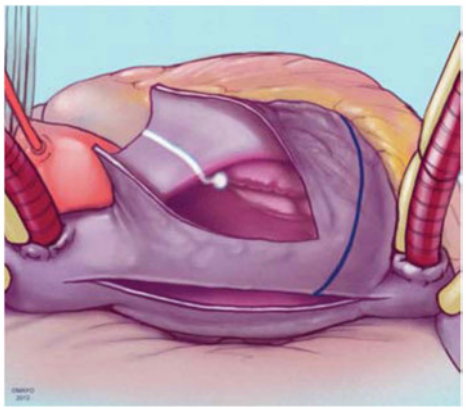
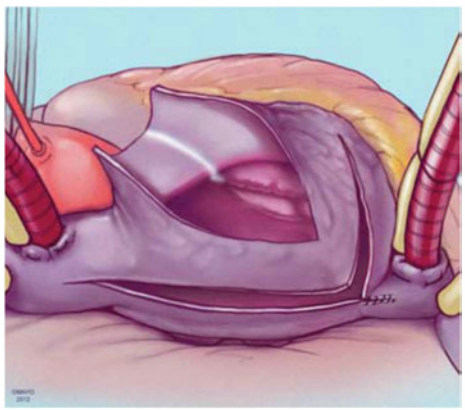


Fig. 65.5. This figure illustrates the left atrial portion of the Cox-maze III procedure, which includes an incision that encircles the pulmonary veins and extends to the amputated left atrial appendage. The left atrial appendage is closed vertically with the line of closure joining the pulmonary vein encircling incision. There is an additional cryolesion (*white line*) that joins the pulmonary vein encircling incision to the mitral valve annulus. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)



A



B

Fig. 65.2. (**A**) Modified "cut-and-sew" Cox-maze III procedure. A second incision extends from the superior to the inferior vena cava at the level of the crista terminalis. Another vertical extension of that incision is made toward the atrioventricular groove. (**B**) In a modification to the original Cox-maze III procedure, we replaced the incision on the medial aspect of the right atrium from the cut edge of the right atrial appendage to the tricuspid valve annulus with a cryolesion. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

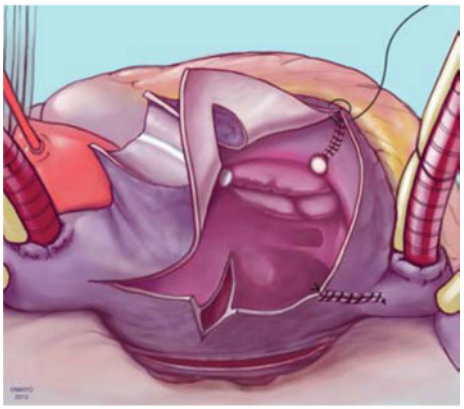


Fig. 65.4. Modified "cut-and-sew" Cox-maze III procedure. The interatrial septum is incised just cephalad to the fossa ovalis. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

with or without left atrial appendage exclusion or excision. We view left atrial appendage ligation as an important step in surgical treatment of AF and never omit this. Epicardial PVI may be useful as an adjunctive procedure to operations that do not include left atriotomy (Fig. 65.7). When the left atrium is opened, as is the case with mitral valve repair or replacement, a bipolar radiofrequency (RF) clamp can be used to isolate the pulmonary veins by connecting the left atriotomy to the stump of the amputated left atrial appendage ("box lesion").

If the patient can be converted to sinus rhythm, it is important to check for exit block to confirm interruption of conduction from the pulmonary veins. However, demonstration of exit block or conduction delay acutely does not guarantee that lesions are transmural and that conduction will not reappear late after operation. Also, it is important to recognize that epicardial application of RF or cryoclamps to the atrial cuffs around the pulmonary vein orifices results in a double layer of atrial tissue to be ablated; with this method, transmurality may be difficult to achieve, especially in a beating perfused heart.

Minimaze Procedure

The term "minimaze" commonly refers to minimally invasive epicardial procedures without the use of cardiopulmonary bypass (Figs. 65.8 and 65.9). Lesion sets include a combination of pulmonary vein encircling lesion, left atrial isthmus lesion to the orifice of the left atrial appendage, and lateral left atrial incision without cryolesions. Wolf describes a minimally invasive procedure for AF performed through bilateral thoracoscopic ports. The operation includes bilateral antrum isolation and partial cardiac denervation. More recently, Sirak and associates describe the "five-box" thoracoscopic maze. In this procedure, bilateral thoracoscopy is performed with extensive dissection of the dome of the left atrium and transverse sinus. The procedure aims at compartmentalization of the posterior left atrium with ablation lines and connecting these lines to the mitral valve annulus, thus interrupting all the possible reentry circuits for AF.

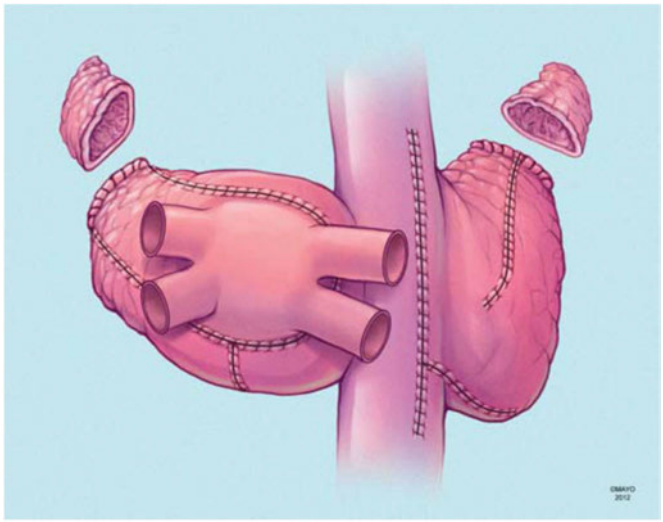


Fig. 65.6. Posterior view of the atria illustrating the completed incisions for the modified "cut-and-sew" Cox-maze III procedure. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

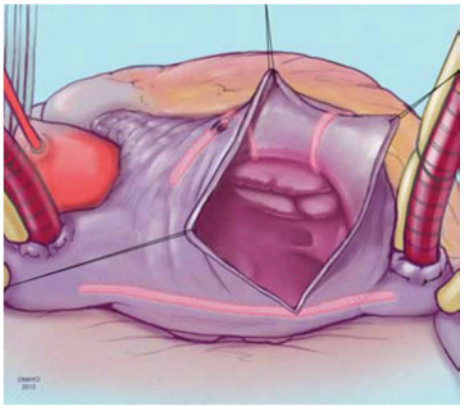


Fig. 65.8. Alternate energy sources can also be used to reproduce right atrial lesions as performed as part of the minimaze procedure. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

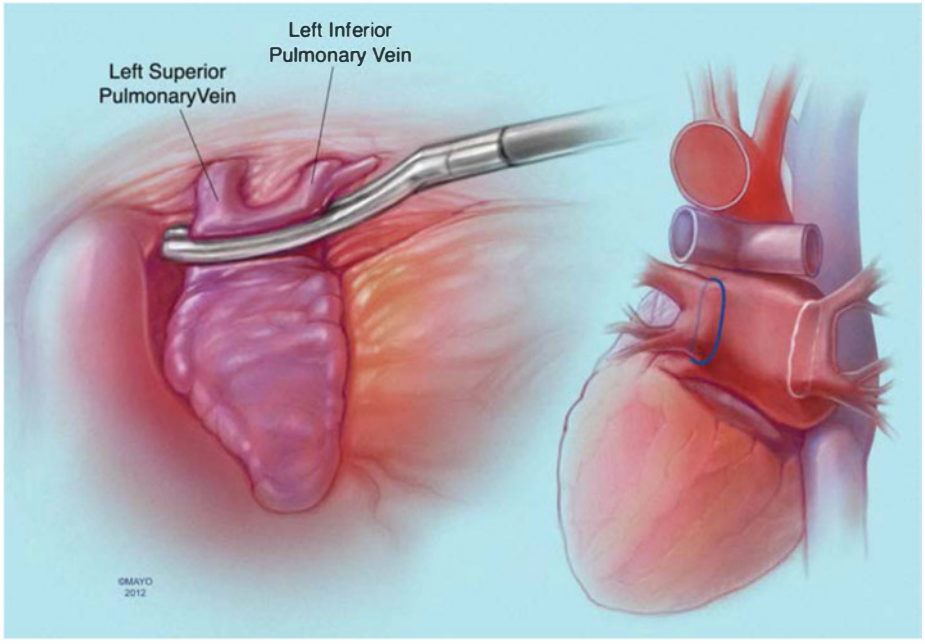


Fig. 65.7. Pulmonary vein isolation can be achieved with alternate energy sources such as the bipolar radiofrequency clamp illustrated here. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

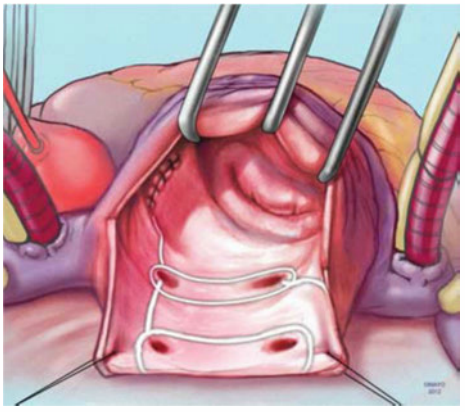


Fig. 65.9. Cryolesions can be used for linear ablation in any lesion pattern. Cryoablation is especially useful for completing encirclement of the left pulmonary veins in a standard maze procedure when exposure of the area is difficult. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Saline-Irrigated Cooled-Tip Radiofrequency Ablation

A "triangle-like" lesion set is created using a saline-irrigated, cooled-tip RF ablation. PVI is performed within the confines of the triangle, as well as left atrial appendage exclusion. One vertex of the triangle meets at the midportion of the posterior mitral valve annulus.

Alternate Energy Sources

The use of alternate energy sources has been increasing in the recent era due to the perceived technical complexity of the classic "cut-and-sew" Cox-maze. Cryoablation can be achieved using either nitrous oxide or argon cryoprobes, and the primary difference between the cooling techniques is the speed at which cooling of tissues occur. A purported advantage of cryoablation is the preservation of the cardiac fibrous skeleton. Cryoablation has been used extensively in clinical practice and appears quite effective in the arrested heart. However, in the beating heart with atria that are blood-filled and perfused, complete transmural lesions may be difficult or impossible to create. Additional cooling time is necessary because of warming of the tissue by the blood interface, and there is a theoretical risk of thrombus formation within the atrium.

RF energy employs alternate current to transfer energy to atrial tissue, and a variety of instruments have been developed including rigid unipolar probes with cooled tips, flexible unipolar probes, and bipolar clamps with and without irrigation. RF ablation can be applied to the epicardial or endocardial surfaces of the atrium in a unipolar configuration. Bipolar RF probes configured as clamps have the advantage of focused discrete lesions, thus, minimizing the potential risk of injury of adjacent structures. Saline irrigation of the tissue

during RF ablation has a cooling effect that prevents char accumulation and increases energy transfer to the tissues. Animal studies suggest a higher success rate of creating transmural lesions with irrigated delivery of RF compared with ablation without irrigation.

Less commonly used alternate energy sources include microwave energy where transmission of electromagnetic waves ablate tissues through the heat produced; with this method, there has been concern about damage to adjacent structures including reported cases of coronary artery stenosis. With laser energy, the tissue is heated directly producing photocoagulation. The transmurality of the lesion as well as the concern for collateral damage are the limiting factors. High-intensity focused ultrasound is promising because of its ability to ablate at specific depth.

Left Atrial Ganglion Ablation

Ectopic impulses originating from the autonomic ganglia adjacent to the pulmonary veins have been implicated in initiating AF. These ganglia are found in the fatty epicardial tissue and have been recently mapped and ablated, which reduces the recurrence of medically refractory AF. The standard locations of these active ganglia have been previously described. The procedure is typically performed through median sternotomy and with the use of cardiopulmonary bypass. Intraoperative mapping is required with 12 V of high-frequency stimulation at a cycle length of 50 milliseconds and a pulse width of 1.5 milliseconds. An active ganglion is identified if stimulation results in a doubling of the R–R interval. All active sites were immediately ablated with an RF pen (Fig. 65.10).

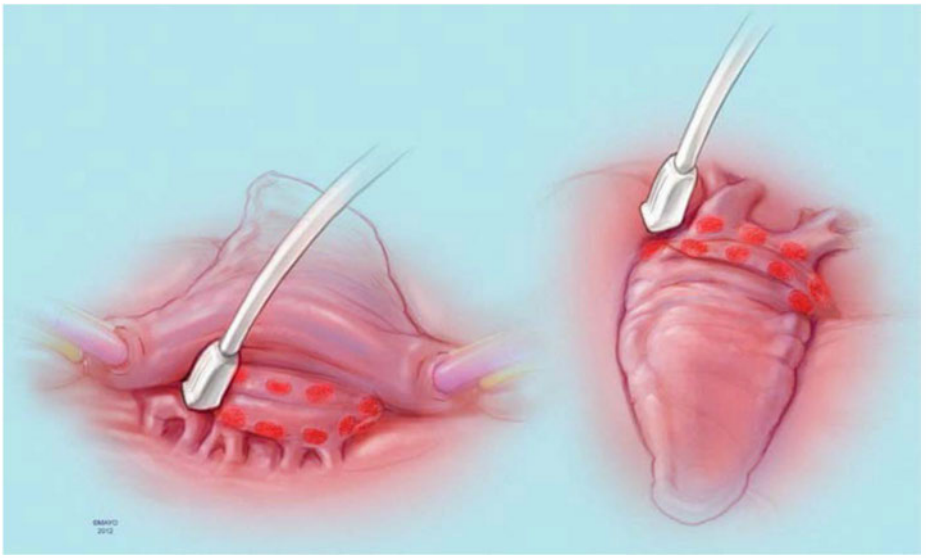


Fig. 65.10. Left atrial ganglion ablation procedure. Ganglia are typically found in the fatty epicardial tissue. Intraoperative mapping is performed and an active ganglion is identified when stimulation results in a doubling of the R–R interval. Active sites can be ablated with radiofrequency energy applied with a pen-like instrument. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Cox-Maze IV (Ablation-Assisted Cox-Maze)

Introduced in 2002, the Cox-maze IV procedure is another modification to the standard "cut-and-sew" maze that serves to further simplify the technique. This is done by replacing most of the incisions with bipolar RF energy and using cryoenergy in addition to achieve the complete biatrial lesion sets. This has the potential advantage of shorter ablation times, can be performed in a less invasive manner and by localization of the ablation lines between clamps, there is minimal collateral damage. However, its durability has been questioned in comparison with the maze III procedure in terms of transmurality (Fig. 65.11).

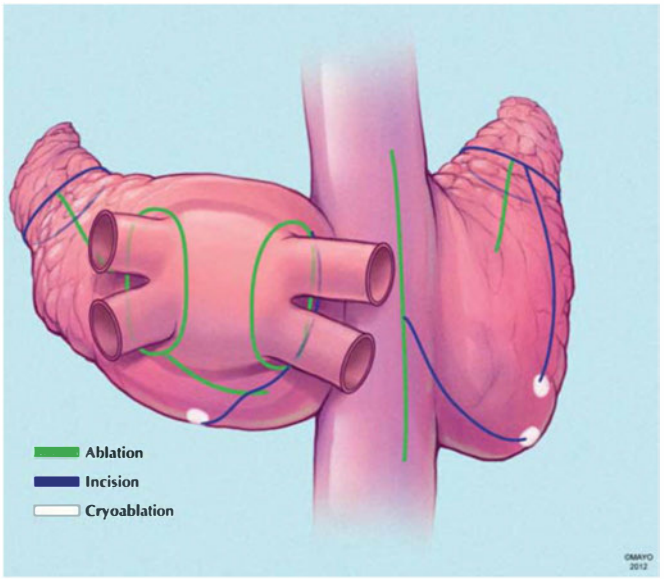


Fig. 65.11. Back view of the atria illustrating the Cox-maze IV procedure in which some of the atrial incisions in the Cox-maze III procedure is replaced with a combination of radiofrequency and cryoablation to expedite the procedure and minimize cardiopulmonary bypass time. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Minimally Invasive Approaches

Many surgeons have used modified lesion sets and alternate energy sources through minimally invasive incisions, including thoracoscopic and robotic-assisted techniques both on- and off-pump. These methods may make the procedure more acceptable to patients and cardiologists, but there is a compromise in regard to effectiveness of ablation (transfer out the lesion sets) and inability to ligate the left atrial appendage when the operation is performed exclusively through the right thorax.

LONE ATRIAL FIBRILLATION

This term was introduced by Evans and Swann in 1954 to describe AF not associated with systemic arterial hypertension or structural heart disease. Other designations used include isolated and "idiopathic" AF,

but it is now known that there are many triggers for lone AF. A surgical procedure for lone AF is appropriate for young patients who have limiting symptoms, especially those who have failed pharmacological treatment or become intolerant of medications. In current practice, most such patients will also have had one or more attempts at catheter ablation for control of AF. Many young patients prefer a curative procedure rather than lifetime treatment with drugs that have bothersome side effects. Surgical treatment of lone AF may also be indicated for patients who have contraindications to systemic anticoagulation, or personal preference to avoid chronic anticoagulation. Other indications for surgical treatment of lone AF include patients who have suffered a thromboembolic stroke while on anticoagulation and those with tachycardia-induced cardiomyopathy that does not respond to medical treatment or catheter ablation.

ATRIAL FIBRILLATION IN PATIENTS HAVING OTHER CARDIAC OPERATIONS

Mitral Valve Surgery

Chronic AF is present in approximately 40% of patients who undergo mitral valve surgery, and interventions on the mitral valve alone fail to restore sinus rhythm in the majority of these patients. Medical therapy, with or without catheter-based ablation, rarely restores and maintains sinus rhythm in these patients because of the chronic enlargement of the left atrium, which acts as a substrate for AF. The pulmonary veins provide a trigger for AF in approximately 90% of patients with paroxysmal arrhythmia; PVI alone would be expected to cure a majority of patients with this type of AF. However, PVI will fail as an ablative procedure in those 10% of patients in whom the pulmonary veins do not contribute to the substrate for AF.

More chronic AF leads to atrial remodeling and the development of macro reentrant pathways that sustain electrical reentry. In patients with chronic AF, the goal of treatment shifts from isolating the trigger of the arrhythmia (pulmonary veins in paroxysmal AF) to ablating the macro reentrant pathways responsible for its maintenance. In this setting, the arrhythmia is not dependent on stimuli from the pulmonary veins, and as such, PVI may be an inadequate treatment. This remodeling is present in a significant portion of patients with mitral valve regurgitation, and especially those that have evidence of left atrial enlargement. Thus, PVI as an adjunct to mitral valve repair in patients with severe mitral valve regurgitation may fail because of macro reentry pathways, the size of the left atrium, and the fact that alternate energy sources, as discussed below, may not create transmural lesions.

Handa et al. reported that the Cox-maze operation is a safe adjunct to mitral valve repair for patients with preoperative AF. In this study, the addition of the Cox-maze procedure was particularly useful in patients with continuous AF of more than 3 months duration preoperatively. Freedom from AF was 82% in patients who underwent combined maze and mitral valve repair compared with 53% in patients who underwent mitral valve repair alone. Morbidity and mortality were not increased by the addition of the maze procedure, and 75% of patients regained sinus rhythm by last follow-up. In this series, only the omission of the maze procedure and the presence of chronic AF were predictors of arrhythmia recurrence. Similar results have been reported by Ad and Cox.

Modifications of the standard maze procedure are commonly used in patients having mitral valve surgery. The use of modified lesion sets and alternate energy sources simplify ablation and, theoretically, minimize the risk of surgery. Some groups advocate only left-sided maze lesions to ablate AF. Although the pulmonary veins may not be the initiating focus for AF in all patients with mitral valve disease, isolating the pulmonary veins still remains a crucial maneuver in any attempt to ablate AF. After the heart has been arrested and a standard left atriotomy is performed, we often isolate the pulmonary veins with two "half-oval" lesions created with a flexible cryoprobe or with an RF clamp. The superior limb of this oval is created from the superior end of the left atriotomy and is carried around the two superior pulmonary veins. Similarly, the inferior limb is then created from the inferior cut edge of the left atriotomy and is carried around the two inferior pulmonary veins and meets the superior lesion. These two lesions are usually joined on the left side of the left-sided pulmonary veins. A connecting lesion is then carried down from this pulmonary vein encircling lesion to the mitral valve annulus. The left atrial appendage cannot be excluded by oversewing the orifice from within the left atrium, by amputating it and oversewing it externally, or by simply ligating it externally.

Another useful method involves amputation of the left atrial appendage after creating the left atriotomy to expose the mitral valve. The open left atrial appendage stump can then serve as the area of joining the cryolesions from above and below the pulmonary veins. Bipolar RF clamps can similarly be used to create each limb of the pulmonary vein encircling lesion. In this instance, for the superior limb of the encircling lesion, one jaw of the clamp is passed epicardially through the transverse sinus, and the other through the left atriotomy and placed endocardially into the stump of the left atrial appendage. The inferior limb of the encircling lesion is created in a similar manner with one jaw placed epicardially and one endocardially with the tip meeting through the stump of the amputated left atrial appendage.

Coronary Artery Bypass and Aortic Valve Procedures

Studies have demonstrated that the addition of the maze procedure to aortic valve procedures or coronary artery bypass grafting (CABG) do not increase the morbidity or the periprocedure risks. Patients undergoing aortic valve procedures or CABG deserve special attention as they represent a group of patients who are less likely to experience preoperative AF and less likely to receive additional treatment for AF. There is compelling evidence that treating AF in these situations is associated with improvement in outcomes. The use of alternate energy sources as cryoenergy or RF ablation may be appropriate under these circumstances due to the concern related to long cardiopulmonary bypass time. Important questions are which category of these patients requires an additional AF ablation and is the lesion set different than in other cardiac pathology?

Evidence-based guidelines for performing adjunctive procedures for AF during cardiac surgery have been published recently by the International Society of Minimally Invasive Cardiac Surgery (ISMICS). These experts recommended addition of surgical AF ablation to increase the chance of return to sinus rhythm, to improve ejection fraction, and also to decrease the risk of thromboembolic complications. The selection of the surgical technique differs according to patient's risk category. With regard to the lesion set, PVI and left atrial appendage exclusion may be appropriate in patients undergoing CABG or aortic valve procedures; however, these lesion sets may not be appropriate for those with an enlarged left atrium or more complex long-standing AF.

Septal Myectomy for Hypertrophic Cardiomyopathy

In patients with hypertrophic cardiomyopathy, AF can result in profound clinical deterioration due to the loss of the atrial component of left ventricular filling. Patients who undergo septal myectomy usually enjoy dramatic relief of symptoms and improved exercise capacity; however, a number of patients will be left with underlying diastolic dysfunction with resultant increased left atrial pressure and dilatation.

The question of whether addition of concomitant surgical ablation of AF at the time of septal myectomy remains a clinical dilemma. It is unclear if surgical intervention for AF would indeed yield superior freedom from AF and improve outcomes to an appreciable extent, mostly because of the underlying substrate (extent of myocardial fibrosis) and pre-existing cardiomyopathic process. In our experience, approximately two-thirds of patients who have AF preoperatively are free from AF after septal myectomy (without surgical AF ablation). Adequate myectomy relieves left ventricular outflow tract obstruction and allows some left ventricular mass regression that may then decrease the risk of recurrent AF.

However, concomitant maze procedure at the time of septal myectomy may be useful for patients with obstructive hypertrophic cardiomyopathy who are extremely symptomatic from their arrhythmia preoperatively, are intolerant to antiarrhythmic medications, or have undergone prior catheter ablations that were unsuccessful. Published reports demonstrate that concomitant AF ablation at the time of septal myectomy is safe, is effective, and does not appear to increase operative mortality or postoperative morbidity. In our experience, concomitant procedures for AF at the time of myectomy improves late freedom from AF among patients with preoperative intermittent and continuous AF, but in patients with continuous AF, late recurrence following myectomy is much greater than this observed after AF ablation during mitral valve procedures. Among patients having septal myectomy and the maze operation, there was, in addition to reduced risk of late arrhythmia, a significant reduction in need for warfarin anticoagulation (maze: 0% vs. no maze: 37%; $P = 0.004$). Importantly, the addition of the Cox-maze procedure in these patients undergoing septal myectomy for obstructive hypertrophic cardiomyopathy does not appear to increase operative morbidity beyond what is expected after an isolated maze procedure.

Congenital Heart Diseases

The most common atrial tachyarrhythmias among adults with congenital heart diseases are AF and flutter, and right-sided cut-and-sew lesions of the Cox-maze III procedure is effective in these patients. With the availability of newer devices such as RF and cryoablation, the procedure time for maze procedure is shortened significantly. Consequently, our current preference is to perform a biatrial maze procedure, particularly when there is chronic AF, left atrial dilation, or concomitant mitral regurgitation. In the presence of atrial flutter, we prefer to add a "right atrial isthmus" lesion, which is the posterolateral tricuspid valve annulus to the coronary sinus to the inferior vena cava. We also make an effort to close the left atrial appendage (Fig. 65.12).

In patients with atrioventricular nodal reentrant tachycardia who underwent unsuccessful ablation in the electrophysiologic laboratory preoperatively, we perform perinodal cryoablation on cardiopulmonary bypass after opening the right atrium and closure of any intracardiac shunt. This involves multiple applications of the cryoprobe around and in the ostium of the coronary sinus, which is then carried anteriorly toward the proximal atrioventricular node until temporary complete heart block is noted, at which time the rewarming is begun; normal atrioventricular conduction returns shortly thereafter.

In our experience, 80% of patients undergoing right-sided maze at the time of congenital heart disease repair had paroxysmal AF, and the most common diagnoses were Ebstein anomaly, isolated atrial septal defect, and tetralogy of Fallot. Approximately 90% of patients were free of AF at the time of hospital dismissal, and approximately 70% manifested sinus rhythm. Permanent pacemakers were required in 15% of these patients; the overwhelming majority of these were for sick sinus syndrome postoperatively. At last follow-up, approximately 75% of patients with preoperative chronic AF and 95% of patients with preoperative paroxysmal AF were free from subsequent atrial arrhythmias.

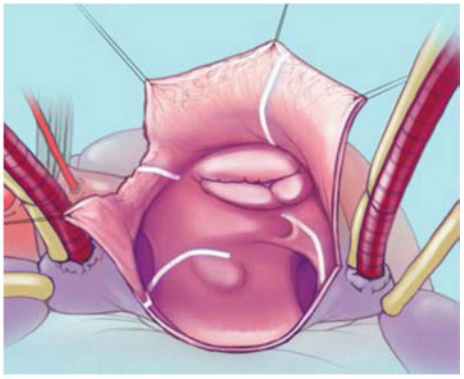


Fig. 65.12. We have used successfully the right-sided cut-and-sew lesions of the Cox-maze III procedure in adults with congenital heart disease, and with the availability of newer devices, such as radiofrequency or cryoablation, the procedure time for the maze procedure is shortened significantly. We prefer to add a "right atrial isthmus" lesion, which is the posterolateral tricuspid valve annulus to the coronary sinus to the inferior vena cava. (With the permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Atrial Fibrillation and Tricuspid Valve Disease

Tricuspid valve disease is present in up to 10% to 50% of those with significant mitral valve pathology. In most patients, concomitant tricuspid regurgitation is associated with elevated pulmonary artery pressure and tends to be functional in etiology. Tricuspid regurgitation may improve after correction of the left-sided pathology; however, it may persist or even worsen. In general, the course is difficult to predict, and reoperation to correct tricuspid regurgitation is associated with high perioperative morbidity and mortality. AF has been identified as a significant predictor for late development of tricuspid regurgitation after mitral valve surgery. In a previous study from our institution, restoration of sinus rhythm after addition of the maze procedure during mitral valve surgery was associated with significant decrease in the progression of late tricuspid regurgitation in comparison to those who continued to have AF postoperatively. Several mechanisms explain the progression of tricuspid regurgitation with the persistence of AF. First, patients in AF are more likely to have right ventricular dilatation with resulting dilatation of the tricuspid valve annulus. Second, AF is associated with higher pulmonary artery pressure late after operation in comparison to patients who are in sinus rhythm.

SPECIAL SITUATIONS

Low Ejection Fraction

AF can lead to cardiomyopathy, and multiple reports have documented that left ventricular dysfunction caused by supraventricular tachycardia can be cured or improved by conversion to sinus rhythm. AF impairs hemodynamic function by several mechanisms. First, AF results in loss

of atrioventricular synchrony and atrial contraction. This may reduce ventricular filling and thereby reduce cardiac output. The consequence of loss of atrial contraction may be especially pronounced in patients with impaired diastolic function as is seen with hypertrophied ventricles, restrictive cardiomyopathy, and mitral valve stenosis. Also, fluctuation in the R–R interval changes the diastolic filling interval, producing a variable stroke volume.

Cardiomyopathy caused by tachycardia is commonly thought to be associated with chronic arrhythmias having rates greater than 120 beats per minute. In our experience, ventricular dysfunction may be associated with resting heart rates considerably lower than this, and further, paroxysmal AF can lead to ventricular dysfunction. Our patients with tachycardia-induced cardiomyopathy having maze operations had a median resting heart rate of 88 beats per minute (range: 58 to 144 beats per minute). Furthermore, among patients with preoperative paroxysmal AF, the median preoperative resting heart rate was 62 beats per minute (range: 47 to 94 beats per minute), and during paroxysmal episodes of arrhythmia, the median heart rate measured was 84 beats per minute (range: 66 to 163 beats per minute). It is important to recognize that resting heart rate is not truly reflective of overall heart rate in patients with AF, because the heart rate response to exercise may vary, and patients with well-controlled resting heart rates may have a rapid ventricular response with minimal activity, and develop tachycardia-induced cardiomyopathy.

The identification of patients with tachycardia-induced cardiomyopathy may be challenging, and careful search for structural heart disease is necessary. The absence of structural anomalies and the documentation of AF before left ventricular dysfunction make the diagnosis of tachycardia-induced cardiomyopathy more likely. Other patients may present with AF and left ventricular impairment, and it is unclear whether the arrhythmia is causative or a consequence of ventricular dysfunction. In such patients, medical management is initiated, and cardioversion should be considered. If the ejection fraction improves when the rate is more adequately controlled, the diagnosis again is more likely.

Prophylactic Cox-Maze

New-onset late AF has been observed after mitral valve repair for degenerative mitral valve disease, and this can occur even without a prior AF history; this risk is greater in patients with advanced age. Those with severe mitral regurgitation and significant functional tricuspid regurgitation have altered right ventricular strain and function. Intervention on coexistent functional tricuspid regurgitation does not reduce the late risk of AF; in fact, the predisposition to late AF may already be set at the time of mitral valve repair. From our prior analysis, the following factors were identified as independent predictors for late AF: (1) advanced age, (2) increased left atrial size (more than 50 mm), (3) moderate preoperative tricuspid regurgitation, and (4) diabetes. Therefore, those with severe mitral regurgitation, left atrial enlargement, and coexistent moderate functional tricuspid regurgitation may be reasonable candidates for consideration of prophylactic maze procedure. This strategy awaits prospective, controlled studies.

POSTOPERATIVE MANAGEMENT

Different protocols including different antiarrhythmic regimens for the postoperative management of patients who have undergone a Cox-maze operation are used. Our drug of first choice is amiodarone; however, it is used selectively in patients who experience atrial or ventricular arrhythmias during hospitalization and is usually continued for 3 months postoperatively. Electrolytes are monitored on a daily basis, especially potassium and magnesium, and both are kept in the high-normal range. Electrical cardioversion is used as needed.

Diuretics are used liberally early after operation with the goal of achieving preoperative weight. Excision of the atrial appendages during the Cox-maze procedure removes an important source of atrial natriuretic peptide, and this may predispose the patient to fluid retention early postoperatively.

Coumadin is used for 3 months postoperatively, but there is no consensus on the need for anticoagulation beyond this interval. Some clinicians prefer to continue Coumadin believing that the risk of thromboembolism is not reduced sufficiently to avoid systemic anticoagulation. Others argue that if AF is eliminated and ventricular function is normal, the risk of thromboembolism from a postoperative patient without a left atrial appendage is very low.

OUTCOMES

The Cox-maze procedure has proved to be the most effective surgical procedure for treating AF and its adverse consequences of irregular rhythm, altered hemodynamics, and increased thromboembolic risk. The indications for its application continue to evolve as new techniques and instruments have been developed. These modifications have simplified surgical ablation of AF and have expanded surgical ablation to additional subgroups of patients as mentioned previously. However, evaluation of outcome for patients who have undergone the maze procedure, as well as new devices and lesion sets, is difficult because of variable methods of reporting success in eliminating AF.

Outcomes of procedures for ablation of AF are influenced by thoroughness of follow-up as well as method of assessment of cardiac rhythm. The electrocardiogram is a "snap-shot" in time and has limited ability to detect those patients that may have transient atrial arrhythmias in the follow-up period. A better method is the Holter monitor, but widespread use for routine follow-up is not feasible. After clinical evaluation and follow-up of rhythm status are obtained, the second difficulty is in how the results of the analysis are reported. "Rhythm at last follow-up" may underestimate the recurrence rate of atrial arrhythmias in the follow-up period and thus overestimate the success of the procedure. Conversely, actuarial methods used to delineate time-related events, "freedom from AF," define any recurrent arrhythmia as a failure of the procedure and thus may underestimate the actual clinical success. Other factors that contribute to confusion in assessing results of surgical treatment of AF are viable terminology (intermittent vs. paroxysmal, etc.) and differing patient populations (lone paroxysmal AF, AF with mitral valve disease, etc.).

The HRS/EHRA/ECAS, in collaboration with the American College of Cardiology, Society of Thoracic Surgeons, and American Heart Association, published an Expert Consensus Statement on catheter and surgical ablation of AF outlining recommendations for personnel, policy, procedures, and follow-up. These guidelines were intended to standardize the design of studies, procedures performed, intensity of follow-up, and reporting success as it pertains to the surgical- or catheter-based treatment of AF. In terms of reporting success, the HRS consensus document recommends that success from AF intervention should be reported as freedom from AF without antiarrhythmic medications.

Cox-Maze III vs. Catheter-Based Ablation

There is debate as to the most effective lesion set during catheter ablation, but an equally important issue is achieving and ensuring transmural lesions. Achieving transmurality can be complicated by several factors such as tissue characteristics (e.g., thickness and fibrosis) and the unpredictability of conditions at the time of ablation. Although conduction block provides a clear endpoint for electrical isolation of the pulmonary veins, reconnection of conduction has been demonstrated in up to 64% of patients during follow-up. The cut-and-sew Cox-maze procedure by its nature ensures a transmural lesion, which we believe is the most important factor in successful electrical isolation of the pulmonary veins.

The data comparing late results of Cox-maze III to catheter-based ablation techniques are scarce. We analyzed the outcome of 97 patients aged 25 to 80 years who underwent an isolated cut-and-sew Cox-maze procedure between 1993 and 2007; this group was matched 1:2 according to age, sex, and AF type with 194 patients who underwent catheter-based ablation for lone AF. The freedom from AF at last follow-up was 82% in those patients who underwent the Cox-maze procedure with discontinuation of the antiarrhythmics, compared with 55% in those who underwent ablation, and 24% of the ablation group required a repeat ablation procedure.

Cox-Maze III vs. Cox-Maze IV

Although the Cox-maze III is considered the gold standard for surgical treatment of AF, many surgeons consider this procedure to be demanding and excessively time-consuming, especially when used in conjunction with other operations. The simplified Cox-maze IV developed at Washington University addresses these issues. The procedure appears to be effective, and in one report comparing 100 patients undergoing the Cox-maze IV with 112 having the Cox-maze III, freedom from AF was 90% following the newer procedure compared with 96% after the Cox-maze III. Also, freedom from AF off antiarrhythmics was slightly higher in Cox-maze III (83%) versus 82% in the Cox-maze IV group, as was freedom from anticoagulation, 86%, and 74% in the Cox-maze III and IV groups, respectively. There was no significant difference between the two groups in the need for a pacemaker.

Mayo Clinic Experience

Success with the standard Cox-maze procedure has varied in published reports. In general, approximately 90% of patients who undergo the Cox-maze operation are free from AF at last follow-up, with new pacemakers required in 10% to 15% of patients. Our experience is similar to others in that operative risk is low, 1.5% overall, which includes patients having concomitant intracardiac repair; risk of operation for isolated AF is $<1\%$, which is comparable to the risk of closure of an atrial septal defect. New permanent pacemakers were required in $<10\%$ of patients, and the indication in almost all was sick sinus syndrome. This incidence of pacing postoperatively is lower than expected based on the original reports of the Cox-maze operation. In the earlier experience, clinicians were hesitant to allow patients to remain in junctional rhythm early postoperatively, but in many such patients, a stable sinus mechanism will return. Thus, some patients may have had pacemakers implanted prematurely. Additionally, technical modifications to the original Cox-maze operation may reduce injury to the sinoatrial node.

Preoperatively, patients are counseled that this operation reliably eliminates AF in most individuals but does not necessarily restore sinus rhythm. In older patients especially, there is an underlying incidence of sick sinus syndrome, and when AF is eliminated, permanent pacemakers may be necessary to manage sinus node dysfunction. Also important is the fact that conduction disturbances may also develop in those patients who require concomitant intracardiac procedures at the time of surgery for AF. Eighty percent of patients undergoing the standard biatrial maze procedure at Mayo Clinic have combined procedures.

In our experience, approximately 90% of patients are free from AF upon dismissal from the hospital. This includes patients with sinus rhythm, paced rhythm, or junctional rhythm with an adequate rate. It is important to recognize that the Cox-maze procedure, like other cardiac operations, predisposes patients to transient AF in the early postoperative period.

Surgical cure of AF appears to be durable, and at last follow-up (median: 34 months), the overall freedom from AF was approximately 85%. When outcome is analyzed in a product limit estimate (Kaplan–Meier), freedom from AF was 76% at 5 years and 51% at 10 years. In our series, late outcome and cure of AF depends on preoperative characteristics. At last follow-up (median: 41 months), 93% of patients with preoperative lone paroxysmal AF were free from their arrhythmia with an actuarial freedom from AF of 90% at 5 years and 64% at 10 years. Patients with preoperative lone chronic AF had 83% freedom from AF at their last follow-up (median: 28 months) with an actuarial freedom from AF of 80% at 5 years and 62% at 10 years. The Cox-maze operation is less durable for patients undergoing combined Cox-maze and mitral valve surgery with 70% of patients free from AF at last follow-up (median: 33 months) and an actuarial freedom from AF of 68% at 5 years and 41% at 10 years (Fig. 65.3).

Although a number of patients in our series had evidence of a junctional rhythm postoperatively, with some subsequently dismissed in this rhythm, we do not routinely utilize antiarrhythmic medications or stimulants, such as theophylline, as have been suggested. A significant number of such patients will regain a stable sinus rhythm, and it has been reported that up to 50 weeks are required until this occurs. Persistent junctional rhythm may reflect sinus node dysfunction, and this will predispose the patient to recurrent arrhythmias and stroke. In these patients, a permanent pacemaker should be considered.

FUTURE DIRECTIONS

New instruments developed to facilitate surgical ablation of AF and new lesion sets may achieve rates of AF control similar to the traditional "cut-and-sew" methods. However, equivalency has not yet been demonstrated, and future comparative studies are necessary. Also, there are many unanswered questions regarding the clinical application of the maze operation in conjunction with other cardiac procedures. Is PVI equally effective as a full Cox-maze procedure for patients with paroxysmal AF and mitral valve disease? Perhaps, the pathogenesis of AF in patients with mitral valve disease and paroxysmal arrhythmia is different from that of patients with chronic AF in the setting of mitral valve disease and a dilated left atrium. Another unresolved issue is whether concomitant left reduction atrioplasty should be performed at the time of valve repair and maze procedure for patients with AF and a dilated left atrium from mitral valve disease. Finally, should a "prophylactic" maze procedure be performed in a patient with preoperative sinus rhythm and a dilated left atrium undergoing surgery for mitral valve disease? The altered atrial tissue in these patients represents an arrhythmogenic substrate, which renders them at high risk (approximately 40%) for the development of postoperative

AF even after mitral valve disease is repaired. Clearly, the risks and benefits should be weighed before an additional procedure is performed, which carries with it the added risk of permanent transvenous pacemaker implantation.

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EDITOR'S COMMENTS

Dr. Schaff and colleagues have given a complete overview of surgical treatment of atrial fibrillation. I would like to focus mostly on the cut-and-sew maze, which they have continued to espouse and do extremely well. Given the mortality for this procedure is <2%, it has been efficacious. The atrial fibrillation is in the cue rate 85% to 90% range as compared with 50% for catheter-based procedures. The real question is which patient will tolerate these procedures. Alternative energy responses are good but not quite as good as the cut-and-sew maze. The difficulty is the more extensive procedures take quite a bit more time and therefore there needs to be appropriate decision making as to which patients will benefit.

There is also a school of thought that solving the mitral valve issue in patients who have mitral regurgitation and atrial fibrillation may have similar results to mitral valve surgery plus maze procedure. This is presently a study being performed by the NHLBI-sponsored Cardiothoracic Surgical Network. This study is nearly closed in terms of patient entry. I believe the results will help us determine which patient will do best with mitral valve surgery alone versus mitral valve surgery plus maze procedure.

ILK

66 Minimally Invasive Treatment of Atrial Fibrillation

Gorav Ailawadi

INTRODUCTION

Atrial fibrillation (AF) develops in nearly 5% of patients in their lifetime and is the most common arrhythmia. An estimated 5 million Americans suffer from AF, and this number is expected to triple by 2050. AF is associated with an increased risk of stroke, thromboembolism, and death. Moreover, AF is an underdiagnosed cause for tachycardia-induced cardiomyopathy. AF is responsible for nearly 26 billion dollars annually. For these clinical and economic reasons, there is great interest in developing effective treatments for AF.

Medical therapy is the first-line therapy for patients with AF but has been shown to have poor success at rhythm restoration. The advent of surgical and ultimately catheter-based ablation technologies has revolutionized approach for this devastating disease.

AF is now classified by the terms paroxysmal, persistent, and long-standing persistent according to the ACC/AHA guidelines. Paroxysmal is characterized by recurrent episodes that last <7 days or convert to sinus without cardioversion. Persistent AF refers to AF that lasts more than 7 days, while long-standing persistent AF is AF lasting more than 1 year. The terms intermittent, chronic, and permanent AF are no longer in use.

The Cox-Maze III procedure (so-called "Cut and Sew Maze") became the gold standard for the cure of AF. Although this procedure has excellent long-term results, it was limited in its adoption due to patient morbidity and complexity for the surgeon. Within the last several years, a number of alternate energy sources were developed to create lesion sets to treat AF, which is a variant of the Cox-Maze III, often termed Cox-Maze IV.

PATHOPHYSIOLOGY OF ATRIAL FIBRILLATION

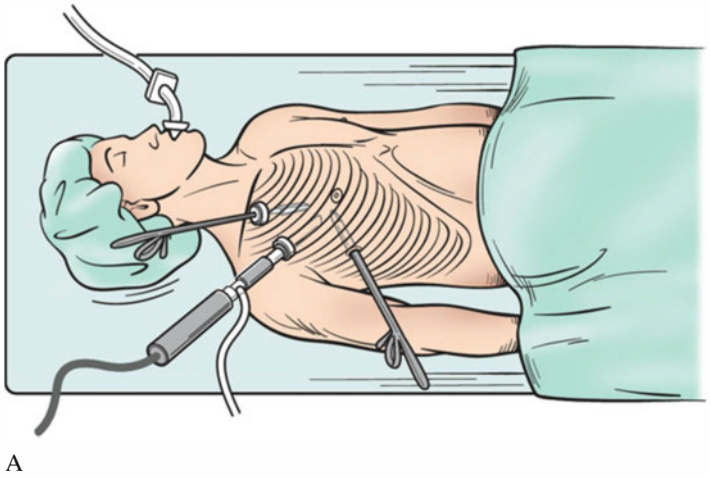
AF is characterized by macro reentrant electrical circuits in both atria. Each patient's electrical circuitry is unique; thus, mapping AF in any given patient can be laborious and challenging. Endocardial mapping has demonstrated that the pulmonary veins and posterior left atrium are the critical anatomic targets for ablation in patients with isolated AF as well as those with concomitant valvular heart disease. Specifically, in patients with paroxysmal AF, the source for reentrant circuit appears to be from the pulmonary veins while in patients with persistent AF, regular and repetitive activation can be identified in the posterior left atrium, the pulmonary veins, and/or the left atrial appendage (LAA). Although routine intraoperative mapping is currently not feasible for guiding intraoperative AF ablation, an anatomic approach based on our understanding of pathophysiology and on empiric results is reasonable. In fact, such an anatomic (rather than map-guided) approach is rapidly becoming the standard for catheter-based ablation of AF. The Cox-Maze III and IV use predetermined lesions/incisions in anatomic locations to interrupt these circuits. Surgical intervention typically includes excision/ligation of the LAA, which may reduce the risk of recurrent AF, thromboembolism, and stroke.

THE COX-MAZE PROCEDURES

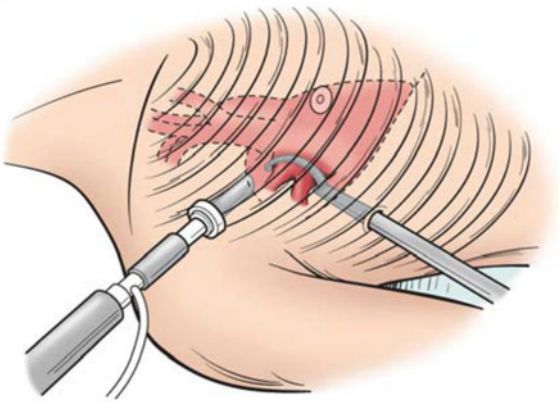
Cox-Maze IV

With recent advances in surgical tools for ablation, there are a number of less invasive approaches to recreate variations of the Cox-Maze III lesion set. The number of cases referred for surgical ablation pales in comparison to those treated via catheter. However, there are several potential advantages to surgical ablation including creation of transmural lesions, direct or endoscopic visualization eliminating the possibility of pulmonary vein stenosis as well as collateral damage to adjacent structures like the esophagus, and the ability to excise or exclude the LAA. Limitations include the inability to replicate the complete Cox-Maze lesion set without opening the heart or performing a concomitant catheter ablation for ablation along the coronary sinus (CS) and cavotricuspid isthmus, as well as poor ability to map for gaps or atrial flutter.

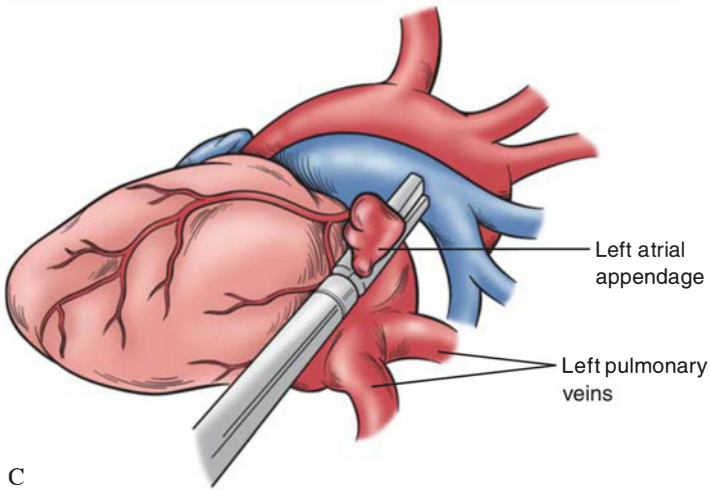
We perform bilateral completely thoracoscopic epicardial ablation followed by endocardial catheter ablation. The patient is placed in supine position with double-lumen endotracheal tube intubation. Three ports are placed in the right chest. Under single-lung ventilation, the right pericardium is opened parallel to the phrenic nerve. The oblique and transverse sinuses are opened bluntly and the right pulmonary veins are encircled using a special dissector. The right veins are ablated using a bipolar radiofrequency (RF) clamp with 3 to 5 successful ablations high on the left atrium. Electrical isolation is confirmed by sensing and pacing the pulmonary veins. The atrial roof and floor lines/connecting lesions are created using a unipolar RF lesions sources from the right side to the left side using thoracoscopic guidance. Unipolar RF can also create a lesion from the superior vena cava (SVC) to the inferior vena cava (IVC) laterally along the right atrium. An SVC encircling lesion is made above the sinus node using bipolar RF. On the left side, the ligament of Marshall is divided after opening the pericardium posterior to the phrenic nerve. The left veins are encircled and ablated using the bipolar RF clamp. The connecting lesions are completed and the LAA is excised with an endoscopic stapler or excluded using commercially available devices (Fig. 66.1). The surgical procedure takes roughly 3 to 4 hours. Patients then undergo catheter procedure at the same setting or at a later date with mitral isthmus/CS ablation and cavotricuspid ablation. The surgical lesions are tested for gaps as well. Our results are fairly consistent with over 85% freedom from AF at 18-month follow-up of antiarrhythmics as tested with 7-day Holter monitor.



A



B



C

Fig. 66.1. (**A**) Thoracoscopic approach for minimally invasive, (**B**) epicardial pulmonary vein isolation with excision of the left atrial appendage. (**C**) Stapled excision of the left atrial appendage after isolation of the left pulmonary veins using a similar technique to that displayed for the right pulmonary veins. Commercially available devices can be used to exclude the LAA.

There are a number of other less invasive methods to recreate some version of the Cox-Maze III lesion set in patients with lone AF. Another common approach is through a port access approach (right mini thoracotomy) using cardiopulmonary bypass via the femoral vessels. The heart is arrested similar to a minimally invasive approach to the mitral valve. Using cryoablation and RF, the lesion set can be replicated after opening the left atrium. If right atrial lesion sets are planned, femoral venous cannula must be in as far as the SVC and the perfusion must be prepared to get air in the venous line. The LAA is oversewn from the inside. With this approach, centers are reported over 80% success in patients with failed catheter ablation.

Cox-Maze III

The traditional Cox-Maze III procedure infrequently performed today is through a median sternotomy. Cardiopulmonary bypass is established using bicaval and ascending aortic cannulation. Coronary artery bypass grafting distal anastomoses are completed prior to the Cox-Maze III procedure, whereas mitral valve surgery is performed after reconstruction of the left atrium. The heart is arrested using antegrade cardioplegia with or without retrograde cardioplegia.

The interatrial groove is opened anterior to the right pulmonary veins and extended around the left atrium above the pulmonary veins similar to a donor heart explant (Fig. 66.2A). Care is taken to stay away from the posterior mitral annulus where the circumflex and CS lie. A stay suture is placed at the left inferior pulmonary vein to mark the anatomy and facilitate atrial reconstruction. The LAA is excised and can either be closed separately or incorporated with closure of the left atrial cuff (Fig. 66.2B, C). As reconstruction is carried around the right side close to the CS, partial thickness bites can help avoid injury to the CS. Along the CS, a 2-minute cryolesion is created from the mitral annulus around P3 toward the CS.

The right-sided incisions begin by opening the mid-right atrium toward the 2 o'clock position on the tricuspid valve (Fig. 66.2D). The interatrial septum is then incised. An additional 2-minute cryolesion is created at the 2 o'clock position of the tricuspid annulus. The right atrial appendage is amputated and bipolar RF clamp is introduced into this stab incision and directed toward the patient's back, creating a lesion in the right atrial appendage that ends 1 cm from the previously placed right atriotomy. The direction of the bipolar RF clamp is then reversed, and a lesion is created toward the tricuspid annulus. The bipolar RF clamp is then used to create a lesion from the right atriotomy to the SVC and to the IVC. Finally, a cryolesion is created on the right atrial isthmus, extending from the tricuspid annulus to the orifice of the IVC. The right atrial incisions are closed and the cross-clamp is removed.



Fig. 66.2. The Cox-Maze III procedure with bipolar radiofrequency application in right atrium. **(A)** The left atrium is opened anterior to the left pulmonary veins, and the pulmonary vein encircling incision is carried to the level of the left inferior pulmonary vein. A stay suture is placed at this point. The left atrial appendage is excised sharply. **(B)** The pulmonary vein encircling incision is completed and then partially closed. **(C)** An incision is created to the mitral annulus, exposing the anterior surface of the coronary sinus. Cryolesions are created at the mitral annulus and coronary sinus. **(D)** There is a single long incision in the right atrium heading from the tricuspid annulus to the fossa ovalis; a cryolesion is created at the tricuspid annulus at the 2 o'clock position. Through this incision, bipolar radiofrequency lesions are created to the superior vena cava and to the inferior vena cava (*dashed lines*). A stab incision is created in the right atrial appendage, and a bipolar radiofrequency lesion is created in the appendage (*dashed line*).

Before weaning the patient from cardiopulmonary bypass, it is prudent to examine the region of left atrial suture lines for bleeding and repair them while on bypass. It is common for patients to require atrial pacing immediately after Cox-Maze III.

Freedom from AF ranges from 75% to 98% at 5 to 10 years following surgery. The most significant risk factors for AF recurrence after a Cox-Maze procedure include (1) duration of preoperative AF longer than 5 years, (2) persistent and long-standing persistent AF, and (3) left atrial enlargement especially >6 cm. The Cox-Maze procedure also has shown to nearly eliminate the risk of late stroke.

PULMONARY VEIN ISOLATION

In patients with paroxysmal AF, the source for AF emanates from the pulmonary veins in over 90% of patients. Thus, pulmonary vein isolation (PVI) is often adequate to treat paroxysmal AF. A variety of energy sources have been employed to perform PVI. The pulmonary veins can be isolated with a single, large, boxlike lesion (Fig. 66.3) or, alternatively, as two separate oval-shaped lesions (Fig. 66.4). Either approach is acceptable. For patients with persistent or long standing persistent AF, the mitral isthmus lesion and lesion to the LAA should be added (Fig. 66.3).

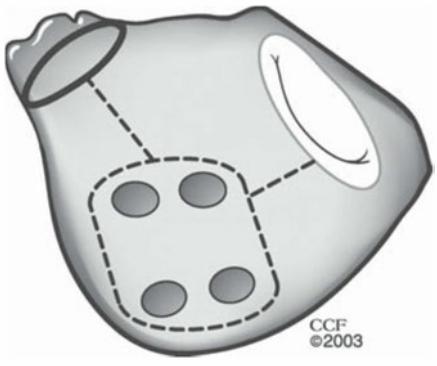


Fig. 66.3. Left atrial lesion set of the Cox-Maze III procedure. *Dashed lines* indicate surgical incisions. The pulmonary veins are encircled by a surgical incision, and there is a connecting incision to the mitral valve annulus. The left atrial appendage is excised, and this incision is connected to the pulmonary vein encircling incision.



Fig. 66.4. Bilateral pulmonary vein isolation with excision of the left atrial appendage. The pulmonary veins are isolated as two separate ovals that include wide areas of left atrial tissue. The left atrial appendage is excised sharply. This can also be done with RF clamps and excision of the appendage as in figure 66.1.

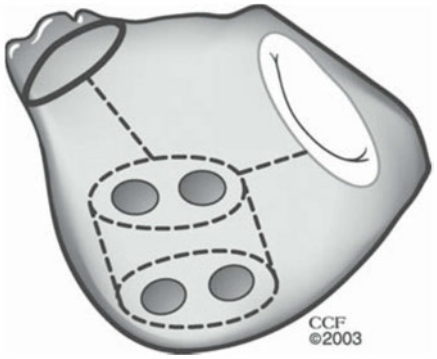


Fig. 66.6. Maze-like lesion set (Cox-Maze IV) created with bipolar radiofrequency and cryothermy (for lesion to the mitral annulus). The lesion to the mitral annulus is created with cryothermy. The left atrial appendage is excised.

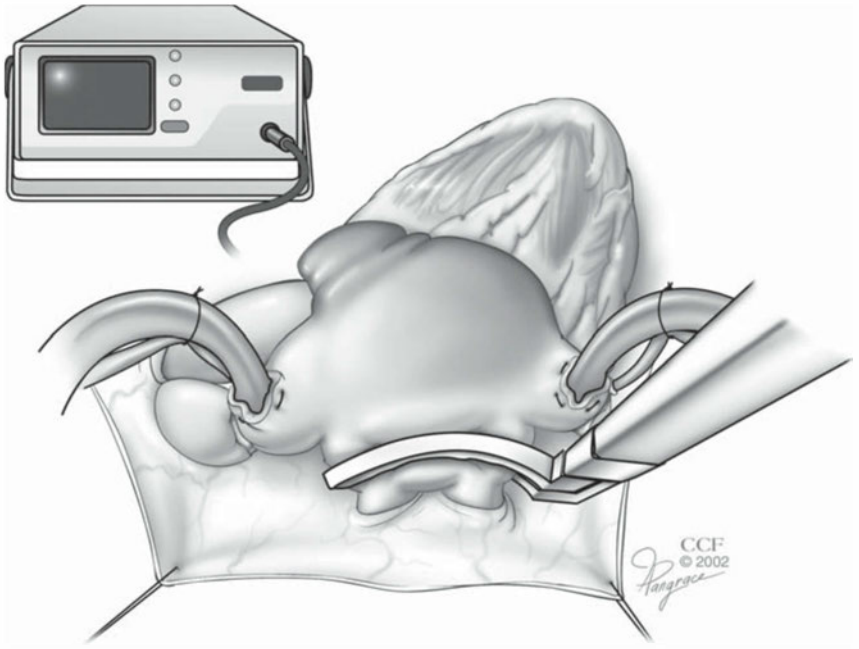


Fig. 66.5. Isolation of the right pulmonary veins with a bipolar radiofrequency clamp (Atricure, Inc., West Chester, OH). Under direct vision, the clamp is placed on the left atrial cuff adjacent to the pulmonary veins. With 3-5 parallel 5- to 15-second applications, the pulmonary veins and a wide left atrial cuff are isolated.

When performed for isolated AF, bilateral thoracoscopic ablation can be performed as described above. PVI is performed with bipolar RF and the veins can be tested for entrance and exit block. During concomitant cardiac surgery, PVI can be performed following sternotomy. It can be done on or off pump. The right superior pulmonary vein is dissected off the right pulmonary artery and the right veins can be encircled with a silastic catheter after opening the oblique sinus. The bipolar clamp is fed around the pulmonary veins. Three to five successful ablations are performed high on the left atrium away from the vein bifurcation (Fig. 66.5). Ablation works best if the interatrial fat pad is opened. The left-sided PVI can also be done off pump but can be challenging. At times, it is easier to perform this on pump or even with the heart arrested. The ligament of Marshall is divided and the left veins are encircled. The bipolar RF clamp is then placed around the left pulmonary veins and ablation performed as high on the left atrium as possible. The LAA can be oversewn, ligated, excised using a stapler, or excluded using commercially available devices. Results with this approach are excellent with 1 year success of 90% freedom from AF.

Other approaches include epicardial box lesion using RF or high-frequency ultrasound. In opening the left atrium, a box lesion can be created using 2-minute cryoablation lesions in a Cox- Maze IV pattern (Fig. 66.6). Similar success rates should be expected.

POSTOPERATIVE MEDICATIONS

Postoperative AF is common and occurs in about 20% to 30%. The first 3 months after ablation is considered a blanking period. If patients develop AF during this time, it does not indicate a failure of the ablation. However, in patients who do not develop AF during this time, they typically remain in sinus rhythm even beyond 3 months. We typically maintain patients after ablation on anticoagulation and antiarrhythmics for 3 to 6 months and gradually wean them off. Typically, patients are monitored with a 7- or 14-day Holter monitor at 6 months and yearly thereafter.

CURRENT CLINICAL ALGORITHM

In patients having concomitant surgery, the plan for ablation depends on the type of AF and the risk of the patient. For patients with paroxysmal AF, there is little risk in performing PVI in most patients. For extremely high-risk patients, or those with severe mitral annular calcification, PVI can add risk, especially ablating the left pulmonary veins as the heart needs to be lifted. The LAA should be excised or oversewn in all these patients. For persistent or long-standing persistent AF patients, a complete lesion set needs to be performed and there is little benefit for PVI alone. Thus, in cases where the left atrium is accessed, a complete lesion set can be performed using cryoablation or RF. In cases where the left atrium is not opened, epicardial RF ablation can be performed. In this case, we base decision to ablate on the longevity of AF, size of the left atrium, and risk to the patient. If patients are unlikely to benefit in this scenario or the risk is additive, then the LAA is excised as the only therapy.

For patients with lone AF, whether paroxysmal or persistent, most patients we see have failed catheter ablation. In these patients, we perform a total thoracoscopic RF ablation with full lesion set as described above. These patients then undergo endocardial testing and ablation of the mitral isthmus and right atrial flutter line (cavotricuspid line).

FUTURE DIRECTIONS

There is great potential in the epicardial treatment of AF for surgeons. Contrary to other areas of cardiology/cardiac surgery, surgical ablation is on the rise. Surgeons must understand the terminology and lesion sets to convince their cardiologists and electrophysiologists to refer patients. Minimally invasive and hybrid approaches are likely going to provide the most durable results at the least invasive approach. Newer technologies for ablation, testing, LAA management, and monitoring will likely lead to improved results. Surgeons must, however, not shy from treating patients with concomitant or lone AF to continue to prove our role in the management of AF.

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EDITOR'S COMMENTS

My partner Gorav Ailawadi has developed a program with our electrophysiologists to perform minimally invasive atrial fibrillation. This will be in contrast to the full cut and sew maze that Dr. Schaff will discuss in another chapter. The bottom line is that this approach does not require a full sternotomy and without a doubt is more appealing to patients as well as to electrophysiologists. The procedure is safe but it certainly takes a while as the technology continues to evolve. The results have been outstanding. It makes sense that this treatment is available for stand-alone atrial fibrillation and I am sure we will continue to be adopted. The results of this procedure are as good if not better as catheter-based ablation.

ILK

67 Surgical Treatment of Hypertrophic Cardiomyopathy

Hartzell V. Schaff

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a relatively common cardiac disease that may be defined as left ventricular (LV) hypertrophy in the absence of an underlying cause such as systemic hypertension or valvular aortic stenosis. Previously, the disorder was referred to as idiopathic hypertrophic subaortic stenosis (IHSS) or asymmetric septal hypertrophy (ASH), but the preferred term is HCM with or without obstruction. Approximately 1 in 500 individuals in the general population will be affected, and this translates into approximately 600,000 people in the United States.

As discussed below, the phenotypes and the clinical presentations of patients with HCM vary widely, and this has led to some confusion in the past about the importance of LV outflow tract (LVOT) obstruction and the indications for surgical treatment. On a molecular level, HCM is an autosomal dominant disease caused by genetic mutations coding for sarcomeric proteins and myofibrils. Over 400 mutations in more than 15 genes have been identified, and double and compound heterozygosity and homozygosity have been reported.

In an individual patient, the occurrence of HCM may be sporadic (20% to 40%) if it represents a new mutation or if there is incomplete penetrance in other family members. Among patients with positive family history, approximately 60% to 70% will have an identifiable HCM mutation by genetic testing. All HCM patients should have a thorough family history and pedigree analysis, preferably by a specialist in medical genetics, to identify and counsel relatives at risk. Genetic testing may be useful to identify specific mutations in the patient and relatives, but there is no clear role for genetic testing in risk stratification for patients known to have HCM.

PATHOLOGY AND PATHOPHYSIOLOGY

The diagnosis of HCM should be considered in any patient who has ventricular hypertrophy without an underlying cause, and most patients with HCM will have a maximal wall thickness ≥ 15 mm. The most common pattern of hypertrophy is diffuse involvement of the ventricular septum, and average maximal LV wall thickness is 20 to 22 mm. In 5% to 10% of patients, LV wall thickness is dramatically increased measuring 30 to 50 mm. Morphology of the septum varies according to age, and older patients with HCM often demonstrate a sigmoid configuration. Among children, increased LV wall thickness is defined as the thickness more than 2 standard deviations above the mean for age, sex, or body size (z score >2). These morphometric distinctions are not rigid, however, as patients have now been identified who are "genotype positive/phenotype negative" and may be considered to have subclinical HCM.

Other patterns of hypertrophy are recognized including mid-ventricular and apical HCM. It has been suggested that as many as one-third of patients with HCM will have segmental wall thickening involving only a portion of the ventricle and overall LV mass may be near normal. Importantly, the phenotype of the disease, the appearance, and severity of hypertrophy does not correlate with genotype, and multiple ventricular morphologies may be found in the same family.

Myocardial fiber disarray is the histologic feature of HCM, and myocytes show large, bizarre nuclei with degenerating muscle fibers and fibrosis. The disorganized whirling of muscle fibers is characteristic of HCM, but myocardial disarray is not pathognomonic for HCM and may be present in myocardium in any conditions in which there is pressure overload.

In the classic form of HCM with obstruction, the mitral valve, especially the anterior leaflet of the valve, moves anteriorly during systole, and the posterior leaflet closes against the mid- and free-edge third of the anterior leaflet instead of at the free edge as occurs normally. The free edge of the anterior leaflet is then displaced upward and narrows the LVOT. Anterior displacement of the valve leaflets produces mitral valve regurgitation (MR) of variable degree, and the jet is oriented in a posterolateral direction. Apposition of the anterior leaflet to the bulging septum produces a contact lesion, a whitish endocardial scar, which is useful in guiding septal myectomy. It is important to note that the occurrence of systolic anterior motion (SAM) of the mitral valve is often dynamic, and provocative maneuvers such as Valsalva, squatting, exercise, and in some cases, adrenergic stimulation may be necessary to elicit SAM, MR, and LVOT obstruction.

Recently, Maron and colleagues reported that mitral valve leaflet length is increased in patients with HCM, an average of 7 mm for the anterior leaflet and 4 mm for the posterior leaflet. Although absolute leaflet length did not correlate with the presence or absence of LVOT obstruction, the authors did find that a ratio of anterior leaflet length to LVOT diameter of >2.0 was associated

with subaortic obstruction. The surgical implications of this finding are unclear, but it was suggested that the elongation of the mitral valve leaflets might constitute a primary phenotypic expression of HCM.

Anomalies of papillary muscles are present in 15% to 20% of patients with HCM who undergo myectomy. These abnormalities include anomalous papillary muscles, direct insertion of papillary muscles into the anterior mitral valve leaflet, fusion of the papillary muscle to the ventricular septum or LV free wall, and accessory muscles and accessory anomalous chordae (false chords). In most cases, these abnormalities do not complicate myectomy or contribute to outflow tract obstruction, but in some patients, anomalous papillary muscles, especially those that insert directly into the body of the anterior leaflet, can contribute to outflow tract obstruction.

Coronary arteries are larger than normal in patients with HCM, and basal coronary flow is increased at rest. In symptomatic patients with HCM, however, coronary flow reserve (CFR) is decreased compared with controls, and phasic flow is abnormal with a greater amount of flow during diastole. Decreased CFR is associated with a reduction in coronary resistance, suggesting that the mechanism is not due to narrowing of intramyocardial small arteries or compression of the microcirculation; indeed, the reduction in CFR in patients with HCM may be the result of near maximal vasodilation of the microcirculation in the basal state.

Atherosclerotic coronary artery disease (CAD) is present in approximately 5% to 15% of patients with HCM depending upon the population studied, and at our clinic, significant CAD was detected in half of the patients who were selected for coronary angiography, and disease was severe in 26%. Survival of HCM patients with obstructive CAD is reduced compared with HCM patients without CAD, and survival is also poorer than patients without HCM who have comparable CAD and normal ventricular function.

Muscular bridging of the left anterior descending (LAD) coronary artery is relatively common and has been identified in 15% of patients with HCM undergoing coronary angiography at our clinic. It is unclear whether bridging of the LAD plays a role in the pathophysiology of the disease and associated symptoms of angina, but for adult patients, risk of death and, in particular, sudden cardiac death are not increased among patients with HCM with myocardial bridging. In contrast, Yetman and coworkers reported that among children with HCM, systolic compression of LAD was present in 285 cases and was associated with a greater incidence of chest pain (60% vs. 19%; $P = 0.04$), cardiac arrest with subsequent resuscitation (50% vs. 4%; $P = 0.004$), and ventricular tachycardia (80% vs. 8%; $P < 0.001$). Myocardial ischemia was postulated to be the cause of this poor outcome.

The most important pathophysiologic feature of HCM is diastolic dysfunction with elevation of the LV end-diastolic pressure, which, in turn, increases left atrial and pulmonary venous pressures. These hemodynamic abnormalities account for the common symptoms of effort dyspnea and limited aerobic capacity. It is understandable, therefore, that atrial arrhythmias, particularly atrial fibrillation (AF), may dramatically worsen diastolic filling when the ventricle is noncompliant and is dependent on atrial contraction.

Elevation of LV end-diastolic pressure is further aggravated by LVOT obstruction and by associated MR. Thus, while surgical myectomy has a minimal immediate effect on the overall LV mass, symptoms related to diastolic dysfunction are immediately improved when outflow gradients and MR are relieved. A secondary effect of septal myectomy on diastolic function is regression of hypertrophy.

A less common mechanism whereby LV hypertrophy led to diastolic dysfunction is the situation where concentric ventricular hypertrophy is so severe that muscle mass encroaches upon the ventricular cavity and reduces the normal cavity size. This is particularly striking in patients with the apical form of HCM where the distal one-third to distal one-half of the left ventricle may be obliterated (during diastole and systole) by muscle. Surgical remodeling by apical myectomy may increase end-diastolic volume and, thus, improve diastolic function.

In the past, there was debate on the importance of outflow tract obstruction as a mechanism for symptoms in patients with HCM. In many instances, outflow tract obstruction is labile, and previously, clinicians questioned whether catheter entrapment accounted for LVOT gradients measured during invasive catheterization. It is now recognized that outflow obstruction is much more common than previously thought. For example, Maron found resting outflow tract gradients of $\geq$50 mmHg in 37% of 320 patients with HCM and, more importantly, found exercise-induced gradients (mean 80 $\pm$ 43 mmHg) in an additional 106 patients. It appears, therefore, that as many as 70% of patients with HCM who come to clinical evaluation will have significant outflow tract obstruction. An important finding in this study was the relative unreliability of the Valsalva maneuver (sensitivity 40%) compared with exercise Doppler echocardiography in detecting these dynamic gradients. Latent obstruction can also be documented during hemodynamic catheterization by isoproterenol challenge, a technique that may be useful in patients who are unable to exercise or in patients in whom reliable Doppler echocardiography cannot be measured.

In patients with obstructive HCM, symptoms of dyspnea and fatigability may also relate to the severity of MR. As is true with LVOT obstruction, associated MR may be dynamic and difficult to appreciate in the resting state. Symptomatic patients with moderate or severe degrees of MR associated with HCM with obstruction are excellent candidates for septal myectomy because the MR and associated symptoms (dyspnea and fatigability) are almost always abolished or significantly improved with relief of LVOT obstruction.

MR that results from SAM is eccentric and directed posterolaterally during late systole. A centrally directed jet should raise suspicion of a primary leaflet abnormality contributing to valve leakage. Rupture of chordae with resultant leaflet prolapse can precipitate congestive heart failure, and hemodynamics may worsen if HCM is not recognized and patients are managed medically with afterload reduction. Preoperative transesophageal echocardiography (TEE) is unnecessary in most patients, but TEE is critically important intraoperatively in assessing the results of myectomy.

CLINICAL PRESENTATION AND NATURAL HISTORY

Many patients with HCM are asymptomatic, but most who come to surgical attention will have limiting symptoms. Typical symptoms caused by obstructive HCM are exertional dyspnea, chest pain (angina), and/or lightheadedness, especially lightheadedness is associated with rapid change in posture. Although some patients present in childhood and adolescence, the most common clinical scenario for patients referred for septal myectomy is the development of symptoms in the fourth, fifth, or sixth decade of life. In most patients, it is the development of symptoms that leads to the diagnosis of HCM, and, thus, it is unknown whether LV outflow gradients existed previously. It seems likely, however, that in most patients, the development of symptoms corresponds to the development of subaortic obstruction. Occurrence of AF can also precipitate symptoms and predispose to systemic embolism, which occurs in 6% of patients. AF is found in 30% of older patients with HCM.

It is interesting to note that approximately one-third of patients with HCM who are symptomatic will have exacerbation of symptoms, predominantly dyspnea or presyncope, after meals. Postprandial symptom exacerbation is associated with higher resting LVOT gradients and advanced clinical symptoms.

In the general population, survival of patients with HCM is similar to survival of individuals without disease, and high mortality in HCM in earlier reports is likely due to excess numbers of high-risk patients included in studies from tertiary referral centers. More recent natural history studies indicate that the annual mortality rate of patients with HCM is approximately 1%, but there are several important subgroups that have higher risk of cardiac death. For example, HCM is the most common cause of sudden death among young athletes.

The importance of LVOT obstruction with regard to late outcome of patients with HCM has been controversial, but there is now substantial evidence that survival of HCM patients with outflow obstruction is reduced in comparison to patients without obstruction. Studies by Maron et al., Autore and associates, and Elliot and coworkers have demonstrated convincingly a strong correlation between resting outflow gradients and late risk of death. In a longitudinal follow-up of 1,101 patients with HCM, Maron et al. reported that patients with outflow tract obstruction (a basal gradient of at least 30 mmHg) had a risk of death from HCM or symptom progression that was more than four times that observed among patients without obstruction (Fig. 67.1). The association of outflow tract obstruction on limiting symptoms and death was independent of other clinical variables. Of note, patients with obstruction and mild symptoms (NYHA class II) were more likely to have progression to severe symptoms or to die from heart failure than asymptomatic patients with cardiomyopathy (Fig. 67.2).

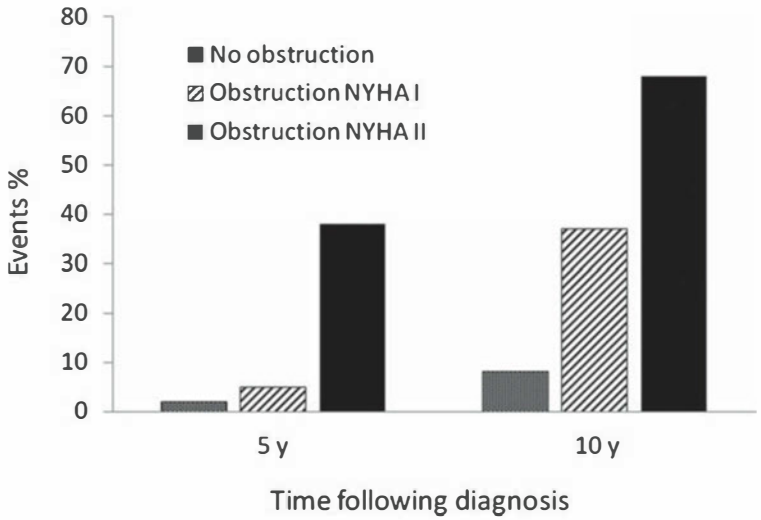


Fig. 67.2. Risk of progression to severe symptoms (New York Heart Association [NYHA] class III or IV) or death from heart failure or stroke. Poorest outcome occurred in patients with obstruction and symptoms at the time of gradient measurement. (Modified from Maron MS, Olivotto I, Betocchi S, et al. Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophic cardiomyopathy. N Engl J Med 2003;348:295-303.)

Elliot et al. reported that risk of sudden death is relatively low (<0.4% per year) in asymptomatic patients with LVOT obstruction and none of the recognized risk factors for SCD, but their study did demonstrate reduced survival among patients with LVOT obstruction and nonsustained ventricular tachycardia, abnormal exercise blood pressure response, a family history of premature sudden death, unexplained syncope, or severe LV hypertrophy.

The prognosis of patients with latent obstruction is less clear, but a study by Vaglio et al. suggests that the clinical course is similar to patients with resting obstruction. In their series, one-quarter of patients required invasive therapy for relief of symptoms, and annual mortality was 2% per year, a rate higher than that reported for patients with HCM and no LVOT obstruction.

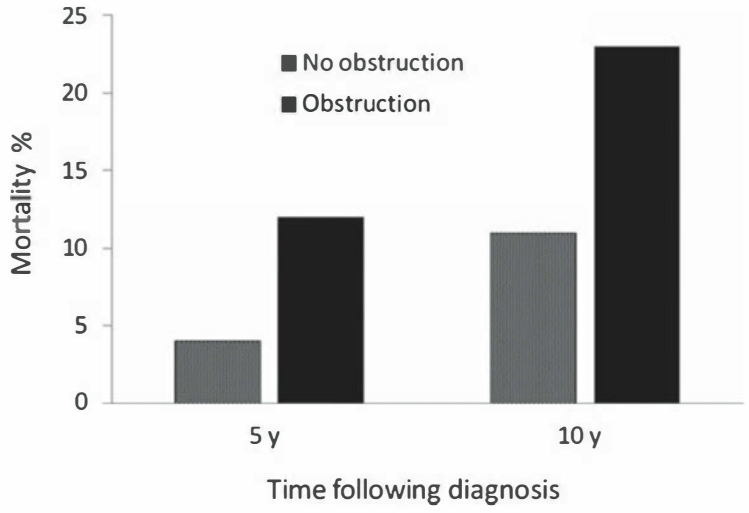


Fig. 67.1. Risk of hypertrophic cardiomyopathy-related death in patients with and without left ventricular outflow tract obstruction (>30 mmHg). (Modified from Maron MS, Olivotto I, Betocchi S, et al. Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophic cardiomyopathy. N Engl J Med 2003;348:295-303.)

Indications for Operation

Septal myectomy is, in general, reserved for patients who continue to have limiting symptoms despite medical treatment. Pharmacologic therapy usually begins with beta-adrenergic blocking agents, which have a negative inotropic effect and may mitigate latent outflow gradients provoked with exercise. Beta-blockers are less effective in reducing high resting gradients. The calcium antagonist, verapamil, has been used for patients with nonobstructive and obstructive HCM with the aim of improving ventricular relaxation and decreasing LV contractility, but hemodynamic and electrophysiologic side effects limit long-term use. Disopyramide has negative inotropic effects and is a type I-A antiarrhythmic agent that can improve symptoms by reducing resting gradients. However, this drug also has side effects including dry mouth and eyes, constipation, and difficulty in micturition. Also, it increases atrioventricular nodal conduction and thus may increase ventricular rate in patients

with AF. It is important to realize that while medications may ameliorate symptoms, drug therapy does not appear to decrease the probability of sudden death. In a recent study of 173 patients who were taking amiodarone, beta-blockers, verapamil, and/or sotalol for the treatment of symptoms, there was no difference in sudden death mortality compared with patients who were on no pharmacologic therapy.

Although guidelines emphasize the importance of preliminary medical treatment before consideration of surgery, there are other important considerations regarding indications for septal myectomy in patients with obstructive HCM. Some patients (Fig. 67.3) have very favorable anatomy with localized hypertrophy in the subaortic septum and relatively normal wall thickness throughout the rest of the ventricle. In these patients, septal myectomy may be the preferred treatment rather than prolonged medical therapy because the results of surgery are so predictably good and relief of outflow tract gradient would be expected to relieve symptoms completely as there is little remaining substrate for diastolic dysfunction. Not infrequently, patients will prefer surgical treatment because of intolerance to side effects of medical treatment, especially side effects of high-dose beta-blockers. A final group of patients who might be considered earlier for surgical myectomy are patients who have high resting gradients (≥50 mmHg) and have a history of prior cardiac arrest or a strong family history of sudden cardiac death.

SURGICAL TECHNIQUES

A standard median sternotomy is preferred to provide adequate access both to the aorta and to the left ventricle. We favor normothermic cardiopulmonary bypass using a single, two-staged venous cannula and cold blood cardioplegia (initial dose of 1,000 to 1,200 ml) for myocardial protection. Adequate exposure of the subaortic septum is critically important, and several maneuvers facilitate the operation. Pericardial sutures are used only on the right side to elevate pericardium toward the surgeon and allow the left ventricle to fall posteriorly in the thorax. Next, an oblique aortotomy is made slightly closer to the sinotubular ridge than is usual for aortic valve replacement, and the incision is carried through the midpoint of the noncoronary aortic sinus of Valsalva to a level approximately 1 cm above the valve annulus. As illustrated in Figure 67.4, the edge of the proximal aorta is held out of the way with stay sutures of 4-0 polypropylene, and a cardiotomy sucker is placed through the aortic valve and used to depress the anterior leaflet of the mitral valve and protect it from injury. The right aortic valve cusp is collapsed against the sinus wall where it will usually stay. A sponge stick is used to depress the right ventricle and rotate the septum posteriorly, orienting the LV outflow anteriorly.

A standard No.10 scalpel blade is used for incision in the septum that begins just to the right of the nadir of the right aortic sinus (Fig. 67.5). The initial incision in the septum is made upward and then leftward over to the anterior leaflet of the mitral valve. Scissors are used to complete excision of this initial portion of myocardium. The area of septal excision is then deepened and lengthened toward the apex of the heart being certain to excise hypertrophied septum beyond endocardial scar. Trabeculations are excised, and the myectomy site is further enlarged with the use of pituitary rongeurs. Adequate septal myectomy usually yields 3 to 12 g of muscle. The use of the sponge stick to depress the heart posteriorly will improve exposure of the distal extent of the myectomy. The aortotomy is closed in two layers with 4-0 polypropylene suture, and the operation proceeds as usual.

This technique described above for more extended myectomy differs from the standard Morrow operation in which parallel incisions create a trough in the septum that extends up to 3 cm from the aortic valve. With the extended myectomy, the wider excision of muscle in the immediate subaortic area improves exposure of the distal extent of the hypertrophied septum, and excision extends up to 7 cm from the aortic valve. Inadequate myectomy results more often from failure to excise sufficient length of the septum (toward the apex) than from inadequate depth of excision. Importantly, the methods described above have proved adequate for all patients with subaortic obstruction, and maneuvers such as anterior leaflet plication are unnecessary and, we believe, potentially harmful. Mitral valve replacement is reserved for patients with intrinsic leaflet abnormalities that cannot be repaired.

To confirm complete relief of the LVOT obstruction, we routinely measure

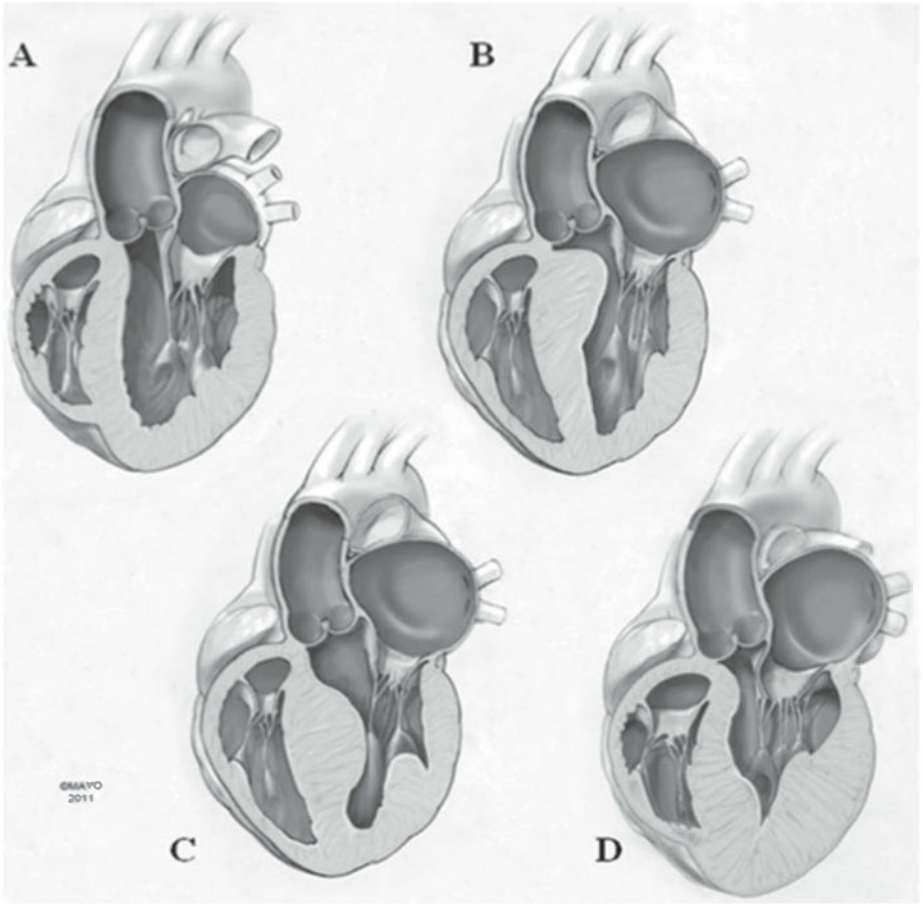


Fig. 67.3. Some anatomic subtypes of hypertrophic cardiomyopathy compared with a normal heart (**panel A**). Patients who predominantly have septal hypertrophy shown in (**panel B**) are good candidates for operation because of the relative lack of thickening in the remaining ventricle and the low likelihood of residual diastolic heart failure.

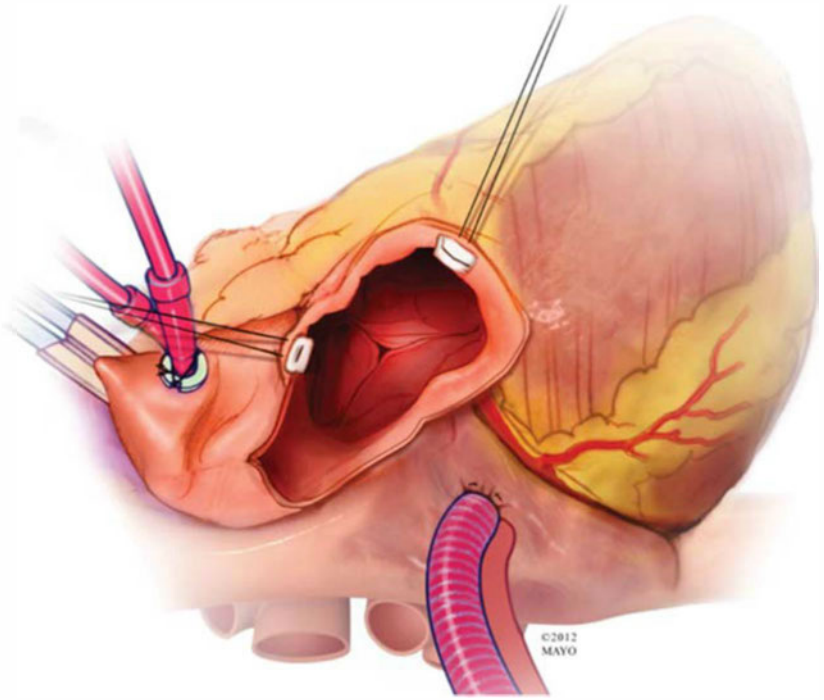


Fig. 67.4. An oblique aortotomy extended into the noncoronary sinus gives the best exposure of aortic valve and subaortic septum.

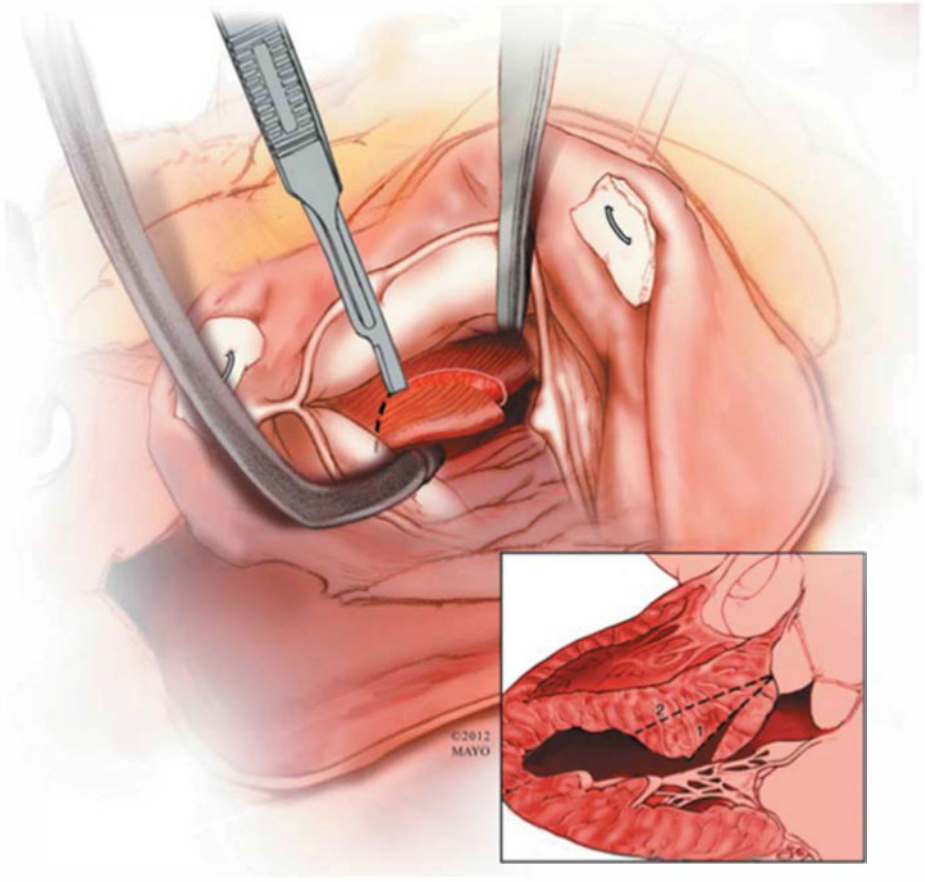


Fig. 67.5. Septal excision begins just to the right of the nadir of the right aortic sinus and extends leftward to the attachment of the anterior leaflet of the mitral valve. The depth of muscle excised is approximately 1.5 to 2.0 cm, but more important than depth is extent of excision toward the apex. This is guided visually by endocardial scar, and as illustrated in the inset, the initial excision is almost always supplemented by excision of another large portion of the more distal septum.

simultaneous aortic and LV pressure by direct needle puncture before and after myectomy. If the resting LVOT gradient is diminished by the effects of anesthesia, premature ventricular beats are induced by stimulating the ventricle to elicit the dynamic gradient produced by the Brockenbrough phenomenon; the postextrasystolic contraction is more forceful due to increased contractility and decreased afterload (Fig. 67.6); the same maneuver is then repeated after myectomy. In some patients, it is necessary to provoke LVOT gradient during surgery with isoproterenol (Fig. 67.7). Intraoperative TEE can identify residual MR, SAM, and any septal defects created by excision of septal muscle.

Severe MR in patients with obstructive HCM is not an indication for mitral valve replacement. Indeed, MR due to SAM is always relieved when outflow tract obstruction is corrected. In patients who have suspected intrinsic mitral valve disease in addition to obstructive HCM, direct mitral valve repair may be necessary, but we prefer first to perform extended septal myectomy and then discontinue cardiopulmonary bypass and reassess mitral valve function by echocardiography. When, in addition to obstructive HCM, there is marked degenerative mitral valve disease producing prolapse with or without ruptured chordae, valve repair is possible in over 85% of patients.

Unroofing of coronary artery bridging is performed in selected cases, particularly in young patients and those who have angina preoperatively. After the identification of the coronary artery distal to the intramyocardial segment, the anterior surface of the vessel is exposed by sharp dissection that is continued proximally until the LAD reemerges onto the epicardium. The divided myocardium over the artery is usually 3 to 5 mm thick. If the course of the artery is so deep that trabeculations of the right ventricular cavity are entered, pledgeted sutures placed deep into the coronary artery repair the opening into the ventricle.

OUTCOMES OF MYECTOMY

In experienced centers, risk of hospital death after isolated septal myectomy for obstructive HCM is very low, <1% (Fig. 67.8), and similar to risk of operation for elective mitral valve repair. As is true with other procedures, perioperative risk of septal myectomy may be higher among very elderly patients (particularly those with severe disabling symptoms associated with pulmonary hypertension), patients with prior myectomy, or those requiring concomitant procedures.

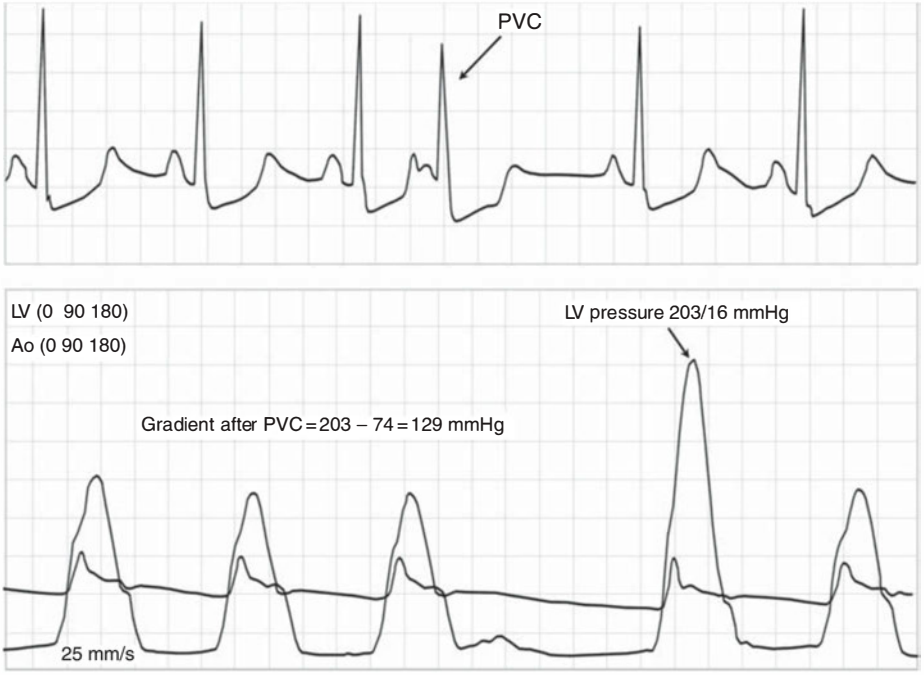


Fig. 67.6. Intracardiac pressures measured before septal myectomy. An augmented left ventricular outflow tract gradient is apparent following a premature ventricular contraction (PVC) induced by stimulating the right ventricle. Ao, aorta; LV, left ventricle. (Modified from Ashikhmina EA, Schaff HV, Ommen SR, et al. Intraoperative direct measurement of left ventricular outflow tract gradients to guide surgical myectomy for hypertrophic cardiomyopathy. J Thorac Cardiovasc Surg 2011;142:53-59.)

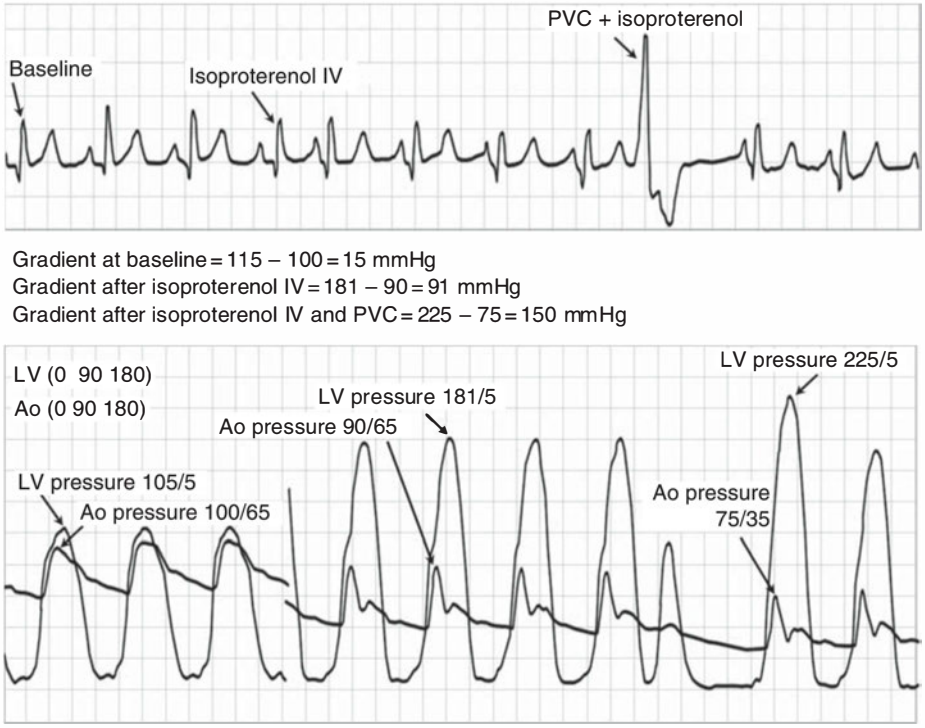


Fig. 67.7. Intracardiac pressures measured before septal myectomy. In patients with latent obstruction who, under general anesthesia, have minimal left ventricular outflow tract (LVOT) obstruction induced by a premature ventricular contraction (PVC), we infuse isoproterenol to augment the gradient, and similar maneuvers are performed after myectomy. Ao, aorta; LV, left ventricle. (Modified from Ashikhmina EA, Schaff HV, Ommen SR, et al. Intraoperative direct measurement of left ventricular outflow tract gradients to guide surgical myectomy for hypertrophic cardiomyopathy. J Thorac Cardiovasc Surg 2011;142:53-59.)

Complications such as complete heart block requiring permanent pacemaker and iatrogenic ventricular septal perforation have become uncommon (≤1% to 2%). Partial or complete left bundle branch block is a frequent finding after surgical myectomy but is not associated with adverse sequelae. However, if the patient has complete right bundle branch block preoperatively, interruption of the left bundle after myectomy increases the risk of complete heart block. This is particularly important in patients who have had alcohol septal ablation prior to operation. Alcohol septal ablation causes right bundle branch block in up to 60% of patients, and in patients who have had previous alcohol septal ablation, the risk of pacemaker insertion following surgical myectomy was 36% compared with 3% for those without prior intervention.

Adequate relief of LVOT obstruction with septal myectomy dramatically improves symptoms in exercise capacity in symptomatic patients with obstructive HCM. In our experience, approximately 90% of severely symptomatic patients have improvement of greater than or equal to two function classes, and relief of outflow gradients by myectomy is equally effective in improving limitation due to dyspnea, angina, or syncope. Importantly, symptomatic benefit of myectomy is directly related to reducing the basal outflow gradient and MR and improving LV systolic and end-diastolic pressures (in more than 90% of patients), which, in turn, may also favorably influence LV diastolic filling and myocardial ischemia. Relief of the gradient may decrease left atrial size and the subsequent risk of developing AF.

Late recurrence of significant resting LV outflow gradients is very uncommon after successful myectomy in either adults or children with obstructive HCM, and this is in contrast to patients who have surgery for the relief of congenital membranous subaortic stenosis. Common causes of recurrent LVOT obstruction and symptoms include limited myectomy at the first operation, mid-ventricular obstruction, and anomalies of papillary muscles. Most often, inadequate myectomy at initial operation is due to failure to extend the myectomy far enough toward the apex of the heart.

As mentioned previously, most patients are referred for septal myectomy because of persistent symptoms despite medical treatment. There is, however, evidence that operation may improve survival of patients with obstructive HCM. In a study from our clinic, Ommen and associates reported that late survival of 289 patients with obstructive

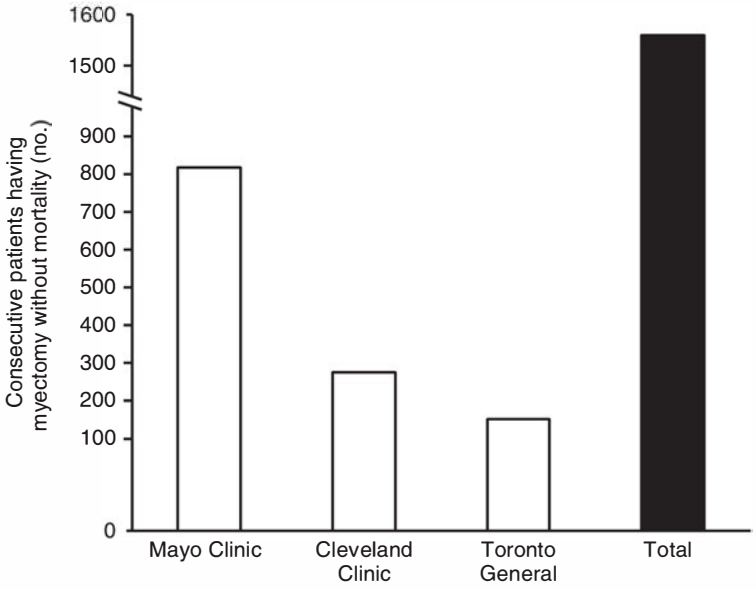


Fig. 67.8. Operative risk of isolated septal myectomy for hypertrophic cardiomyopathy is <1% and approaches 0 in centers with broad experience with the procedure. (Modified from Maron BJ. Surgical myectomy remains the primary treatment option for severely symptomatic patients with obstructive hypertrophic cardiomyopathy. Circulation 2007;116:196-206.)

HCM undergoing septal myectomy was 98%, 96%, and 83%, 1, 5, and 10 years postoperatively, and this survival was similar to that of an age- and sex-matched U.S. population and similar to survival of HCM patients without obstruction. In the same study, myectomy patients had superior survival free from all-cause mortality ($P < 0.001$), HCM-related mortality ($P < 0.001$), and SCD ($P = 0.003$) compared with unoperated HCM patients with obstruction (Fig. 67.9), and on multivariable analysis, performance of myectomy had a strong, independent association with survival (hazard ratio 0.43; $P < 0.001$).

Recent studies also suggest mechanisms whereby relief of LVOT gradients might improve survival. The extent of LV hypertrophy is an important determinant of survival in patients with HCM, and successful myectomy results in some regression of LV hypertrophy. Thus, patients with the obstructive form of HCM may have an additive burden of ventricular hypertrophy caused by pressure overload that is relieved by successful myectomy. Although the severity of LV hypertrophy is a determinate of survival in unoperated patients, LV mass and wall thickness are not predictors of late survival in HCM patients following myectomy.

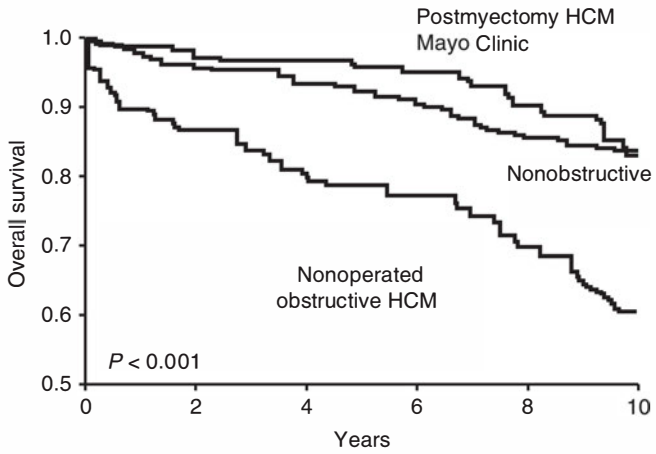


Fig. 67.9. Survival free from all-cause mortality in three subgroups of patients with HCM: Mayo Clinic surgical myectomy ($n = 289$), nonoperated with LVOT obstruction ($n = 228$), and nonobstructive ($n = 820$). Myectomy versus nonoperated obstructive hypertrophic cardiomyopathy, $P < 0.001$; myectomy versus nonobstructive hypertrophic cardiomyopathy, $P = 0.8$. (Modified from Ommen SR, Maron BJ, Olivotto I, et al. Long-term effects of surgical septal myectomy on survival in patients with obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol 2005;46:470-476.)

Relief of LVOT obstruction may also reduce the risk of serious arrhythmias. McLeod and associates reviewed patients with HCM who had received ICDs, and during a median follow-up of 4.5 years, 12 patients (17%) in the nonmyectomy group and only one patient (2%) in the myectomy group had appropriate ICD discharges (Fig. 67.10). The average annualized event rate was 4.3% per year in the nonmyectomy group, compared with 0.24% per year following myectomy ($P = 0.004$). Thus, surgical myectomy and relief of outflow gradient are associated with a marked reduction in the incidence of appropriate ICD discharges and risk of SCD.

Latent obstruction is the term used to describe low resting LVOT gradients (0 to 30 mmHg) that increases to over 50 mmHg with provocation by Valsalva maneuver, exercise, inhalation of amyl nitrite, or infusion of catecholamines. Indeed, as many as one-third of patients with HCM and exertional symptoms will have low resting LVOT gradients. Septal myectomy should be considered in patients with latent obstruction because studies have shown that the degree of functional improvement and late survival in this subgroup of patients are similar to the clinical improvement experienced by patients who have successful operation for relief of high resting gradients (Fig. 67.11).

Selective infarction of the septum by infusing alcohol into the septal branches of the LAD can relieve LVOT obstruction, and there are obvious advantages to the patient of avoiding surgical incision and hospitalization. In a review of alcohol septal ablation from 42 published studies (2,959 patients), resting LVOT gradient decreased from 65 to 16 mmHg ($P < 0.001$), basal septal diameter reduced from 21 to 14 mm ($P < 0.001$), and hospital mortality was 1.5%. However, the incidence of complete heart block requiring permanent pacemaker was 11%, and other important complications included coronary dissection (1.8%), pericardial effusion (0.6%), ventricular fibrillation (2.2%), stroke (1.1%), new right bundle branch block (46%), and new left bundle branch block (6.5%). Also, symptoms persisted in 11% of patients, redo alcohol ablation was required in 6.6%, and surgical myectomy was necessary in 2.0%. Thus, while this procedure is less invasive, it has significant morbidity, and residual gradients and incidence of conduction abnormalities are higher than that observed following septal myectomy.

Some patients with obstructive HCM may not be candidates for alcohol septal

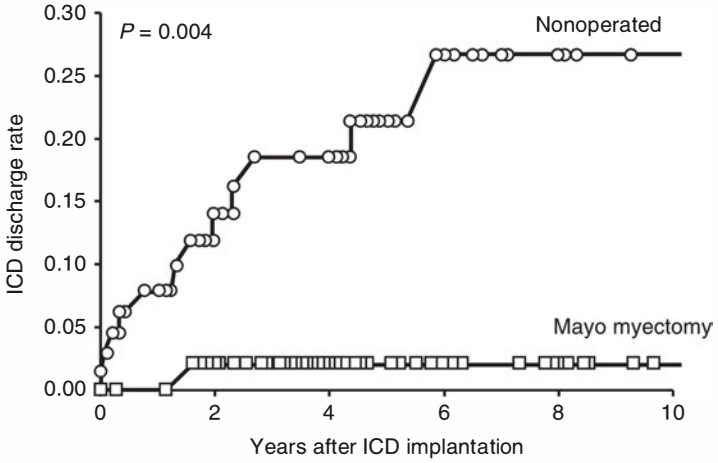


Fig. 67.10. Cumulative rates of ICD discharges in hypertrophic cardiomyopathy (HCM) patients with obstruction who had myectomy at Mayo Clinic and those who did not have myectomy. Average annualized implantable cardioverter defibrillator discharge rates of 0.24% in the myectomy group, compared with 4.3% in the nonmyectomy group ($P = 0.004$). (Modified from McLeod CJ, Ommen SR, Ackerman MJ, et al. Surgical septal myectomy decreases the risk for appropriate implantable cardioverter defibrillator discharge in obstructive hypertrophic cardiomyopathy. Eur Heart J 2007;28:2583-2588.)

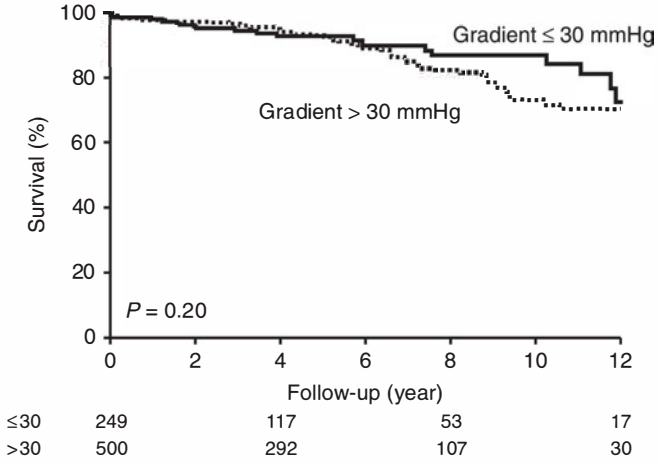


Fig. 67.11. Overall survival of patients after isolated transaortic septal myectomy is similar for patients with latent obstruction (≤30 mmHg) preoperatively compared with those with more severe obstruction (>30 mmHg). (Modified from Schaff HV, Dearani JA, Ommen SR, et al. Expanding the indications for septal myectomy in patients with hypertrophic cardiomyopathy: results of operation in patients with latent obstruction. J Thorac Cardiovasc Surg 2012;143:303-309.)

ablation because of unfavorable coronary anatomy. Also, concern has been raised about the potential long-term risk of sudden death related to arrhythmogenic substrate from the resulting septal scar. Cardiac magnetic resonance imaging demonstrates that in most patients, alcohol septal ablation causes transmural tissue necrosis, located more inferiorly in the basal septum than myectomy; the infarct usually extends into the right ventricular side of the septum and sometimes spares the basal septum leading to residual gradients at follow-up. A recent report from a registry of 874 patients (average age 55 years) who underwent alcohol septal ablation describes postprocedure survival of 86% and 74% at 5 and 9 years, considerably lower than the 96% and 83% of 5- and 10-year survival of surgical patients. In our practice, alcohol septal ablation is considered an option for patients who because of advanced age, infirmity, and/or comorbid conditions have increased risk for operation. For most patients, however, septal myectomy remains the gold standard for patients with HCM and severe LV outflow obstruction who are refractory to maximal medical therapy.

SPECIAL CONSIDERATIONS

There are few guidelines for the management of asymptomatic patients with HCM. Patients without symptoms may be discovered to have HCM during the course of evaluation of other cardiopulmonary disease, as a result of pedigree analysis of affected family members, or through genetic testing precipitated by identification of HCM in a relative. Negative inotropic drugs and beta-blockers may be useful for associated comorbid conditions such as systemic hypertension, but there is no evidence that these medications alter the course of HCM. Asymptomatic patients should be cautioned to avoid extreme exercise and should have thorough evaluation for risk factors for SCD. For patients who have no obstruction and no symptoms, surgery is not a consideration. For asymptomatic patients with HCM and very severe LVOT obstruction, however, decision-making is more difficult. Formal exercise testing with measurement of peak oxygen consumption may be helpful in uncovering significant cardiopulmonary limitation in patients who minimize symptoms. Established guidelines caution against septal reduction therapy in asymptomatic patients, but septal myectomy may be appropriate in selected young patients who have appropriate anatomy, very high resting and exercise gradients, and strong family history of premature cardiac death.

Sudden cardiac death is a special concern in HCM, and there is debate on the appropriate use of ICDs for primary and secondary prevention of SCD. Variables that appear to identify patients at increased risk of arrhythmic events include prior cardiac arrest or sustained ventricular tachycardia, a family history of premature sudden death due to HCM, repetitive nonsustained ventricular tachycardia on ambulatory Holter monitoring, massive ventricular hypertrophy (wall thickness >30 mm), and hypotensive response to exercise. Other findings such as myocardial bridging in young patients or myocardial fibrosis detected by cardiac MRI may also increase the risk of sudden death.

Apical HCM is a relatively rare morphologic subtype of HCM that is more common in Asia than in the West. It has a benign course in most patients, but diastolic dysfunction leading to heart failure may occur. In many of these patients, diastolic heart failure appears to be due to very small LV end-diastolic volume related to encroachment on the LV cavity by the hypertrophied apical muscle. The diastolic heart failure in selected symptomatic patients with small LV cavities may have improved LV remodeling achieved by myectomy through an apical incision (Figs. 67.12 and 67.13).

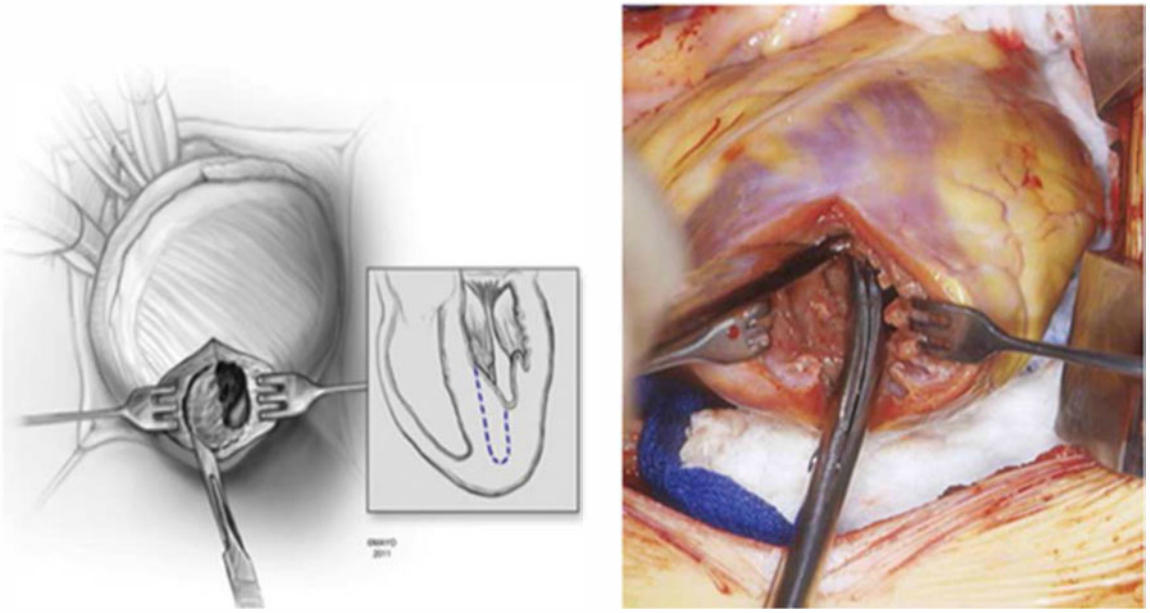


Fig. 67.12. A transapical ventriculotomy is used for apical myectomy as illustrated here. The incision is made over the dimple of the left ventricular apex and far enough from the left anterior descending coronary artery so that the incision can be closed without compromising coronary blood flow.

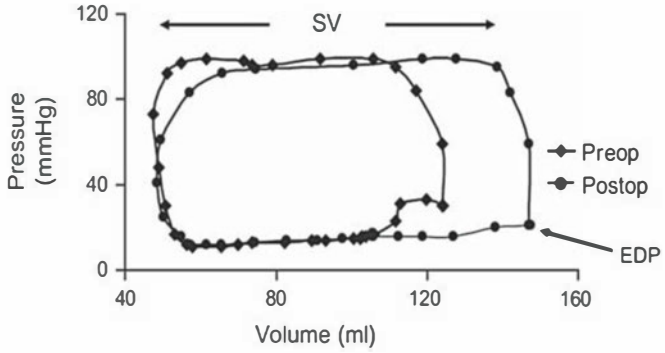


Fig. 67.13. These left ventricular pressure–volume loops obtained before and after apical myectomy demonstrate improvement in diastolic function and stroke volume in a patient with severely symptomatic apical hypertrophic cardiomyopathy. EDP, end-diastolic pressure; SV, stroke volume.

Other patients with HCM who may be surgical candidates are patients with mid-ventricular obstruction. In its pure form, mid-ventricular obstruction occurs because of hypertrophy of the papillary muscles and mid-septum, which come into contact during systole. The resulting high intracavitary gradients can cause exertional-related dyspnea and angina, as is seen with subaortic obstruction. Also, ventricular arrhythmias may occur in approximately 20% of patients with mid-ventricular obstruction who develop apical aneurysms. A key differentiating feature separating these patients from patients with typical subaortic obstruction is the lack of SAM of the mitral valve apparatus with mid-ventricular obstruction. Relief of mid-ventricular obstruction improves symptoms, and in our experience, the best surgical approach is a transapical ventriculotomy. Through this incision, the hypertrophied ventricular septum is easily exposed, and endocardial scar guides the extent of myectomy. Also, it is relatively simple to shave the anterolateral and/or posteromedial papillary muscles, which contribute to the mid-ventricular obstruction.

Children with HCM are usually asymptomatic, and the overall annual mortality for pediatric HCM beyond the first year of life is relatively low, approximately 1% per year. However, symptomatic HCM and HCM-related SCD can occur in childhood. As is true with adults, symptomatic children with obstructive HCM can be dramatically improved with extended transaortic myectomy, but there are special considerations in operating on infants and young children. Exposure of the subaortic septum may be difficult when the aortic diameter is small, and extreme care should be taken to avoid injury to the aortic valve cusps and to the mitral valve leaflets. In our experience with myectomy in 127 children, there has been no operative mortality, but injury to the aortic valve producing valvular regurgitation occurred in 6% of patients; fortunately, valve repair was possible, and no patient required replacement at the time of myectomy.

There is general consensus that an ICD is strongly warranted for secondary prevention of SCD in patients with prior cardiac arrest or sustained and spontaneously occurring ventricular tachycardia, and should be considered for primary prevention in patients with multiple clinical risk factors and in selected patients with a single major risk factor such as a history of SCD in a close relative. In patients who receive a device for secondary prevention, the yearly rate of appropriate ICD discharge is 10%; for HCM patients with ICDs implanted for primary prevention, the rate is 4% per year. For patients who need an ICD and are referred for septal myectomy, we prefer to delay device implantation until the third or fourth day postoperatively and avoid the potential for lead dislocation if the ICD is placed immediately preoperatively.

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EDITOR'S COMMENTS

I doubt many people have near the experience that Dr. Schaff has in the surgical treatment of hypertrophic cardiomyopathy. Dr. Schaff has given us a thorough review and his analysis of results as well as surgical therapy is outstanding. I think there are two important points to take from this discussion. The most important is the length of the myectomy, maybe more important than the depth of myectomy. As Dr. Schaff has noted first that the failure to do a long enough myectomy can lead to recurrences. Second, he has noted that mitral regurgitation can be helped with both the myectomy and occasionally concomitant mitral repair. He reserves mitral valve replacement to those patients who have intrinsic mitral valve disease. I think these are critical points and his approach should essentially lead to good results.

ILK

68 Cardiac Tumors

Bo Yang, Himanshu J. Patel, Francis D. Pagani, and Richard L. Prager

HISTORICAL BACKGROUND

Realdo Columbus first described a cardiac tumor as an anatomic finding in Padua, Italy, in 1559. Centuries later, in 1931, the first classification system similar to what is in current use was reported by Yater. In his monograph, he reported nine cases of primary cardiac tumors from pathologic examination. However, it was not until 1934 that the first antemortem diagnosis of a cardiac tumor (sarcoma) was made by Barnes, who used electrocardiography and a biopsy of a lymph node. Twelve years later, Mahaim's classic paper described over 400 cardiac tumors.

The era of operative treatment of cardiac neoplasms was ushered in by Beck in 1936, when he successfully removed a teratoma located on the right ventricular (RV) surface. Bahnson is credited with the removal of the first right atrial myxoma with inflow occlusion, but the patient died postoperatively on day 24. Following the advent of cardiopulmonary bypass in 1953, left-sided intracardiac neoplasms were successfully removed first by Crafoord in Sweden in 1954. By 1964, the removal of 60 intracardiac neoplasms had been reported. The addition of cardiac echocardiography allowed easier antemortem diagnosis, and thus resulted in an increase in the number of tumor resections.

EPIDEMIOLOGY AND CLASSIFICATION

Cardiac neoplasms can be divided into primary and secondary types. Primary tumors are typically more common, but the overall incidence is still quite low, at 0.15% to 0.2% in autopsy series. Most primary cardiac tumors are benign (70%; Table 68.1). More than half of the primary cardiac neoplasms in adults are myxomas.

Malignant primary cardiac tumors (Table 68.2) are more frequently seen in adults than in children. Of malignant cardiac tumors, metastatic malignancies (Table 68.3) comprise the majority of those noted. Virtually every neoplasm has been shown to metastasize to the heart. The most frequent primary malignancies are leukemic neoplasms (54%), melanoma (34%), and bronchogenic carcinoma (10%), with others including sarcoma, breast, and esophageal carcinoma. Primary cardiac malignancies are uncommon, and most frequently are sarcomas.

MYXOMAS

Myxomas are the most frequently seen adult cardiac neoplasm. They usually occur sporadically but have been reported in autosomal-dominant inherited forms in 5% of cases. The typical sporadic tumor is seen in women aged 30 to 60 years and is solitary in nature. The familial form is more likely seen in younger patients, often male, and multicentric in nature. The two types can be differentiated on the basis of DNA ploidy, with the familial type having an abnormal ploidy. Carney complex is an autosomal X-linked inheritance characterized by cardiac myxoma, cutaneous pigmentous lentigines, and primary pigmented adrenocortical disease with hypercortisolism.

Myxomas are typically located in the atria (most commonly on the limbus of the fossa ovalis) (Fig. 68.1A) but can arise in the ventricles. Multiple myxomas with involvement of different heart chambers have been reported but are very rare. The tumors are variable in their gross pathologic appearance (Fig. 68.1B), and can be papillary or smooth, pedunculated, or sessile, but are often quite friable. They are usually white or yellow and may be covered with thrombus. On cut sections, they frequently contain areas of hemorrhage. They are typically 5 to 6 cm in size but have been reported to reach up to 15 cm. Histopathologically, they have an acid mucopolysaccharide matrix and contain smooth muscle cells, capillaries, and reticulocytes. Calcification is reported more frequently in right atrial myxomas. These tumors typically grow outward into the cardiac chambers and rarely invade into the walls of the heart. However, those tumors that are biatrial in location are thought to be a result of bidirectional growth of the myxoma because they are usually attached to the same point on the atrial wall. Usually, they are limited to the subendocardial region at their base. The subendocardial multipotential mesenchymal cell is considered the precursor cell for myxoma, thus accounting for the variable cell types seen in the tumor.

The natural history of myxomas is of rapid growth. Although these tumors are considered to be benign, there have been reports of extensive local extension, as well as metastatic spread. The familial type is more likely to be recurring and more aggressive.

The clinical presentation patterns of myxomas relate to their potential to cause obstruction (congestive heart failure, atrial fibrillation, fatigue, and syncope), embolization, and constitutional symptoms (myalgias, fevers, arthralgias, and weakness). Occasionally, they can present with evidence of infection, with a syndrome not unlike infective endocarditis. Physical findings can include signs of right- or left-sided congestive failure, the early diagnostic "tumor plop," or diastolic rumbles.

The workup of a suspected myxoma includes echocardiography as the most useful test for diagnosis. Although surface echocardiograms can identify the pathology in most cases, the transesophageal echocardiogram (Fig. 68.1C) gives the best images and provides details regarding the location and attachment areas for tumors even as small as 2 mm. This aids in planning the operative approach. We also request left heart catheterizations to aid in the diagnosis of coronary disease in patients older than the age of 45 years or even younger if they have significant risk factors or symptoms. Computed tomography (CT) and magnetic resonance imaging (MRI) are rarely utilized at our center for suspected myxomas because in the setting of a myxoma, they typically do not add additional information. However, if there is suspicion that the

Table 68.1 Benign Cardiac Neoplasms

Myxoma
Papillary fibroelastoma
Lipoma
Teratoma
Rhabdomyoma
Pheochromocytoma
Fibromas
Atrioventricular node mesothelioma
Neurofibroma
Lymphangioma
Granular cell tumor

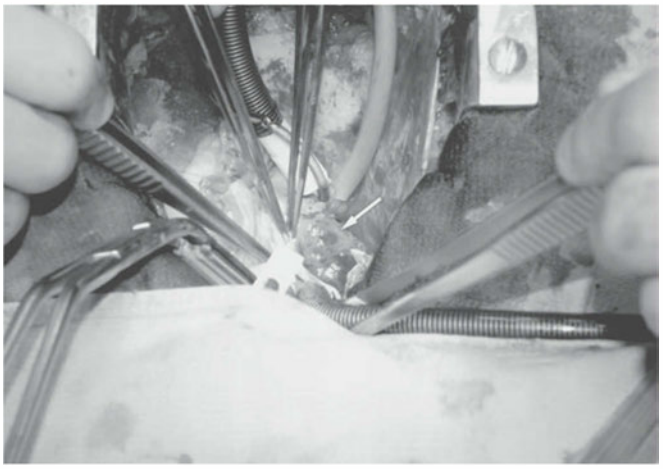
Table 68.2 Malignant Primary Cardiac Neoplasms

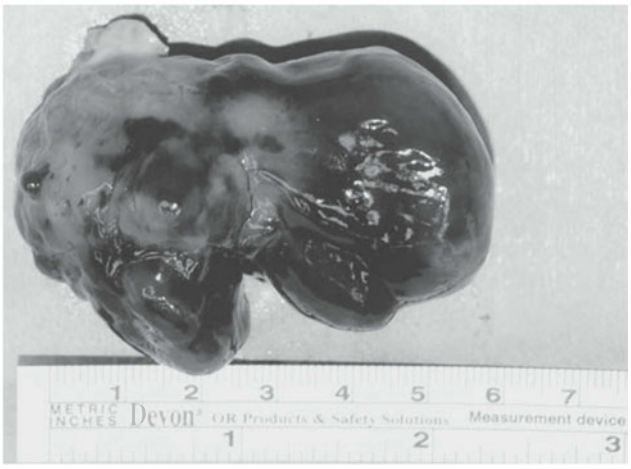
Angiosarcoma
Rhabdomyosarcoma
Liposarcoma
Malignant mesothelioma
Malignant fibrous histiocytoma
Lymphoma
Malignant teratoma
Malignant pheochromocytoma
Thymoma
Osteosarcoma
Synovial sarcoma

Table 68.3 Metastatic Cardiac Neoplasms

Leukemia
Bronchogenic carcinoma
Melanoma
Sarcoma
Breast cancer
Esophageal cancer
Ovarian cancer
Prostate cancer
Renal cell carcinoma (hypernephroma)
Lymphoma

A





B

C

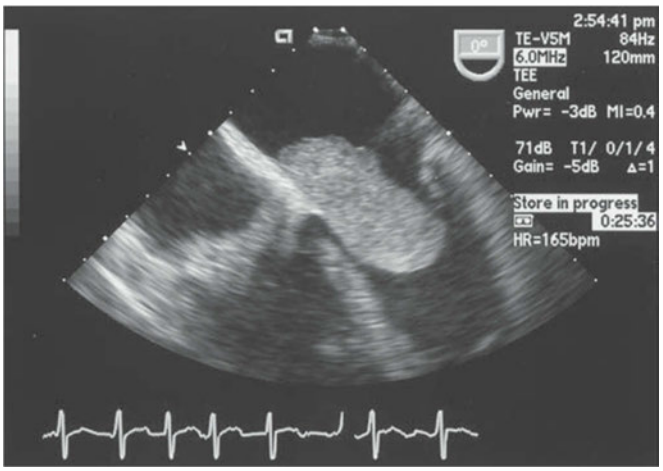


Fig. 68.1. (**A**) Typical operative approach using median sternotomy and cardiopulmonary bypass using a bicaval inflow technique. The exposure of the tumor is via a biatrial approach, and the tumor (*white arrow*) is readily delivered via the left atriotomy. (**B**) Gross pathologic appearance of this myxoma, which consists of large, mottled-tan hemorrhagic tissue, somewhat gelatinous and myxoid, measuring 6 cm in maximal dimension. (**C**) Transesophageal echocardiogram of this tumor in vivo. Note the "ball-valve" obstruction of the mitral valve caused by the tumor. The attachment is to the left atrial wall, along the fossa ovalis.

tumor may not be a myxoma, cardiac-gated CT scans or cardiac-gated MRI may better delineate the extent of involvement of adjacent structures by the tumor. It is important to exclude endocarditis as a cause of cardiac mass.

The indication for operation in patients with a myxoma is the presence of one. Given the risk of embolization (8% to 10%) or obstruction, we typically perform the operation in an urgent manner following the establishment of diagnosis. These patients are usually operated on during the same hospitalization. Initial attempts are made to diurese the patients while obtaining necessary workup, but the surgery is typically not delayed. Anticoagulation with heparin is important to

institute during the time of evaluation because it is felt to decrease the risk of embolization.

Operative Approach

Median sternotomy is the incision of choice. However, in this era of minimally invasive surgery, other options include anterolateral thoracotomy (fourth intercostal space), partial sternotomy (with a "J" or "T" extension), or in female patients, a submammary incision. We utilize a "no-touch" technique to minimize the risk of embolization until the aortic cross-clamp is in place. Cannulation for cardiopulmonary bypass is performed with bicaval inflow, and if the tumor is in the typical location in the left atrium, left ventricular venting through the left superior pulmonary vein is not performed, to avoid dislodging tumor material. The patient's temperature is allowed to drift down to 32 to 34°C, and the heart is arrested with cold blood antegrade cardioplegia. We often use a biatrial approach, having exposed the left atrium through the interatrial groove. The right atrial incision is a standard one, approximately 1 cm parallel to the atrioventricular groove. This exposure usually results in adequate immediate visualization of the tumor (Fig. 68.1A). If the tumor is small, it may be delivered through the left atriotomy with minimal manipulation. When it is larger, gentle application of pressure to the RV outflow tract area may aid in exposing the stalk of the tumor. The tumor is then removed with a 0.5- to 1-cm margin of tissue, taking care not to injure the mitral annulus, the area of conduction tissue, and the tricuspid annulus. The defect is closed primarily if the stalk is ≤1 cm and with a patch (pericardium or Gore-Tex) if the stalk is larger. It should be emphasized that adequate resection with negative margins is the key in resection of this tumor to avoid the risk of recurrence. A copious amount of saline is then used to irrigate the ventricular cavity, and the atria are closed in a routine manner. After deairing the heart, the patient is weaned from cardiopulmonary bypass.

If the tumor is in the right atrium, cannulation may be done in a bicaval manner or with a superior vena cava cannula and a femoral vein cannula. A standard right atriotomy is made, and the tumor is then resected. If the tumor extends to the entrance of the inferior vena cava into the pericardium, deep hypothermic circulatory arrest may be needed to aid in complete resection.

Operative mortality is typically <5%, with the majority of deaths occurring in the older patient population (>70 years of age). Morbidity can be related to embolic phenomena (e.g., stroke). Atrial fibrillation is commonly seen both preoperatively and postoperatively in these patients and may require anticoagulation (given the raw surface of the atria in this scenario). Patients who underwent myxoma resection had survival characteristics that were not significantly different from that of an age- and gender-matched population, whereas those who underwent resection of fibromas, papillary fibroelastomas (PFE), lipomas, and other benign tumors had significantly poorer survival characteristics. Recurrence is estimated to be <5% in patients with sporadic myxomas, but up to 25% in patients with the familial form. Therefore, all patients should continue to get routine echocardiographic follow-up after myxoma resection.

PAPILLARY FIBROELASTOMAS

These benign tumors typically occur on valves or adjacent endocardium and comprise 7% to 10% of cardiac neoplasms. They are usually asymptomatic but can present with embolic phenomena (stroke) or with obstruction of coronary arteries (myocardial ischemia, infarction, or sudden death). PFE usually affects the anatomically contiguous structures of the aortic valve, the left ventricle outflow tract, and the anterior leaflet of the mitral valve. Their gross pathologic appearance consists of a frondlike mass with multiple projections, and they are typically small. Primary surgical excision is recommended for symptomatic PFEs, with left heart side location and with a specific picture suggestive for possible embolization. The indication in asymptomatic patients is not clearly defined. If the tumor is mobile, or larger than 1 cm or the patient is undergoing open heart surgery, the tumor should be resected. Asymptomatic patients with small nonmobile lesions (<1 cm) should be closely followed up with echocardiography. An interval of 4 weeks is advisable after an intracerebral hemorrhage. Surgical resection can be achieved by complete excision without any inclusive margin of normal tissue. A simple valve-sparing shave excision is adequate in majority of patients, whereas valvular repair or replacement is required when leaflet is damaged or restricted. Every effort should be made to repair rather than replace the valve.

Lipomas

Lipomas can occur at any age in any location within the pericardium. These tumors are typically slow growing and present with obstructive or compressive symptoms. Grossly, these tumors are like lipomas seen elsewhere in the body, being well-encapsulated and containing mature adipocytes. Resection is indicated in the presence of symptoms. Tumors found incidentally are resected at the time of other cardiac operations, assuming that the addition of this procedure confers minimal added risk. These tumors are not thought to recur, so a large negative margin is likely not necessary.

When these tumors exist in a nonencapsulated form in the interatrial septum, the condition is called lipomatous hypertrophy of the interatrial septum. This entity is often difficult to separate from neoplasm, but the addition of MRI allows identification of masses with fat, intense signals. Very little is known about the natural history of this entity, and typical indications for resection have not been identified but may include arrhythmias occurring with this pathology. If this lesion is found intraoperatively or preoperatively on transesophageal echocardiography, resection is not felt to be necessary.

Rhabdomyoma

The most frequently seen tumor in children is rhabdomyoma (45% to 60% of cardiac tumors in children vs. <1% of cardiac tumors in adults). It will often present in the first few days of life with symptoms of obstruction. The diagnosis is established on echocardiography. This neoplasm is usually multicentric, occurring with equal frequency in either ventricular chamber. Tuberous sclerosis is sought for because greater than one half of patients with rhabdomyoma have this hereditary disorder. Partial or complete spontaneous regression has been observed, more frequently in younger patients or those with smaller tumors. Surgery was recommended only for symptomatic patients with significant intracardiac obstruction and hemodynamical change, respiratory compromise, or significant risk of systemic embolization. For those patients in which complete resection is not achievable, relief of the obstruction and restoration/preservation of sufficient heart function is the primary goal. The prognosis is poor if they have associated tuberous sclerosis.

Fibromas

Fibromas are cardiac tumors that present often in children. They are usually located in the ventricular chambers and are solitary. Presenting symptoms are secondary to obstruction or arrhythmias. Because these tumors can calcify, they are occasionally

identified on chest X-rays. Complete surgical resection is the goal for cure, but debulking can be associated with long-term survival.

CARDIAC PHEOCHROMOCYTOMAS

Less than 2% of pheochromocytomas occur in the thoracic cavity. Those occurring in the intrapericardial location are often located in the dome of the left atrium. The diagnosis is suspected on clinical manifestations and laboratory confirmation of catecholamine oversecretion. Localization is obtained with the ^{131}I-metaiodobenzylguanidine (MIBG) scan. A CT scan is the next most appropriate test for identifying the extent of involvement of adjacent structures. We also obtain transesophageal echocardiograms in all patients to evaluate the cardiac chambers and valves, as well as to obtain information about the extent of tumor. Finally, cardiac catheterization is performed in selected patients to identify significant coronary disease. Management first consists in controlling the hyperadrenergic syndrome (α-blockade and if necessary intravenous hydration, followed by β-blockade if needed). The tumors are then removed using cardiopulmonary bypass. Complete resection is the goal.

CARDIAC PSEUDONEOPLASMS

There are five reactive and "pseudoneoplastic" tumors of the heart, which mimic true neoplasms either by clinical imaging or microscopically, including inflammatory myofibroblastic tumor (IMT), hamartoma of mature cardiac myocytes, mesothelial/monocytic incidental cardiac excrescences, calcified amorphous tumor (CAT), and lipomatous hypertrophy of the atrial septum. Surgical resection is recommended for IMTs and CATs. Castleman disease is a poorly understood, benign lymphoproliferative disease that may behave in a malignant manner, depending on the structures it invades, including the heart and great vessels. Surgical resection is recommended for unicentric lesion.

PRIMARY MALIGNANT CARDIAC NEOPLASMS

Primary cardiac malignancies are rare and almost always a type of sarcoma. In order of decreasing frequency, the types include angiosarcoma (Fig. 68.2), rhabdomyosarcoma, malignant mesothelioma, and fibrosarcoma. They can arise in any location and are often disseminated widely at the time of diagnosis. The symptoms are dependent on the location and the extent

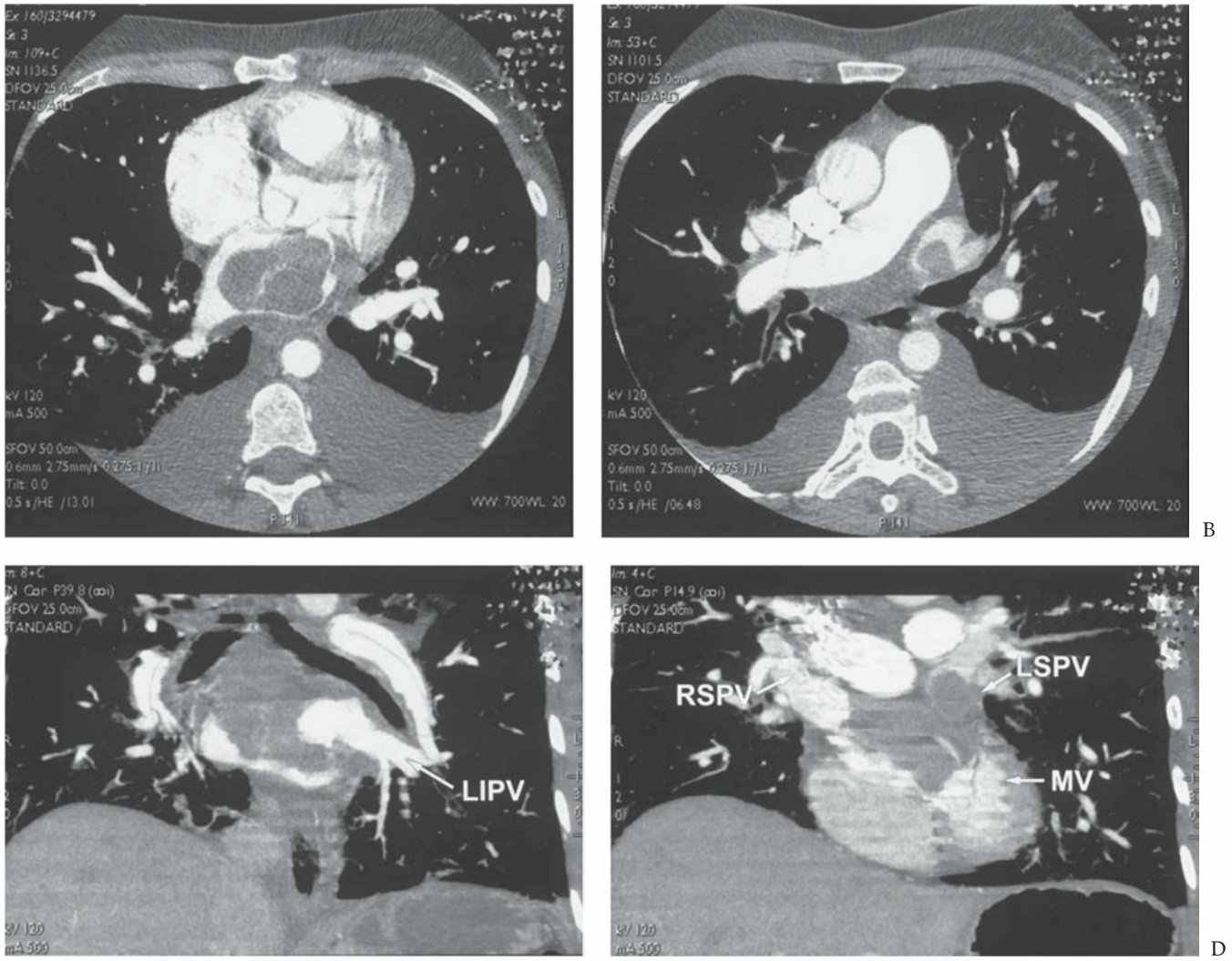


Fig. 68.2. Selected cuts of a cardiac-gated thoracic computed tomography scan demonstrating this large angiosarcoma arising from the left atrial wall. (**A**) Neoplasm filling approximately 80% of the left atrial volume. (**B**) Tumor extension into the left superior pulmonary veins. (**C** and **D**) Sagittal and coronal views, respectively, each showing extension into the pulmonary veins. LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; MV, mitral valve; RSPV, right superior pulmonary vein.

of the tumor, and seem not to be related to the tumor histology. For the left heart sarcomas, most patients present with shortness of breath and dyspnea on exertion due to the obstruction of intracardiac blood flow. The right heart sarcoma tends to be bulky, grow in a more exophytic manner, be more infiltrative, and metastasize earlier, but not usually cause heart failure. The preoperative workup includes a search for extracardiac spread. CT scan and PET/CT scan are excellent modalities to assess the primary tumor and metastasis. Tissue diagnosis is very helpful in planning operation, but sometimes it is very difficult to obtain especially from those left-sided tumor. Operative therapy is reserved for those in whom metastases are absent, and some centers advocate adjuvant chemotherapy. If tumors are not resectable, chemotherapy and radiation therapy may be considered.

Overall, the prognosis of cardiac sarcomas is very poor with median survival of 6 months. The median survival was improved to 12 months with surgery and 25 months with complete surgical resection. The prognosis is dependent on the anatomical location, extent of tumor, and completeness of resection of the tumor rather than the tumor histology. The cardiac group at Methodist Debakey Heart and Vascular Center introduced the surgical technique of cardiac explanation, ex vivo tumor resection, cardiac reconstruction, and subsequent cardiac reimplantation and cardiac autotransplantation for the left heart sarcomas. They reported that this approach improved the completeness of local resection, decreased local recurrence, and extended patient's survival. For the right heart sarcomas, the complete resection can include the superior vena cava, a short segment of inferior vena cava, the entire right atrium, the tricuspid valve, the right coronary artery, and up to about 30% of the RV muscle mass if the RV function is normal.

There have been recent reports of orthotopic heart transplantation as a therapeutic option in patients with cardiac neoplasms (sarcoma, pheochromocytoma, lymphoma, and myxoma). In malignant tumors, reported median survival has been 12 months. However, this remains a controversial option, given the scarcity of donor organs, the need for rapid treatment in patients after presentation, and, finally, the unknown effect of immunosuppression on future malignant and metastatic potential.

METASTATIC CARDIAC NEOPLASMS

Metastatic tumors typically involve the pericardium, followed by epicardial, myocardial, and endocardial involvement in decreasing frequency. The mode of metastasis is dependent on primary tumor. Melanoma, sarcoma, and pulmonary malignancies spread by hematogenous routes. Direct extension is frequently seen in association with pulmonary, breast, thymic, and esophageal cancers.

Symptoms occur infrequently in metastasis, but when they do occur, they usually result from pericardial effusions. The therapy in this setting is a palliative subxiphoid pericardial drainage procedure. This procedure often results in durable symptomatic relief. Some authors advocate a formal "pericardial window" procedure to allow drainage into the left chest. We believe that this approach is not significantly different from simple subxiphoid drainage because the left lung often adheres to the "window," thus eliminating the possibility of continuous drainage of pericardial fluid into the left chest. Moreover, this procedure may require general anesthesia (with double-lumen intubation), whereas a subxiphoid approach can be performed under local anesthesia.

Myocardial metastases from neuroendocrine tumor (NET) occur very rarely with an incidence of 4% of NET. Midgut NET is most often described to be associated with cardiac metastases. Cardiac metastases can be located in the myocardium of the left ventricle, right ventricle, biventricle, or in the ventricular septum too. Malignant arrhythmia or cardiac arrest can develop if the tumor infiltrates the conductive structures of the heart, such as the AV node and His bundle. Echocardiography and MRI are most frequent image modalities. PET/CT is more sensitive for detection of metastases in general and will more likely contribute to earlier detection of cardiac metastases. More than 50% of those patients can have carcinoid syndrome due to systemic effect of circulating vasoactive peptides, mainly serotonin and may develop carcinoid heart disease for the same reason. The cardiac involvement is characterized by progressive fibrosis and smooth muscle proliferation of right-sided valvular and endocardial structures leading to impaired function of the tricuspid and pneumonic valves due to thickening and regurgitation, which can eventually lead to RV failure. Surgical resection of the metastatic NET should be considered if the primary site is controlled and the metastatic tumor is resectable. If the carcinoid valve disease is developed, valve replacement is indicated.

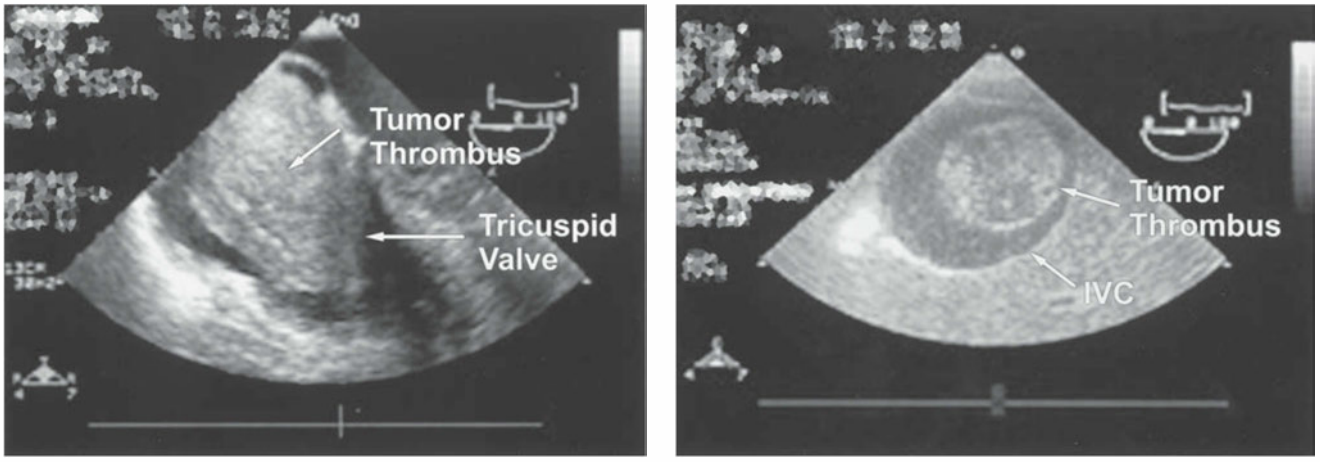
RENAL CELL CARCINOMAS

Renal cell carcinomas (hypernephromas) are an important group of metastatic tumors to the heart. About 5% of all hypernephromas present with direct vena caval extension to the right atrium (Fig. 68.3A). In such cases, symptoms of caval obstruction, including lower body anasarca, gastrointestinal intolerance, and ascites predominate. If complete resection is performed and there is an absence of distant metastasis, 5-year survival can be 75%. Other tumors that extend from the abdomen into the right atrium include hepatic, adrenal, and gynecologic tumors.

Our approach to these tumors is a multidisciplinary one with urologists or surgical oncologists. Preoperative workup includes chest, abdomen, and pelvic CT scanning, transesophageal echocardiography, and left and right heart catheterization in selected patients. Tumors that are confined to Gerota fascia with extension into the vena cava or right atrium without regional lymph node or distant metastasis have the best prognosis, but only distant metastasis is an absolute contraindication to surgery.

The patients are prepared and draped from the neck to the upper thighs. The abdominal team starts with an exploratory laparotomy and mobilization of the kidney. After the kidney is mobilized and the retroperitoneum is hemostatic, a median sternotomy is performed and the patient is placed on cardiopulmonary bypass. The patient is cooled to 18°C, and neuroprotective agents (Solu-Medrol, pentobarbital, and mannitol) are administered. The ascending aorta is cross-clamped, and cardioplegia is delivered in an antegrade manner. Bypass is discontinued, and the right atrium is incised with an extension down to the inferior vena cava. We typically do not use adjunctive retrograde cerebral perfusion because resection of the tumor is accomplished usually in <20 minutes. In a combined manner, the abdominal and thoracic teams resect all tumor and thrombus. Bypass is then initiated, the cross-clamp is removed, and the patient is then rewarmed. The incisions in the vena cava and right atrium are closed. After adequate rewarming, the patient is weaned from bypass.

Another approach without profound hypothermia and circulatory arrest is used by the Stanford group. The immediate



A B

Fig. 68.3. Two cuts of a transesophageal echocardiogram of a renal cell carcinoma with tumor thrombus extension into the right atrium. (**A**) Tumor thrombus extending up the inferior vena cava. (**B**) Tumor thrombus encroaching onto the tricuspid valve.

infrahepatic cava is exposed and controlled. A short opening is made on the IVC and the tumor is extracted out from right atrium and IVC if possible. The infrahepatic cava is then clamped and the IVC is repaired. If the caval tumor does not just slip out, but requires an endarterectomy approach to dissect it from the cava, then an aortic occlusion balloon is inserted through the femoral artery and parked above the celiac. The patient is cooled to 25°C. The aorta is occluded above the celiac artery to avoid blood from hepatic veins running into the operative field, and upper body circulation is maintained with cardiopulmonary bypass. After the tumor is resected from right atrium and IVC, and the cava is repaired, the aortic occlusion balloon is deflated, and the lower body circulation is resumed

SUMMARY

Operative therapy of cardiac neoplasms generally comprises a small portion of all cardiothoracic surgical practice. These pathologic entities often require considerable clinical judgment for both diagnosis and treatment. However, because a significant percentage of these tumors can be treated with good results, they often provide surgeons a chance to dramatically alleviate symptoms in patients debilitated by these neoplasms.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Dr. Yang and colleagues have given us an extensive approach to cardiac tumors. I am in full agreement with their analysis. I would like to focus on atrial myxomas. I am in total agreement with the authors on avoiding embolization. We use a no-touch technique and use myocardial protection prior to removing the atrial myxoma. As the authors have noted these are gelatinous tumors and must be handled with care. Typically, we do not grasp the tumor but rather the atrial wall near where the tumor is attached. By doing this, one can fully resect the tumor without any fear of embolization. We have also found it helpful to invert the left atrial appendage externally to push the tumor out to avoid embolization. These procedures are critically important and one should be able to minimize the risk with the no-touch technique.

ILK

69 Acute Pulmonary Embolus

Salil G. Shah, Joshua N. Baker, and Thoralf M. Sundt

It is ironic that the heart–lung machine is rarely used today in the treatment of the very condition that inspired its creation: acute pulmonary embolism (PE). Dr. Gibbon was inspired to his life's work as a research resident at the Massachusetts General Hospital in 1931 as he sat at the bedside of a young woman dying of a massive PE. Have we wrongfully relinquished this part of our surgical heritage?

Acute PE is a remarkably common phenomenon, particularly among hospitalized individuals. The majority of acute pulmonary emboli result in little morbidity, and most are likely entirely unrecognized. Massive PE, however, may lead to hemodynamic collapse and death via a combination of mechanical obstruction of central right ventricular outflow and a peripheral vasospastic response in the remaining unobstructed vasculature. Although such massive emboli represent a relatively small proportion of acute events, they account for significant mortality and morbidity each year by virtue of the frequency with which PE occurs.

Why then is surgical embolectomy an uncommon procedure in most practices? The 2011 American Heart Association Scientific Statement on the management of massive and submassive PE discusses catheter-based embolectomy and thrombolysis more extensively than surgical embolectomy, with the surgical approach being reserved only for patients with contraindication to thrombolysis or in whom thrombolysis has failed. It should be noted that while the guidelines state that "… surgical embolectomy is reasonable for patients… who remain unstable after receiving fibrinolysis," massive bleeding can be anticipated to complicate any invasive procedure until normal coagulation is restored. The guidelines of the British Thoracic Society published in 2003 are even more dismissive of the role of surgical intervention, with only thrombolysis and "invasive approaches (thrombus fragmentation and IVC filter insertion)" entertained as options for the management of massive PE. Surgical embolectomy, it was noted, was not an option as it was available in few centers.

Why has there been reluctance to offer surgical embolectomy? Apart from logistical issues regarding on-site cardiac surgical support, published mortality rates for acute pulmonary embolectomy traditionally have ranged from 20% to 60%, making it difficult to argue that the surgical results were any better than the natural history. A closer look, however, suggests that the hesitancy to proceed to the operating room may itself be part of the problem. Preoperative cardiac arrest roughly doubles the mortality rate, and the time interval between diagnosis and operation has similarly been shown to have a profound effect on surgical risk. This becomes, of course, a self-perpetuating problem with many physicians, on the basis of the reported high risk of the procedure, reluctant to refer patients for surgery unless profound hemodynamic instability is present. It is only fair to add, however, that technically inadequate surgical embolectomy is likely a contributor to excessive mortality as well.

A critical assessment of the data suggests that there is, in fact, a small but definite place for surgical embolectomy in the management of massive PE. Furthermore, with the remarkable advances in the biocompatibility of extracorporeal membrane oxygenation (ECMO) technology in recent years, the cardiac surgeon, it may be argued, should once again be center stage as part of a multidisciplinary team evaluating all treatment options up front when a patient suffers massive PE. As surgeons we must, of course, take the responsibility for true "around-the-clock" availability and for performing the lowest risk procedure with the best possible results. We aim here to review our current approach to this condition in our hospital including a small number of technical points that are important for both minimizing the intraoperative insult to the struggling right ventricle and optimizing the relief of outflow obstruction. With such an approach, we believe that others can achieve the kind of results reported by Leacche and colleagues in 2006 with a 6% 30-day mortality rate among 47 patients undergoing embolectomy and 1- and 3-year survival rates of 86% and 83%, respectively, or those of Kadner and colleagues with an 8% overall mortality rate including a 28% mortality among patients experiencing a preoperative cardiac arrest.

DIAGNOSIS

In most instances, the diagnosis of acute PE will have already been made by the time the surgeon is called. Still, it is important to be familiar with current diagnostic modalities, as frequently the patient is being concurrently evaluated for aortic dissection or acute coronary syndrome. Of course, a clinical history consistent with deep vein thrombosis, such as the postoperative orthopedic patient in a long-leg cast, a history of malignancy or pelvic surgical patient after prostatectomy or hysterectomy, or trans-oceaning air travel can be quite helpful. Physical examination is seldom enlightening; however, biomarkers may be revealing; serum D-dimer enzyme-linked immunosorbent assay is almost universally elevated in the presence of acute pulmonary embolus and is frequently used in emergency rooms as a screening test, while elevation in cardiac biomarkers including troponin I and T, and natriuretic peptides have been shown to have negative prognostic impact with the concurrent diagnosis of PE. Nuclear scintigraphy has been almost entirely replaced by helical computerized tomographic (CT) scanning (Fig. 69.1). An adequate scan requires an appropriately timed bolus of contrast material, and a scan performed for other reasons may not image the pulmonary arteries adequately to rule out the diagnosis. Conversely, while CT scanning has the added advantage of providing information about other intrathoracic pathology, a contrast bolus timed appropriately for PE may be inadequate to rule out aortic dissection. The place of nuclear scintigraphy lies principally with

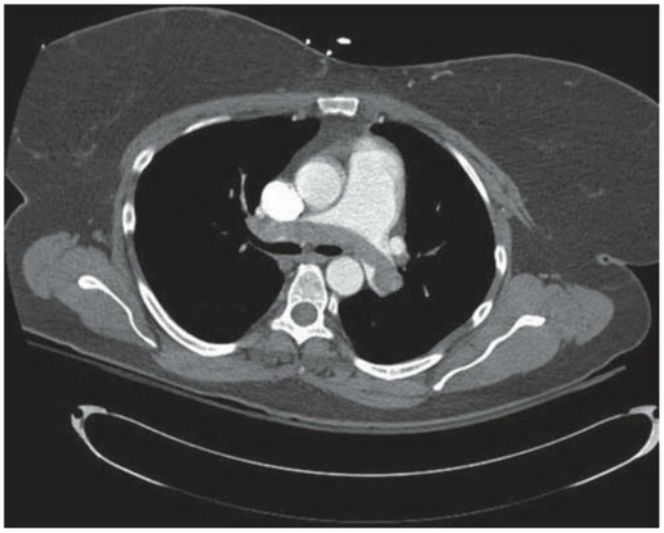


Fig. 69.1. Computed tomography demonstrating a "saddle" pulmonary embolus.

the evaluation of pregnant women and patients allergic to contrast agents or with a marginal renal function.

An increasingly important recent diagnostic tool from a surgical standpoint is echocardiography (Fig. 69.2). Right ventricular function can be rapidly assessed in the emergency room or ICU by transthoracic echo, whereas embolic material may be identified in the pulmonary vasculature or, far more importantly, in the atria as "embolus in transit," including those traversing a patent foramen ovale. Certainly, transesophageal echo should be performed in the operating room if at all possible.

INDICATIONS FOR SURGERY

When called to evaluate the patient at the bedside for surgical intervention, it is important to remember that, in addition to simple mechanical obstruction, the pathophysiology of pulmonary hypertension in acute PE entails the release of serotonin from platelets, histamine from tissues, and circulating thrombin. Hypoxia due to ventilation/perfusion mismatch and increased dead space will also worsen pulmonary vasoconstriction. Accordingly, oxygenation should be optimized before determining that a patient must go to the operating room.

It is widely accepted that systemic hypotension despite inotropic support is an indication for aggressive intervention—surgical, catheter-based, or lytic—as is persistent, refractory hypoxemia. The difficulty, however, is that operative risk is markedly elevated once the patient is in profound shock or suffers cardiac arrest. Accordingly, the challenge is to stratify patients early with regard to risk of poor outcome with anticoagulant therapy alone. In 2005, Sukhija and colleagues showed a reduction in in-hospital mortality among patients with right ventricular dysfunction who received timely pulmonary embolectomy, and multiple other studies have demonstrated the association of right ventricular dysfunction to both short and long-term mortality. Accordingly, the 2011 AHA scientific statement recommends swift intervention either with fibrinolysis or invasive intervention for those with right ventricular dysfunction.

Perhaps because of their wide availability and familiarity with their use in the context of treating acute coronary syndromes and more recently stroke, thrombolytics have taken center stage in the aggressive treatment of the unstable patient with massive PE. It is important to remember, however, that use of thrombolytics is not without risk. In the International Cooperative PE Registry, the incidence of cerebral events with thrombolytics was 3%. Furthermore, Thabut's meta-analysis of thrombolytic studies reported in 2002 found no improvement in mortality rate when they were used in unselected patients as compared with heparin, but an almost twofold increased risk of hemorrhage (risk ratio 1.76, confidence interval 1.04 to 2.98). To be fair, among the four registries documenting the outcomes of patients with PE (Mappet, ICOPER, PIETE, and EMPEROR), there is suggestion of a decrease in mortality among patients with hemodynamic compromise who are treated with thrombolytics although the numbers are small. There are also a few randomized control trials, mostly in intermediate-risk patients, which show a reduction in pulmonary artery pressures and reduction in thrombus burden albeit with no survival benefit except in high-risk patients.

Catheter embolectomy is another option, and one that is undergoing rapid evolution. Endovascular techniques include clot fragmentation, clot aspiration, and rheolytic therapy. The clinical efficacy of percutaneous therapy alone (Rotares, Aspirex, Angiojet) has been reported to range from 81% to 92% and when combined with thrombolytic therapy to range from 91% to 100% depending on the modality. The mortality rate associated with these interventions, however, has been 16%

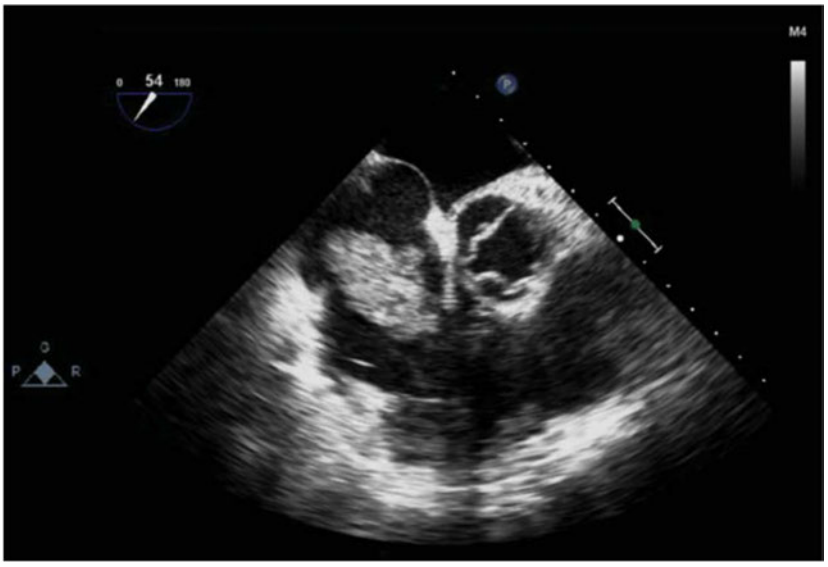


Fig. 69.2. Transesophageal echocardiography demonstrating embolus in transit.

to 30%. They are also technically challenging and require a team very familiar with the use of these proprietary devices. While they have gained more widespread acceptance, they are still not universally available. The two most current options are the AngioVac (Vortex Medical Albany, NY), a modified bypass circuit with a movable suction cannula and a series of filters within the circuit that works well for proximal thrombus or embolism, and the EKOS catheter (EKOS Corp, Bothell, WA) that provides for directed thrombolysis and ultrasonic acceleration of clot lysis. Both of these devices show promise, but the data are sparse regarding clinical efficacy currently.

It has been our view that surgical intervention performed before hemodynamic collapse has an operative risk equivalent to or lower than that of thrombolytic therapy in most cases. Surgical pulmonary embolectomy is performed safely on a warm, perfused, beating heart with a minimal pump run in most cases. Surgery is clearly the option of choice when there is clot in transit present in the right atrium or, even more compelling, trapped in a patent foramen ovale or atrial septal defect. Operative intervention may be contraindicated, however, in the patient in full arrest who cannot be pharmacologically resuscitated or in whom thrombolytics have been recently administered. In these settings, we have found the institution of percutaneous ECMO remarkably effective with gratifying cases of successful weaning from support and discharge from the hospital with intact cardiovascular and neurologic function. While this is far from the standard of care, results have been so encouraging as to stimulate us to question whether our own surgical paradigm should be inverted with placement of the patient with massive PE and right ventricular dysfunction on ECMO prior to any sort of surgical or catheter-based intervention rather than viewing ECMO as a support for those who fail to come off bypass after surgical embolectomy. Given the aforementioned data concerning increased morbidity and mortality in patients with right ventricular dysfunction, there is argument for rapid decompression of the heart to allow recovery of myocardial function.

For patients not requiring surgical intervention, heparin is the cornerstone of therapy. Individuals with heparin-induced thrombocytopenia may require the use of a direct thrombin inhibitor like argatroban or hirudin analogs. In either case, oral warfarin will be indicated for 3 to 6 months.

ANATOMIC CONSIDERATIONS

The anatomy of the pulmonary vasculature should be familiar to all cardiothoracic surgeons. What may be less well appreciated, however, is the remarkable access to lobar vessels via median sternotomy. All lobar and segmental vessels can be accessed via separate incisions in the right and left pulmonary arteries from within the pericardial space as one would during pulmonary thromboendarterectomy. This is an important point as a true saddle embolus lodged at the pulmonary bifurcation without distal fragments is relatively uncommon. More often, a large amount of embolic material is impacted in the orifices of the lobar vessels and will be inaccessible, particularly on the right side, via an incision limited to the main pulmonary artery.

OPERATIVE PROCEDURE

Pulmonary embolectomy is accomplished on normothermic cardiopulmonary bypass without cardioplegic arrest to minimize further insult to the right ventricle. Bicaval cannulation with tapes permits complete removal of thrombus bilaterally under direct vision. There are some older reports of pulmonary embolectomy under simple inflow occlusion, and this remains an option if cardiopulmonary bypass is unavailable; however, a much more complete job can be performed with the aid of the pump without risking hypoperfusion of end organs during the procedure.

Once the decision has been made to proceed with surgical intervention, the patient should be expeditiously transported to the operating room. Routine anesthetic monitoring including an arterial blood pressure line and an internal jugular introducer should be placed, but insertion of a pulmonary artery catheter at the beginning of the case should be avoided. Intraoperative transesophageal echocardiography is a routine in our institution and greatly facilitates intraoperative decision-making particularly with regard to exploration of the right atrium and evaluation for clot in transit.

The groin vessels should be prepped into the field in case postoperative ECMO is necessary. A full sternotomy permits complete inspection of the right atrium. Of note, on more than one occasion, we have observed poor venous return from the inferior vena cava line only to find clot in transit impacted in the cannula orifice. For this reason, the superior vena cava cannula is placed first so that partial bypass may be initiated if clot is dislodged from the inferior vena cava. Jakob has even advocated routine massage of the lower extremities and abdomen with open aspiration of the inferior vena cava using cardiotomy suckers to extract additional material in transit.

Normothermic cardiopulmonary bypass is instituted, and tapes are passed around the superior vena cava and inferior vena cava. If a patent foramen ovale or paradoxic embolus in transit has been identified by transesophageal echo, a brief episode of cardioplegic arrest will be necessary and should be performed first to permit maximal reperfusion. In such instances, opening the right and left atria via a Dubost incision through both atrial-free walls and the septum will assure control of the distal clot (Fig. 69.3). After the patent foramen ovale and left atrium have been closed, the cross-clamp should be removed to permit maximum reperfusion.

If no patent foramen ovale is identified, it is preferable to perform the entire procedure without cardioplegic arrest. Clot in transit in the right atrium can easily be removed with the heart beating (Figs. 69.4 to 69.6). If right atrial exploration is not required, a bullet-tip sucker can be dropped into the right atrium via a stab wound with a purse-string suture to scavenge coronary sinus return and reduce the

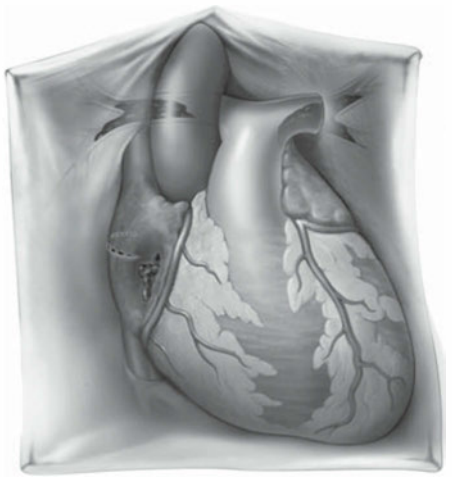


Fig. 69.3. Paradoxical embolus in transit can be identified by transesophageal echo. When it is identified, the left atrium must be opened before making any attempt to remove the thrombus. This may be accomplished via simultaneous standard right and left atriotomies or, as shown here, a Dubost incision extending vertically across the right atrial free wall and Waterston's groove into the left atrium. This permits the surgeon to open the septum into the patent foramen while controlling the thrombus. This should be performed with a brief episode of cardioplegic arrest at the beginning of the procedure, providing the remainder of the bypass run for the struggling right heart to reperfuse and recover.

amount of blood passing through the right ventricle and into the pulmonary artery.

Clot in the pulmonary arteries should be removed via the same incisions used for the relief of chronic pulmonary thromboembolic disease during pulmonary thromboendarterectomy. Embolic material lodged in the left lung should be removed under direct vision via a pulmonary arteriotomy beginning in the main pulmonary artery and extending out onto the left pulmonary artery to the level of the pericardial reflection or just beyond (Fig. 69.7). Via this arteriotomy, the clot can be removed with stone forceps. Note that care must be taken not to grasp the bifurcation itself, as the pulmonary artery can be easily torn. We also pass soft endotracheal suction catheters down the artery orifices while massaging the lungs to remove any small clot that has migrated distally (Fig. 69.8). This is frequently productive. Balloon-tipped embolectomy catheters *should not* be passed as the pulmonary vessels are extraordinarily fragile and rapidly taper in diameter, making rupture of the vessels a very real possibility.

The incision in the right pulmonary artery is made between the aorta and superior vena cava again as one would when doing a pulmonary thromboendarterectomy for chronic disease. With a blunt-bladed retractor separating the aorta and superior vena cava, the transverse portion of the right pulmonary artery can be readily accessed (Fig. 69.9). A linear incision first in the posterior pericardium overlying the vessel and then in the vessel itself provides ready access to all of the lobar and

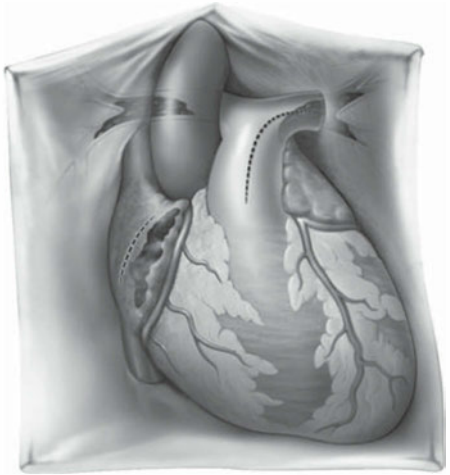


Fig. 69.4. If embolus in transit is identified in the right atrium, this can be extracted via a standard right atrial incision without cardioplegic arrest. This approach provides optimal protection of the right ventricle during the procedure.

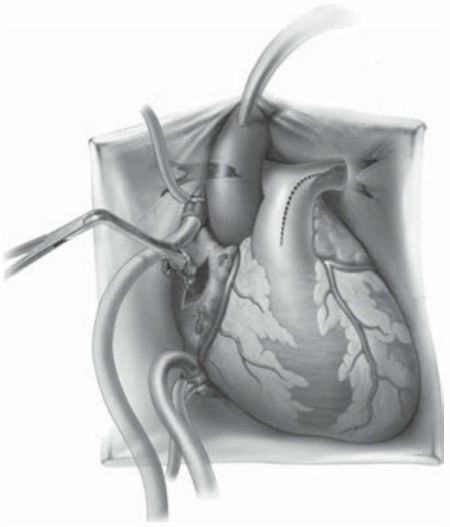


Fig. 69.5. Common duct stone forceps are ideally suited for extraction of the thrombus.

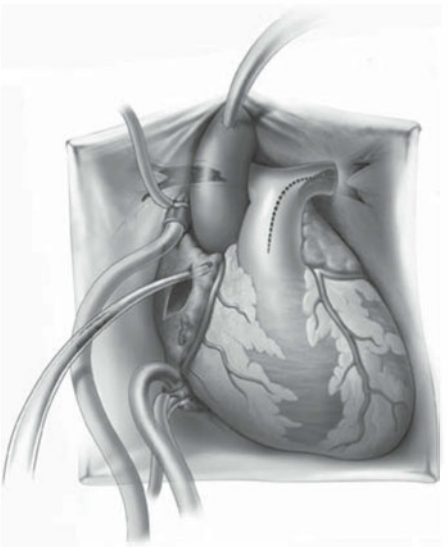


Fig. 69.6. Any fragments of clot are easily extracted using a wide-mouthed sucker.

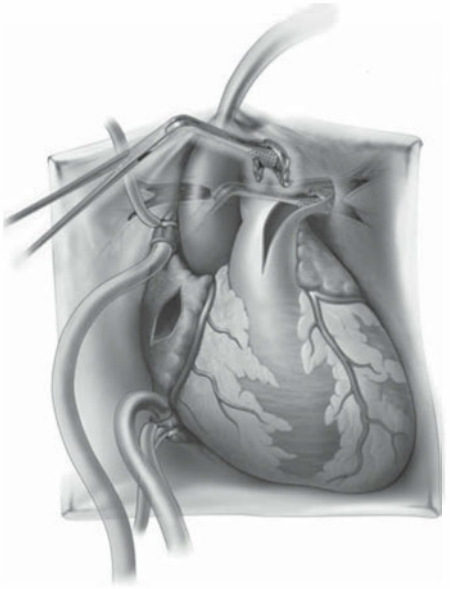


Fig. 69.7. Thrombus in the left pulmonary artery is accessed via an incision beginning in the main pulmonary artery. Adequate access permitting direct visualization of the segmental vessels requires extension of the incision onto the left pulmonary artery itself. This may require division of the pericardial reflection over the ventral surface of the pulmonary artery. Some caution in the application of electrocautery is appropriate superiorly to prevent injury to the recurrent nerve. Again, common duct stone forceps are ideal for extraction of thrombus under direct vision.

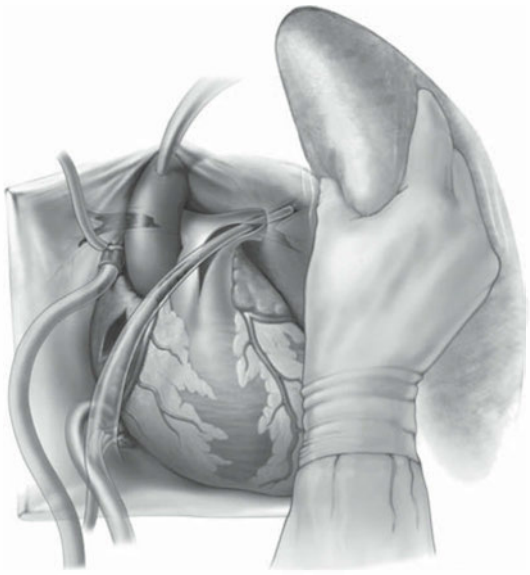


Fig. 69.8. More distal fragments of thrombus can be cleared with a wide-mouthed suction catheter or flexible endotracheal suction catheter. Massage of the lungs can be performed to milk thrombus proximally for removal. Balloon-tipped embolectomy catheters should not be introduced. The pulmonary vasculature is fragile, and the risk of rupture with such devices is real.

segmental vessels. If one has not previously performed an arteriotomy such as this, one will be surprised how central the takeoff to the right upper lobe is. The right upper lobe branch, right middle lobe branch, and the segmental vessels to the right lower lobe can be directly inspected. Again, clot is removed under direct vision using stone forceps (Fig. 69.10). This is followed with a flexible suction catheter and massage of the lungs (Fig. 69.11). These arteriotomies are easily closed with running 4-0 Prolene (Fig. 69.12). Pericardial patch augmentation is seldom necessary. The patient is then weaned from cardiopulmonary bypass, with inotropic support for the right ventricle routine. A pulmonary artery thermodilution catheter can now be passed.

The final step in the procedure is insertion of an inferior vena cava filter via a purse-string suture on the right atrial appendage (Fig. 69.13). There is general agreement that an inferior vena cava filter is indicated in patients with contraindications to anticoagulation, if PE has recurred while on anticoagulant therapy, or if PE is sufficiently severe that recurrent PE would be life-threatening. Although such filters do not prevent thrombosis and some argue that they are themselves a nidus of thrombosis, it is our practice to insert a filter in all patients with chronic disease as well as anyone having an embolus severe enough to require emergent surgery.

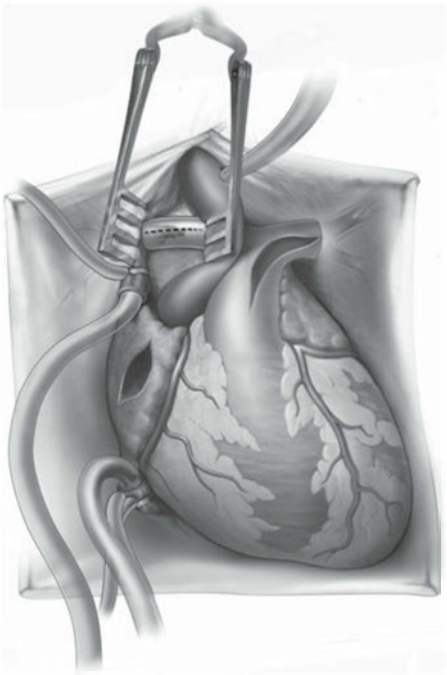


Fig. 69.9. The right pulmonary artery can be accessed directly between the aorta and superior vena cava.

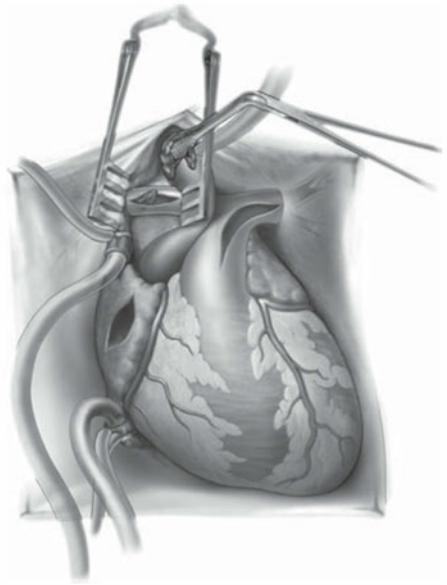


Fig. 69.10. This permits removal of thrombus again under direct vision.

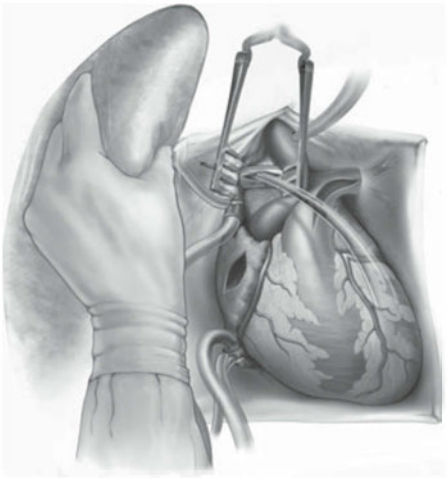


Fig. 69.11. The use of stone forceps is followed by the use of suction devices.

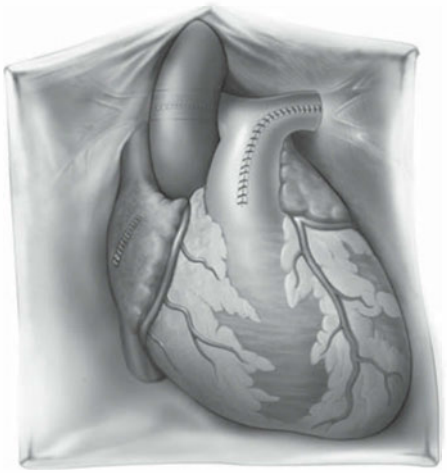


Fig. 69.12. Incisions are closed primarily.

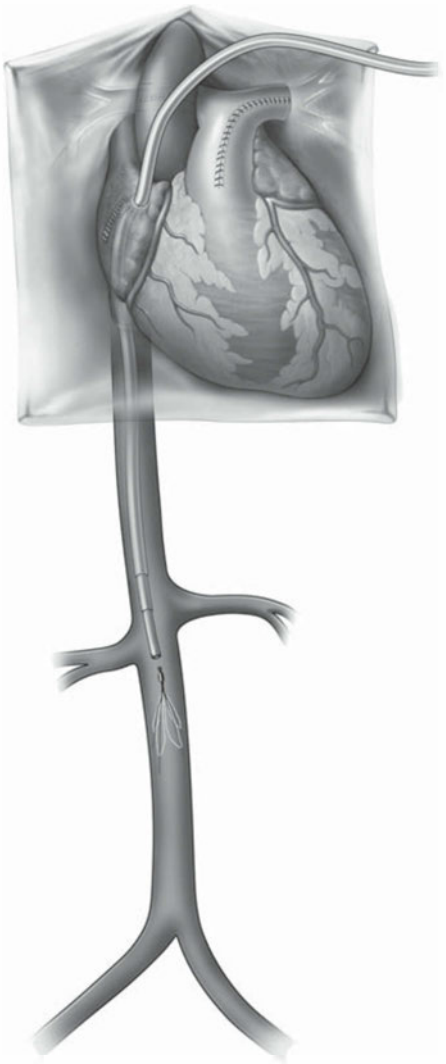


Fig. 69.13. An inferior vena cava filter may be placed via a purse-string suture in the right atrial appendage.

POSTOPERATIVE CONSIDERATIONS

Postoperatively, these patients frequently require significant inotropic support while the right ventricle recovers from surgery or, in some instances, venoarterial ECMO support to unload the right ventricle allowing for faster recovery while supporting systemic circulation. With biocompatible circuits, our threshold for such support has diminished. This mode may also allow for more rapid lung recovery, as well. These patients should be aggressively anticoagulated postoperatively with heparin followed by warfarin for 6 months.

A search should certainly be made for factors predisposing these patients to PE, and efforts should be made toward secondary prevention. Surgeons should take part in encouraging patients in secondary

prevention of modifiable factors such as obesity, tobacco abuse, use of oral contraceptives, or postmenopausal hormone replacement. If no causes are identified, consideration should be given to a search for occult malignancy. Consultation with a hematologist and systematic search for a prothrombotic state is routine. If no treatable cause is identifiable or patients have evidence of a hypercoagulable state, warfarin therapy is indicated for life.

SUGGESTED READINGS

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EDITOR'S COMMENTS

Sundt and colleagues have discussed surgical treatment of acute pulmonary embolus. As the authors have mentioned that cardiac surgery was born around the concept, but for whatever reason we tend to only be called when the patient is terminal. Certainly, acute pulmonary embolectomy in the patients who have already arrested is futile due to anoxic cerebral injury. The question, therefore, is the sweet spot where the surgical approach is preferred. Certainly in those patients who have not arrested the mortality for this procedure is surprisingly low.

My belief is that this procedure is best used when there is a need for mechanical support while performing an embolectomy. There are patients who are hemodynamically challenged and echo shows right ventricular strain. These patients cannot wait for the outcome of thrombolytic therapy or anticoagulation. In these situations, urgent/semielective pulmonary embolectomy will work beautifully. Unfortunately, we are called when things are too late. I think institutions that do best with this procedure are those where all the clinicians are aware of the multiple modalities of treating pulmonary embolus.

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70 Chronic Pulmonary Thromboembolism and Pulmonary Thromboendarterectomy

Michael M. Madani and Stuart W. Jamieson

Pulmonary thromboendarterectomy (PTE) for the treatment of chronic thromboembolic pulmonary hypertension (CTEPH) is an uncommon surgical procedure; however, it is the only curative option that provides immediate and permanent cure for this devastating disease. The condition is remarkably underdiagnosed, and as a result the procedure is uncommonly applied. Patients with chronic pulmonary hypertension secondary to thromboembolic disease may present with a variety of debilitating cardiopulmonary symptoms. However, once it is diagnosed, there is no curative role for medical management, and surgical removal of the thromboembolic material is the only therapeutic option.

The exact incidence of pulmonary embolism (PE) is unknown, but there are some valid estimates. Acute PE is the third most common cause of death (after heart disease and cancer). Approximately, 75% of autopsy-proven PEs are not detected clinically. It is estimated that PE results in approximately 700,000 symptomatic episodes in the United States yearly. The disease is particularly common in hospitalized elderly patients. Of the hospitalized patients who develop PE, 12% to 21% will die in the hospital, and another 24% to 39% die within 12 months. Thus, approximately 36% to 60% of the patients who survive the initial episode live beyond 12 months, and they may present later in life with a wide variety of symptoms.

The mainstay of treatment of patients with deep vein thrombosis (DVT) and acute PE is medical management. In general, cardiac surgeons rarely intervene in hospitalized patients who suffer a massive embolus that causes life-threatening acute right heart failure and severe hemodynamic compromise. In contrast, the only treatment for patients with chronic pulmonary thromboembolic disease is the surgical removal of the disease by means of PTE. Medical management in these patients is only palliative, and surgery by means of transplantation is an inappropriate use of resources with less than satisfactory results.

The prognosis for patients with pulmonary hypertension is poor, and it is worse for those who do not have intracardiac shunts, since the shunt will reduce right-sided pressures. Thus, patients with primary pulmonary hypertension and those with pulmonary hypertension secondary to pulmonary emboli fall into a higher risk category than those with Eisenmenger syndrome and have a higher mortality rate. In fact, once the mean pulmonary pressure in patients with thromboembolic disease reaches $\geq$50 mmHg, the 3-year mortality approaches 90%.

Surgical preferences are dependent on both the principal disease process and the reversibility of the pulmonary hypertension. With the exception of thromboembolic pulmonary hypertension, lung transplantation is the only effective therapy for patients with end-stage pulmonary hypertension. Although it has been performed less frequently in the last few years, pulmonary transplantation is still used in some centers as the treatment of choice for patients with thromboembolic disease. However, a true assessment of the effectiveness of any therapy should take into account the total mortality after the patient has been accepted and put on the waiting list. Thus, the mortality for transplantation (and especially double-lung or heart–lung transplantation) as a therapeutic strategy is much higher than is generally appreciated because of the significant loss of patients awaiting donors. Bearing in mind, in addition, the long-term use of immunosuppressants with their associated side effects, the higher operative morbidity and mortality, the inferior prognosis even after successful transplantation, and the long waiting period, one can see that transplantation is clearly an inferior alternative to PTE and should be considered an inappropriate and outdated form of therapy.

INCIDENCE

Determining an accurate incidence of CTEPH is almost impracticable. Most patients with this condition do not have a clear history of DVT or PE. Furthermore, the majority (about 75%) of autopsy-proven PEs are not clinically diagnosed. This makes the exact incidence of this disease even more difficult to determine than that of acute PE. A conservative estimate only considers patients who do have an acute PE and survive the episode. There are approximately 600,000 such patients every year in the United States. The incidence of chronic thrombotic occlusion in the population depends on what proportion of patients fails to resolve acute embolic material. Recent studies have shown that of these patients, up to 3.1% will have symptomatic CTEPH at 1 year and 3.8% will have it at 2 years. If these figures are correct, and if one counts only patients with symptomatic acute pulmonary emboli, approximately 18,000 to 22,000 individuals would progress to CTEPH in the United States each year. However, because many (if not most) patients diagnosed with chronic thromboembolic disease have no antecedent history of acute embolism, the true incidence of this disorder must be much higher.

Regardless of the exact incidence or the circumstances, it is clear that both acute embolism and its chronic relation, fixed chronic thromboembolic occlusive disease, are much more common than generally appreciated and are seriously underdiagnosed. Calculations extrapolated from mortality rates and the random incidence of major thrombotic occlusion found at autopsy support a postulate that more than 100,000 people in the United States currently have pulmonary hypertension that could be relieved by operation.

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PATHOLOGY AND PATHOPHYSIOLOGY

Most cases of chronic pulmonary emboli arise from previous acute embolic episodes, even though the majority of individuals with chronic pulmonary thromboembolic disease are unaware of a past thromboembolic event and give no history of deep venous thrombosis. Why some patients have unresolved emboli is not certain, but a variety of factors must play a role, alone or in combination.

For instance, the volume of acute embolic material may simply overwhelm the lytic mechanisms, and the total occlusion of a major arterial branch may prevent lytic material from reaching, and therefore dissolving, the embolus completely. Furthermore, repetitive emboli may not be able to be resolved. Other causes may relate to the fact that the emboli may be made of substances that cannot be resolved by normal mechanisms (already well-organized fibrous thrombus, fat, or tumor), and the lytic mechanisms themselves may be abnormal. In some groups, patients may actually have a propensity for thrombus or a hypercoagulable state.

In general, after the clot becomes wedged in the pulmonary artery, one of two processes occurs: (1) the clot may proceed to canalization, producing multiple small endothelialized channels separated by fibrous septa (i.e., bands and webs) or (2) it may continue to form a solid mass of dense, fibrous connective tissue, without canalization, totally obstructing the arterial lumen.

In addition, chronic indwelling central venous catheters and pacemaker leads are sometimes associated with pulmonary emboli. Less frequent causes include tumor emboli; tumor fragments from stomach, breast, and kidney malignancies have been demonstrated to cause chronic pulmonary arterial occlusion. Right atrial myxomas may also fragment and embolize.

Whatever the predisposing factors to residual thrombus within the vessels, the genesis of the resultant pulmonary vascular hypertension is more complex than generally appreciated. With the passage of time, the increased pressure and flow as a result of redirected pulmonary blood flow in the previously normal pulmonary vascular bed can create a vasculopathy in the small precapillary blood vessels similar to the Eisenmenger syndrome.

Factors other than the simple hemodynamic consequences of redirected blood flow are also probably involved in this process. For example, after a pneumonectomy, 100% of the right ventricular output flows to one lung, yet little increase in pulmonary pressure occurs, even with follow-up to more than a decade. In patients with thromboembolic disease, however, we frequently detect pulmonary hypertension even when <50% of the vascular bed is occluded by thrombus, and not uncommonly as early as a few months to 1 year after the initial episode. It thus appears that an array of sympathetic neural connections and hormonal changes may be responsible for setting off pulmonary hypertension in the initially unaffected pulmonary vascular bed. This process can occur with the initial occlusion in either the same or the contralateral lung.

Regardless of the cause, the evolution of pulmonary hypertension as a result of changes in the previously unobstructed bed is serious because this process may lead to an inoperable situation. Accordingly, with our accumulating experience in patients with thrombotic pulmonary hypertension and superior surgical outcomes, we have increasingly been inclined toward early operation so as to avoid these changes.



CLINICAL PRESENTATION

There are no specific signs or symptoms associated with pulmonary hypertension as a result of chronic pulmonary thromboembolism, which explains the degree of underdiagnosis with this condition. The most common symptom associated with thromboembolic pulmonary hypertension, as with all other causes of pulmonary hypertension, is exertional dyspnea. Generally, this dyspnea is out of proportion to any abnormalities found on clinical examination. Like complaints of easy fatigability, dyspnea that initially occurs only with exertion is often attributed to anxiety or being "out of shape." In patients with more advanced disease and higher pulmonary artery pressures, syncope or presyncope (lightheadedness during exertion) is another common symptom.

Nonspecific chest pains occur in approximately 50% of patients with more severe pulmonary hypertension. Hemoptysis can occur in all forms of pulmonary hypertension and probably results from abnormally dilated vessels distended by increased intravascular pressures. Peripheral edema, early satiety, and epigastric or right upper quadrant fullness or discomfort may develop as the right heart fails (cor pulmonale). Some patients with chronic pulmonary thromboembolic disease present after an additional small acute pulmonary embolus that may produce acute symptoms of right heart failure. A careful history will bring out symptoms of dyspnea on minimal exertion, easy fatigability, diminishing activities, and episodes of angina-like pain or lightheadedness. Further examination reveals the signs of pulmonary hypertension.

The physical signs of pulmonary hypertension are the same no matter what the underlying pathophysiology. Initially, the jugular venous pulse is characterized by a large A wave. As the right heart fails, the V wave becomes predominant. The right ventricle is usually palpable near the lower left sternal border, and pulmonary valve closure may be audible in the second intercostal space. Occasional patients with advanced disease are hypoxic and slightly cyanotic. Clubbing is an uncommon finding.

As the right heart fails, a right atrial gallop is usually present, and tricuspid insufficiency develops. Because of the large pressure gradient across the tricuspid valve in pulmonary hypertension, the murmur is high pitched and may not exhibit respiratory variation. These findings are quite different from those usually observed in tricuspid valvular disease. A murmur of pulmonic regurgitation may also be detected.



DIAGNOSIS

To ensure the diagnosis in patients with chronic pulmonary thromboembolism, a standardized evaluation is recommended for all patients who present with unexplained pulmonary hypertension. This workup includes a chest radiograph, although a large number of patients present with a relatively normal chest radiograph, even in the setting of high degrees of pulmonary hypertension. When abnormal, the chest X-ray may show either apparent vessel cutoffs of the lobar or segmental pulmonary arteries, or regions of oligemia suggesting vascular occlusion. Central pulmonary arteries are also typically enlarged, and the right ventricle may be enlarged without enlargement of the left atrium or ventricle (Fig. 70.1).

The ventilation–perfusion lung scan is the essential test for establishing the diagnosis of unresolved pulmonary thromboembolism. An entirely normal lung scan excludes the diagnosis of both acute and chronic, unresolved thromboembolism. The usual lung scan pattern in most patients with pulmonary hypertension is either relatively normal or shows a diffuse nonuniform perfusion. When subsegmental or larger perfusion defects are noted on the scan, even when matched with ventilatory defects, pulmonary angiography is appropriate to confirm or rule out thromboembolic disease.

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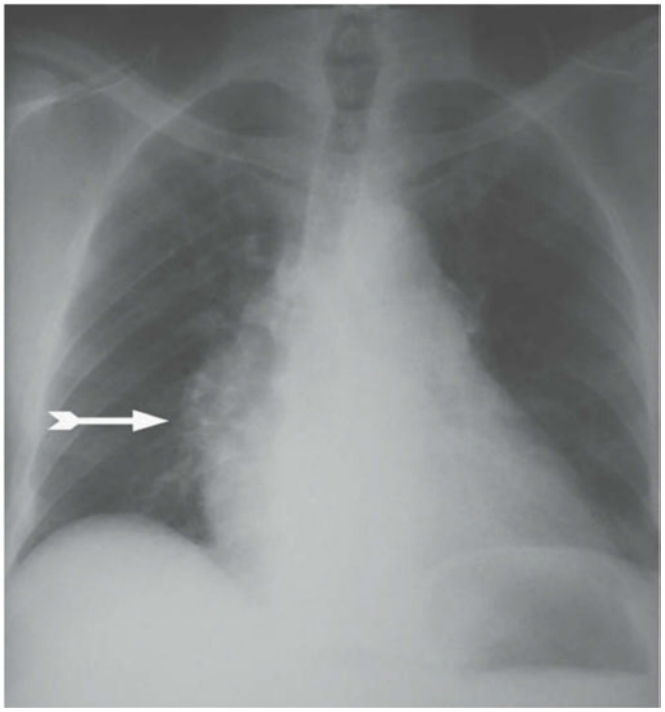


Fig. 70.1. Chest radiograph in a patient with chronic thromboembolic pulmonary hypertension. Note the bilaterally enlarged hilar shadows (*arrow*), prominent right atrium and superior vena cava, and small left atrium.

Pulmonary angiography is the gold standard for the diagnosis of chronic pulmonary thromboembolism. In addition to identifying the level of obstruction and providing a surgical roadmap, right heart catheterization can be performed in the same setting, measuring right heart parameters and evaluating the degree of pulmonary hypertension and pulmonary vascular resistance (PVR). Organized thromboembolic lesions do not have the appearance of the intravascular filling defects seen with acute pulmonary emboli, and experience is essential for the proper interpretation of pulmonary angiograms in patients with unresolved, chronic embolic disease. Typically, organized thrombi appear as unusual filling defects, webs, or bands or as completely thrombosed vessels that may resemble congenital absence of the vessel (Fig. 70.2). Organized material along a vascular wall of a recanalized vessel produces a scalloped or serrated luminal edge. Because of both vessel-wall thickening and dilation of proximal vessels, the contrast-filled lumen may appear relatively normal in diameter. Distal vessels demonstrate the rapid tapering and pruning characteristic of pulmonary hypertension.

In recent years, higher resolution computed tomography (CT) scans of the chest have been used more frequently in the diagnosis of PE. The presence of large clots in lobar or segmental vessels generally confirms the diagnosis. In addition, in rare situations where occlusion of main pulmonary arteries is present or there are concerns of external compression, CT scans can be helpful to differentiate thromboembolic disease from other causes of pulmonary vascular obstruction such as mediastinal fibrosis, lymph nodes, and tumors. CT scanning has the disadvantage, however, that it may miss distal disease.

Pulmonary angiography remains the gold standard for the diagnosis and for planning the operative approach. In addition to pulmonary angiography, patients older than 45 years of age undergo coronary arteriography and other cardiac investigation as necessary. If significant disease is found, additional cardiac surgery is performed at the time of PTE.

Pulmonary angioscopy may be performed in patients where the differentiation between the primary pulmonary hypertension and distal small-vessel pulmonary thromboembolic disease is difficult. The pulmonary angioscope is a fiberoptic telescope that is placed through a central line into the pulmonary artery. The tip contains a balloon that is then filled with saline and pushed against the vessel wall. A bloodless field can thus be obtained to view the pulmonary artery wall. The classic appearance of chronic pulmonary thromboembolic disease by angioscopy consists of intimal thickening, with intimal irregularity and scarring, and webs across small vessels. The presence of embolic disease, occlusion of vessels, or thrombotic material is diagnostic.

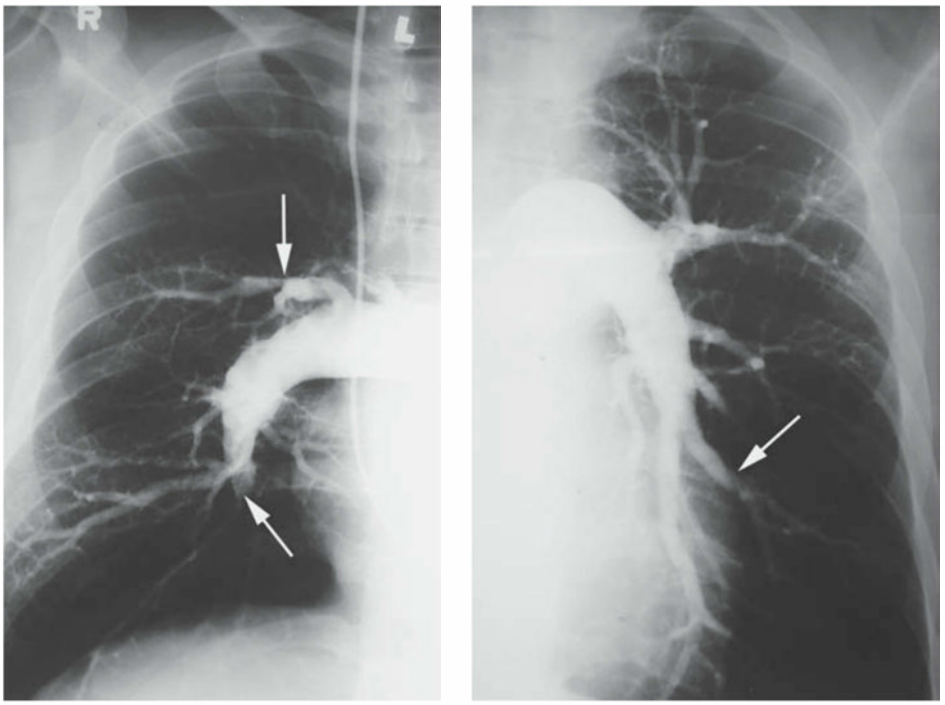


Fig. 70.2. Right and left pulmonary angiograms demonstrate intraluminal filling defects, abrupt cutoffs of branches, and bands (*white arrows*). Note lack of filling to the periphery.

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ALTERNATIVE TREATMENTS

Medical therapy for CTEPH is of limited value and is palliative at best. There are a wide variety of pharmacologic agents in recent use for the treatment of primary pulmonary hypertension. These include calcium-channel blockers such as diltiazem and nifedipine, prostacyclins such as epoprostenol (Flolan, Remodulin), prostacyclin analogs, endothelin-receptor antagonists (Tracleer), and nitric oxide. However, thromboembolic disease represents a mechanical obstruction that is not amenable to drug therapy.

Right ventricular failure is generally treated with diuretics and vasodilators, and although some improvement may result, the effect is generally transient because the failure will not resolve until the obstruction is removed. Similarly, the prognosis is unaffected by medical therapy, which should be regarded as only supportive. However, because of the bronchial circulation, pulmonary embolization seldom results in tissue necrosis. Surgical endarterectomy, therefore, will allow distal pulmonary tissue to be used once more in gas exchange.

Chronic anticoagulation represents the mainstay of the medical regimen. Anticoagulation is primarily not only used to prevent future embolic episodes but also serves to limit the development of thrombus in regions of low flow within the pulmonary vasculature. Inferior vena caval filters are used routinely to prevent recurrent embolization. If caval filtration and anticoagulation fail to prevent recurrent emboli, immediate thrombolysis may be beneficial, but lytic agents are incapable of altering the chronic component of the disease.

The only other surgical alternative for these patients is transplantation. However, we consider transplantation to be inappropriate for the treatment of this disease and believe it is an outdated form of surgical management for CTEPH. Considering the mortality and morbidity rates of patients on the waiting list, the higher risk of the operation, and the lower survival rate (approximately 80% at 1 year at experienced centers for transplantation vs. at least 95% for pulmonary endarterectomy), we believe that PTE is the superior choice. Furthermore, pulmonary endarterectomy appears to be permanently curative, and the issues of a continuing risk of rejection and immunosuppression are not present.

PULMONARY THROMBOENDARTERECTOMY

The first successful pulmonary "thromboendarterectomy" was performed by Allison in 1960, and took place in a patient who 12 days before had a thigh injury that had led to a pulmonary embolus. Allison used a sternotomy and surface hypothermia, but only fresh clots were removed. Since then, there have been many occasional reports of the surgical treatment of chronic pulmonary thromboembolism, but most of the surgical experience in pulmonary endarterectomy has been reported from the University of California, San Diego (UCSD), Medical Center. Braunwald commenced the UCSD experience with this operation in 1970, which now totals <2,800 cases. The operation to be described, using deep hypothermia and circulatory arrest, is now our standard procedure.

Indications

When the diagnosis of thromboembolic pulmonary hypertension has been firmly established, the decision for operation is made based on the severity of symptoms and the general condition of the patient. Early in our pulmonary endarterectomy experience, Moser and colleagues pointed out that there were three major reasons for considering thromboendarterectomy: hemodynamic, alveolo respiratory, and prophylactic. The hemodynamic goal is to prevent or ameliorate right ventricular compromise caused by pulmonary hypertension. The respiratory objective is to improve respiratory function by removing a large ventilated but unperfused physiologic dead space. The prophylactic goal is to prevent progressive right ventricular dysfunction or retrograde extension of the obstruction, which might result in further cardiorespiratory deterioration or death. Our subsequent experience has added another prophylactic goal: the prevention of secondary arteriopathic changes in the remaining patent vessels.

The ages of the patients in our series have ranged from 7 to 86 years. An ideal patient will have a severely elevated PVR level at rest, the absence of significant comorbid disease that is unrelated to right heart failure, and the appearances of chronic thrombi on angiogram that appear to be in balance with the measured PVR level. Exceptions to this general rule, of course, occur.

Some patients (increasingly in our practice) have discordance between the angiographic appearance and a very high PVR. This indicates that there may be secondary arteriopathic changes, but provided that the basis for the elevated PVR is thromboembolic we will still accept these patients for operation, though they might not have an optimal result. Although most patients have a PVR level in the range of 800 dynes/second/cm^{-5} and pulmonary artery pressures less than systemic, the hypertrophy of the right ventricle that occurs over time makes pulmonary hypertension to suprasystemic levels possible. Therefore, many patients (perhaps 30% in our practice) have a level of PVR in excess of 1,000 dynes/second/cm^{-5} and suprasystemic pulmonary artery pressures. There is no upper limit of PVR level, pulmonary artery pressure, or degree of right ventricular dysfunction that excludes patients from operation.

We have become increasingly aware of the changes that can occur in the remaining patient (unaffected by clot) pulmonary vascular bed subjected to the higher pressures and flow that result from obstruction in other areas. Therefore, with the increasing experience and safety of the operation, we are tending to offer surgery to symptomatic patients whenever the angiogram demonstrates thromboembolic disease. A rare patient might have a PVR level that is normal at rest and elevated with minimal exercise. This is usually a young patient with total unilateral pulmonary artery occlusion and unacceptable exertional dyspnea because of an elevation in dead-space ventilation. Operation in this circumstance is performed to reperfuse lung tissue, reestablish a more normal ventilation/perfusion relationship (thereby reducing minute ventilatory requirements during rest and exercise), and preserve the integrity of the contralateral circulation. If one has not been previously implanted, an inferior vena caval filter is routinely placed several days before the operation.

OPERATIVE TECHNIQUES

Principles

Although the essential techniques of PTE are quite similar to those of other open-heart operations, there are several guiding principles for this procedure. The disease is almost always bilateral, although the volume of chronic thromboembolic material may vary significantly between the two lungs. Furthermore, for pulmonary hypertension to be a major factor, both pulmonary arteries must be substantially involved. The surgery is therefore always bilateral. The only reasonable approach to both pulmonary arteries is through a median sternotomy incision. Historically, there were many reports of unilateral operation, and occasionally this is still performed, in inexperienced centers, through

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a thoracotomy. However, the unilateral approach ignores the disease on the contralateral side, subjects the patient to hemodynamic jeopardy during the clamping of the pulmonary artery, and does not allow good visibility because of the continued presence of bronchial blood flow. In addition, collateral channels develop in chronic thrombotic hypertension not only through the bronchial arteries but also from diaphragmatic, intercostal, and pleural vessels. The dissection of the lung in the pleural space via a thoracotomy incision can, therefore, be extremely bloody. The median sternotomy incision, apart from providing bilateral access, avoids entry into the pleural cavities and allows the ready institution of cardiopulmonary bypass.

Cardiopulmonary bypass is essential not only to ensure cardiovascular stability when the operation is performed but also to allow cooling of the patient for periods of circulatory arrest. Exceptional visibility of the pulmonary vasculature is required. A bloodless field is an absolute requirement to define an adequate endarterectomy plane and then to follow the pulmonary endarterectomy specimen deep into the subsegmental vessels. Because of the copious bronchial blood flow usually present in these cases, periods of circulatory arrest are necessary to ensure perfect visibility. Again, there have been sporadic reports of the performance of this operation without circulatory arrest. However, it should be emphasized that although endarterectomy is possible without circulatory arrest, a complete and full endarterectomy is not. We always initiate the procedure without circulatory arrest, and depending on the collateral flow, a variable amount of dissection is possible before the circulation is stopped, but never complete dissection. The circulatory arrest periods are limited to 20 minutes, with restoration of flow between each arrest. With experience, the endarterectomy usually can be performed with a single period of circulatory arrest on each side.

A true endarterectomy in the plane of the media must be accomplished. It is essential to appreciate that the removal of visible thrombus is largely incidental to this operation. Indeed, in most patients, no free thrombus is present; on initial direct examination, the pulmonary vascular bed may appear normal to an inexperienced eye. The early literature on this procedure indicates that thrombectomy was often performed without a complete endarterectomy, and in these cases, the pulmonary artery pressures did not improve, often with the resultant death of the patient.

Preparation and Anesthetic Considerations

The operative preparation and anesthetic concerns are for the most part quite similar to those for any open-heart procedure. Routine monitoring for anesthetic induction includes a surface electrocardiogram, cutaneous oximetry, and radial artery pressure lines. After induction, a pulmonary artery catheter and a transesophageal echocardiographic probe are also inserted. We generally include a femoral arterial line for more accurate assessment during rewarming and on discontinuation of cardiopulmonary bypass. It is quite common for these patients to develop some degree of peripheral vasoconstriction after hypothermic circulatory arrest, thereby making radial pressure readings inaccurate. The femoral line is generally removed in the intensive care unit when the two readings are correlated.

Electroencephalographic recording is routinely performed to ensure the lack of cerebral activity before circulatory arrest is induced. The patient's head is enclosed in a cooling jacket, and cerebral cooling is begun after the initiation of bypass. Temperature measurements are made of the esophagus, tympanic membrane, urinary catheter, rectum, and blood (through the Swan–Ganz catheter). If the patient's condition is stable after the induction of anesthesia, up to 500 mL of autologous whole blood is withdrawn for later use, and the volume deficit is replaced with crystalloid solution.

Operative Techniques

After a median sternotomy incision is made, the pericardium is incised longitudinally and attached to the wound edges. Typically, the right heart is enlarged, with a tense right atrium and a variable degree of tricuspid regurgitation. There is usually severe right ventricular hypertrophy. These patients are generally quite sensitive to any manipulation of the heart, and with critical degrees of obstruction, the patient's condition may become quite unstable.

Anticoagulation is achieved with the use of beef-lung heparin sodium (400 units/kg, intravenously) administered to prolong the activated clotting time beyond 400 seconds. Full cardiopulmonary bypass is instituted with high ascending aortic cannulation and two caval cannulae. These cannulae must be inserted into the superior and inferior vena cavae sufficiently to enable subsequent opening of the right atrium. The heart is emptied on bypass, and a temporary pulmonary artery vent is placed in the midline of the main pulmonary artery 1 cm distal to the pulmonary valve. This will mark the beginning of the left pulmonary arteriotomy.

When cardiopulmonary bypass is initiated, surface cooling with both the head jacket and the cooling blanket is begun. The blood is cooled with the pump-oxygenator. Cooling generally takes 45 minutes to 1 hour. When ventricular fibrillation occurs, an additional vent is placed in the left atrium through the right superior pulmonary vein. This prevents atrial and ventricular distension from the large amount of bronchial arterial blood flow that is common with these patients.

Generally, the primary surgeon starts the operation on the patient's left side. During the cooling period, some preliminary dissection can be performed, with full mobilization of the right pulmonary artery from the ascending aorta. The superior vena cava is also fully mobilized. The approach to the right pulmonary artery is made medial, not lateral, to the superior vena cava (Fig. 70.3). All dissection of the pulmonary arteries takes place intrapericardially, and neither pleural cavity should be entered. An incision is then made in the right pulmonary artery from beneath the ascending aorta out under the superior vena cava and entering the lower lobe branch of the pulmonary artery just after the takeoff of the middle lobe artery (Fig. 70.4). It is important that the incision stays in the center of the vessel and continues into the lower rather than the middle lobe artery.

A modified cerebellar retractor is placed between the aorta and superior vena cava. Upon opening the pulmonary artery, a varying degree of loose thrombus may be present. The material is then removed to ensure good visualization of the vascular bed. It is most important to recognize, however, that first, an embolectomy without subsequent endarterectomy is quite ineffective regardless of the size of thromboembolic material, and second, in most patients with chronic thromboembolic hypertension, direct examination of the pulmonary vascular bed at operation generally shows no obvious embolic material. Therefore, to the inexperienced or cursory glance, the pulmonary vascular bed may well appear normal even in patients with severe chronic embolic pulmonary hypertension.

When the patient's temperature reaches 20°C, the aorta is cross-clamped and a single dose of cold cardioplegic solution (1 L) is administered. Additional myocardial protection is obtained by the use of a cooling jacket. The entire procedure is now

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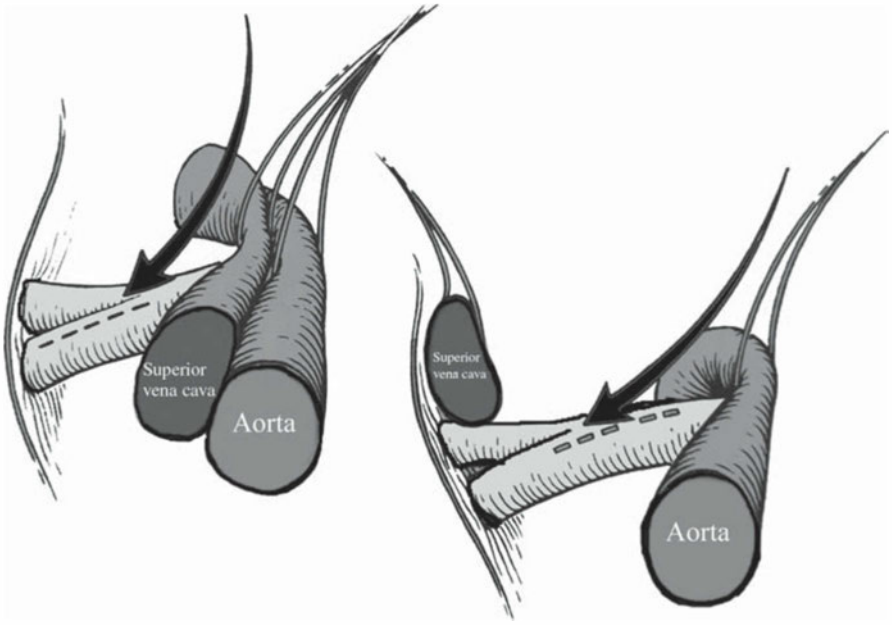


Fig. 70.3. Surgical approach to the right pulmonary artery. The approach is medial to the superior vena cava, between the superior vena cava and the aorta. An approach lateral to the superior vena cava provides a restricted view.

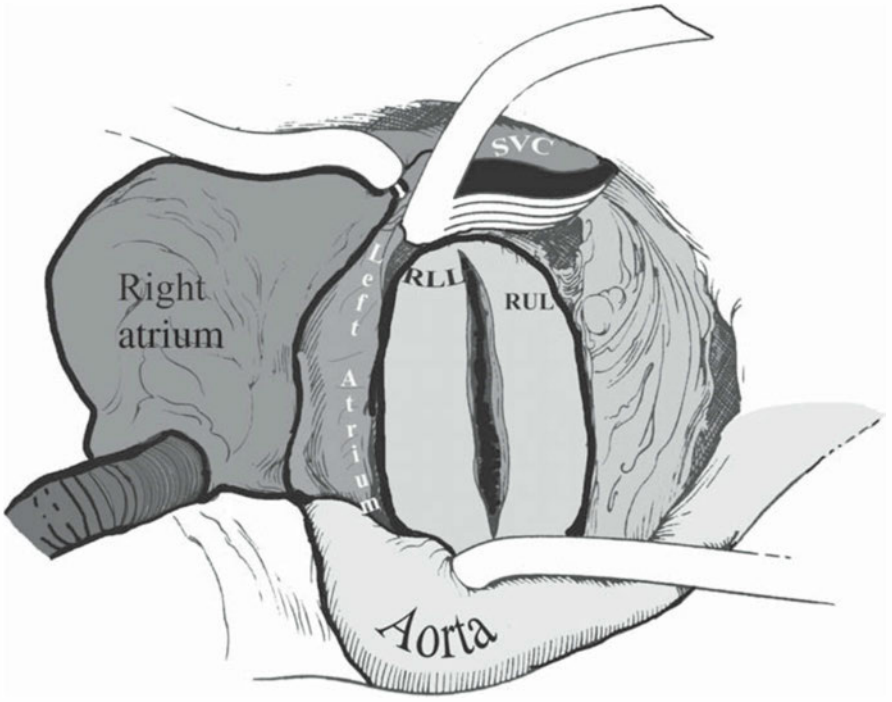


Fig. 70.4. Exposure to the right pulmonary artery. Note how the incision is made between the superior vena cava (SVC) and the aorta. In addition, the right upper lobe (RUL) and the right lower lobe (RLL) takeoffs are shown.

performed with a single aortic cross-clamp period with no further administration of cardioplegic solution.

If the bronchial circulation is not excessive, the endarterectomy plane can be found during this early dissection. However, although a small amount of dissection can be performed before the initiation of circulatory arrest, it is unwise to proceed unless perfect visibility is obtained because the development of a correct plane is essential. Recognizing the plane is perhaps the most crucial and technically challenging part of the operation.

When blood obscures direct vision of the pulmonary vascular bed, thiopental is administered (500 mg to 1 g) until the electroencephalogram becomes isoelectric. In most cases, the electroencephalogram is already isoelectric when the core temperature reaches 20°C. Circulatory arrest is then initiated, and the patient undergoes exsanguination. All monitoring lines to the patient are turned off to prevent the aspiration of air. Snares are tightened around the cannulae in the superior and inferior venae cavae. It is rare that one 20-minute period for each side is exceeded. Although retrograde cerebral perfusion has been advocated for total circulatory arrest in other procedures, it is not helpful in this operation because it does not allow a completely bloodless field, and with the short arrest times that can be achieved with experience, it is not necessary.

A microtome knife is used to develop the endarterectomy plane posteriorly because any inadvertent egress in this site could be repaired readily or simply left alone. Dissection in the correct plane is critical because if the plane is too deep, the pulmonary artery may perforate, with fatal results, and if the dissection plane is not deep enough, inadequate amounts of the chronically thromboembolic material will be removed, leaving the patient with residual pulmonary hypertension.

After the plane is correctly developed, a full-thickness layer is left in the region of the incision to ease subsequent repair (Fig. 70.5). The endarterectomy is then performed with an eversion technique. Because the vessel is everted and subsegmental branches are being worked on, a perforation here will become completely inaccessible and invisible later. This is why the absolute visualization in a completely bloodless field provided by circulatory arrest is essential. It is important that each subsegmental branch is followed and freed individually until it ends in a "tail" beyond which there is no further obstruction. Residual material should never be cut free; the entire specimen should "tail off" and come free spontaneously.

After the right-sided endarterectomy is completed, circulation is restarted, and the arteriotomy is repaired with a continuous 6-0 polypropylene suture. The hemostatic nature of this closure is aided by the nature of the initial dissection, with the full thickness of the pulmonary artery being preserved immediately adjacent to the incision.

After the completion of the repair of the right arteriotomy, the surgeon moves to the patient's right side. The pulmonary vent catheter is withdrawn, and an arteriotomy is made from the site of the pulmonary

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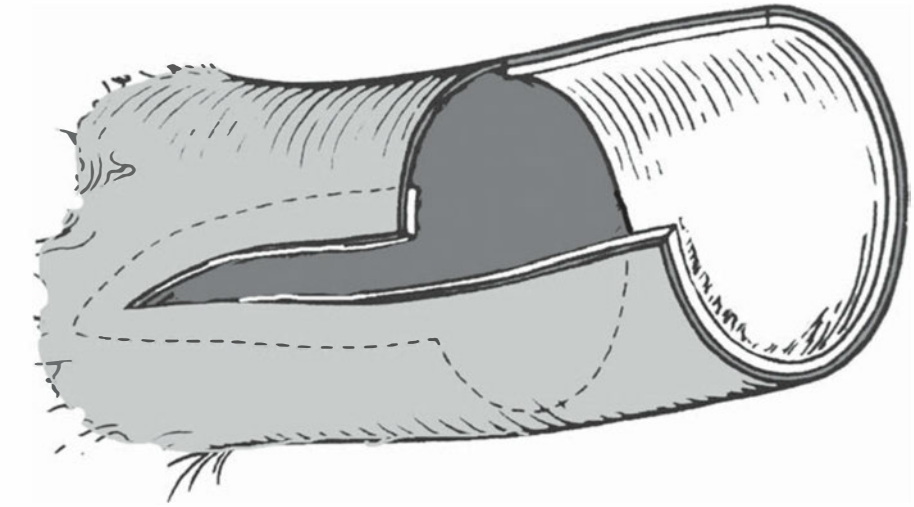


Fig. 70.5. The plane of dissection is first raised posteriorly and continued toward the edge of the arteriotomy but trimmed short in the region of the incision. This results in a full-thickness artery for subsequent closure.

vent hole laterally to the pericardial reflection, avoiding entry into the left pleural space. Additional lateral dissection does not enhance intraluminal visibility, may endanger the left phrenic nerve, and makes subsequent repair of the left pulmonary artery more difficult (Fig. 70.6).

The left-sided dissection is virtually analogous in all respects to that accomplished on the right. The duration of circulatory arrest intervals during the performance of the left-sided dissection is subject to the same restriction as the right.

After the completion of the endarterectomy, cardiopulmonary bypass is reinstituted and warming is commenced. If the systemic vascular resistance level is high, nitroprusside is administered to promote vasodilation and warming. The rewarming period generally takes approximately 90 minutes, but it varies according to the body mass of the patient.

The pulmonary artery is then closed, and the pulmonary vent is replaced. The right atrium is then opened and examined. Any intra-atrial communication is closed. Although tricuspid valve regurgitation is invariable in these patients and is often severe, tricuspid valve repair is not performed unless there is a structural defect. Right ventricular remodeling occurs within a few days, with the return of tricuspid competence. If other cardiac procedures are required, such as coronary artery or mitral or aortic valve surgery, these are conveniently performed during the systemic rewarming period. Myocardial cooling is discontinued after all cardiac procedures have been concluded. The left atrial vent is removed, and the vent site is repaired. All air is removed from the heart, and the aortic cross-clamp is removed.

When the patient has rewarmed, cardiopulmonary bypass is discontinued. Dopamine hydrochloride is routinely administered at renal doses, and other inotropic agents and vasodilators are titrated as necessary to sustain acceptable hemodynamics. The cardiac output is generally high, with a low systemic vascular resistance. Temporary atrial and ventricular epicardial pacing wires are placed.

Despite the duration of extracorporeal circulation, hemostasis is readily achieved, and the administration of platelets or coagulation factors is generally unnecessary. Wound closure is routine. A vigorous diuresis is usual for the next few hours, also a result of the previous systemic hypothermia.

There are four broad types of pulmonary occlusive disease related to thrombus that can be appreciated, and we use the following classification: type I disease (approximately 20% of cases of thromboembolic pulmonary hypertension; Fig. 70.7) refers to the situation in which major vessel clot is present and readily visible on the opening of the pulmonary arteries. As mentioned earlier, all central thrombotic material has to be completely removed before the endarterectomy. In type II disease (approximately 60% of cases; Fig. 70.8), no major vessel thrombus can be appreciated. In these cases only thickened intima can be seen, occasionally with webs, and the endarterectomy plane is raised in the main, lobar, or segmental vessels. Type III disease (approximately 20% of cases in our series; Fig. 70.9) presents the most challenging surgical situation. The disease is very distal and confined to the segmental and subsegmental branches. No occlusion of vessels can be seen initially. The endarterectomy plane must be carefully and painstakingly raised in each segmental

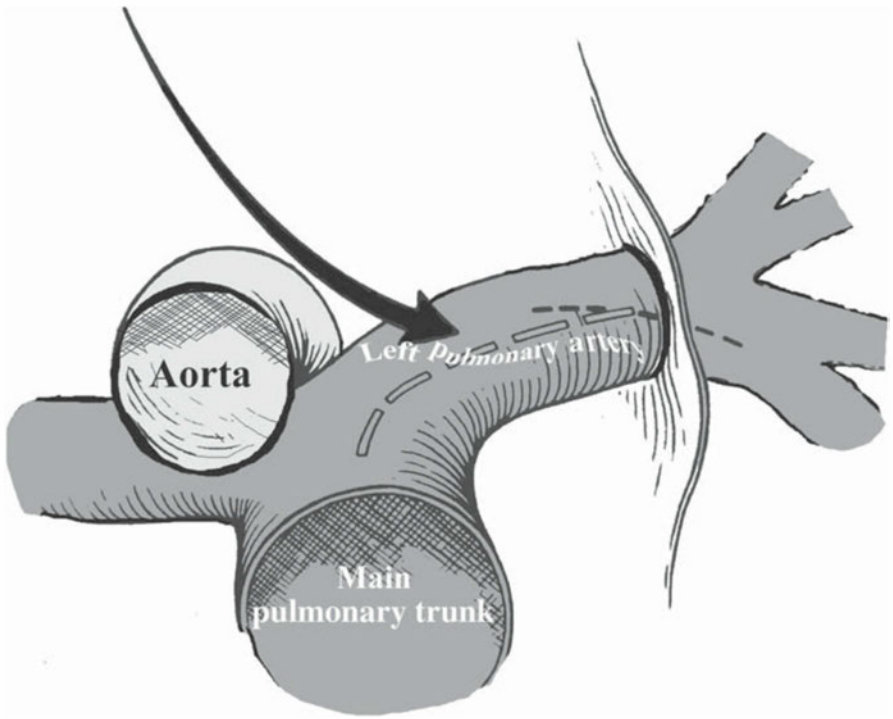


Fig. 70.6. Surgical approach to the left pulmonary artery. The approach begins in the mid-portion of the main artery and carried past the takeoff of the left upper lobe. The incision stays in the midline of the vessel as far as the pericardial reflection. A more distal incision provides limited view.

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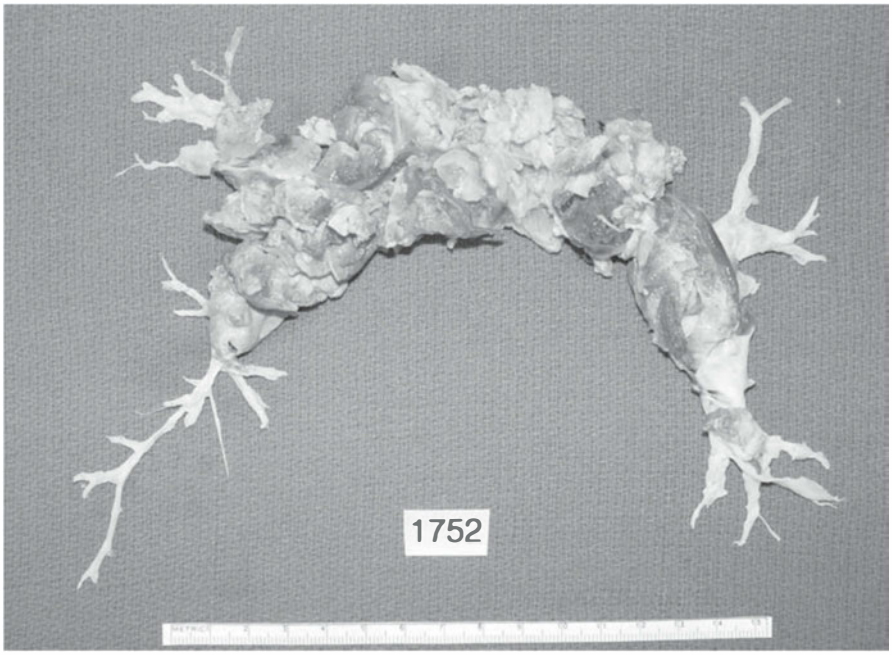


Fig. 70.7. Surgical specimen removed from a patient with type I surgical classification. Note that the removal of fresh thromboembolic material leaves behind a large amount of obstructive disease.

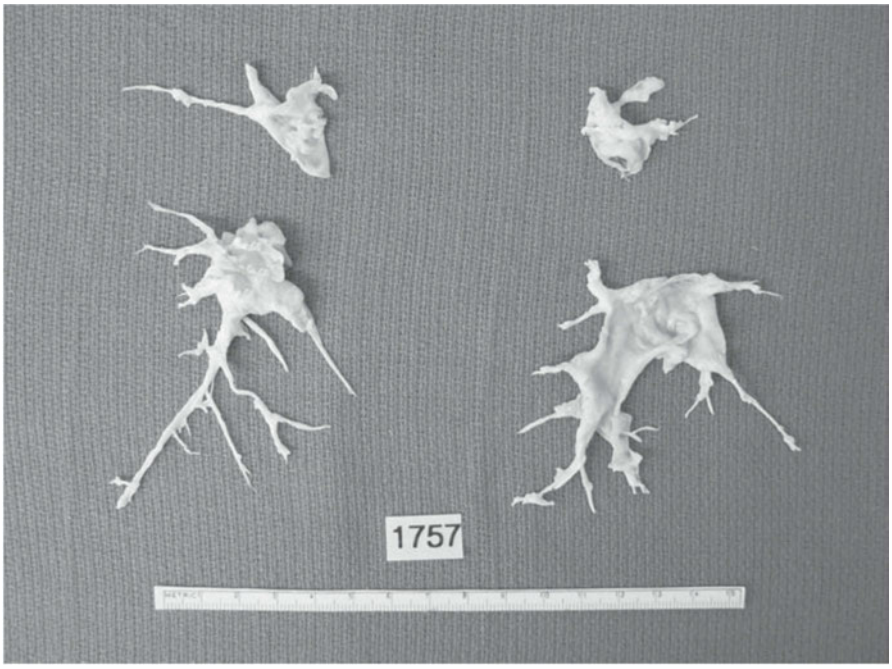


Fig. 70.8. Surgical specimen removed on right and left from a patient with type II surgical classification. Note how the specimen ends in "feathered tails" in each of the branches.

and subsegmental branch. Type III disease is most often associated with presumed repetitive thrombi from indwelling catheters (such as pacemaker wires) or ventriculoatrial shunts. Type IV disease (Fig. 70.10) does not represent primary thromboembolic pulmonary hypertension and is inoperable. In this entity, there is intrinsic small-vessel disease, although secondary thrombus may occur as a result of stasis. Small-vessel disease may be unrelated to thromboembolic events ("primary" pulmonary hypertension) or occur in relation to thromboembolic hypertension as a result of a high-flow or high-pressure state in previously unaffected vessels similar to the generation of Eisenmenger syndrome. We believe that there may also be sympathetic "cross-talk" from an affected contralateral side or stenotic areas in the same lung.

POSTOPERATIVE CARE

Although much of the postoperative care is common to that of the more ordinary open-heart surgery patients, there are some important differences. Meticulous postoperative management is essential to the success of this operation. All patients are mechanically ventilated for at least 24 hours, and all patients are subjected to a maintained diuresis with the goal of reaching the patient's preoperative dry weight within 24 hours.

The electrocardiogram, systemic and pulmonary arterial and central venous pressures, temperature, urine output, arterial oxygen saturation, chest tube drainage, and fluid balance are monitored. A pulse oximeter is used to continuously monitor peripheral oxygen saturation. Management of cardiac arrhythmias and output and treatment of wound bleeding are identical to those for other open-heart operations. In addition, higher minute ventilation is often required early after the operation to compensate for the temporary metabolic acidosis that develops after the long period of circulatory arrest, hypothermia, and cardiopulmonary bypass. Extubation should be performed on the first postoperative day, if possible.

Complications

Patients are not only subject to all complications associated with open-heart and major lung surgery (arrhythmias, atelectasis, wound infection, pneumonia, mediastinal bleeding, etc.) but also may develop complications specific to this operation. One such complication is the development of a "reperfusion response." This is a specific complication that occurs in most patients to some degree and is related to localized pulmonary edema. Reperfusion injury is defined as a radiologic opacity seen in the lungs within 72 hours of pulmonary endarterectomy. This unfortunately loose definition may, therefore, encompass many causes, such as fluid overload and infection.

True reperfusion injury that directly adversely impacts the clinical course of the patient now occurs in approximately 8% to 10% of patients. In its most dramatic form, it occurs soon after operation (within a few hours) and is associated with profound desaturation. Edema-like fluid, sometimes with a bloody tinge, is suctioned from the endotracheal tube. Frank blood from the endotracheal tube, however, signifies a

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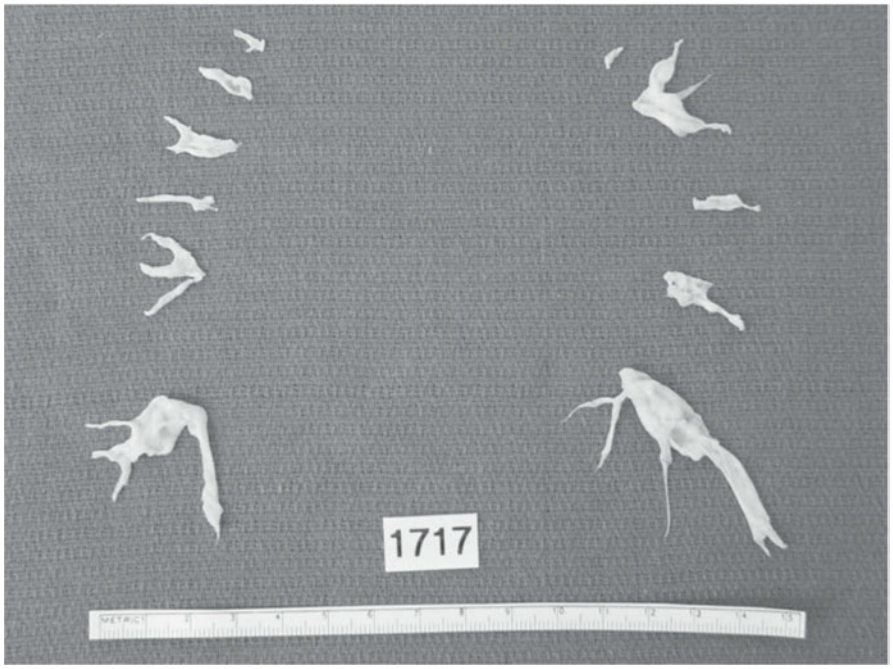


Fig. 70.9. For this specimen, the plane of dissection was raised at each segmental level. The surgical classification is type III.

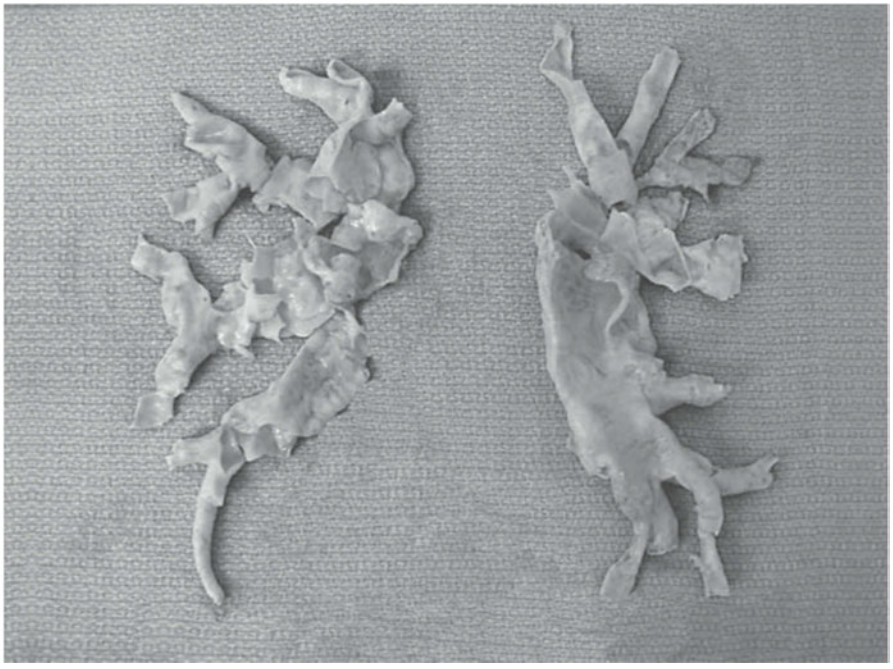


Fig. 70.10. The surgical specimen from a patient with a surgical classification of type IV. Note that each branch ends abruptly in "trousers" rather than "feathered tails." This patient's postoperative hemodynamic numbers were unchanged despite the impressive appearance of the endarterectomy specimen.

mechanical violation of the blood–airway barrier that has occurred at operation and stems from a technical error. This complication should be managed, if possible, by identification of the affected area by bronchoscopy and balloon occlusion of the affected lobe until coagulation can be normalized.

One common cause of the reperfusion pulmonary edema is persistent high pulmonary artery pressures after operation when a thorough endarterectomy has been performed in certain areas, but there remains a large part of the pulmonary vascular bed affected by type IV change. However, the reperfusion phenomenon can also be encountered in patients after a seemingly technically perfect operation with complete resolution of high pulmonary artery pressures. In these cases, the response may be one of reactive hyperemia after the revascularization of segments of the pulmonary arterial bed that have long experienced no flow. Other contributing factors may include perioperative pulmonary ischemia and conditions associated with high-permeability lung injury in the area of the now denuded endothelium. Fortunately, the incidence of this complication is very much less common now in our series, probably as a result of the more complete and expeditious removal of the endarterectomy specimen that has come with the large experience over the last decade and the recognition that an aggressive diuresis is required postoperatively.

Management of the "Reperfusion Response"

Early measures should be taken to minimize the development of pulmonary edema with diuresis, maintenance of the hematocrit levels, and the early use of ventilatory peak end-expiratory pressure. Once the capillary leak has been established, treatment is supportive because reperfusion pulmonary edema will eventually resolve if satisfactory hemodynamics and oxygenation can be maintained. Careful management of ventilation and fluid balance is required. The hematocrit is kept high (32% to 36%), and the patient undergoes aggressive diuresis, even if this requires ultrafiltration. The patient's ventilatory status may be dramatically position sensitive. The inspired oxygen fraction (FiO_2) level is kept as low as is compatible with an oxygen saturation of 90%. A careful titration of positive end-expiratory pressure is carried out, with a progressive transition from volume-limited to pressure-limited inverse ratio ventilation and the acceptance of moderate hypercapnia. The use of steroids is discouraged because they are generally ineffective and may lead to infection. Infrequently, inhaled nitric oxide at 20 to 40 parts per million can improve the gas exchange. On occasion, we have used extracorporeal perfusion support (extracorporeal membrane oxygenator or extracorporeal carbon dioxide removal) until ventilation can be resumed satisfactorily, usually after 7 to 10 days.

CONCLUSION

It is increasingly apparent that pulmonary hypertension caused by chronic PE is a relative common condition that is

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under-recognized and carries a poor prognosis. Medical therapy is ineffective in prolonging life and only transiently improves the symptoms. The only therapeutic alternative to PTE is lung transplantation. The advantages of thromboendarterectomy include a lower operative morbidity and mortality and excellent long-term results without the risks associated with chronic immunosuppression and chronic allograft rejection. The mortality for thromboendarterectomy at our institution is now in the range of 3%, with sustained benefit. These results are clearly superior to those for transplantation in both the short and long terms.

Although PTE is technically demanding for the surgeon, requiring careful dissection of the pulmonary artery planes and the use of circulatory arrest, excellent short- and long-term results can be achieved. Successive improvements in operative technique developed over the last two decades now allow pulmonary endarterectomy to be offered to patients with an acceptable mortality rate and excellent anticipation of clinical improvement. With this growing experience, it has also become clear that unilateral operation is obsolete and that circulatory arrest is essential.

We have performed >2,800 pulmonary endarterectomies at the University of California, San Diego, Medical Center, with almost all of these carried out since 1990. With our growing experience, we are now able to offer this procedure to some very high-risk patients with a total overall mortality rate of about 3%. The vast majority of patients enjoy a dramatic hemodynamic improvement postoperatively. A reduction in pulmonary pressures and PVR to normal levels and a corresponding improvement in pulmonary blood flow and cardiac output are generally immediate and sustained. In general, these changes can be assumed to be permanent. Whereas before the operation, >95% of the patients are in New York Heart Association (NYHA) functional class III or IV, at 1 year after the operation, 95% of patients are in NYHA functional class I or II. In addition, echocardiographic studies have demonstrated that with the elimination of chronic pressure overload, right ventricular geometry rapidly reverts toward normal. Right atrial and right ventricular enlargement regresses. Tricuspid valve function returns to normal within a few days as a result of restoration of tricuspid annular geometry after the remodeling of the right ventricle, and therefore tricuspid repair is not part of the operation.

Even with increased awareness of this disease, coupled with our continuous effort in spreading our understanding and knowledge, the principal difficulty remains that this is an under-recognized condition. Increased understanding of both the prevalence of this condition and the possibility of a surgical cure should avail more patients of the opportunity for relief from this debilitating and ultimately fatal disease.

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EDITOR'S COMMENTS

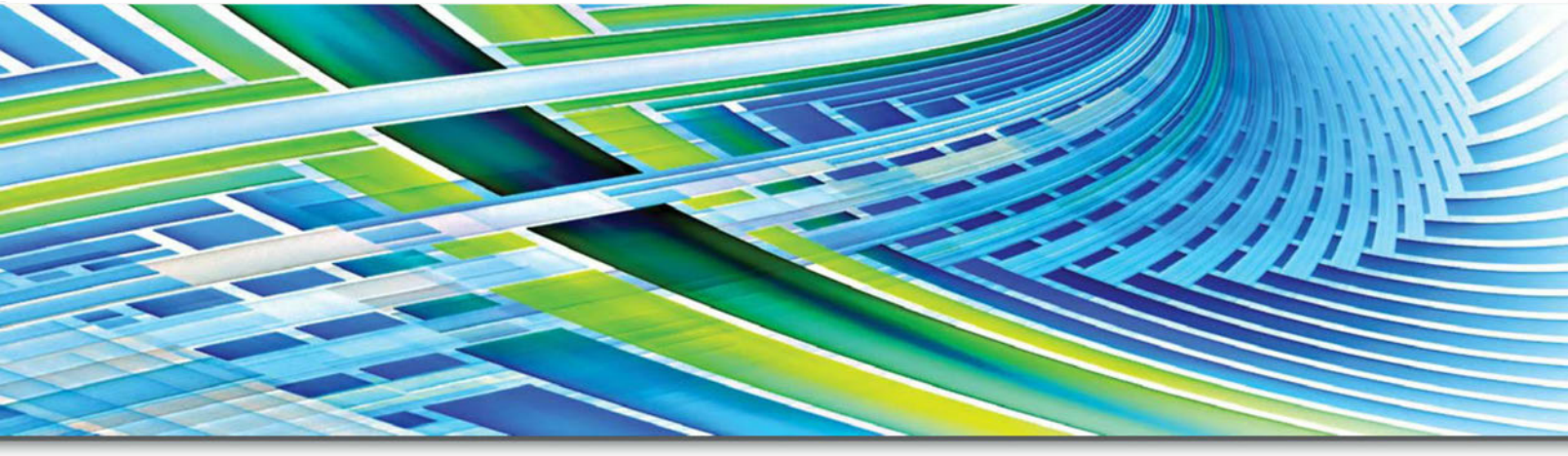
Clearly, these authors have the largest experience with this disease. This has been treated at their institution for the last 40 years and they have had outstanding results. They have made the operation doable with the use of deep hypothermia and circulator arrest. They demonstrate that it is very important to do a complete thromboendarterectomy to get the results that are necessary. The technical points include interpericardial pulmonary arteriotomy and being certain that right planes are dealt with and are critical to the procedure being done safely. They also discuss several complications such as reperfusion injury. We have been interested in this complications for a long time, particularly following lung transplantation. As noted, this is often short lived and they have modalities that need to be available in these patients, such as ECMO, to make certain that long-term survival is achieved.

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Congenital Cardiac Surgery III



71 Anatomy and Classification of Congenital Heart Disease

Paul M. Weinberg

INTRODUCTION

The hallmark of congenital heart disease is the variability in spatial relationships as well as interconnections among the various cardiac and vascular structures. To appreciate these types of abnormalities, it is first necessary to define certain terms that may have different meanings from those used to describe normal anatomy. When cardiac chambers are described, the terms right and left refer only to morphological characteristics which will be further elaborated upon below. They do not refer to the right–left frame of reference in the body. For example, right ventricle refers to a particular cardiac structure with specific morphological features, regardless of its location in the body. When the right–left frame of reference is intended, the terms right-sided or left-sided are used.

It is also important to recognize which structures are, by definition, part of a chamber and which are frequently associated with that chamber. For example, the normal right ventricle includes the infundibulum or outflow tract portion of the heart, whereas there are some hearts in which the infundibulum is partially or entirely related to the left ventricle (anatomically corrected malposition or double outlet left ventricle). Therefore, the presence of infundibulum does not define the right ventricle, but rather is usually associated with it.

Another concept that is central to a morphological approach to congenital heart disease is the recognition of the difference between anatomical, physiological, and surgical diagnoses. This chapter will concern itself entirely with anatomical descriptions and diagnoses. Some of the terms can also be used in a physiological sense; it is vital that one cannot make the mistake of using them interchangeably. For example, anatomically, transposition of the great arteries means that the aorta arises above the morphological right ventricle and the pulmonary artery above the left ventricle. This is true regardless of whether the right ventricle receives desaturated blood from the body or fully saturated blood from the lungs. Transposition physiology, however, refers to any anatomical situation in which the pulmonary artery receives blood of a higher saturation than the aorta. If a term is used without further clarification, it should always refer to anatomical diagnoses, whereas physiologic terms should always be stated as such, for example, "transposition physiology" or "physiological mitral stenosis."

CLASSIFICATION OF CONGENITAL HEART DISEASE

Aims and Basic Principles

The purpose of a classification system is to permit the identification of all examples of anatomy, physiology, and surgery of congenital heart disease in a way that permits storage and retrieval from computerized databases. The aim of a classification system is really to classify entities, not names. As Shakespeare said, "What's in a name? That which we call a rose by any other name would smell as sweet." (Romeo and Juliet II, ii, 43). There seems to be a broad consensus on what the *entities* are. Rather than trying to win over everyone to the best nomenclature, classification systems should focus on the following five principles: (a) organization, (b) economy, (c) accuracy, (d) precision, and (e) quantification.

Organization is essential with any comprehensive classification system. Simple alphabetical listing of diagnoses is unhelpful because of the absence of a standardized nomenclature. Instead diagnoses should be grouped by the organization of the heart itself according to a systematic method. In this way, all possible diagnoses related to a given portion of the heart can be viewed at one time and the appropriate one selected. Because of the fact that there are different schools of thought as to organization, it is unlikely that a single standardized nomenclature will be developed in the near future. One approach to this problem is the one adopted by the International Society for Nomenclature of Paediatric and Congenital Heart Disease. This multidisciplinary group including pediatric cardiologists, cardiac surgeons, and morphologists has created the International Paediatric and Congenital Cardiac Code, which essentially maps terms from one nomenclature system to those of another so that entities will have a single numerical code or group of codes irrespective of the original terms. Inherent in this approach is acceptance of the notion that different groups may use different organizational approaches to categorize entities and may thus "arrive" at the same diagnostic entity by way of different paths. For example, our group would categorize tricuspid atresia as a form of abnormal atrioventricular alignment—specifically an abnormal atrial-to-atrioventricular valve (AVV) connection, whereas another group might think of the same entity as a form of single ventricle or univentricular heart. The path is not as important as assuring that a given entity is always coded in the same way, regardless of the approach. Because of this, definitions of entities are essential. *Economy* refers to saving time of the physician who determines which diagnoses to use and saving computer space with the avoidance of redundant information. This is accomplished through mutual exclusivity. Diagnoses are grouped within organizational categories into subgroups where diagnoses are mutually exclusive—only one diagnosis within that subgroup is possible. For example, only one atrial situs, one ventricular situs, one great artery situs, one atrial-to-AVV connection, one AVV-to-ventricle connection, and one ventriculo arterial alignment abnormality are possible. Additional diagnoses that are thought to relate a group of similar abnormalities are superfluous, and if ultimately shown to be incorrect (i.e., abnormalities not similar), can actually be counterproductive. Provision of redundant information during data

entry is wasteful of time as well as computer storage space.

Accuracy means that a given diagnosis has the same meaning every time it is used. The meaning does not change with context. In other words, a particular anatomical ventricular septal defect (VSD) has the same designation regardless of ventriculoarterial alignment and great artery position. The atrial situs designation is the same whether there is dextrocardia or levocardia. Dependence on context complicates data retrieval, since a given diagnosis could have different meanings depending on what other diagnoses are present in each case.

Precision is the ability to classify fine anatomical detail if the information is available. Nonspecific diagnoses should only be used if more precise information is lacking. For example, a plain chest roentgenogram may indicate a right aortic arch (not further specified), but magnetic resonance imaging (MRI) or angiography may show a right aortic arch with retroesophageal diverticulum. A dilution of information would occur if only "right aortic arch" were used as the diagnosis. Whereas if one were searching for all right aortic arches, general as well as specific entities could be included.

Quantification is the option to assign an indication of severity to certain diagnoses. Sometimes the severity of a lesion is as important as the anatomical detail. While an epidemiologist may consider all VSDs of a particular type equally significant, the clinician may wish to distinguish small VSDs from large. In many cases, the designation is arbitrary but may still be useful.

Implementation—The Segmental Approach

The segmental approach to diagnosis in congenital heart disease was developed by Van Praagh to provide a systematic method for describing congenital heart defects. It fits the basic principles outlined above. Important features that distinguish Van Praagh's segmental approach from many other systems include the following: (1) diagnoses are based on morphological analysis. While physiological correction of defects is the primary goal of cardiac surgery, it is also clear that attention to the underlying morphology is important as it influences surgical approach as well as long-term function. (2) The system is not situs-dependent; each segment is diagnosed independently. Situs-dependent systems suffer from two shortcomings. If the situs is found to be different than initially thought, all of the *dependent* diagnoses change as well. If the situs is ambiguous or if the downstream connections are not one-to-one, as in double inlet ventricles, the concept of a situs-dependent system breaks down. (3) There is a hierarchy of diagnoses (Fig. 71.1). Those entities at the top do not change the diagnoses or nomenclature of those below, but they have greater significance: internal organization is more important than a stenotic valve in terms of classification. (4) There is a logical sequence to the analysis and description of the heart.

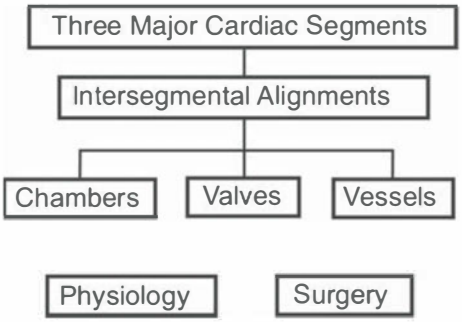


Fig. 71.1. Segmental approach hierarchy. Schematic hierarchy of cardiac diagnoses based on the segmental approach to diagnosis of Van Praagh.

The cardiovascular system is thought of as having three major segments—viscero atrial situs, ventricular loop, and great artery situs. The situs or spatial organization of each of the segments can be described *independently* using morphological features of the chambers and vessels. Adjacent segments are related to each other by intersegmental alignments. The three major segments can be thought of as the blueprints of each of the floors of a three-story house. Each can be understood individually without reference to the other two, hence independence. The intersegmental alignments are analogous to stairways that connect adjacent floors.

Once segments and intersegmental alignments have been determined, individual chamber, valve, and vessel diagnoses are made. Continuing the architectural analogy, chamber, valve, and vessel abnormalities are like the furnishings of individual rooms.

Cardiac Segments

Viscero atrial situs describes the arrangement of the asymmetrical abdominal viscera and vessels, the lungs, and the atria. There are three forms of viscero atrial situs: solitus, inversus, and ambiguus. Situs solitus is characterized by a right-sided liver, left-sided stomach, and right-sided inferior vena cava (IVC). The atria display characteristic right atrial morphology of the right-sided appendage (described specifically in the "Chambers, Valves, and Vessels" section), left atrial morphology of the left-sided appendage, IVC entering the right-sided atrium, and septum primum attached to the left side of septum secundum. Situs inversus means the mirror image of solitus, viz., left-sided liver and IVC, right-sided stomach, right atrial appendage on the left, left atrial appendage on the right, IVC entering left-sided atrium, and septum primum attached to the right side of septum secundum. Situs ambiguus has characteristics of both situs solitus and situs inversus in the same person. For example, a liver that spans the abdomen from right to left (sometimes referred to as a "midline" liver) being both right-sided and left-sided has characteristics of both solitus and inversus. Similarly, bilateral morphologic right or bilateral morphologic left atrial appendages mean one solitus and one inversus. In the same way, a left-sided abdominal IVC that crosses to the right in the liver and enters a right-sided atrium with right atrial appendage morphology has features of both situs inversus (abdominal) and situs solitus (atrial). All these examples of combinations of situs solitus and situs inversus are termed situs ambiguus. This is the viscero atrial situs seen in the abdominal heterotaxy syndrome. Most, but not all, of these patients have splenic abnormalities: asplenia or polysplenia. The terms bilateral right sidedness and bilateral left sidedness are conveniences for remembering the constellation of abnormalities frequently seen with asplenia (bilateral right sidedness, since the normal right side has no spleen) or polysplenia (bilateral left sidedness, since the normal left side has a spleen, so multiple spleens is like multiple left sides), but do not carry the force of viscero atrial situs since there are numerous exceptions to the symmetry implied by those terms.

Ventricular loop is the designation for the situs of the ventricles. There are two types: D loop and L loop. Unlike the atria that are virtually always side-by-side, the ventricles, while frequently so, may be oriented antero posteriorly or supero inferiorly, still displaying one of two internal spatial organizations. Therefore, a designation independent of a simple right–left frame of

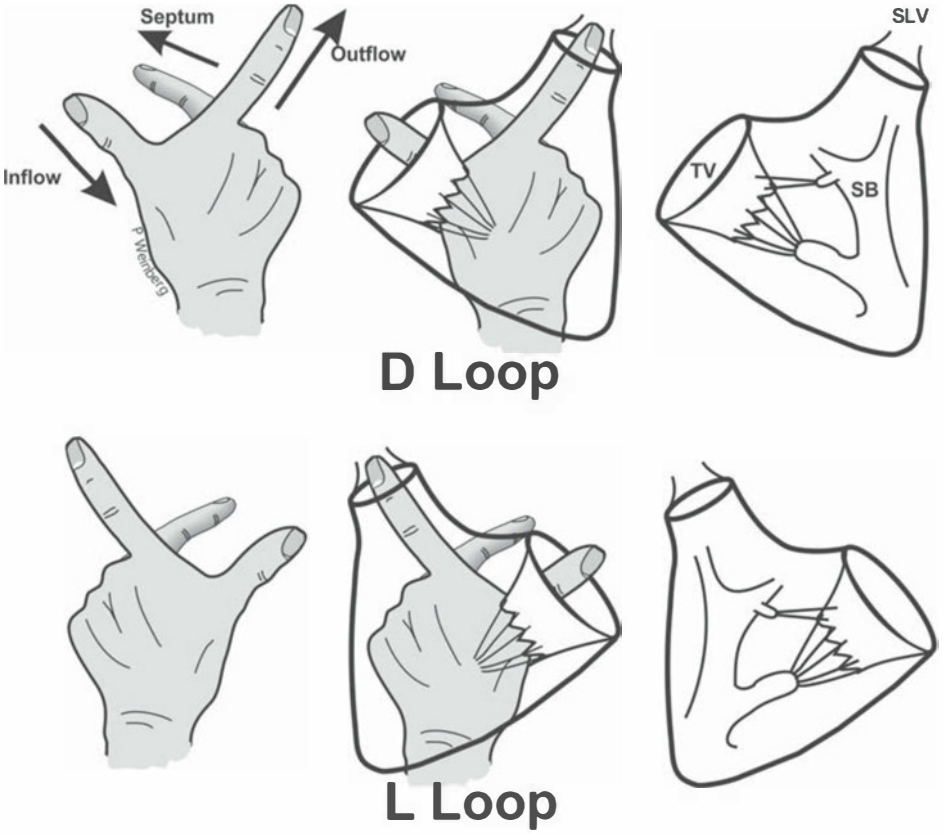


Fig. 71.2. Ventricular loop and handedness. Ventricular loop or situs is determined by the internal organization of the ventricles. Right ventricular organization shown schematically at the right is compared with the right hand; thumb, index finger, and middle finger held mutually orthogonally and representing ventricular inflow, outflow, and septum, respectively. D loop, right-hand organization of the right ventricle; L loop, left-hand organization of the right ventricle; SB, septal band; SLV, semilunar valve; TV, tricuspid valve.

reference is necessary. Just as it is possible to distinguish a right hand from a left hand without seeing the attachment to the rest of the body, that is, without a right–left frame of reference, so too it is possible to distinguish the two stereoisomers of ventricular organization. By focusing on ventricular inflow, outflow, and septum, it is possible to attribute chirality or handedness to the ventricular organization (Fig. 71.2). If one holds his hand with the thumb, index finger, and middle finger mutually orthogonally, with the thumb representing ventricular inflow, the index finger, outflow, and the middle finger pointing to the septum, a right hand in the morphologic right ventricle indicates a D loop arrangement of ventricles. If the left hand fits the right ventricle, there is an L loop. Just as a right hand can always be distinguished from a left, so too D loop ventricles should not be mistaken for L loop ventricles regardless of the position of the heart or ventricles in three-dimensional space. What if the right ventricle does not have all three components: inflow, outflow, and septum, for example, tricuspid atresia has no right ventricular inflow? The ventricles are always concordant with each other having adjacent inflows, adjacent outflows, and a septum common to both. Because of this a right-handed right ventricle is matched with a left-handed left ventricle. Similarly, a left-handed right ventricle would go with a right-handed left ventricle. Therefore, in the case of tricuspid atresia one would simply determine the handedness of the left ventricle and interpret the ventricular loop as just described.

Great artery situs refers to the spatial arrangement of the semilunar valves and great arteries. A special spatial arrangement exists when the great arteries are normally aligned with the ventricles (see intersegmental connections below). Thus, a special segmental description is given in those cases where the pulmonary artery arises above the right ventricle and the aorta normally above the left ventricle. If the two great arteries spiral about each other in a clockwise manner (viewed from the ventricles) so that the ascending aorta crosses the right pulmonary artery, this is designated solitus normal great arteries. If they spiral counterclockwise with the ascending aorta crossing the left pulmonary artery, they are termed inversus normal great arteries. For all other cases—transpositions and malpositions described under intersegmental connections—the great artery segment is called D if the aortic valve is to the right of the pulmonary valve, L if the aortic valve is to the left of the pulmonary, and A if aortic valve is directly anterior (i.e., same sagittal plane) to the pulmonary.

Segmental notation: The purpose of the segmental approach to diagnosis is not only to have designations for the various spatial arrangements of the cardiac structures but also to have a concise way of communicating the basic organization of the heart when also listing the various intersegmental connections and chamber, valve, and vessel abnormalities mentioned below. This shorthand description of the cardiac organization is the segmental notation. Van Praagh conceived of each heart as representing a subset of all the possible spatial arrangements that could occur in nature. He borrowed mathematical set notation—braces { }—to accomplish this. Each heart can be described as a three-member subset of the whole with the first member of the set representing viscero-atrial situs, the second, ventricular loop, and the third, great artery situs. Furthermore, the possibilities for each member of the set are designated by their first letter in capital form with the individual members separated by commas (Fig. 71.3). Therefore, the viscero atrial segment can be described by S, I, or A for situs solitus, inversus, or ambiguus. Similarly, the second member, ventricular loop, is represented by D or L for D loop or L loop organization of the ventricles. The great artery notation is S or I for solitus normal or inversus normal if the pulmonary artery arises above the right ventricle and aorta normally above the left ventricle, or D, L, or A for aortic valve to the right, left, or directly anterior relative to the pulmonary valve. In cases where the information is insufficient to determine the particular segment, that member of the subset may be represented by the letter X.

In the cases of normal arrangement of cardiac chambers and vessels, the segmental notation is {S,D,S} for viscero-atrial situs solitus, ventricular D loop, and solitus normal great arteries. These would also be the segments in classical tetralogy of Fallot, and the same segmental notation would apply to classical tricuspid atresia with normally aligned great arteries. Thus, it can be seen that the segmental description does not describe abnormalities of chambers, valves, or vessels nor does it describe

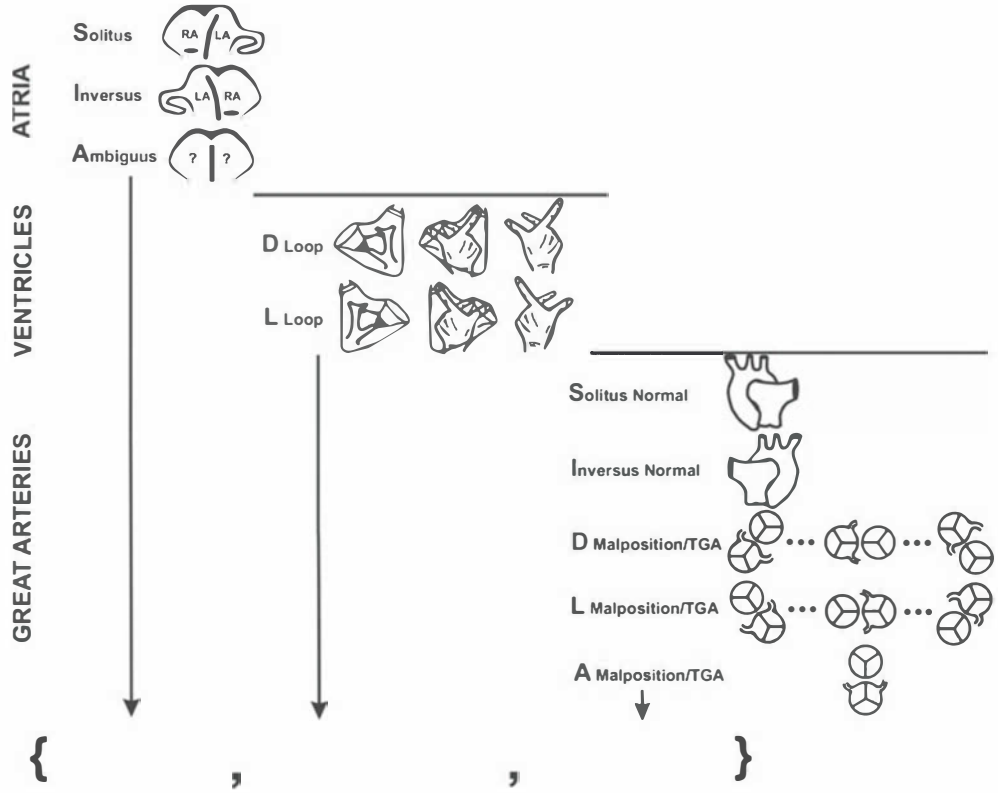


Fig. 71.3. Segmental notation. Segmental notation consists of a three-member set in braces { }. The first member is the viscero atrial situs—solitus, inversus, or ambiguus; the second is the ventricular loop—D or L; and the third is the great artery situs—solitus normal, inversus normal, D, L, or A transposition or malposition. The meaning of those terms is discussed in the text and is represented diagrammatically in the figure. Great artery situs diagrams with malposition or transposition of the great arteries in the lower right represent the two semilunar valves viewed from above; the aorta is shown with coronary arteries. LA, left atrium; RA, right atrium; ?, ambiguous morphology, a combination of solitus and inversus.

intersegmental connections (except for normally aligned great arteries). Classical complete transposition of the great arteries has segments {S,D,D}, since the atria and ventricles are normally arranged, but in the absence of normally aligned great arteries the segmental notation indicates that the aorta is to the right of the pulmonary artery at the level of the semilunar valves. The same segmental description could apply to a case of double outlet right ventricle with the aorta to the right of the pulmonary artery. Classical physiologically corrected transposition of the great arteries has segments {S,L,L} indicating normal (solitus) arrangement of the atria but a ventricular L loop plus aorta to the left of pulmonary artery. Transposition of the great arteries {S,D,L} has situs solitus of viscera and atria, ventricular D loop but the transposed aorta is to the left of the pulmonary artery, rather than to the right as in the {S,D,D} form. This difference in segments does not imply different connections but rather an altered spatial arrangement, which has implications for associated abnormalities (e.g., hypoplasia of the right ventricle, strad-

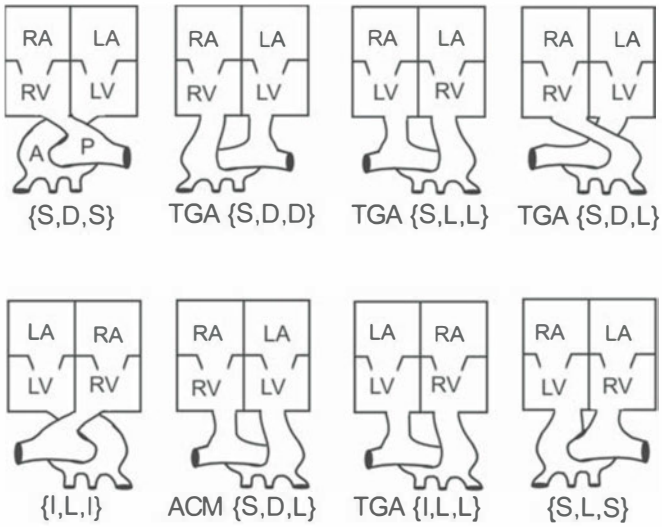


Fig. 71.4. Examples of selected anomalies and their segmental notations. Box diagrams of the heart showing the position of the atria—right atrium (RA) and left atrium (LA), the ventricles—right ventricle (RV) and left ventricle (LV), and the great arteries—aorta (A) and pulmonary artery (P). The upper left diagram represents normal spatial relations and the lower left, normal relations in situs inversus of viscera and atria. The three examples of transposition of the great arteries (TGA) in the upper row are discussed in the text. The case of anatomically corrected malposition (ACM) in the lower row has the aorta arising abnormally from a left-sided infundibulum above the left ventricle. Transposition of the great arteries {I,L,L}, physiologically complete transposition in situs inversus; {S,L,S}, isolated ventricular inversion: physiologically transposition but with normal ventriculo arterial alignment.

dling tricuspid valve) as well as for surgical approaches. Figure 71.4 shows examples of spatial arrangements of atria, ventricles, and great arteries with their corresponding segmental notation. The segmental designation is a fundamental part of the overall cardiac diagnosis and is not meant to be a parenthetical, that is, extraneous or explanatory, supplement. Thus, it is *incorrect* to substitute parentheses () or square brackets [] for the curly braces {} used in mathematical set notation.

INTERSEGMENTAL ALIGNMENTS

Each pair of adjacent segments has a discrete set of intersegmental alignments. Under the principle of *economy* described above, intersegmental alignment diagnoses are mutually exclusive entities.

Atrioventricular alignments are really a combination of atrial-to-AVV connection and AVV-to-ventricle connection. There are four basic types of *atrial-to-AVV connection*: two AVVs, one from each atrium; one common AVV from both atria; right AVV atresia, that is, no AVV from the right-sided atrium but one AVV from the left-sided atrium; and left AVV atresia. Each atrial-to-AVV connection has several possible *AVV-to-ventricle connections:* with two AVVs, each may enter one ventricle (normal), one or both may enter two ventricles (straddling AVV[s]), or both may enter one ventricle (double inlet ventricle) (Fig. 71.5); with a common AVV, it may connect to both ventricles equally (balanced), unequally (unbalanced), or only one ventricle (common inlet) (Fig. 71.6); with AVV atresia, the other valve may enter the ipsilateral ventricle, the contralateral ventricle, or both ventricles (straddling AVV) (Fig. 71.7).

Ventriculo arterial alignment names do not necessarily reflect anatomical reality! The ventricles are connected to the great arteries via the infundibulum or AV canal. (The normal right ventricle is connected to the pulmonary artery via the infundibulum, while the left ventricle is connected to the aorta via the AV canal, hence mitral–aortic fibrous continuity.) However, unlike the atrioventricular alignments, most classification systems have not used this relationship in developing their nomenclatures. Instead, ventriculo arterial alignments tend to be based on a system in which each great artery is "assigned" to one, and only one, ventricle. When the ventricular septum is intact, the assignment is straightforward. In the presence of the commonly seen relatively large VSD near the great arteries, the assignment can be problematic. All such classification schemes have, therefore, adopted certain rules governing assignment of great arteries to ventricles. We use the rules shown pictographically in Figure 71.8 that can be stated as follows: (1) The pulmonary artery is assigned to a ventricle if the pulmonary valve lies more than 50% above it. (2) Since the aorta *normally* may lie predominantly above the ventricular septum rather than above the left ventricular cavity, mitral valve-to-aortic valve fibrous continuity, if present, determines assignment of the aorta to the left ventricle, regardless of how far the aorta extends above the right ventricle in the setting of a large VSD. Mitral valve is defined as that valve on the side of the left ventricle or its remnant (compared with the right ventricle or its remnant). For the purposes of assigning aorta to a ventricle, obviously, if there is only one ventricle and no remnant of the other, the point is moot. (3) If mitral–aortic fibrous continuity is not present, the 50% rule, as described for the pulmonary artery above, applies. (4) With conotruncal anomalies and pulmonary atresia, it may not be feasible to assign the pulmonary artery to either ventricle; in those cases only the aorta is assigned: right ventricular aorta with pulmonary atresia or left ventricular aorta with pulmonary atresia. Using these rules, the ventriculoarterial alignments are named according to Figure 71.8.

As with any system of more or less arbitrary rules, there are potential problems or "exceptions." Here are a few. If there is tenuous mitral–aortic fibrous continuity through a tiny VSD so that the aorta arises virtually exclusively above the right ventricle, it is assigned to the right ventricle. If there is no mitral valve but a common AVV instead, the above rules apply to the portion of the common AVV attached to the left ventricle, as if it were a mitral valve. Finally, if there is no mitral valve due to AVV atresia, the 50% rule applies but the aorta, if assigned to the left ventricle is considered normally aligned unless there is clearly subaortic infundibular muscle. Thus, typical hypoplastic left heart with mitral atresia is considered to have normally aligned great arteries. In the case of left ventricular aorta with pulmonary atresia, as long as there is mitral to aortic fibrous continuity solitus normal—S—or inversus normal—I—great arteries are considered appropriate designations.

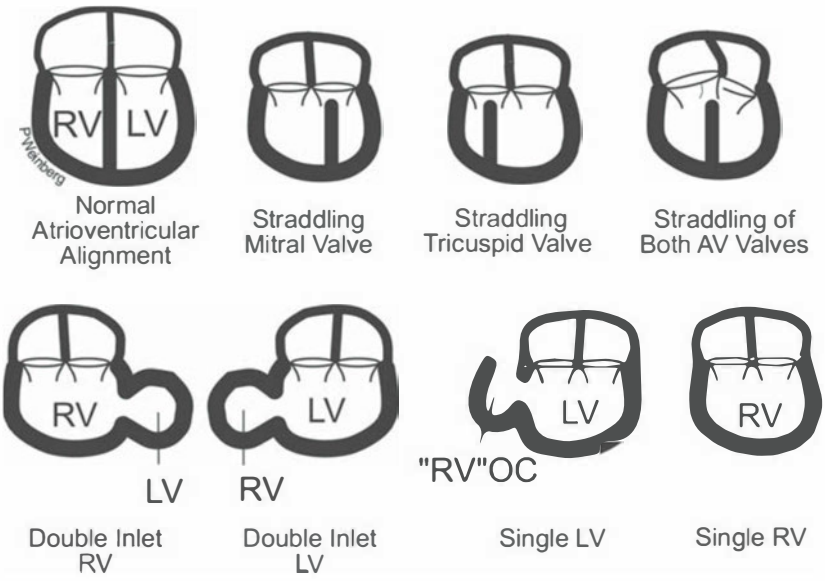


Fig. 71.5. Atrioventricular alignments with two atrioventricular valves. All possible ways for two atrioventricular valves, one from each atrium, to connect with ventricles. (Shown here only for D loop ventricles; comparable connections for L loops exist, keeping in mind that the tricuspid valve is left sided and the mitral right sided. Similarly, the right ventricular outlet chamber ["RV"OC], not a ventricular sinus, is left-sided in an L loop.) LV, left ventricle; RV, right ventricle.

Chamber, Valve, and Vessel Abnormalities

An exhaustive description of all chamber, valve, and vessel abnormalities is not within the scope of this chapter. The following sections indicate the characteristics that define the various structures and list some of the surgically important anatomical abnormalities.

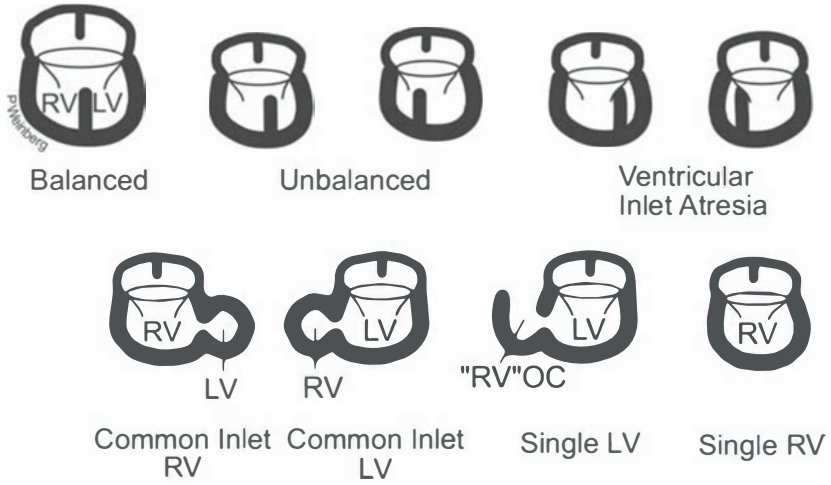


Fig. 71.6. Atrioventricular alignments with a single, common atrioventricular valve. All possible ways for a common atrioventricular valve, one valve from both atria, to connect with ventricles. (Shown here only for D loop ventricles; comparable connections for L loops exist. Right ventricular outlet chamber ["RV"OC], not a ventricular sinus, is left-sided in an L loop.) In ventricular inlet atresias, a leaflet of the common atrioventricular valve, seals off the would-be "septal defect," the only entrance into the ventricle. LV, left ventricle; RV, right ventricle.

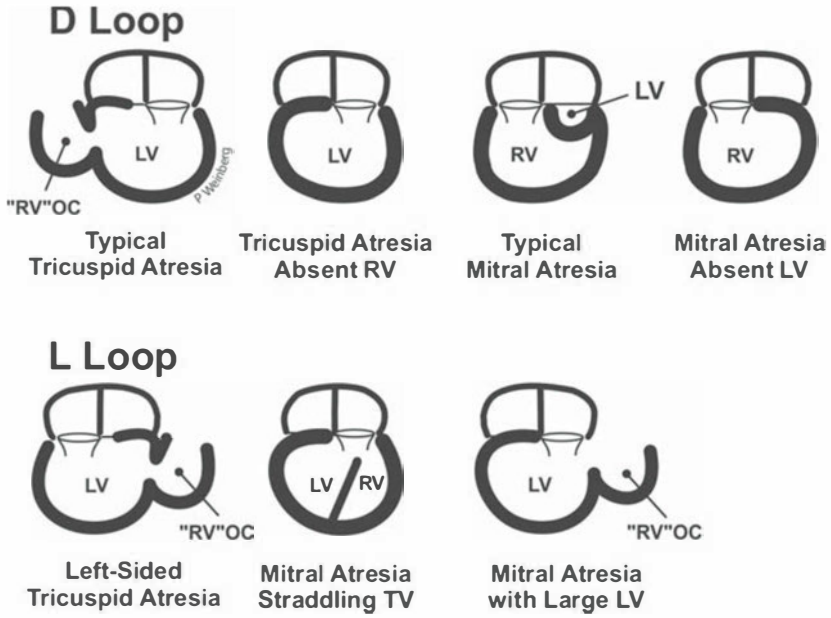


Fig. 71.7. Atrioventricular alignments with right or left atrioventricular valve atresia. Several possible ways for the valve contralateral to the atretic one, to connect with ventricles. While other segmental combinations may exist, some, such as tricuspid atresia with straddling mitral valve, have not been reported. (Reports in the literature with that diagnosis are actually examples of right-sided mitral atresia with straddling left-sided tricuspid valve based on ventricular loop and the association of canal type/inlet ventricular septal defect only with straddling tricuspid valve.) LV, left ventricle; RV, right ventricle; "RV"OC, right ventricular outlet chamber.

Venae cavae and coronary sinus: The superior vena cava (SVC) connects veins from the upper portion of the body to the heart. Normally, there is a single, right-sided SVC that enters the right atrium, but single SVCs can connect to a coronary sinus (CoS) or to the left atrium. Bilateral SVCs may be present, most commonly with a right SVC connecting to the right atrium and a left SVC connecting to a CoS and ultimately draining to the right atrium. However, each may connect to its ipsilateral atrium as in heterotaxy, or rarely, both may connect to the same atrium. A venous structure that is in a similar position to the SVC but connects a pulmonary vein confluence to the innominate vein is not considered to be an SVC because it does not connect to the heart. Rather, the term vertical vein or connecting vein is appropriate.

The IVC enters the right atrium guarded medially by the left venous valve, which blends with the septum primum portion of the atrial septum, and laterally by the right venous (Eustachian) valve. The anterior commissure of these two venous valves forms the tendon of Todaro, which points to the AV node. The IVC is almost always single at the level of the heart. (Double IVC refers only to duplication of the infrarenal portion.) While two IVC-like vessels may enter the heart either in the same atrium or one in each atrium, one vessel almost invariably enters only hepatic veins. Because the IVC is nearly always single at atrial level, it is used as a strong indicator of the right atrium (see below). However, there are two potentially ambiguous situations that render this unacceptable for all cases: the so-called absent IVC and an IVC which enters both atria via a so-called sinus venosus-type atrial septal defect (ASD) of the inferior caval variety. In absent IVC, the renal-to-hepatic portion of the IVC is absent so that the veins of the lower portion of the body (except the hepatic veins) are drained by the azygos vein to a SVC. Only the hepatic veins connect directly to the right atrium. In the IVC type of sinus venosus ASD, the right atrium cannot be defined by IVC entrance since it straddles the atrial septum.

The CoS usually enters the right atrium near the entrance of the IVC (Fig. 71.9). Its entrance is partially covered by the Thebesian valve that may be an extension of the Eustachian valve or a separate structure. The AV node may extend into the mouth of the CoS. If there is SVC connection to the CoS, its orifice will be quite large and could be mistaken for an ASD or occasionally for a common atrium. A large CoS ostium may also occur with anomalous connection of pulmonary veins or, rarely, hepatic veins to the CoS. Uncommonly, the CoS ostium may enter the left atrium and rarely there may be no patent CoS ostium, in which case the CoS drains into an SVC remnant (vein of Marshall), which drains blood superiorly to the innominate vein. Surgical or catheter device occlusion of the vein of Marshall in this situation results in coronary venous obstruction.

The morphological *right atrium* is characterized by a broad-based appendage which, on the exterior, blends almost imperceptibly with the venous portion of the atrium. Anderson has pointed out that this is more definitively seen from the interior

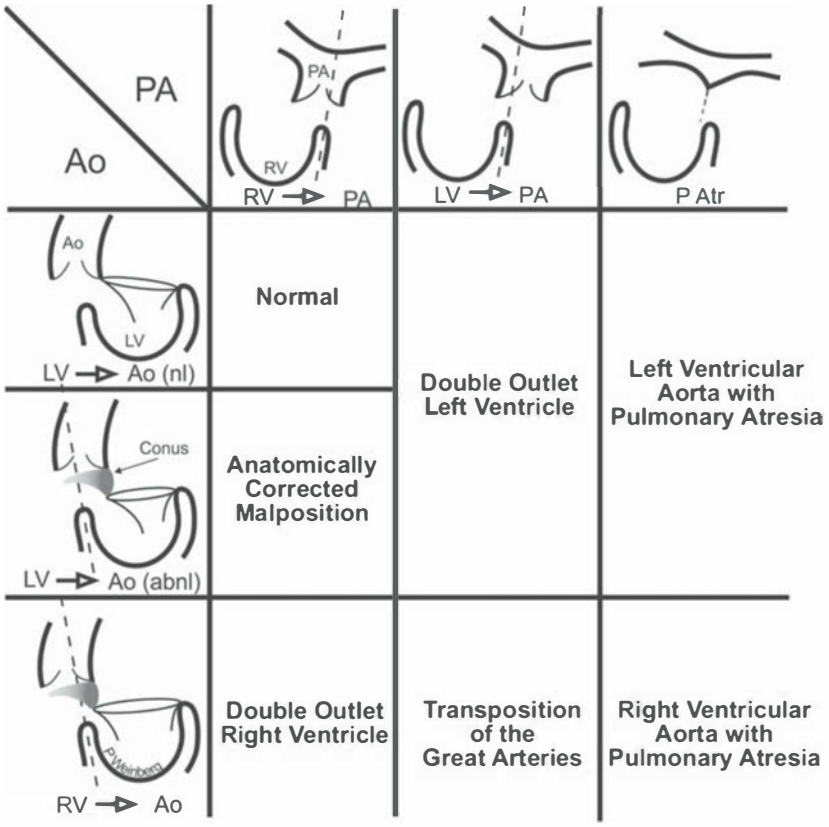


Fig. 71.8. Ventriculo arterial alignments. The rules for assigning great arteries to ventricles discussed in the text are shown diagrammatically. For the pulmonary artery across the top row, the pulmonary artery is said to arise from the right ventricle if its valve is more than 50% above the right ventricle. It is assigned to the left ventricle if more than 50% above that. If there is pulmonary atresia (P Atr) without clear evidence of association with either ventricle, no assignment is made (far *right column*). For the aorta (Ao), the rules are depicted in the far left column. The Ao is assigned to the LV in a normal fashion (Ao (nl)) if there is mitral valve-to-aortic valve fibrous continuity, regardless of the degree to which the aorta may be situated above the right ventricle. If there is no mitral-aortic fibrous continuity such as occurs with subaorticconus, the aorta is assigned to the left ventricle but as an abnormal alignment (Ao [abnl]) if it arises more than 50% above left ventricle, or is assigned to right ventricle if more than 50% above that. Note that mitral aortic discontinuity does not necessarily imply the presence of subaorticconus, since there could be tricuspid valve–aortic valve fibrous continuity. The ventriculo-arterial alignments for each of the combinations of aortic and pulmonary assignment are given in the corresponding boxes.

aspect as pectinate muscles extending all the way to the crux of the heart. The fact that pectinate muscles comprise a large portion of the effective right atrial cavity makes that chamber more compliant than the left atrium whose appendage is more isolated from the main cavity. Septum secundum or the superior limbic band of the atrial septum is seen in front of septum primum or the flap valve of the foramen ovale from the right atrial aspect. Typically, the IVC enters the right atrium with the exception of those situations noted above under *Venae cavae*. The SVC and CoS, being much more variable, cannot be used to define the right atrium. When the SVC does enter the right atrium, the crista terminalis forms the lateral aspect of the entrance internally. The external aspect of this structure is the sulcus terminalis in which lies the sinus node.

The *atrial septum* has three portions (Fig. 71.9), most easily appreciated in the newborn but still identifiable in the adult heart: septum primum, septum secundum, and canal septum. The septum primum or flap valve of the foramen ovale is the thin central portion of the septum. In the fetus and preterm infant, it is so thin as to be transparent. At term, it is typically translucent and becomes somewhat thicker with advancing age but is always thinner than the other two components of the septum. It is this structure that is torn by balloon septostomy. Defects in or deficiency of septum primum are called ostium secundum ASDs. Septum secundum, or the superior limbic band, is a thick muscular ridge at the superior aspect of the atrial septum immediately medial to the SVC (if one enters the atrium directly). The septum primum usually has two points of attachment onto the left atrial side of septum secundum with the space between the two being the foramen ovale. The superior aspect of septum primum (between the two points of attachment) forms a half-moon shape. The third portion of the atrial septum is the atrioventricular canal septum, a thick muscular tissue anterior to septum primum extending to the level of the AVVs and actually continuing on to become the canal or the so-called inlet portion of the ventricular septum. It is the canal septum that is deficient or absent in cases of common atrioventricular canal including the ostium primum ASD. Other apparent ASD are actually vessel ostia. Sinus venosus atrial septal defects are examples of either right pulmonary vein straddling atrial septum and/or a vena cava straddling the septum. CoS ostium-type ASD occurs when the CoS is unroofed into the left atrium. ASD types are shown in Figure 71.10.

By definition, the *tricuspid valve* is the right-sided AVV in a D loop or the left-sided AVV in an L loop. Because of the marked variability in morphology of the AVVs and the propensity for AVV morphology to be influenced by the ventricle into which they enter, the above definition is used to avoid ambiguity in such circumstances as double inlet ventricles in which both AVVs may appear morphologically similar. Having thus defined tricuspid valve, it is nonetheless worth describing the commonly seen morphology. The classical three leaflets are anterior, posterior, and septal. While the name implies three leaflets, this is not always the case. Frequently, the newborn has only two distinct leaflets with no commissure between the anterior and posterior leaflets. The most characteristic feature of the tricuspid valve is the similarity of depth (annulus-to-free edge) of all leaflets.

The morphological *right ventricle* has broad or coarse and relatively parallel trabeculations. If an AVV is present, there are chordal attachments onto the face (not simply the crest) of the ventricular septum as well as onto the free wall. There is typically a single large papillary muscle attached to the septal band by the moderator band.

Ventricular septum: The right ventricular septal surface, like the rest of the right ventricle, shows coarse, parallel trabeculations. A large trabeculation called the septal band appears like an appliqué on top of the septum. At its inferior aspect, the septal band

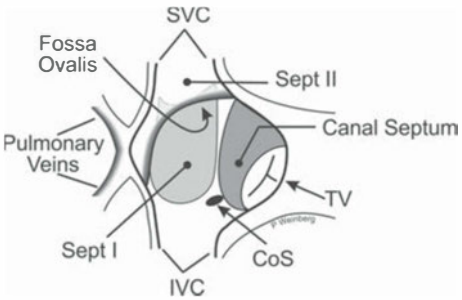


Fig. 71.9. Right atrial septal surface. The opened right atrium viewed in anatomical position from the right lateral aspect shows the various components of the atrial septum. CoS, coronary sinus; IVC, inferior vena cava; Sept I, septum primum, the flap valve of the foramen ovale; Sept II, septum secundum, the superior limbic band; SVC, superior vena cava; TV, tricuspid valve.

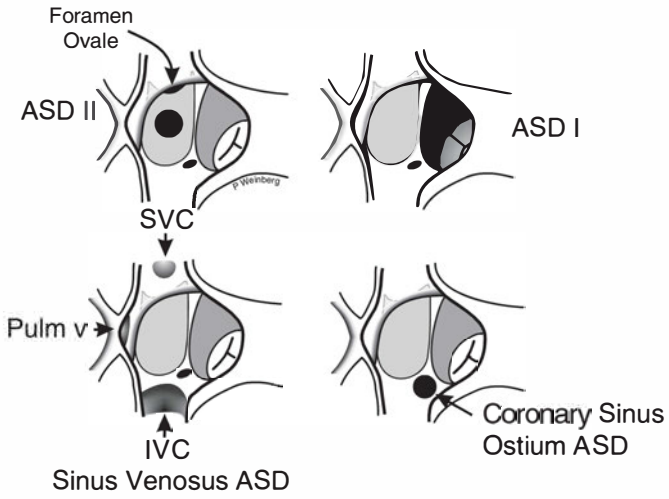


Fig. 71.10. Atrial septal defects. Diagrams of the four major types of atrial septal defect in the same format as in Figure 71.9. The defects are shown in *black*. The ostium secundum atrial septal defect (ASD II) is a defect in septum primum. The ostium primum atrial septal defect (ASD I) is a failure of formation of the canal septum. The sinus venosus type has three varieties: the superior vena cava type where the superior vena cava straddles the atrial septum, the pulmonary vein type where one or more right pulmonary veins straddle, and the inferior vena cava type where the inferior vena cava straddles. Pulmonary vein and superior vena cava types are frequently combined into a single defect. The coronary sinus ostium atrial septal defect is a communication between the atria when the coronary sinus is partially or completely unroofed.

becomes the moderator band and that, in turn, the main papillary muscle of the right ventricle. The superior portion of the septal band has the shape of the letter "Y." The papillary muscle of the conus or muscle of Lancisi is usually at or slightly posterior to the fork of the "Y." The infundibular septum normally fills the "Y" of the septal band. However, in cases of infundibular (conal) septal malalignment or hypoplasia the "Y" is empty, that is, there is a VSD in its place, and the infundibular septum is not part of the ventricular septum. The right bundle branch runs along the septal band between the upper two-thirds and the lower one-third, and extends onto the moderator band to the base of the main papillary muscle. The septal–moderator band complex marks the boundary between the sinus portion of the right ventricle below and the outflow tract above. Pathological extension of the septal-moderator band complex can divide the two, resulting in subpulmonary stenosis—anomalous muscle bundle or the so-called double-chambered right ventricle.

The left ventricular septal surface is smooth over the one-half to two-thirds basilar portion and has fine interdigitating trabeculations over the apical one-third to one-half. The left bundle branch penetrates the septum from the right ventricular side and divides into an anterior and a posterior fascicle, which run along the smooth portion of the left ventricular septal surface before traveling around the ventricular free wall or across the left ventricular cavity to the bases of the two left ventricular papillary muscles.

One interesting aspect of the ventricular septum is that despite what the name implies, it is not necessarily located entirely between the ventricular cavities. In cases where one ventricular sinus is hypoplastic (e.g., aortic atresia or pulmonary atresia with intact ventricular septum) or absent (e.g., tricuspid atresia, double-inlet left ventricle), a structure with the morphology of the ventricular septum lies between one ventricle and either an outlet chamber or the exterior of the heart. Hence, ventricular septum does not always imply two well-developed ventricular sinuses. Thus, a defect in that structure is a VSD even if the two chambers joined are not full fledged ventricles, as in double-inlet (single) left ventricle with right ventricular outlet chamber.

VSDs occur in five morphological types based on location relative to landmarks in the ventricular septum, most easily recognized from the right ventricular septal surface (Fig. 71.11). This can be thought of as the "Geographical" classification. The key landmarks are the septal band, the muscular or trabecular septum, the infundibular septum, and the annulus of the tricuspid valve. Conoventricular VSDs are located between the conal (infundibular) septum and the muscular or trabecular septum but with the conal septum still situated in the "Y" of septal band. This is in the general location of the membranous ventricular septum but is often larger than the 1 to 3 mm diameter of the normal membranous septum. It can be separated from the membranous septum by a small amount of muscle. This type of VSD is frequently partially covered by a *portion* of the septal leaflet of the tricuspid valve. Conal septal malalignment VSDs, or malalignment for short, are located *in* the "Y" of the septal band because the infundibular septum is displaced out of the "Y." The AV canal-type VSD, also called inlet VSD is located beneath the *entire* length of the AVV (either tricuspid or common AVV) that opposes or intersects the ventricular septum. The defect comes up to the valve, that is, there is no muscle between the defect and the valve. (VSDs with interposed muscle between defect and AVV are considered posterior muscular; see below.) Conal septal hypoplasia VSDs are, like malalignment defects, located in the "Y" of septal band, but without malalignment. Any remnant of the infundibular septum is normally aligned with the septal band, but there is actual deficiency of the infundibular septum. All other VSDs are termed muscular. Muscular VSDs are located in the muscular or trabecular septum in any of the following general areas: postero inferior to main portion of septal band, midmuscular; inferior aspect of muscular septum, "inferior muscular"; beneath septal leaflet of tricuspid valve but with muscle bar separating defect from valve annulus, posterior muscular; distal to the moderator band, apical muscular; anterior to main portion of septal band, anterior muscular. Muscular VSDs often have

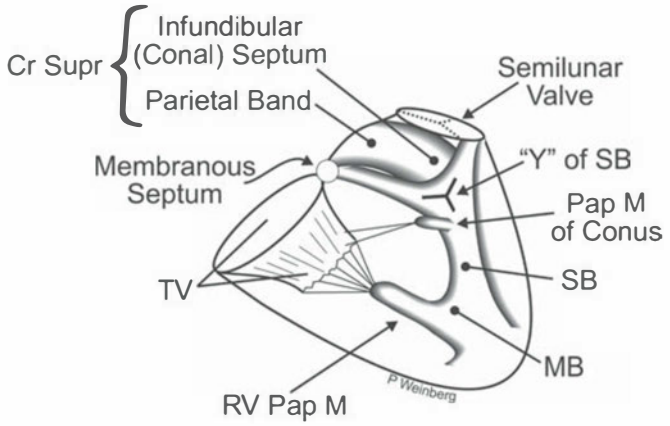


Fig. 71.11. Right ventricular septal surface. The opened right ventricle viewed in anatomical position from a right anterior oblique aspect shows the landmarks used to describe the location of ventricular septal defects. Cr Supr, crista supraventricularis, which is the composite of the infundibular (conal) septum between the two outflows and the parietal band, which, if present, forms part of the right ventricular free wall; MB, moderator band; Pap M of Conus, papillary muscle of the conus/muscle of Lancisi; RV Pap M, main papillary muscle of the right ventricle; SB, septal band; TV, tricuspid valve.

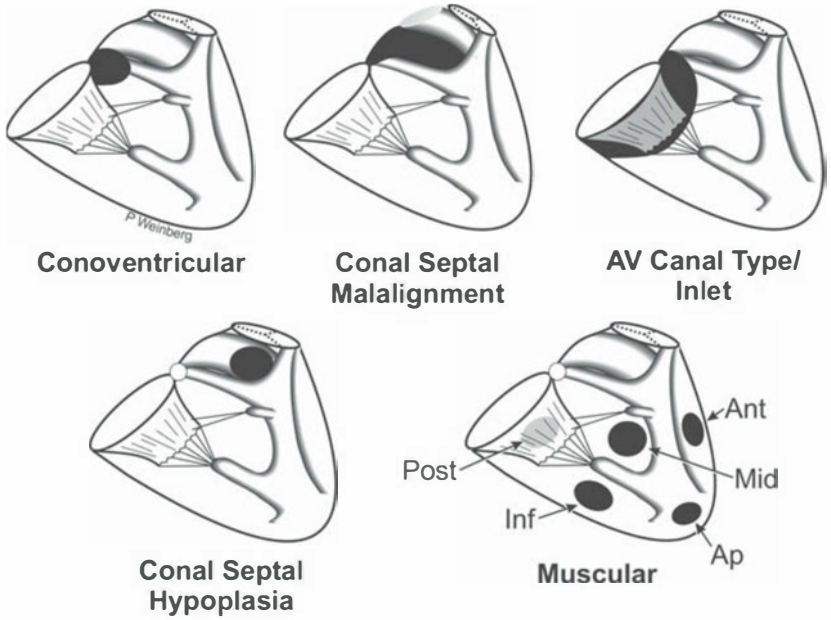


Fig. 71.12. Ventricular septal defects. Diagrams of the five major types of ventricular septal defect in the same format as in Figure 71.11. Ant, anterior muscular VSD; Ap, apical muscular VSD; Inf, inferior muscular VSD; Mid, midmuscular VSD; Post, posterior muscular VSD.

the appearance of multiple defects when viewed from the right ventricle, or when viewing angiography or color Doppler, due to the lattice-like trabeculations of the right ventricular septal surface. However, such defects are usually single on the left ventricular side. Thus, the term multiple VSDs could mean multiple streams from a single muscular VSD or multiple VSD types. This should be specified whenever the term "multiple VSDs" is used. The five main types of VSDs are shown in (Fig. 71.12). In addition to these five main types, there are combinations of some of the above: malalignment–conal septal hypoplasia, malalignment–canal type, malalignment–canal-type-conal septal hypoplasia, conoventricular–muscular.

In keeping with the principle of accuracy—independence from context—the definitions of the various VSDs are uninfluenced by associated conotruncal or other malformations. This is why the terms subpulmonary, subaortic, beneath both (doubly committed), or beneath neither (uncommitted) are not acceptable by themselves, because a subaortic defect with normally aligned great arteries is in the same anatomical location as a subpulmonary defect with transposition of the great arteries. (These terms may be used in a supplementary manner with an anatomical term to specify great artery location in double outlet right or left ventricles.)

There are other classifications of VSDs beside the geographical approach. One is the "Borders" approach popularized by Anderson. This approach emphasizes the constitution of the borders over the location within the septum. Thus, most VSDs are described as perimembranous, subarterial, or muscular if a border of the VSD is made up by the area of tricuspid valve–aortic valve fibrous continuity, one or both semilunar valves, or only muscle, respectively. It is important to understand that these two approaches are not simply different but equivalent names. Rather, they represent different frames of reference, so that perimembranous VSDs include all canal type or inlet VSDs, some, but not all conoventricular VSDs (some are muscular), some but not all malalignment VSDs, and some but not all conal septal hypoplasia VSDs. In the geographical approach, the borders are mentioned as descriptions but are not the primary elements of classification. Similarly, in the Borders approach, secondary terms such as perimembranous inlet, perimembranous outlet are used, although we believe this is contrary to the goal of economy.

The *pulmonary valve* is normally a trileaflet valve with the infundibular septal commissure facing the aortic valve and all three commissures equally spaced around the valve's circumference. Pulmonary valvar stenosis, not part of a conotruncal anomaly, is typically a domed valve with all three commissures rudimentary and a central orifice, whereas pulmonary stenosis with a conotruncal anomaly is usually bicuspid or unicuspid with at least a rudimentary septal commissure.

The *pulmonary arteries* refer to the main and proximal right and left branch pulmonary arteries. These, in turn, supply the peripheral pulmonary arteries. The morphological right pulmonary artery courses anterior to the right mainstem, eparterial, bronchus. The morphological left pulmonary artery crosses over the left mainstem (hyparterial) bronchus. Absent right or left pulmonary artery means absence of the proximal pulmonary artery, not the entire pulmonary arterial tree. In such cases, the peripheral pulmonary artery may be supplied by a ductus arteriosus,

bronchial artery, other systemic collateral vessel, or, rarely, a coronary artery. True pulmonary arteries always arise anterior to the tracheo bronchial tree. Therefore, a confluence behind the trachea is always some form of systemic collateral vessel.

Pulmonary veins, numbering anywhere from three to five, drain blood from the lungs and normally connect to the morphological left atrium. Any other connection is considered "anomalous pulmonary venous connection," regardless of the ultimate destination of the pulmonary venous blood. Hence, partial or total anomalous pulmonary venous connections are anatomical terms, whereas partial or total anomalous pulmonary venous return are physiological diagnoses, describing the destination of the blood, which may or may not be concordant with their anatomical counterparts.

The morphological *left atrium* is best characterized by a discreet, narrow-based appendage, distinct from the venous portion. Pectinate muscles, being confined to the appendage portion, do not come near the crux of the heart as do the pectinate muscles in the morphological right atrium. As mentioned previously, the absence of the accordion-like pectinate muscles from the main cavity of the left atrium renders it less compliant than the morphological right atrium. This has implications for pressure changes engendered by AVV regurgitation, especially that which occurs acutely.

The *mitral valve* is defined as the left AVV in a D-loop heart and the right AVV in an L-loop. The rationale for this approach is described above under tricuspid valve. The usual mitral valve morphology is a bileaflet valve with the anterior leaflet deeper (from annulus to leaflet edge) than the posterior or mural leaflet. The anterior leaflet is shorter in circumferential length than the posterior leaflet.

The morphological *left ventricle* is characterized by fine, interdigitating trabeculations, and absence of AVV chordal attachments to the ventricular septal surface. The left ventricle usually has two papillary muscles—anterolateral and posteromedial—arising from the ventricular-free wall. The left ventricle does not usually have an infundibulum; however, this is not by the way of definition. That is, the left ventricle, as defined, can give rise to the infundibulum. It must be emphasized that a left ventricular infundibulum, if present, is on the same side of the ventricular septum as the left ventricular sinus. When the infundibulum is on the opposite side of the ventricular septum as described above under "ventricular septum," it must be presumed to be a *right* ventricular infundibulum.

The *aortic valve*, like the pulmonary valve described above, is normally a trileaflet valve with evenly spaced commissures. Valvar stenosis is almost always due to a bicuspid or unicuspid valve with one or two rudimentary commissures, respectively. Unicuspid valves have a funnel shape with an eccentric orifice. The commissure between the left coronary and noncoronary cusps is nearly always well developed. Rarely, aortic stenosis can be from a dysplastic tricuspid aortic valve.

The *aorta* arises above the aortic valve and supplies at least one of the brachiocephalic vessels. Unlike the pulmonary artery that may be atretic or even absent, the ascending aorta is virtually always patent down to the level of the valve even if the valve is atretic, because it carries at least coronary blood flow. The aortic arch is said to be left or right according to which bronchus it crosses over, regardless of the side of its origin from the heart. However, arch sidedness can be inferred very reliably from the fact that the first arch vessel usually contains the carotid artery opposite the side of the arch. However, rare exceptions necessitate more complete evaluation such as MRI or CT scan if arch sidedness is crucial to be known prior to surgery.

There are three main *coronary arteries*—right (ventricular), anterior descending, and (left ventricular) circumflex—and one minor coronary—infundibular or conal branch. The coronaries follow the ventricles. Thus, the right ventricular coronary can be right- or left-sided just as the right ventricle can be. Normally, the anterior descending and circumflex coronaries arise from a single vessel named left (ventricular) main coronary artery. Similarly, the right and conal branch arise from a single vessel, the right coronary. A common variant is for the right and conal branch to arise from separate orifices from the same sinus of valsalva. However, other combinations are possible, particularly with conotruncal anomalies in which the spatial relationship between the aorta and the ventricles differs from normal. Hence, the anterior descending can arise from the conal branch of the right coronary as in some cases of tetralogy of Fallot, or the right and the (left ventricular) circumflex can arise from one vessel and the anterior descending from the other, as occurs in about one-quarter of D-transposition of the great arteries cases.

Conotruncal anomalies are defects involving the outflow tract (infundibular) portion of the heart, including the infundibular septum, the proximal great arteries, and the semilunar valves (Fig. 71.13). These anomalies are determined by the relative amounts of infundibular (conal) muscle beneath each

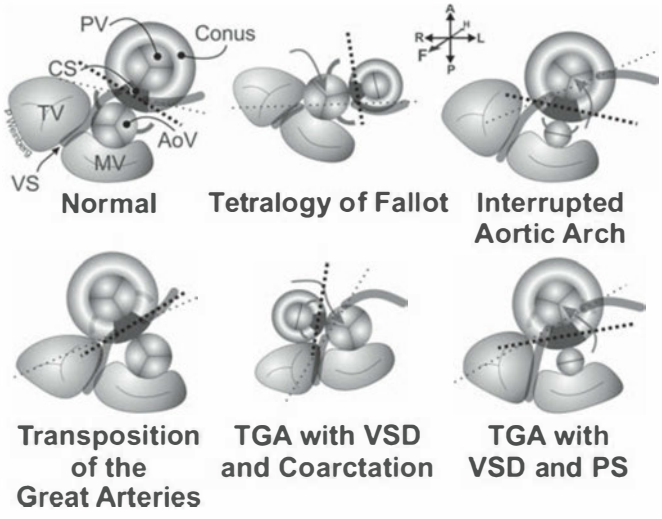


Fig. 71.13. Conotruncal anomalies. Diagrammatic representation of normal conotruncus and representative conotruncal anomalies as if viewed from the ventricles looking back toward the tricuspid valve and mitral valve and forward toward the two outflows. The aortic valve is indicated by accompanying coronary arteries. A coarse dotted line shows the plane of the infundibular or conal septum for comparison with a fine dotted line indicating the plane of the ventricular septum. Note the leftward anterior deviation of the conal septum in tetralogy of Fallot relative to normal, the rightward anterior deviation in transposition of the great arteries with ventricular septal defect and coarctation, the posterior leftward deviation in interrupted aortic arch, and the posterior and rightward deviation in transposition of the great arteries with ventricular septal defect and subpulmonary stenosis. The large arrows represent the malalignment ventricular septal defects associated with displacement of the infundibular or conal septum relative to the "Y" of septal band indicated by the gap in the VS. PV, pulmonary valve. Orientation directions: A, anterior; F, feet; H, head; L, left; P, posterior; R, right.

semilunar valve *plus* the degree of expansion of each outflow tract. The normal conotruncus has subpulmonary but not subaorticconus with full expansion of both outflow tracts. A poorly expanded subpulmonary conus with no subaorticconus is the tetralogy conotruncus. A normally expanded subpulmonary conus with poorly expanded subaortic area without subaortic conus is the interrupted aortic arch conotruncus. Subaortic conus without subpulmonary conus is typically seen with transposition of the great arteries. When the subaortic conus is poorly expanded, the conotruncus is that seen in transposition-VSD-coarctation. With poorly expanded subpulmonary area in the setting of subaortic conus only, the conotruncus is characteristic of transposition-VSD-subpulmonary stenosis. Other conotruncal anomalies include bilateral conus with or without outflow obstruction and bilaterally deficient conus.

It is important to recognize the difference between conotruncal anomalies and ventriculo arterial alignments. There are three factors that determine ventriculoarterial alignment: the relative amounts of subsemilunar infundibulum (conus)—the conotruncal anomaly, the association of infundibulum with one or both ventricles, and ventricular size. A subarterial conus carries the corresponding great artery anterior and superior, which tends to place it above the right ventricle. However, association of the infundibulum with left ventricle and/or hypoplasia of the right ventricle can vitiate the above tendency and result in association of the great artery in question with the left ventricle. Similarly, hypoplasia of the left ventricle can result in association of either or both great arteries with the right ventricle, irrespective of subvalvar conus. Thus, nearly any conotruncus can be seen with nearly any ventriculo arterial alignment. Remember that the conotruncal anomaly is anatomical fact, whereas the ventriculo arterial alignment is an intellectual construct to describe the proximity of each great artery to a ventricle.

Cardiac Malpositions

The heart is normally located within the thorax, in the mediastinum, extending predominantly into the left chest, although it crosses the midline slightly into the right chest. Locations outside this usual one are called cardiac malpositions. They fall into two broad categories: intrathoracic and extrathoracic. Intrathoracic malpositions include dextrocardia, heart predominantly in the right chest; mesocardia, predominantly midline; extreme levocardia, entirely in the left chest, that is, more leftward than normal; absent pericardium, in which part or all of the heart lies in an abnormal position within one of the chest cavities; and pericardo diaphragmatic defect, in which the heart is raised up above herniated liver, as if the heart were perched on a "pitcher's mound." Extracardiac malpositions include ectopia cordis and thoracopagus conjoined twins.

Syntax for Cardiac Diagnoses

The convention for listing diagnoses is to use the following order:

Cardiac syndromes (e.g., hypoplastic left heart syndrome, heterotaxy syndrome with asplenia).

Cardiac malpositions (dextrocardia).

Atrioventricular alignment abnormalities.

Ventriculo arterial alignment abnormalities.

Segmental notation.

Chamber, valve, and vessel abnormalities beginning at the arterial end of the heart and proceeding to the venous end.

Arrhythmias and physiological diagnoses.

Surgical diagnoses and residua in chronological order.

For example, one case showing all of the above headings could be "heterotaxy syndrome with asplenia, dextrocardia, common atrioventricular canal, double-outlet right ventricle {A,D,D} with mirror image right aortic arch, pulmonary stenosis, total anomalous pulmonary venous connection to the ductus venosus, persistent left SVC to left-sided atrium, ectopic atrial rhythm, status post-modified (polytetrafluoroethylene tube graft) left Blalock–Taussig shunt, status post-Fontan operation with residual left pulmonary artery stenosis." In cases where some of the above headings are normal, those are simply omitted. For example, one could have "ventricular septal defect and persistent left superior vena cava to coronary sinus," or "tricuspid atresia with ventricular septal defect and pulmonary stenosis." One must always include the segmental notation if there is a ventriculo arterial alignment abnormality. It is also useful to include the segmental notation when there is a cardiac malposition even if the segments are normal, because the segmental description clarifies the type of malposition instead of using additional, and frequently ambiguous, descriptions such as dextroposition and dextroversion.

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EDITOR'S COMMENTS

Complex forms of congenital heart disease are being seen with increasing frequency in most centers for congenital cardiac surgery in part because the relative success with surgical palliative interventions for congenital heart disease has improved and surgical therapy is being more widely applied to malformations that in the past were associated with very high mortality. Thus, it becomes increasingly important for cardiac surgeons and cardiologists to be able to communicate effectively the anatomic features of these complex defects. The anatomic classification scheme devised by Van Praagh and described in this chapter has advantages for creating a consistent description of certain features of the heart. Thus, the use of the Van Praagh classification system can accurately convey information about the situs of the heart, the situs or isomerism of the ventricles, and the position of the aortic origin. It must be emphasized, however, that these classification systems are primarily *anatomic*. Physiologic and surgical definitions and definitions of common complex malformations that have come into the terminology of congenital heart disease but are anatomically ambiguous must also be taken into account in any classification scheme or description of cardiac malformations. Thus, cardiac syndromes such as hypoplastic left heart syndrome, heterotaxy syndromes, and cardiac malpositions should also be used in descriptive terminology of cardiac malformations. These syndromes have surgical implications and can make communication simpler. The segmental notation scheme of Van Praagh then is complementary information. A hierarchical scheme of description is important because certain features are ambiguous. For example, double-outlet right ventricle with transposition of the great arteries is a relative misnomer because either double-outlet right ventricle or transposition should be the primary diagnosis and the location of the great arteries can be described separately. Transposition of the great arteries has certain anatomic and certain physiologic abnormalities separate from double-outlet right ventricle.

Certain phrases have become commonly used that may be anatomically somewhat inaccurate but remain part of the nomenclature. For example, corrected transposition of the great arteries, isolated ventricular inversion, the Taussig–Bing heart, complete transposition of the great arteries, and AV canal are diagnoses that may be embryologically inaccurate, not anatomically descriptive, or may have physiologic connotations. Additional descriptors such as the concordancy or discordancy of AV or ventricular–arterial connections may be important. Although AV alignment and ventricular arterial alignment abnormalities and segments are defined in the Van Praagh system, the common conventional usage of concordant and discordant relationships may also aid in communication.

It is also important to recognize that the embryologic development of the heart is not completely understood, and specific gene defects identified with congenital heart malformations are rare. Thus, the anatomic classification system, surgical syndromes, and embryologic development are not necessarily consistent.

It is obviously fundamentally important that cardiologists and surgeons dealing with congenital heart disease be able to communicate accurately about cardiac diagnoses and anatomy. The multiple classification systems and descriptions cloud the ability to create databases that can accurately collect data on large numbers of patients internationally with congenital heart disease. Because of this problem, there has been a very active international effort among cardiologists, surgeons, and pathologists to create standard nomenclature for congenital heart disease diagnoses and surgery and to cross-map these diagnoses to other databases, such as the European Congenital Heart Database. The Herculean efforts involved in creating these common nomenclatures has now provided a standardized list of anatomic diagnoses and operative procedures and enable data acquisition internationally and benchmarking of outcomes for congenital heart disease from one congenital heart center to another.

As noted by Dr. Weinberg, the International Society for Nomenclature of Pediatric and Congenital Heart Disease (ISNPCHD) has created the international pediatric and congenital cardiac code (IPCCC). This code maps terms from one nomenclature system to another so that entities have a single code or group of codes regardless of the original terms used. This approach recognizes that different groups continue to use different morphologic nomenclature systems. The importance of this work is hard to underestimate since having the ability to have all nomenclature systems cross code to one another such that data entry can be uniform across multiple different databases around the world is critical in establishing large multinational databases to collect information on the prevalence, significance, and outcomes in complex congenital heart disease. As databases become more and more important for our activities, these remarkable efforts of committed individuals to standardize and cross-reference data systems are of extraordinary importance.

Many different anatomic classification schemes can describe similar entities as has been noted in this chapter by Dr. Weinberg. The classification of a "simple" congenital defect such as ventricular septal defect can be completely different between different morphologic schemes. For example, the geographical scheme Dr. Weinberg describes is quite different from the border scheme described by Robert Anderson. Nevertheless, these schemes result in similar descriptions of the types of ventricular septal defects that are encountered and each might be considered complementary for consideration by surgeons.

TLS

72 Echocardiographic Evaluation of Congenital Heart Disease

Meryl S. Cohen and Jack Rychik

Methodologies using ultrasonic imaging have been developed over the last three decades that allow for accurate and timely assessment of the form and the function of the heart. Echocardiography is noninvasive and portable; thus, it has become the imaging modality of choice in the initial diagnostic evaluation of the patient suspected of having congenital or acquired heart disease. Structural abnormalities as well as alterations in flow and hemodynamics may be quickly and easily identified with minimal disturbance to the patient. Unlike angiography, in which images of opacified blood are radiographically exhibited and in which structural form is assumed from the visualization of nonopacified regions, echocardiography allows for direct, real-time imaging of cardiac structures based on their ultrasonic reflective properties. Multiple advantages are inherent in this diagnostic tool. Echocardiography is safe, portable, and radiation free. Views and sweeps of the heart from different angles and positions may be performed freely to conceptually reconstruct a three-dimensional (3D) image. Presently, 3D echocardiography can also be performed to better define valve abnormalities as well as define all of the rims of atrial septal defect (ASD) and ventricular septal defect (VSD). Serial assessments can be performed at different points in time or used continuously as a monitoring tool during procedures or interventions. Familiarity with the principles used to generate the echocardiographic image and its limitations will aid in the appropriate interpretation and clinical application of ultrasound in the surgical management of infants and children with congenital heart disease.

PRINCIPLES OF ULTRASOUND PHYSICS AND APPLICATIONS

Ultrasonic Frequencies

Ultrasonic energy is generated by the delivery of electrical impulses to piezoelectric crystals, which resonate at a set frequency. The range of ultrasound frequencies used for conventional cardiac imaging is 2.0 to 10 MHz. The choice of a particular frequency for imaging is based on its tissue penetration and resolution characteristics. High-frequency ultrasound is dissipated quickly in tissue and can be propagated only for short distances but allows for greater resolution of structure, whereas low-frequency ultrasound penetrates greater distances before attenuation but resolution is sacrificed. This concept is illustrated by the formula: wavelength = velocity/frequency, where the velocity of ultrasound in biologic tissue is constant at 1,540 m/sec. To ultrasonically resolve two points in space, they must be the distance of at least one wavelength from each other; hence, the higher the frequency, the smaller is the wavelength and the greater is the ability to resolve points that are in close proximity. For example, using 2.0-MHz ultrasound, one can resolve two points that are a minimum of 0.78-mm apart. If the objects are any closer, they will not be resolved and will appear as one. At 7.5 MHz, two points can be resolved at a minimum of 0.21-mm apart, hence resolution is greater. In practice, high-frequency transducers are chosen for use in newborns and small children to maximize resolution, whereas in older children and adolescents lower frequencies are used to maximize penetration. Higher resolution can also be used in transesophageal probes because penetration is not a major obstacle.

The Doppler Principle

In 1842, Christian Johann Doppler described the change in frequency of energy emission of an object in motion in relation to its velocity of motion toward or away from a stationary observer as follows:

$$F_d = 2VF_o(\cos Y)/c$$

where F_d is the frequency shift, F_o is the emitting frequency, Y is the angle of incidence between the direction of motion of the object and the emitted frequency, V is the velocity of motion of the object, and c is the velocity of the energy in the medium (a constant). In the setting of reflected ultrasound, this principle can be used to assay for the velocity of blood flow moving through the chambers of the heart. Rearranging the equation, we obtain

$$V = F_d F_o(\cos Y)c/2$$

Hence, the velocity of blood flow can be derived from the Doppler frequency shift of reflected ultrasound from moving blood if the emitting frequency and the angle of incidence between the direction of motion of blood and the interrogating ultrasound beam are known. In the clinical setting, it is cumbersome to measure the angle of incidence between flow direction and the ultrasound beam. Every effort is therefore made to align the ultrasound beam parallel to the direction of blood flow; angle Y is thereby assumed to be 0 (cosine of $Y = 1$). Doppler velocity assessments based on this assumption continue to be valid for angles of incidence of $\leq$20 degrees because the cosine of 20 remains close to unity; however, at angles $>$20 degrees this assumption is not valid, and Doppler-derived velocities may be underestimated.

Information relating to velocity and direction of blood flow can be obtained by several methods including pulsed-wave, continuous-wave, or color Doppler techniques. Pulsed-wave and continuous-wave Doppler quantify the blood flow velocity. In the pulsed-wave Doppler technique, ultrasound crystals fire pulses of energy and then stop to "listen" for reflected sound. This technique permits the spatial determination of velocity by allowing for interrogation of flow within a selected region of interest. Distance is calculated from the time it takes for reflected ultrasound to return to the transducer during the listening phase. Pulsed-wave Doppler is limited by its inability to assess peak velocities when there are significant disturbances of flow and velocities are high. Once a region of disturbed flow is identified, continuous-wave

Doppler may be applied to determine the peak velocity in the region. Continuous-wave Doppler emits and listens simultaneously. Thus, continuous-wave Doppler will assess all velocities within a line of interrogation—velocities within as well as proximal to and distal to the site of interest. Combining pulsed-wave and continuous-wave Doppler when assessing disturbed flow patterns is ideal to give the location of the turbulence and the maximum velocity.

Color Doppler is a pulsed-wave method in which flow within a region is assigned a color based on velocity and direction and is displayed as an overlay onto the two-dimensional image. By convention, flow toward the transducer is designated as varying shades of red, whereas flow away from the transducer is blue. Color flow is a pictorial indicator of blood flow direction and velocity but does not quantify the maximum velocity of a flow jet.

Modified Bernoulli Theorem

Based on the concept of exchange of potential energy into kinetic energy, the velocity of flow between cardiac structures can be used to calculate the pressure difference, thereby providing hemodynamic information. Bernoulli showed that the difference in potential energy, or pressure, between two sites is equal to the kinetic energy loss in addition to the energy losses caused by inertial and frictional forces. If the loss of energy because of inertia or friction is assumed to be minimal, as can be done when assessing flow across a discrete, short segmental narrowing (such as a valve or VSD), then these contributing variables may be ignored and the modified Bernoulli formula can be applied to calculate the pressure difference between upstream point 1 (proximal point) and downstream point 2 (distal point):

$$P_1 - P_2 = 4(V_2^2 - V_2^2)$$

If the proximal blood flow velocity is ≤1.0 m/sec, as is the case within most of the structures of the normal heart, the formula can be further simplified to

$$\text{Pressure difference} = 4V_2^2.$$

Hence, the pressure difference across an area of discrete stenosis can be calculated based on the peak velocity across the region of narrowing obtained by Doppler echocardiography (Fig. 72.1A, and 72.1B). Systolic pressure gradients derived from peak velocity data reflect the peak instantaneous pressure gradient, which usually occurs during the upstroke of systole and not at peak systole. This is why Doppler-derived gradients may be higher than cardiac catheterization gradients in which peak-to-peak pressure gradients are measured. Lesions in which the time for reaching peak pressure is delayed in one chamber relative to the other will add to a further exaggeration of differences between the catheter-derived and Doppler-derived gradients. For example, in aortic stenosis, peak ascending aorta pressure is reached much later than peak left ventricular pressure. Hence, whereas the peak-to-peak (catheter) gradient may be 40 to 50 mmHg when comparing the two time-delayed peaks, the peak instantaneous (Doppler echocardiography) gradient, which will likely occur during early systole when aortic pressure is low and left ventricular pressure is rapidly on the rise, may be up to 30 to 40 mmHg higher, providing an echo-derived gradient of 70 to 80 mmHg. The mean gradient, which can be calculated from the echocardiogram by integrating the sum of the peak instantaneous gradients within the systolic cycle, is most likely the best reflection of the afterload work imposed on the ventricle; however, clinical correlates were developed in the era before echocardiography, and the standard for grading and treatment of valvular stenosis of a congenital cause today is still based on the peak-to-peak catheter-based gradient assessment.

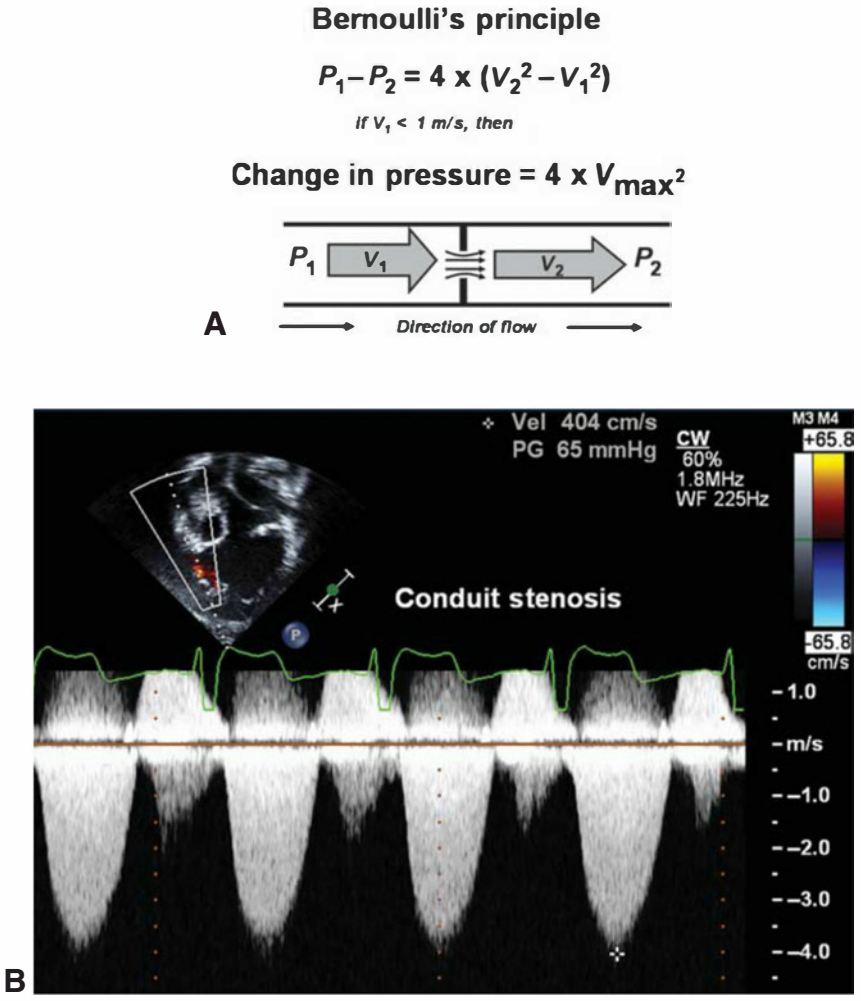


Fig. 72.1. (**A**) The Bernoulli principle defines the relationship between kinetic energy (velocity) and potential energy (pressure drop) across a discrete stenosis such that inertial and frictional forces can be ignored. (**B**) Practical example of the use of the Bernoulli principle. Continuous-wave Doppler measurement across a stenotic right ventricular to pulmonary artery conduit in a patient with L-transposition of the great arteries after atrial switch operation and Rastelli procedure. The peak velocity is 4 m/sec generating a peak instantaneous gradient across the conduit of 65 mmHg.

APPROACH TO IMAGING CHILDREN WITH UNREPAIRED CONGENITAL HEART DISEASE

Echocardiographic evaluation of a child with potential heart disease should be performed in a standardized, systematic manner with identification of all segments of the anatomy from multiple planes. In addition to the identification of all structural defects, physiologic information via the Doppler methodologies is also obtained. To facilitate the study, patients ranging from 3 months to 3 years of age should generally be sedated. A complete echocardiographic study consists of two-dimensional views and sweeps (incremental views obtained while rotating the transducer through a plane or on an axis), starting with subcostal imaging in which the liver is used as an acoustic window to the heart and ending with the suprasternal windows (Table 72.1). These standardized views and tomographic sweeps are acquired in a sequential manner during the pediatric echocardiogram, and synthesized into a comprehensive picture of cardiac anatomy, function, and blood flow (Fig. 72.2A–72.2G).

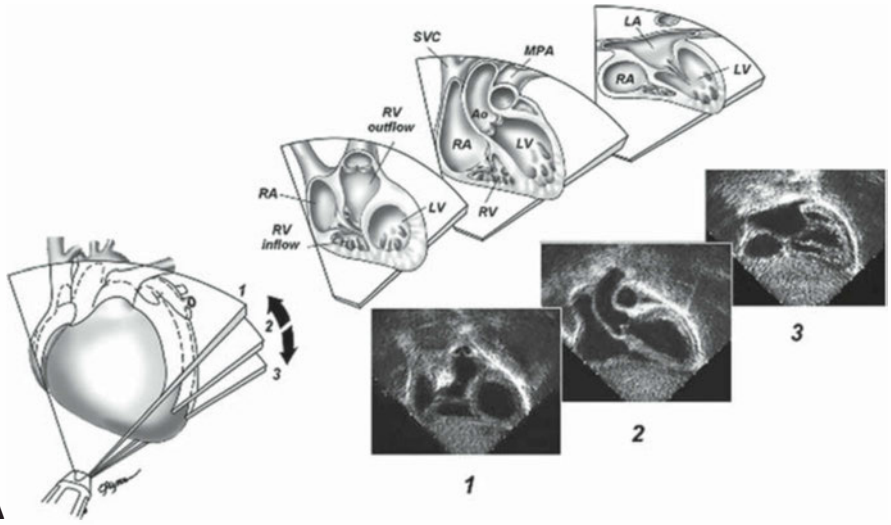
Abdomen and Situs

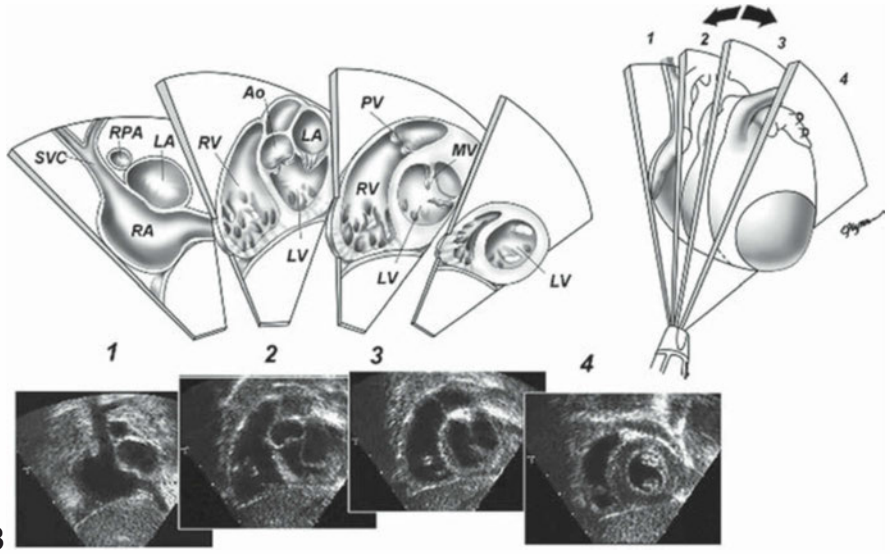
The echocardiographic study commences with a determination of situs. A transverse or coronal view of the abdomen just below the sternum at the level of the diaphragms displays the position of the liver, stomach, inferior vena cava, and descending aorta. In situs solitus, the following should be noted: (1) the liver and the inferior vena cava traversing throughout the body of the liver are to the right of the spine; (2) the stomach is to the left of the spine; and (3) the aorta is retroperitoneal and lies just anterior and slightly to the left of the spine (Fig. 72.3). In situs inversus, the mirror image is found, whereas in heterotaxy syndrome (asplenia or polysplenia), the liver is often in the midline, and the inferior vena cava and aorta may be juxtaposed on the same side of the spine either to the right or the left. In heterotaxy syndrome, systemic venous anatomy is clinically important as the majority of associated cardiac defects require single-ventricle palliation. In polysplenia, the inferior vena cava may be interrupted at the infrarenal level, with no inferior vena cava seen at the level of the diaphragms. In this case, a dilated azygos vein should be sought in the retroperitoneal space adjacent to the spine, which carries inferior vena caval blood to the superior vena cava (Fig. 72.4). A dilated superior vena cava helps confirm this finding. In addition to interruption of the inferior vena cava, the hepatic venous drainage into the atria should be identified. Occasionally, the hepatic veins drain to both atria particularly when there is a common atrium. The size of the hepatic veins should also be noted because inordinate dilation may be a sign of (1) right atrioventricular valve insufficiency, stenosis, or atresia or (2) infradiaphragmatic total

Table 72.1 Views and Sweeps for Echocardiographic Imaging of the Child with Suspected Congenital Heart Disease

Imaging window	Transducer position	Angling	Structures best visualized
Transverse abdominal view	Below the xiphoid process	Posterior, at a straight line perpendicular to the spine	Diaphragms, liver, stomach, aorta, IVC
Frontal subcostal sweep	Below the xiphoid process	Posterior to anterior in a coronal plane, parallel to long axis of patient	Atrial appendages, atrial, vent, and great arterial relationships
Left axial oblique sweep	Below the xiphoid process	45 degrees left oblique from frontal starting from right shoulder sweeping toward apex of the heart	Pulmonary veins, atrial sept, and large portion of vent sept on-end, LV outflow tract, and aorta
Sagittal subcostal sweep	Below the xiphoid process	At 90 degrees from the frontal, sweeping from right to left	SVC, IVC, atrial sept, conal sept, muscular and apical vent sept
Right anterior oblique view	Below the xiphoid process	45 degrees right oblique from frontal	RV inflow and outflow, conal septum, pulmonary artery
Apical "four-chamber" view	Apex of the heart, sixth intercostal space, medial to the left anterior-axillary line	Superiorly aimed toward the right shoulder	Atrioventricular relationship, mitral and tricuspid valves, muscular and apical sept
Apical "two-chamber" view	Rotate 90 degrees clockwise from apical "four-chamber"	Superiorly aimed toward the right shoulder and anteriorly	Left atrium, mitral valve, LV inflow, vent sept, LV outflow, aorta
Parasternal long-axis sweep	Third–fourth intercostal space, just to the left of the sternum	Plane cuts through long axis of the heart from right shoulder to the left hip. Sweep is from direction of right hip to left shoulder	Tricuspid valve, RV inflow, LV inflow and outflow, vent sept, RV outflow, and pulmonary valve
Parasternal short-axis sweep	Rotate 90 degrees clockwise from long-axis sweep	Plane cuts through short axis of the heart starting at level of great vessels. Sweep is from cephalad to caudad, toward the apex	Aortic and pulmonary valves, RV inlet and outlet, mitral valve apparatus, anterior portion of muscular sept, apical sept
Suprasternal frontal view	Suprasternal notch	Parallel to long axis of the patient, aimed caudad	SVC and innominate vein, arch sidedness, branch pulmonary arteries, pulmonary veins
Suprasternal sagittal view	Suprasternal notch	Perpendicular to long axis of the patient, aimed caudad	Aortic arch, descending aorta, left pulmonary artery

IVC, inferior vena cava; LV, left ventricle; RV, right ventricle; sept, septum; SVC, superior vena cava; vent, ventricular.





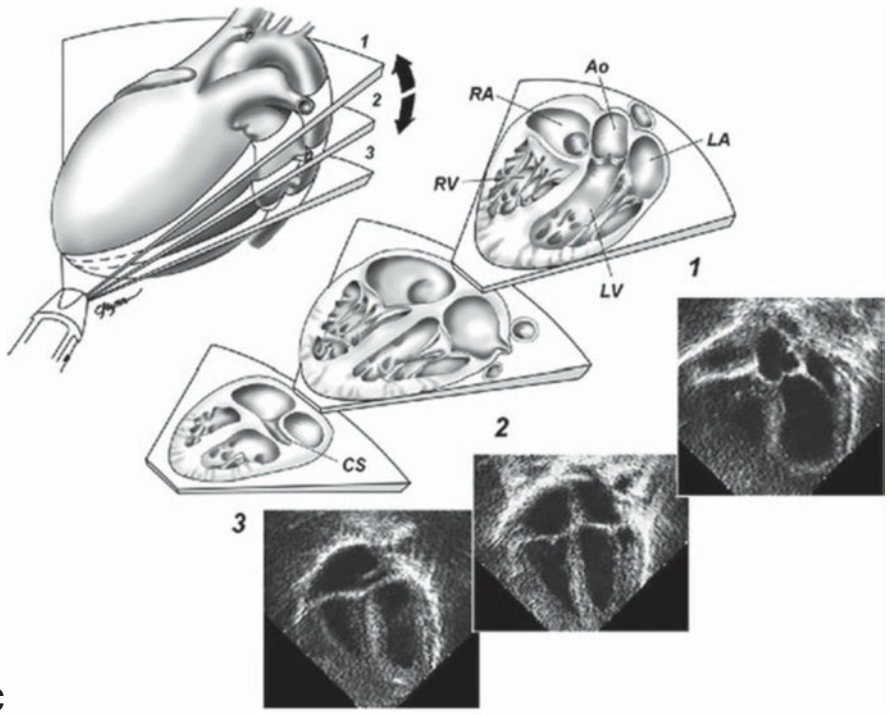
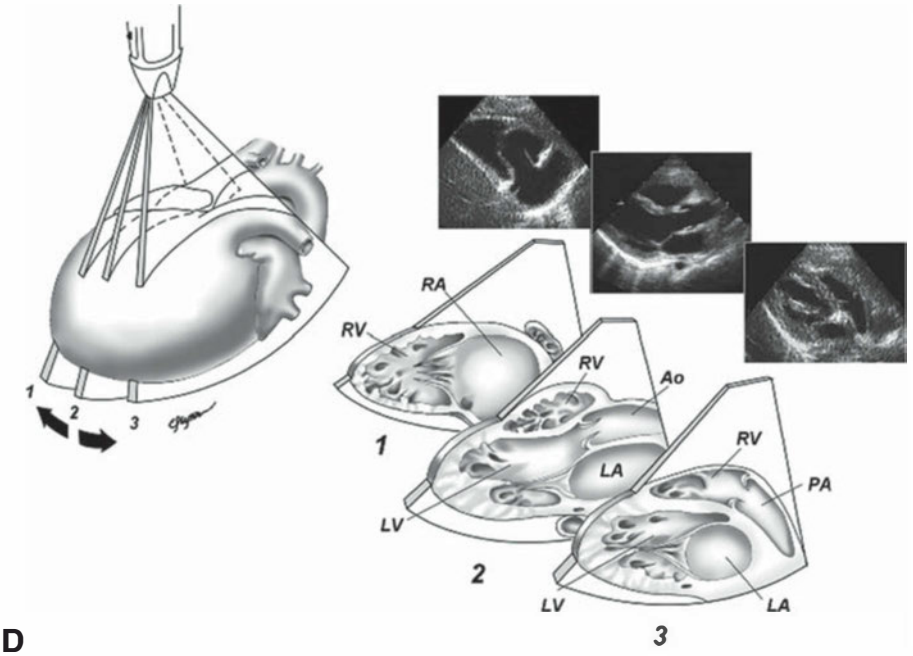
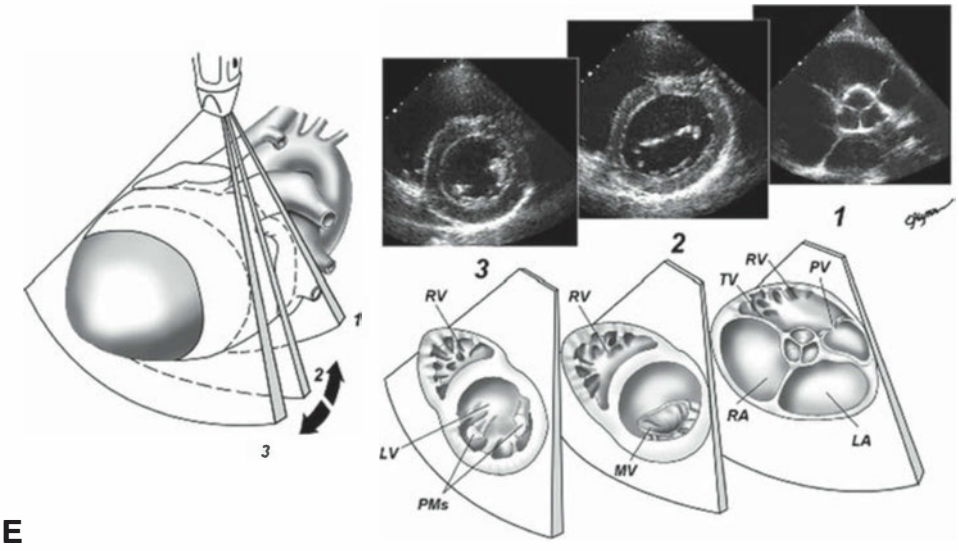


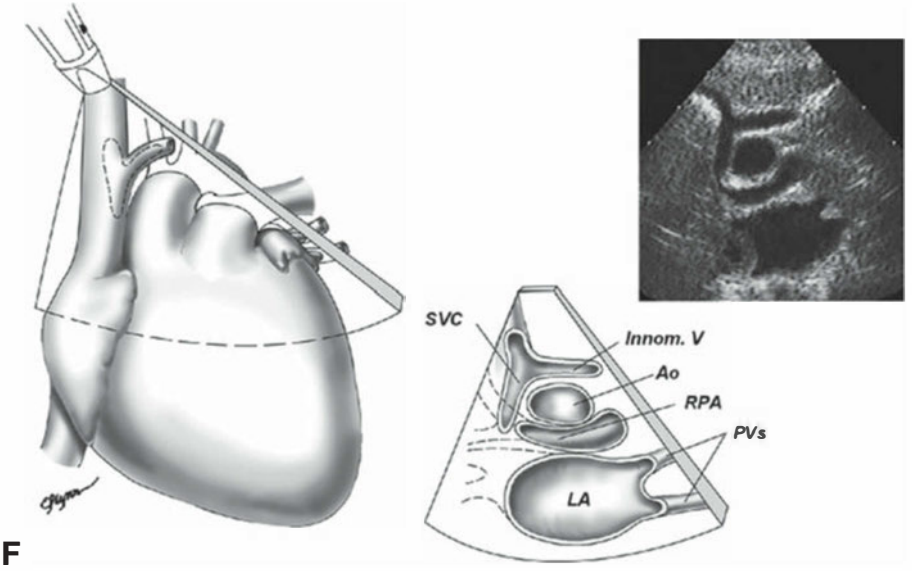
Fig. 72.2. (**A**) Subcostal frontal sweep. Coronal plane with sweep from anterior (position 1) to posterior (position 3). At position 1, structures visualized include the right ventricular outflow tract and the apex of the left ventricle. At position 2, the left ventricular outflow tract is seen. At position 3, the right and left atria and atrial septum are highlighted. Ao, aorta; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava. (All tomographic sweep and view figures are reproduced with permission from Lai WW et al. Guidelines and standards for performance of a pediatric echocardiogram: a report from the Task Force of the Pediatric Council of the American Society of Echocardiography. J Am Soc Echocardiogr 2006;19:1413-1430.) (**B**) Subcostal sagittal sweep. Sagittal plane sweep from right to left. At position 1, the entry of the superior vena cava is seen. At position 2, the right ventricle inflow and outflow is visualized. At position 3, the short axis of the left ventricle and the right ventricle cavity wrapping around anteriorly are seen, and at position 4, the apex of the left ventricle. Ao, aorta; LV, left ventricle; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava. (**C**) Apical four-chamber view demonstrates the atria, ventricles, and atrioventricular valves. Posterior angulation visualizes the coronary sinus; anterior angulation visualizes the left ventricular outflow tract and proximal aorta. Ao, aorta; CS, coronary sinus; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. (**D**) Parasternal long axis views with sweep from rightward/inferior (position 1), which focuses on the tricuspid valve and right ventricle inflow to leftward/superior (position 2) with visualization of the right ventricle outflow and pulmonary valve. At position 2, the mitral valve, left ventricle inflow, left ventricle outflow tract, and aortic valve in long axis are well seen. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle. (**E**) Parasternal short-axis sweep from superior (position 1) to inferior (position 3). Structures viewed in sequence in this sweep include the aortic valve in cross section with the identification of valve leaflets, course of the coronary arteries, conal septum, pulmonary valve, ventricular septum, mitral valve, papillary muscles, muscular septum, right ventricle cavity, and left ventricle cavity. LA, left atrium; LV, left ventricle; MV, mitral valve; PMs, papillary muscles; RA, right atrium; RV, right ventricle; TV, tricuspid valve. (**F**) Suprasternal frontal view. Structures well seen include the innominate vein, superior vena cava, pulmonary arteries, and pulmonary vein entry into the left atrium. Ao, aorta; Innom V, innominate vein; PVs, pulmonary veins; RPA, right pulmonary artery. (**G**) Suprasternal arch view, also known as the "candy cane view" as the ascending and descending aorta are displayed with the origin of cephalic vessels. Ao, aorta; Innom V, innominate vein; RPA, right pulmonary artery.



D



E



F

Fig. 72.2. (*Continued*)

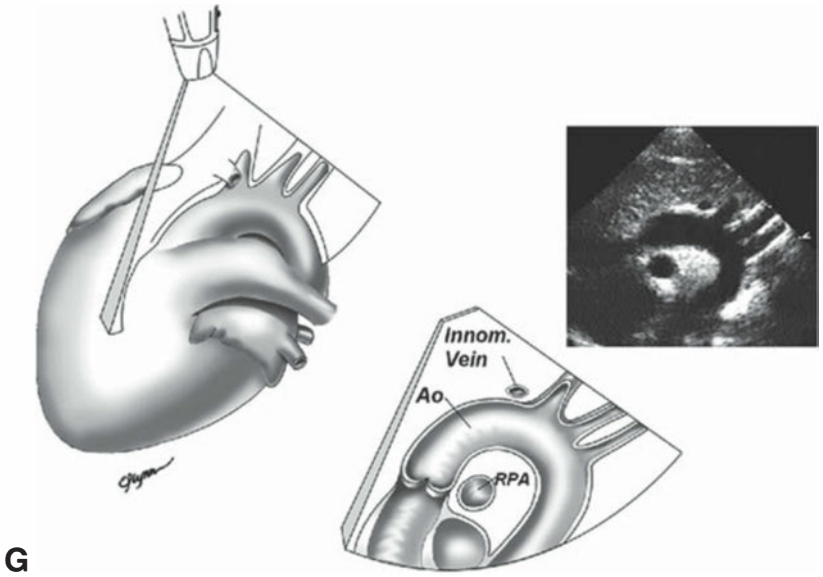


Fig. 72.2. (*Continued*)

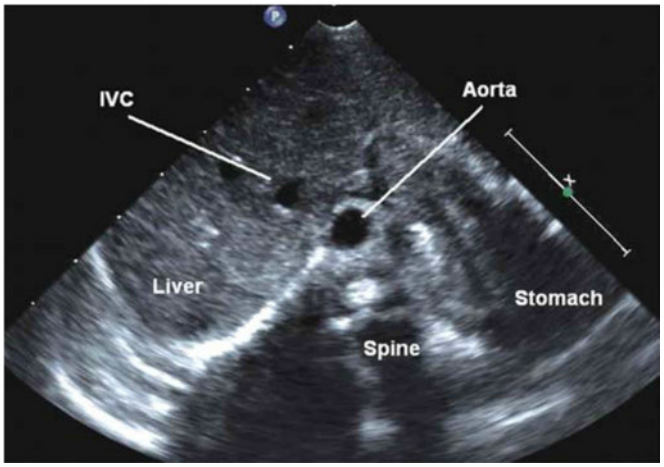


Fig. 72.3. Coronal view used to determine situs. Situs solitus or normal situs is seen with the aorta to the left of the spine and the inferior vena cava (IVC) running through the liver on the right.

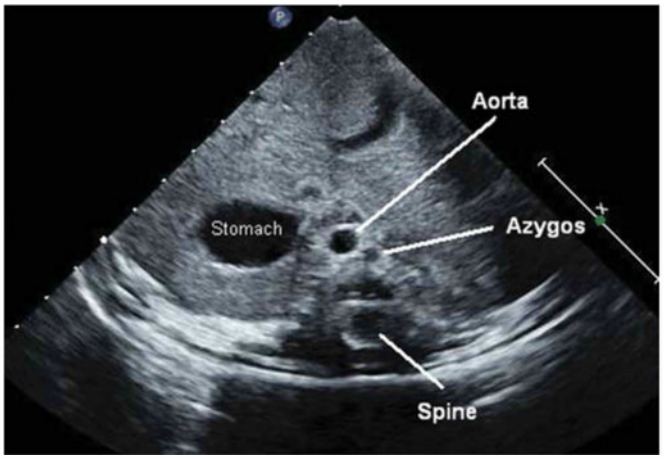


Fig. 72.4. Coronal view in a patient with polysplenia type of heterotaxy syndrome. The stomach is left-sided, the aorta runs to the right along the spine, and there is interruption of the inferior vena cava such that the azygos runs adjacent to the aorta and inserts into the left superior vena cava (not seen).

anomalous pulmonary venous connection (TAPVC). A left superior vena cava has no hemodynamic significance in the structurally normal heart. Typically, this vessel crosses anterior to the left pulmonary artery and enters the coronary sinus posteriorly. However, bilateral superior venae cavae are common in heterotaxy syndrome. Identification of this anomaly becomes important particularly if single-ventricle palliation is planned (Fig. 72.5). Without a bridging vein between the superior venae cavae, a bilateral bidirectional superior cavopulmonary anastomosis is necessary in single-ventricle palliation; when a large enough bridging vein is present, the left superior vena cava can be ligated rather than connected to the pulmonary artery. In heterotaxy syndrome, the left superior vena cava may empty directly into the left atrium or by way of an unroofed coronary sinus resulting in a right-to-left shunt.

Atria, Ventricles, and the Atrioventricular Connection

Identification of atrial and ventricular morphology is an important part of the echocardiographic examination. The atria are identified by their appendage morphology as well as the attachments of the septum primum to the septum secundum. The atria should not be defined by the connected venous structures because they can be variable (i.e., anomalous pulmonary venous connection to the right atrium, and left superior vena cava to left atrium). The right atrial appendage has a wide inlet and a broad-based triangular appearance; the

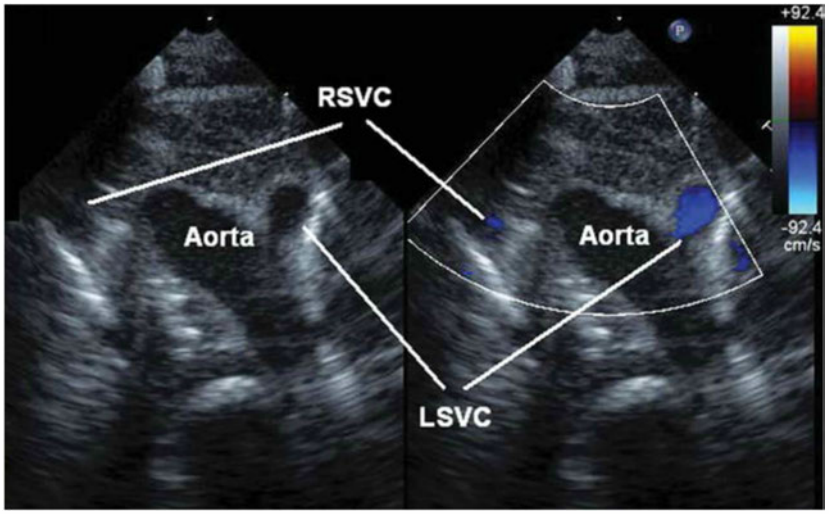


Fig. 72.5. Suprasternal frontal view in color compare mode of a patient with bilateral superior venae cavae. The right superior vena cava (RSVC) and the left superior vena cava (LSVC) are seen running along each side of the mediastinum. Blue flow in the color view demonstrates that the flow is directed toward the heart (away from the probe). There is no evidence of a bridging vein in this patient. The LSVC drains to a coronary sinus (not shown).

left atrial appendage has a narrow inlet and a long, fingerlike appearance. In heterotaxy syndrome, the atrial appendages may be mirror image; thus, identification of the right and left atria can be challenging.

The right and left ventricle have anatomic features that distinguish them, and these can usually be seen by echocardiography barring circumstances of severe hypoplasia or hypertrophy. The right ventricle is identified by the presence of (1) coarse septal trabeculations, (2) a single papillary muscle, (3) a trileaflet atrioventricular valve, and (4) chordal attachments to the ventricular septum. The left ventricle is identified by the presence of (1) smooth trabeculations along the septal surface, (2) two papillary muscles, and (3) no chordal attachments to the ventricular septum. When the ventricles are "D-looped" (concordant atrioventricular connection), the right and left atria connect normally to their respective right and left morphologic ventricles. When the ventricles are "L-looped" (discordant atrioventricular connection), the right atrium connects to a morphologic left ventricle and the left atrium connects to a morphologic right ventricle. In an L-looped heart, Ebstein's anomaly of the left-sided right ventricle is frequently seen.

THE ATRIAL SEPTUM AND ATRIAL COMMUNICATIONS

The atrial septum is comprised of three components; septum primum, septum secundum, and canal septum. Additionally, the sinus venosus septum is adjacent to the atrial septum.

1. Ostium secundum ASD is a deficiency in the septum primum. These defects are usually located centrally, may be of varying size, and, if small, may be difficult to distinguish from a patent foramen ovale (Fig. 72.6). A defect measuring >6 mm in diameter on a subcostal long axial oblique or sagittal sweep should be considered one of significance. Defects <5 mm may be considered a large patent foramen ovale and may spontaneously become smaller over time. Occasionally, even an anatomically small defect may be physiologically significant (large left-to-right shunt), and surgical or catheter-directed closure may be indicated. Identification of the rims of an ostium secundum ASD have become important in the era of device closure. Some defects have minimal rim at the level of the venae cavae or pulmonary veins making surgical closure a better option.
2. Ostium primum ASD is part of the spectrum of common atrioventricular canal defects. It is also called an incomplete or partial atrioventricular canal defect. In addition to the atrial septal deficiency, there is by definition a "cleft" in the left atrioventricular valve and possibly a VSD of varying sizes. The cleft is where the superior and inferior bridging leaflets meet at the ventricular septum. Valve regurgitation in this region is quite common. The atrial septal deficiency is typically large, located inferiorly and anteriorly, and best seen in the subcostal left axial oblique sweep and in the apical view.
3. Coronary sinus septal defects have the same physiologic effect as an ASD. There can be partial or complete unroofing of the coronary sinus with obligatory left-to-right shunting through the ostium into the right atrium. Often, the coronary sinus ostium is enlarged. It can be challenging to diagnose a coronary sinus septal defect when there is a left

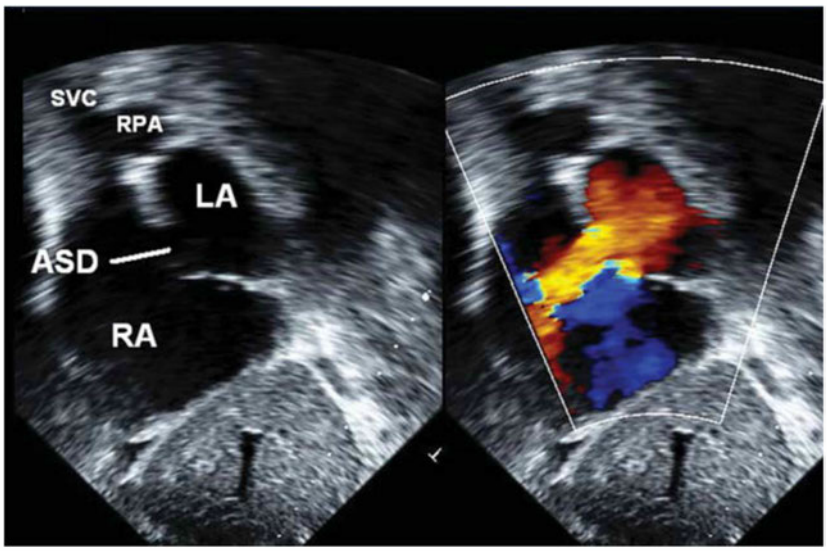


Fig. 72.6. Subcostal, left anterior oblique view in color compare mode demonstrating a moderate-sized ostium secundum-type atrial septal defect with left-to-right flow demonstrated in the color panel. ASD, atrial septal defect; LA, left atrium; RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.

superior vena cava to coronary sinus (thus, the coronary sinus is enlarged already) or in patients with complex congenital heart disease. Contrast injection can sometimes be helpful to make the diagnosis.

4. Sinus venosus defects are in fact not true ASDs because the sinus venosus portion of the heart is not a component of the true atrial septum. However, from a clinical standpoint, the defect results in a left-to-right shunt at the atrial level and causes the same right ventricular overload seen in other ASDs. Sinus venosus defects are generally large and usually occur in association with the superior vena cava; less frequently, these defects are associated with the inferior vena cava. A superior defect is located at the orifice of the superior vena cava into the right atrium, hence echocardiographically, the defect creates the appearance of the superior vena cava overriding the atrial septum (Fig. 72.7). The right upper pulmonary vein is often either overriding the defect or connected anomalously to the superior vena cava. In an inferior defect, the inferior vena cava overrides the septum and the flow may be directed toward the left atrium resulting in cyanosis. The right lower pulmonary vein may drain anomalously in this setting.

Because it is a thin structure and false image drop-out is possible, the atrial septum should always be imaged in multiple planes perpendicular to the ultrasound beam. When an ASD is present, the right atrium, right ventricle, and pulmonary arteries will often appear dilated, and pulmonary artery flow velocities may be elevated in relation to the right-sided volume load. Echocardiography generally provides all of the information that is sufficient to refer patients for catheter-directed or surgical intervention.

THE VENTRICULAR SEPTUM AND VENTRICULAR COMMUNICATIONS

The ventricular septum is a complex 3D structure made up of tissue of varying morphologies as well as embryologic origin; hence, there are a variety of VSDs. In some cases, multiple defects can be seen in the same patient.

1. The conoventricular or perimembranous VSD is the most common and is located in the region adjacent to the septal leaflet of the tricuspid valve and just under the aortic valve (Fig. 72.8). It may be of varying sizes and in fact may be partially filled in with tricuspid valve tissue, forming an "aneurysm." The presence of aneurysm tissue on echocardiographic examination may indicate a high likelihood for spontaneous diminution or closure. Prolapse of the right coronary cusp of the aortic valve may occur over time, particularly with a small defect, and may be an indication for surgery.
2. Displacement of the conal or infundibular septum results in a malalignment-type VSD. Malalignment defects are generally large and unrestrictive and do not undergo spontaneous closure. With anterior displacement of the conal septum, the aorta overrides the ventricular septum and the right ventricular outflow tract becomes smaller resulting in tetralogy of Fallot (TOF). The conal septum can also be displaced posteriorly, impinging on the left ventricular outflow tract. This type of defect is commonly associated with arch obstruction or interruption.
3. Deficiency of the infundibular septum may occur with or without displacement, resulting in a conoseptal hypoplasia or doubly committed subarterial VSD (Fig. 72.9). In contrast to the more common conoventricular or perimembranous defect, this VSD is located just under the pulmonary valve. The proximity of this defect to the aortic valve annulus makes it a substrate for prolapse of the right aortic cusp with deformity of the aortic valve architecture and subsequent aortic insufficiency. Though these defects may be small, they do not close spontaneously.
4. An atrioventricular canal or inlet-type VSD may be part of the common atrioventricular canal anomaly in which a primum ASD and a common atrioventricular valve are present. In rare cases, a canal-type VSD occurs without common atrioventricular canal defect but with an associated conotruncal anomaly such as double-outlet right ventricle (DORV). A canal-type VSD is usually large and located posteriorly at the "inlet" portion of the right ventricle. In the common AV canal spectrum, the VSD may be filled in with valve tissue, which may partially or completely occlude the defect.
5. A muscular VSD may exist in the anterior trabecular zone or in the middle portion, apex, or posterior aspect of the septum. Muscular defects are rimmed circumferentially by muscular tissue and may be of varying sizes. These defects may also undergo spontaneous closure over time, particularly if they are small. In the presence of a large VSD such as in TOF, an additional small or moderate-sized muscular defect has been reported in up to 5% to 10% of cases and may be difficult to detect because flow is preferentially across the larger defect. Once the larger defect is closed, the smaller defects may then play a greater physiologic role and cause morbidity related to increased left-to-right shunt in the postoperative period. Hence, a careful search for additional muscular defects, particularly in

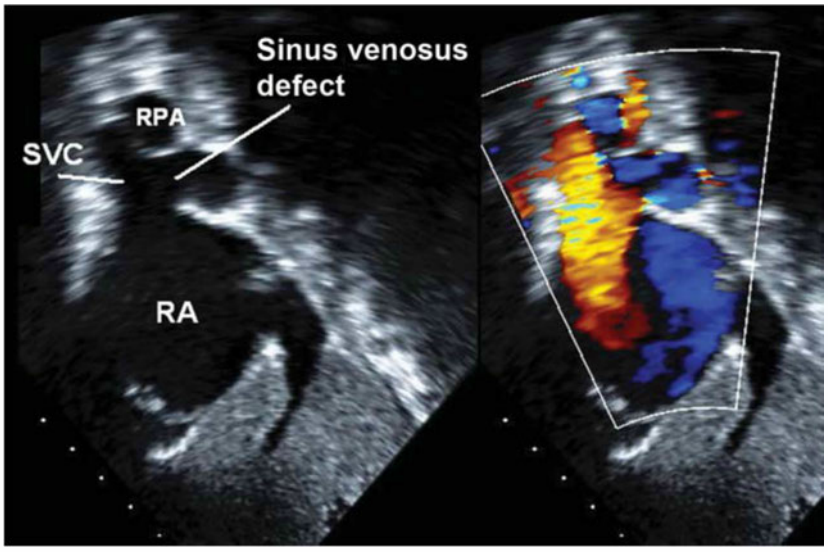


Fig. 72.7. Subcostal sagittal view in color compare mode demonstrating a superior vena cava (SVC)-type sinus venosus defect. The SVC appears to override the atrial septum and flow across the defect can be seen. The right pulmonary veins are not seen in this image but connect anomalously to the SVC. RA, right atrium; RPA, right pulmonary artery.

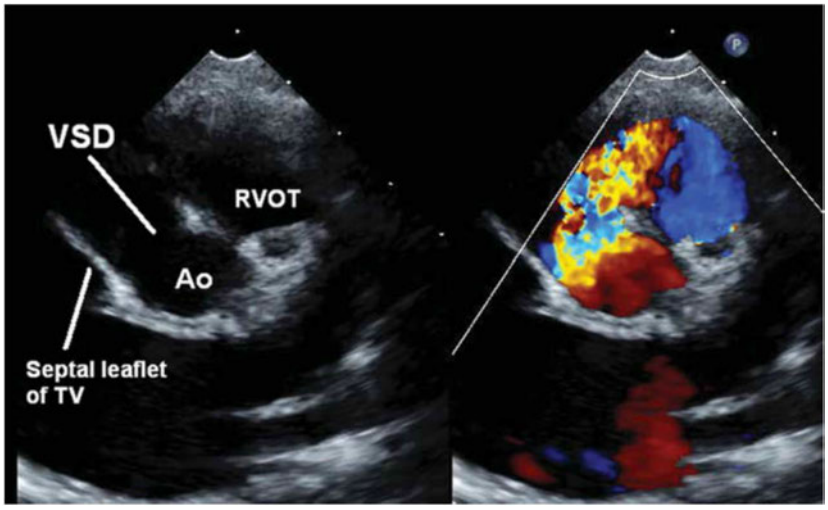


Fig. 72.8. Parasternal short-axis view in color compare mode demonstrating a large conoventricular (perimembranous) ventricular septal defect (VSD) located adjacent to the septal leaflet of the tricuspid valve (TV). Ao, aorta; RVOT, right ventricular outflow tract.

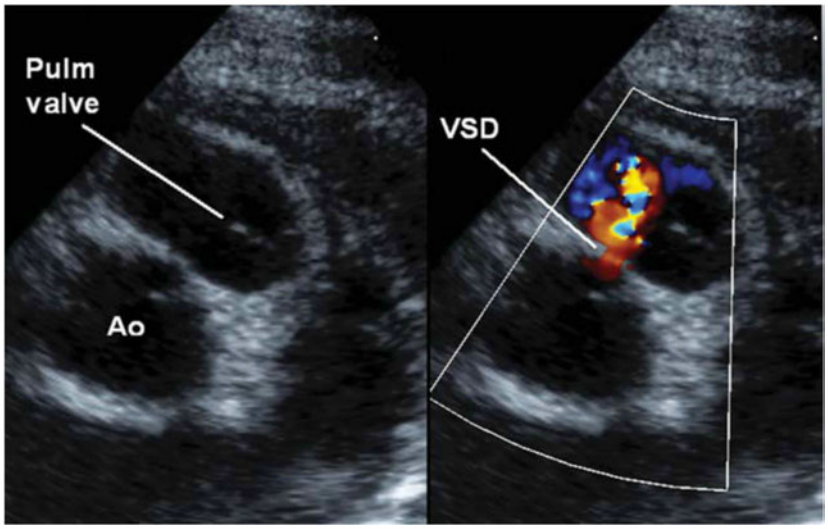


Fig. 72.9. Parasternal short-axis view in color compare mode demonstrating a conoseptal hypoplasia or doubly committed subarterial ventricular septal defect (VSD), which is located just under the pulmonary valve. Ao, aorta; Pulm valve, pulmonary valve.

the anterior trabecular zone of the right ventricle, should be undertaken in all patients with a large VSD.

A part of the assessment of the physiologic effect of a VSD is an estimate of the right ventricular pressure. Assessing the velocity of tricuspid regurgitation will allow for the calculation of the peak gradient between the right ventricle and the right atrium. By adding the value of right atrial pressure, which in the absence of direct monitoring may be assumed to be 5 to 10 mmHg, one can obtain an estimate of right ventricular pressure. In addition, a less accurate but somewhat useful method of assessing right ventricular pressure is to measure peak velocity across a native VSD or a residual defect after repair. Because the Doppler technique measures the peak instantaneous pressures within the cardiac cycle, a right bundle–branch block, common after closure of a VSD, may create a period in early systole in which a large pressure gradient is present, and a high peak velocity will be obtained. This may create the spurious finding that the right ventricular pressure is low. Cardiac catheterization is at times indicated before referral for surgery of patients with VSDs. If a defect appears anatomically large on echocardiography in conjunction with signs and symptoms of congestive heart failure and if there is no suspicion of pulmonary vascular disease (i.e., the patient is <6 months of age), cardiac catheterization is unnecessary and repair should be undertaken. Invasive catheter assessments are indicated when calculation of the degree of shunt helps in decision-making and in circumstances in which a calculation of the vascular resistances is useful.

ABNORMAL ATRIOVENTRICULAR CONNECTION

In atrioventricular discordance, the right atrium is aligned with the left ventricle and the left atrium is aligned with the right ventricle. The most common defect with atrioventricular discordance is congenitally corrected transposition of the great arteries (ccTGA) where there is also ventriculoarterial discordance. With these anatomic findings, the circulation remains in series ("congenitally corrected"). ccTGA is frequently associated with Ebstein-like malformation of the tricuspid valve with associated tricuspid regurgitation (Fig. 72.10). Additional cardiac defects are common such as VSD, pulmonary outflow obstruction, or systemic outflow obstruction. These findings often lead to early presentation, although presentation in adulthood has been reported. Surgery to align the left atrium to the left ventricle and the left ventricle to the aorta requires a "double switch" approach including an atrial switch along with an arterial switch or a Rastelli-like procedure if a VSD and pulmonary stenosis is present.

SINGLE-VENTRICLE LESIONS

Single-ventricle lesions or univentricular hearts are defined as anatomical abnormalities with a single-ventricular chamber or a large dominant ventricle with a diminutive opposing ventricle that cannot support its circulation. Functional single ventricles include patients with lesions such as hypoplastic left heart syndrome (HLHS, Fig. 72.11), pulmonary atresia with intact ventricular septum, and tricuspid atresia. True anatomic single ventricles typically have two atrioventricular valves (though one may be atretic) or a common atrioventricular valve connected to a single ventricle. The most classic example of true single ventricle is double-inlet left ventricle (Fig. 72.12). The single-ventricular chamber can usually be recognized as being a right or a left ventricle by its anatomic features. Important echocardiographic features that require elucidation in patients with

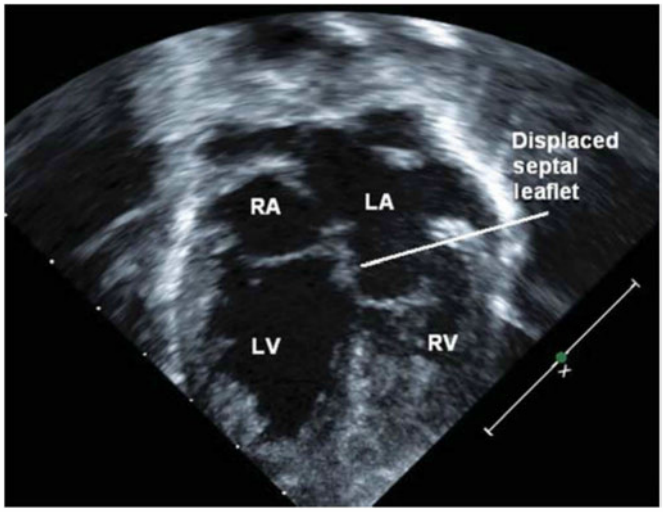


Fig. 72.10. Apical four-chamber view demonstrating congenitally corrected transposition of the great arteries. The discordant atrioventricular alignment is seen with the right atrium (RA) connected to the smooth-walled left ventricle (LV) and the left atrium (LA) connected to the trabeculated right ventricle (RV). The left-sided tricuspid valve is mildly displaced into the ventricle or Ebstein-like, which is a common association with this lesion.

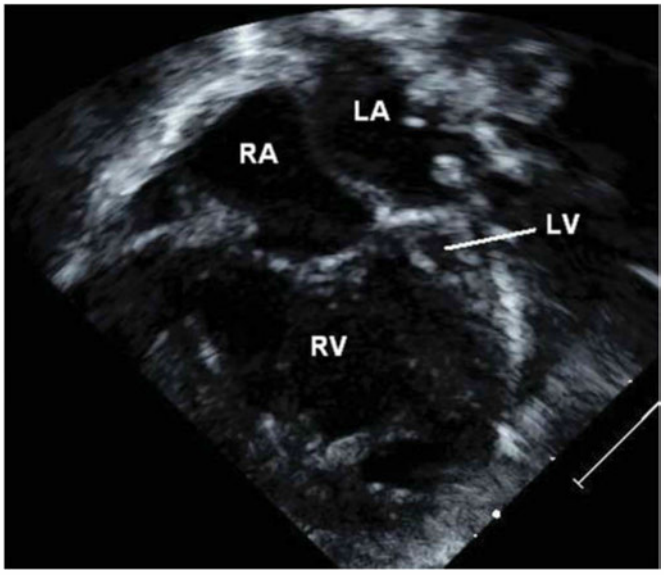


Fig. 72.11. Apical four-chamber view demonstrating hypoplastic left heart syndrome. The left ventricle (LV) is quite diminutive. There is aortic atresia in this case (not seen). LA, left atrium; RA, right atrium; RV, right ventricle.

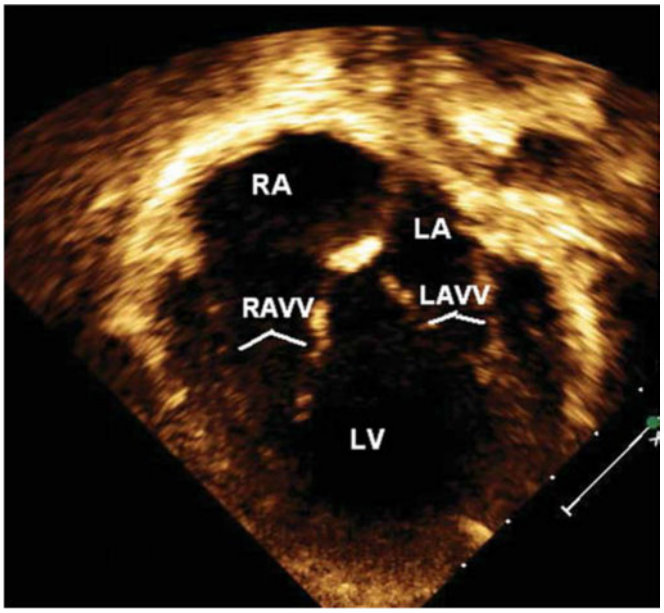


Fig. 72.12. Apical four-chamber view demonstrating a double-inlet left ventricle. The right ventricle is an outlet chamber (not seen in this figure) and the atrioventricular valves are designated as right (RAVV) and left (LAVV) rather than tricuspid and mitral. LA, left atrium; LV, left ventricle; RA, right atrium.

single-ventricle lesions include (1) systemic and pulmonary venous anatomy, (2) anatomy and function of the atrioventricular valve(s), (3) anatomy and function of the semilunar valve(s) and outflows, (4) arch sidedness, and (5) assessment for coarctation of the aorta. In conditions of one atrioventricular valve being hypoplastic or atretic, it is also important to determine if there is an adequate atrial communication to prevent high pressure in the affected atrium. In single-ventricle lesions, there may be one atretic or obstructed great vessel, or both may be unobstructed. Bilateral outflow tract obstruction is usually fatal in fetal life.

Anatomic features of single-ventricle lesions are important to help determine if a neonatal surgery is required. Neonatal surgeries include Norwood procedure (classically for HLHS), modified Blalock–Taussig shunt placement for patients with pulmonary atresia or severe pulmonary stenosis, or pulmonary artery banding for those with unobstructed pulmonary outflow. Some patients have just enough pulmonary outflow obstruction that an initial surgery is not required. After neonatal surgery, staged palliation is similar for all patients with single-ventricle lesions. By 4 to 7 months, bidirectional cavopulmonary anastomosis is performed followed by the Fontan procedure at 2 to 4 years of age. Important features of the echocardiographic evaluation of patients after staged palliation include (1) evaluation of atrial septal communication, (2) assessment of presence and severity of atrioventricular valve regurgitation, (3) assessment of ventricular performance, (4) assessment of semilunar valve function, (5) assessment of aortic arch (if reconstruction has been performed), and (6) reassessment of venous anatomy to identify a left superior vena cava or pulmonary venous anomaly.

Ventriculoarterial Connections and Conotruncal Anomalies

Conotruncal anomalies involve defects of the infundibulum (conus) and the great arteries. The conus is a smooth-walled region of muscle normally situated beneath the pulmonary valve, raising it above the level of the aortic valve and positioning it anterior and to the left of the aorta. When

the conotruncal anatomy is normal, the aortic valve annulus is in fibrous continuity with the mitral valve. This relationship is easily visible in many different echocardiographic planes. Conotruncal defects arise when displacement of the infundibular septum is present and when the normal relationship of the great arteries is disturbed. Isolated aortic and pulmonary valve disease will not be covered in this chapter.

TETRALOGY OF FALLOT

TOF is the most common form of conotruncal anomaly. In TOF, there is a large anterior malalignment VSD with displacement of the infundibular septum into the right ventricular outflow tract (Fig. 72.13). In addition, distal pulmonary artery stenosis and hypoplasia are present. The key parts of the echocardiographic evaluation include an examination of (1) the nature and degree of the infundibular and pulmonary arterial narrowing, (2) the branch pulmonary artery architecture, (3) the presence of additional muscular defects, (4) the location and course of the coronary arteries, and (5) aortic arch anatomy to assess for right aortic arch and/or vascular ring. If all five aforementioned points of anatomy are well delineated by echocardiography, diagnostic cardiac catheterization is not necessary prior to surgical intervention. If TOF occurs in association with a right aortic arch, genetic testing to identify 22q11 deletion is highly recommended.

There is a wide spectrum within the diagnosis of TOF. TOF with pulmonary atresia has a wide range of anatomical presentations including plate-like atresia of the pulmonary valve with well-developed and confluent pulmonary arteries, discontinuous pulmonary arteries (Fig. 72.14), and the most severe form with markedly hypoplastic pulmonary arteries and aortopulmonary collateral vessels perfusing various lung segments. By echocardiography, assessment for confluence of the branch pulmonary arteries can be challenging. Identification of a ductus arteriosus in the setting of tetralogy with pulmonary atresia generally indicates a less severe form of the disease. It is important to identify coronary anomalies in these patients. In approximately 5% to 10% of cases, the anterior descending artery arises from the right coronary artery and courses across the infundibulum in the region where a surgical incision may be anticipated. If this coronary variant is present, an infundibulotomy or transannular patch may not be possible and a right ventricle to pulmonary artery conduit may be considered as a surgical option. The other common variant of TOF is absent pulmonary valve syndrome, whereby the pulmonary valve is hypoplastic but essentially unguarded with dysplastic and hypoplastic leaflets. On echocardiography, there is usually not only significant pulmonary stenosis but also severe pulmonary regurgitation. The branch pulmonary arteries tend to be markedly enlarged in this clinical setting. TOF can also be seen associated with common atrioventricular canal defect. This is seen typically in children with Down syndrome.

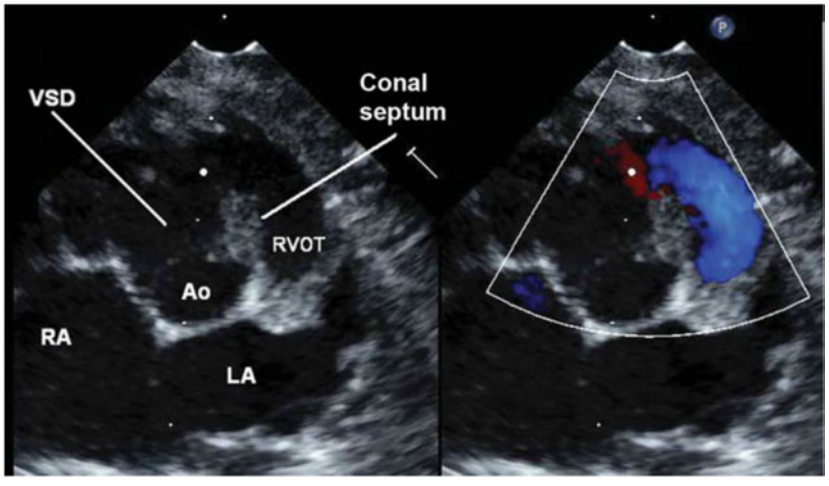


Fig. 72.13. Parasternal short-axis view in color compare mode demonstrating a patient with tetralogy of Fallot. There is anterior displacement of the conal (infundibular) septum with a large malalignment ventricular septal defect (VSD) and impingement on the right ventricular outflow tract (RVOT). Ao, aorta; LA, left atrium; RA, right atrium.

TRANSPOSITION OF THE GREAT ARTERIES

TGA is a defect in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (Fig. 72.15). Preoperative echocardiographic evaluation should include determination of (1) the nature and size of the interatrial communication as a source of mixing, (2) the presence or absence of left ventricular outflow tract obstruction, (3) the relative position of the great arteries to one another, and (4) the coronary artery anatomy, in order to anticipate the technique of coronary reimplantation at the time of arterial switch repair. TGA can be seen with intact ventricular septum or in association with any type of VSD. With malalignment-type VSD, outflow tract obstruction is common. In the classic association of TGA with posterior malalignment VSD and pulmonary stenosis, a set of surgical procedures can be performed with the goal of baffling the left ventricle to the aorta. In most circumstances, echocardiographic evaluation is sufficient to refer infants with this lesion for surgery without cardiac catheterization. Balloon atrial septostomy, which may be needed to increase the degree of intercirculatory mixing and raise the arterial oxygen level, can be done under fluoroscopic guidance or under echocardiographic guidance at the bedside.

DOUBLE-OUTLET RIGHT VENTRICLE

DORV is not a specific congenital malformation but rather refers to an abnormal ventriculo arterial alignment, whereby both great vessels arise from the right ventricle (Fig. 72.16). By most criteria, both great vessels must sit more than 50% over the right ventricle to be defined as DORV. Typically, there is a bilateral conus with myocardium separating both great vessels from the atrioventricular valves; however, other conal anatomy may be present. Associated lesions dictate a wide spectrum of variability in the physiologic presentation of this lesion. A VSD is almost always present, and in addition there may be (1) left ventricular hypoplasia with mitral stenosis or atresia (physiologically similar to HLHS), (2) infundibular narrowing of the pulmonary outflow tract with VSD flow directed toward the aorta (physiologically similar to TOF), or (3) infundibular narrowing of the aortic outflow tract with VSD flow directed toward the pulmonary artery (physiologically similar to TGA and typically called the Taussig–Bing anomaly). The key points

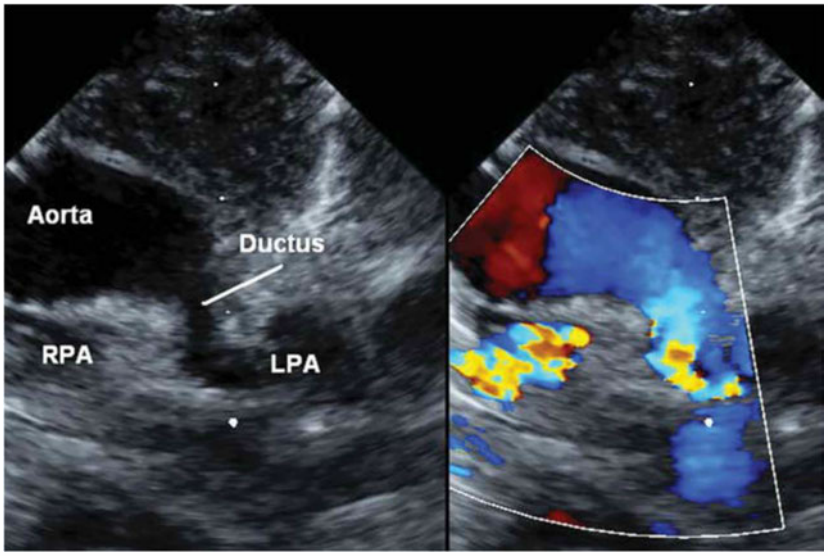


Fig. 72.14. Suprasternal view in color compare mode of a patient with tetralogy of Fallot with pulmonary atresia and discontinuous pulmonary arteries. The left pulmonary artery (LPA) is fed by a left ductus arteriosus arising from the underside of the aorta (the ductus seen in the figure). The right pulmonary artery (RPA) is fed by a right ductus arteriosus arising from the innominate artery (not shown). The distance between the pulmonary arteries is seen.

that must be delineated echocardiographically are (1) the relative position of the great arteries one to the other, (2) the presence or absence of conus beneath either of the great vessels and the associated degree of outflow obstruction, (3) direction of flow from the VSD to the great vessels, (4) the location of the VSD, which may be under one or both great vessels or may be remote (canal type or muscular type), (5) the size of the VSD, which may be restrictive, and (6) chordal attachments of the AV valves, which may be to the infundibular septum or across the VSD. DORV is one of the most challenging defects to image because of the 3D complexity. In some cases, additional imaging with 3D echocardiography, cardiac magnetic resonance, or cardiac catheterization may be helpful in anticipation of surgical reconstruction.

TRUNCUS ARTERIOSUS

Truncus arteriosus is a conotruncal defect characterized by the following criteria: a solitary great vessel arises from the heart giving rise to the aorta, at least one pulmonary artery and at least one coronary artery. Almost invariably, there is a malalignment-type VSD. The truncal valve is often abnormal; truncal stenosis and/or regurgitation is common (Fig. 72.17A). Though a tricuspid truncal valve is the most common anatomic subtype, other variations can occur including (in order of frequency), quadricuspid, bicuspid, and rarely valves with five leaflets (Fig. 72.17B). Approximately 25% of patients with truncus arteriosus have a right aortic arch and it can be associated with 22q11 deletion. There have been rare reports of truncus arteriosus with single ventricle. Truncus arteriosus has two classifications: the Colette Edwards and the Van Praagh classification. We describe here the Van Praagh classification.

1. *Type 1*: It is the most common type. There is partial absence of the aorto pulmonary artery septum: the main pulmonary artery arises from the common trunk giving rise to the branch pulmonary arteries.
2. *Type 2*: There is a total absence of the aorto pulmonary septum, and the

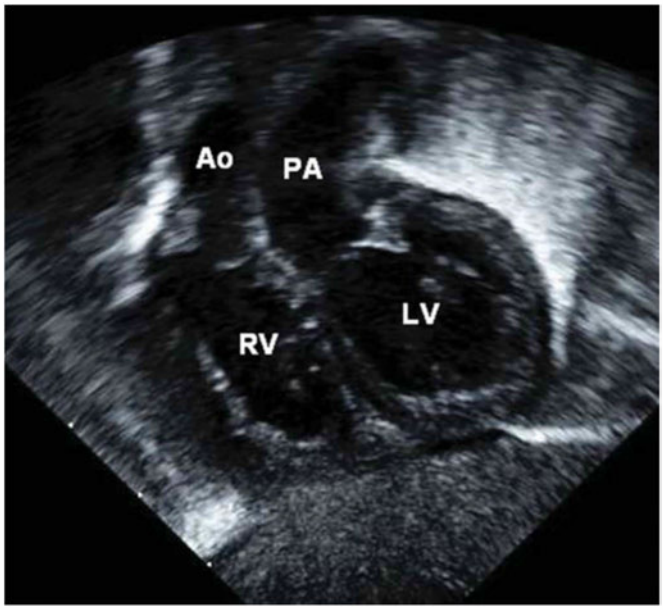


Fig. 72.15. Subcostal left anterior oblique view of a patient with D-looped transposition of the great arteries demonstrates that the aorta (Ao) arises from the right ventricle (RV) and the pulmonary artery (PA) arises from the left ventricle (LV). The outflow tracts and great vessels are parallel to each other rather than crossing, a hallmark of the anomaly.

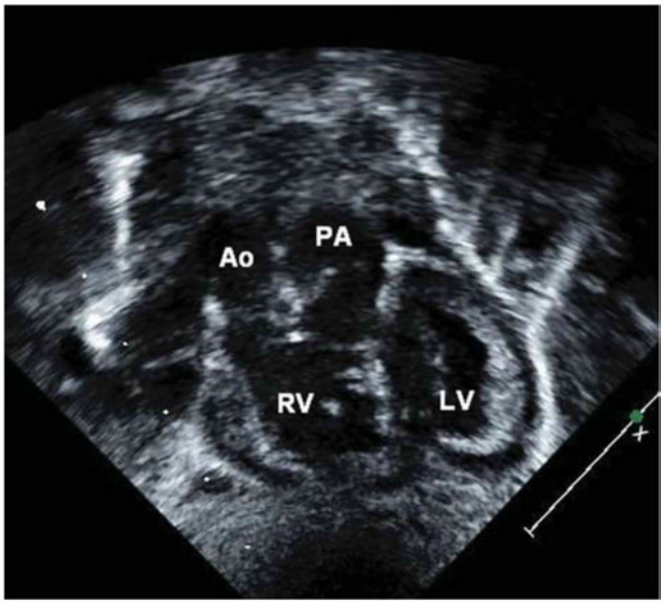


Fig. 72.16. Subcostal left anterior oblique view of a patient with double-outlet right ventricle demonstrating that both the aorta (Ao) and pulmonary artery (PA) predominantly arise from the right ventricle (RV). The ventricular septal defect is subpulmonary in this case. LV, left ventricle.

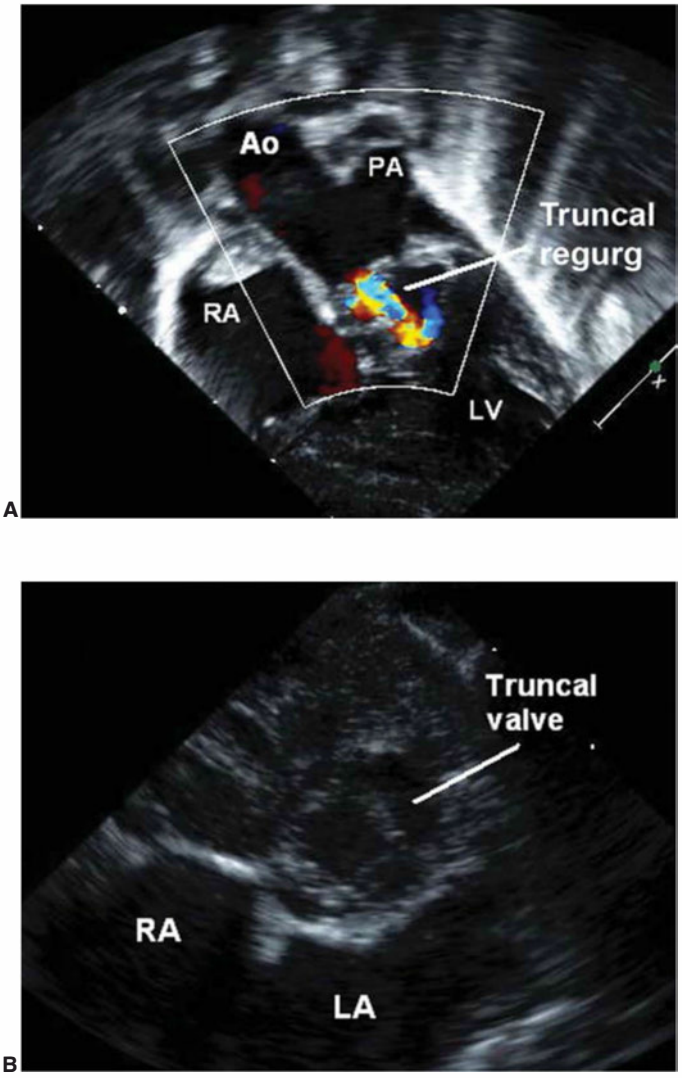


Fig. 72.17. (**A**) Subcostal left anterior oblique view of a patient with type 1 truncus arteriosus. The aorta (Ao) and pulmonary artery (PA) arise from the same trunk. The truncal valve in this patient is thickened and doming with truncal regurgitation seen (truncal regurg). LV, left ventricle; RA, right atrium. (**B**) Parasternal short-axis view in the same patient demonstrating that the truncal valve is thickened and quadricuspid. LA, left atrium.

pulmonary arteries arise directly from the posterior aspect of the common trunk.

3. *Type 3*: It is the rarest type. One of the pulmonary arteries arises directly from the trunk and the other arises directly from a collateral supply (usually, the ductus arteriosus). This is also known as truncus with absent pulmonary artery.
4. *Type 4*: Truncus arteriosus in association with interruption of the aortic arch. The trunk gives rise to the ascending aorta, the ductus arteriosus, and the pulmonary arteries. The transverse arch is absent. The ductus provides blood to the descending aorta. The interruption is usually type B (between the left carotid and the left subclavian artery).

In general, truncus arteriosus can be diagnosed by echocardiography without the need for additional imaging prior to referral for surgery. Assessment includes the (1) definition of the type of truncus arteriosus, (2) evaluation of truncal valve anatomy and function, (3) determination of the origin of both pulmonary arteries, and (4) assessment of arch sidedness.

Venous Connections and Extracardiac Vessels

A complete echocardiographic evaluation includes visualization of the pulmonary venous connections, the systemic venous connections, and aortic arch. All four pulmonary veins should be identified entering the left atrium from the subcostal planes and/or suprasternal frontal plane.

PULMONARY VENOUS ANOMALIES

TAPVC occurs when all four pulmonary veins join as a confluence and connect to a cardiac structure other than the left atrium. There are three subtypes including:

(1) *Supracardiac*: A pulmonary venous confluence connects by the way of a vertical vein to the innominate vein or to a superior vena cava.
(2) *Cardiac*: A pulmonary venous confluence connects either directly to the right atrium or to a coronary sinus (Fig. 72.18).
(3) *Infracardiac*: A pulmonary venous confluence connects to a vertical vein that travels through the diaphragm and eventually connects to the ductus venosus or inferior vena cava.

The mediastinum should be carefully interrogated for left-sided vertical venous flow. TAPVC with obstruction is generally a surgical emergency. Thus, quick identification of the pulmonary venous confluence adjacent to the left atrium and the venous drainage is important for prognosis. The mixed type of TAPVC (pulmonary veins connect to multiple other structures) appears to carry a higher mortality than other forms. In addition, TAPVC in association with single ventricle generally has a poor prognosis; recurrent pulmonary venous obstruction is common. Echocardiography can usually identify the connection of all pulmonary veins and the location of the confluence in relation to the left atrium. The etiology of the pulmonary venous obstruction, if present, can also be identified by echocardiography using color and spectral Doppler. An atrial communication (which shunts exclusively from right atrium to left atrium in the setting of TAPVC (Fig. 72.18) is a typical associated finding. When obstruction is severe, the left-sided structures may appear hypoplastic because of compression from the distended right ventricle.

AORTIC ARCH ANOMALIES

Aortic arch sidedness is determined by identifying each of the branches arising off the arch from proximal to distal. In a left aortic arch, the first vessel arising off the arch is the right innominate artery bifurcating into the right carotid and subclavian arteries, followed by the left carotid, and then left subclavian arteries. In a suprasternal frontal sweep, if the first vessel off the arch is the left innominate artery, the arch is right-sided. The branching pattern of the first vessel should be noted to

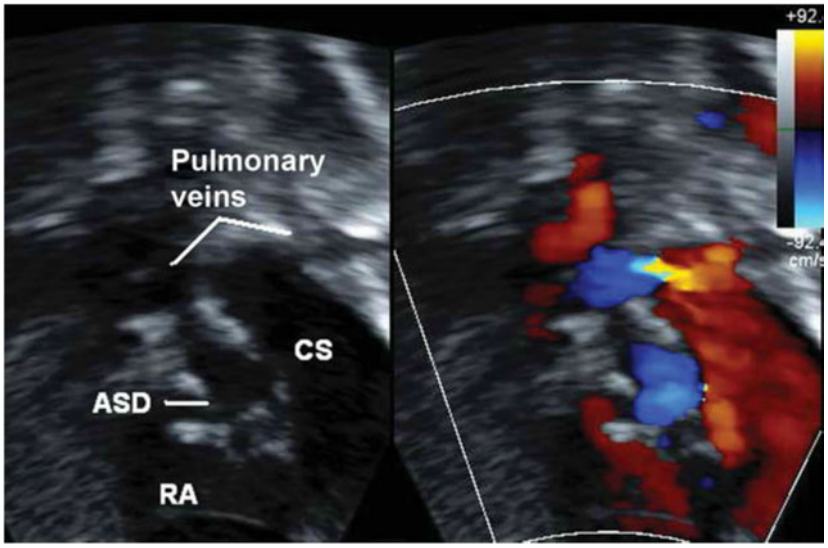


Fig. 72.18. Subcostal left anterior oblique view in color compare mode of a patient with total anomalous pulmonary venous connection to the coronary sinus. The coronary sinus (CS) is markedly dilated with a dilated ostium. The pulmonary veins entering the CS have a "whale's tale" appearance that is typical of this disease. The atrial septal defect (ASD) shunts exclusively right to left as is shown by the blue jet crossing into the left atrium. The flow in the CS does not appear to be obstructed based on the color scale. RA, right atrium.

be sure that it bifurcates into a carotid and subclavian artery. If it does not bifurcate, the possibility of an aberrant subclavian artery should be entertained. A right aortic arch with an aberrant left subclavian artery is a substrate for a potential vascular ring. Double aortic arch can also be suspected on echocardiography. However, if one arch is atretic, the diagnosis is more challenging. Any patient with a right aortic arch without mirror imaging branching should be suspected of having a vascular ring. Such patients should undergo further investigation, usually using cardiac magnetic resonance imaging.

PATENT DUCTUS ARTERIOSUS

Patency of the ductus arteriosus may be an isolated defect or in association with other lesions, whereby its function is to augment the systemic or pulmonic circulation. Doppler sampling of the flow patterns in the ductus arteriosus aids in understanding the physiology. In an isolated patent ductus arteriosus or when associated with lesions in which there is impediment to pulmonic flow (TOF, pulmonary atresia), the direction of flow will be from the aorta to the pulmonary artery (left to right) continuously throughout the cardiac cycle (Fig. 72.19). In lesions in which severe left-sided obstruction is present (critical aortic stenosis, coarctation of the aorta, HLHS), the role of the ductus arteriosus is to support the systemic circulation; hence, flow in systole will be from the pulmonary artery to the aorta (right to left); reversed flow into the pulmonary circulation will be observed during diastole because of lower pulmonary vascular resistance relative to systemic vascular resistance. Right-to-left flow across the ductus without an associated lesion is an ominous sign reflecting the presence of abnormally elevated pulmonary vascular resistance.

INTRAOPERATIVE TRANSESOPHAGEAL ECHOCARDIOGRAPHY AND POSTOPERATIVE IMAGING

As a result of the miniaturization of ultrasound probes, transesophageal echocardiographic (TEE) imaging may be performed in infants as small as 2 kg in weight. TEE may be utilized before and after surgical repair in the operating room. Preoperative TEE may identify previously unrecognized additional lesions. However, it is most often used to assess valve and ventricular function that can then be compared with postoperative studies. Postoperative imaging provides instantaneous information in the operating room prior to chest wall closure. TEE is useful to assess for residual lesions after closure of a septal defect, repair of an outflow tract obstruction, and repair of an atrioventricular valve or a semilunar valve. TEE can also identify myocardial dysfunction after cardiopulmonary bypass. Quantitative hemodynamics such as pressure gradients and degrees of valvular insufficiency, however, must be interpreted cautiously in the operating room because body temperature, cardiopulmonary bypass, inotropes, and an open chest wall may be variables that transiently influence the vascular resistances and/or myocardial mechanics. Information obtained in this setting may not always predict the patient's steady-state outcome under different conditions in the intensive care unit or beyond.

Conventional transthoracic echocardiography has been shown to be effective in detecting residual lesions in the majority

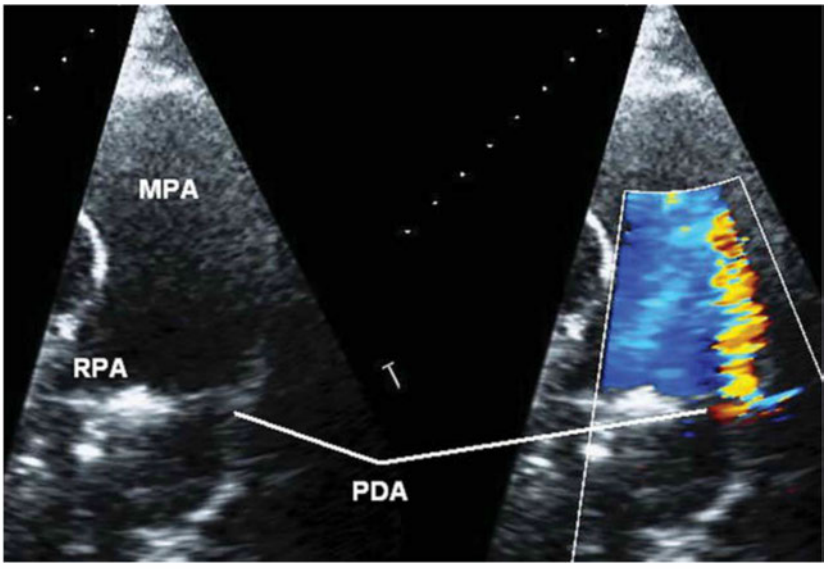


Fig. 72.19. A high parasternal (ductal) view in color compare mode demonstrating a moderate-sized patent ductus arteriosus (PDA) shunting blood from the aorta to the pulmonary artery. The left pulmonary artery is out of the plane of the image. MPA, main pulmonary artery; RPA, right pulmonary artery.

of children who have had cardiac surgery. In addition to assessment for residual lesions, postoperative echocardiography can help identify ventricular dysfunction and important pericardial and pleural effusions. It is standard of care to perform a postoperative echocardiogram on patients who have undergone cardiac surgery to identify these potential complications.

It is also important to emphasize that TEE findings in the immediate postoperative period as the patient is separating from cardiopulmonary bypass may identify less or more severe residual disease than that seen several days after repair. The hemodynamic state as the anesthetized patient emerges from circulatory arrest and cardiopulmonary bypass is quite different than that in the awake child in the intensive care unit. Assessment of residual left atrioventricular valve regurgitation after atrioventricular canal repair is an excellent example of this phenomenon. Often, the regurgitation appears mild in the operating room, but is more severe on the study performed before hospital discharge. Correlation of the intraoperative assessment of the severity of neoaortic regurgitation after the Ross procedure with postoperative assessment is also poor.

THREE-DIMENSIONAL ECHOCARDIOGRAPHY

Three-dimensional echocardiography has emerged in the past decade as an important imaging tool in patients with congenital heart disease. Three-dimensional echocardiography captures a thick sector of echocardiographic information which can then be cropped in any angle to obtain unique views of the heart. Septal defects can now be seen as would be seen on an autopsy specimen, in relation to all of the surrounding structures. Atrioventricular valves can also be viewed and the image can be cut into planes readily recognized by surgeons (Fig. 72.20). In some cases, the etiology of valve regurgitation may be identified using this technique. The resolution of the image remains an obstacle especially in those with poor two-dimensional acoustic windows. Recently, a TEE probe has been developed that has 3D capability. The resolution is superior to transthoracic 3D and the probe can be used to help guide catheter-directed interventions such as ASD device closure. At present, the 3D TEE probe is large and can only be used in patients >40 kg.



Fig. 72.20. Three-dimensional image of a "cleft" in the left atrioventricular valve of a patient with common atrioventricular canal defect (*arrow*). The superior and inferior bridging leaflets meet at the ventricular septum.

SUGGESTED READINGS

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EDITOR'S COMMENTS

An understanding of the standard views and methods of evaluation by echocardiography and a good understanding of echocardiographic imaging are important for congenital cardiac surgeons not only for preoperative diagnosis but also for postoperative evaluation of surgical repairs. Intraoperative echocardiography, usually by the transesophageal technique, has now become a common source of information on the suitability of cardiac repairs. Although in some centers echocardiography is used selectively to evaluate VSD patches for the presence of residual leaks or for evaluation of residual valvular insufficiency after AV canal repairs or tricuspid valvuloplasties, in other centers echocardiography is used routinely in all cardiac cases to assess the quality of the operative intervention and to rule out any additional hemodynamic lesions. (RM Ungerleider, JA Kislo, WJ Greeley, et al. Echocardiography during congenital heart operations, experience from 1000 cases. Ann Thorac Surg 1995;60:S539.) The hypothesis associated with the use of intraoperative echocardiography in the immediate evaluation of postoperative anatomy is that if residual lesions are identified, then addressing these at the initial operation would improve the morbidity or mortality in the postoperative period. Our experience with intraoperative echocardiography for assessing AV canal defects has shown that its routine use is associated with a significant reduction in the rate of reoperation for AV canal defects over time because significant residual valvular regurgitation can be addressed promptly. This approach has also decreased the risk of pulmonary hypertensive problems after repair. (CE Canter, TL Spray, CB Huddleston, E Mendeloff. Intraoperative evaluation of atrioventricular septal defect repair by color flow Doppler echocardiography. Ann Thorac Surg 1997;63:592.) The argument certainly can be made that intraoperative TEE is an integral part of any congenital cardiac operation as more complex repairs are being performed frequently in newborns and infants.

Recent echocardiographic techniques, including three-dimensional imaging, have improved the ability to provide anatomic information on mechanisms of valvar regurgitation and more three-dimensional

(*continued*)

EDITOR'S COMMENTS (*continued*)

geometric information that can aid in guiding cardiac surgical repair.

There have been multiple studies looking at use of postoperative transesophageal echocardiography that have shown that the identification of residual lesions in the operating room with prompt repair decreases the overall morbidity and mortality of cardiac operations. Thus, the use of postoperative transesophageal echocardiography in the operating room prior to decannulation from cardiopulmonary bypass is becoming routine in most centers except in the most straightforward of congenital heart defects where the anatomy is simply visualized at the time of the operation (secundum ASD, patent ductus arteriosus).

The argument certainly can be made that intraoperative transesophageal echocardiography is now an integral part of any cardiac operation as more complex repairs are being performed frequently in newborns and infants. As noted in this chapter, recent echocardiographic techniques, including three-dimensional imaging, have improved the ability to provide anatomic information on mechanisms of valvar regurgitation and more three-dimensional geometric information that can aid in guiding cardiac surgical repair.

As noted by the authors in this chapter, postoperative transthoracic echocardiography can show lesions that are different from those seen in the operating room environment. It is frequent for the magnitude of AV valve regurgitation to be greater postoperatively than in the operating room under conditions of general anesthesia with significant afterload reduction. Thus, in the majority of cases, a predischarge transthoracic echocardiogram is performed to evaluate the overall surgical repair.

As echocardiography has now become more available and portable and with the use of small portable probes, ultrasound-guided care in the Cardiac Intensive Care Unit is becoming increasingly standard. It is common for patients after cardiac surgical procedures to have multiple echocardiograms performed during their intensive care unit course to evaluate myocardial function, rule out residual shunts, assess diaphragm function and the presence of pleural or pericardial effusions, and to interrogate for evidence for thrombosis or valvular vegetations. There has been an increasing interest in training cardiac intensivists in the use of cardiac ultrasound for routine postoperative intensive care unit management and this trend is certainly likely to continue. Ultimately, echocardiography will become a standard monitoring tool in the cardiac intensive care unit, which will be used with increasing frequency.

TLS

73 Cardiac Magnetic Resonance in Congenital Heart Disease

Mark A. Fogel and Yoav Dori

INTRODUCTION AND GENERAL COMMENTS

Cardiac magnetic resonance (CMR) has made incredible advances in the past 10 years and has become integrated into the management and care of the patient with congenital heart disease (CHD) in the 21st century. Its role in pre- and postoperative management of the patient with CHD is a useful adjunct to echocardiography and cardiac catheterization, adding significant clinical information which the other imaging modalities cannot and many times, supplanting the need for those techniques. Many times, this information can change the care of the child. In most instances, all anatomic data necessary for diagnosis and much of the physiologic information needed can be obtained through CMR. With all the choices today with regard to imaging type, choosing the correct modality or combination of modalities is important.

Table 73.1 displays the distinct advantages CMR has over both echocardiography and cardiac catheterization as well as its limitations. For example, because patient size plays an important role in the ability of echocardiography to visualize structures, CMR has a distinct advantage in the older child, adolescent, and adult. On the other hand, patients with pacemakers, in general, and some types of coils and stents cannot be placed in the scanner due to patient safety issues (although patients with pacemakers can be studied in selected patients in highly specialized centers). Some coils can give major artifacts. There is a lot to sort out in the choice of imaging modalities.

Anatomically, in complex CHD, the overlapping of structures in angiography or the "sweeps" utilized in echocardiography may not be sufficient to conceptualize the total anatomic picture. CMR is inherently a three-dimensional (3D) imaging technique and as such, whether they are a stack of steady-state free precession (SSFP) images obtained in contiguous, parallel slices, or a 3D slab obtained with a gadolinium (CMR contrast agent) injections, software can reformat the volumetric data set in any desired plane to demonstrate the salient points of the anatomy. Even "curved plane's" are utilized clinically. Volume rendering of the 3D slab can create a 3D image which can be rotated, sliced, and manipulated in a myriad of ways to view the anatomy. Routine use of "dynamic" 3D contrast-enhanced CMR, where a 3D image slab can be obtained in just over a second, adds temporal information to a formerly static technique; gadolinium is injected in the periphery and its path is visualized as the bolus courses through the cardiovascular system (similar to a peripheral injection of iodinated contrast dye and fluoroscopy). The technique of T_2 prepared SSFP utilizing the navigator technique, generally utilized for coronary imaging, can be utilized at times for 3D imaging of the entire thoracic cardiovascular tree.

Another strength of CMR lies in its ability to delineate physiology and function in 3D. Using through plane phase-contrast velocity mapping, flows in any vessel rather than simply velocities (as in echocardiography) are obtained. Using cine imaging, ventricular volumes rather than simple linear dimensions (as in echocardiography) can be measured; CMR is the "gold standard" for measuring ventricular volumes and mass. In addition, a CMR image is typically averaged over multiple heartbeats (unlike echocardiography and angiography, a single CMR image can be averaged over two to many hundreds of heartbeats) and therefore, the functional analysis by CMR reflects many heartbeats embedded in the image yielding a much more realistic reflection of the physiologic state of the patient. In echocardiography, for example, the physician would have to perform this averaging "in his head"; it would be as if an echocardiographer would need to measure 75 Doppler velocities to be the equivalent of one velocity by CMR. It should be noted, however, that "real time" as well as "interactive" CMR is in routine clinical use, and this allows for instantaneous imaging typical of echocardiography and cardiac catheterization. Exercise and functional CMR makes much use of this technique.

In addition, CMR has the ability to "tag" myocardial tissue to calculate regional wall motion and strain, similar to speckle tracking by echocardiography and "tag" blood to visualize its course. Furthermore, myocardial velocimetry, the application of phase-contrast velocity mapping, is similar to Doppler tissue imaging by echocardiography where myocardial velocities can be measured. The advantage of both these CMR techniques over echocardiography, once again, is in its ability to determine all these parameters in 3D.

In a third broad category of CMR ability, tissue characterization plays a major role (the other two categories being anatomy and physiology/function). Viability, also called delayed enhancement, allows for identification of myocardial scar tissue and surgical patches after injection of gadolinium and a delay of approximately 10 minutes. In contrast to an injection and a subsequent delay in imaging, first pass myocardial perfusion imaging utilizing gadolinium can be used in conjunction with adenosine and is equal to nuclear techniques, which involve ionizing radiation and have less tissue contrast and resolution ("dynamic" 3D contrast-enhanced CMR mentioned above can be used to assess regional lung perfusion). Using a whole host of CMR techniques, cardiac tumors can be evaluated highly accurately (including delayed enhancement) and the "Lake Louis Criteria," which utilizes T_2 imaging for myocardial edema, T_1 imaging before and after gadolinium injection for hyperemia/capillary leak and delayed enhancement for myocardial scar, has a high sensitivity and specificity for the diagnosis of myocarditis. T_2^* imaging is a technique that allows for the determination of iron deposition in the myocardium, useful in disease states such as thalassemia and sickle cell disease to monitor and alter chelation therapy.

Table 73.1 CMR—Advantages and Limitations

Advantages over catheterization
Noninvasive
No ionizing radiation
No contrast agent needed to delineate cavities and lumina
Measure flow in parallel circuits
No overlapping structures
Advantages over echocardiography
No limit to patient size
Not limited by patient "windows"
No artifacts from calcification, surgical patches, or prosthetic valves
Compared with TEE, it is noninvasive
Identify areas of perfusion defects
Advantages over both echocardiography and catheterization
Averages functional data over hundreds of heartbeats
3D imaging of anatomy after Routine acquisition
4D flow imaging
Ability to magnetically "tag" myocardial tissue and blood (calculation of myocardial strain, wall motion, and visualizing velocity profiles)
Ability to assess velocity at different points across the ventricular cavity or blood vessel
Does not rely on geometric assumptions to calculate mass, volume, etc.
Identify areas of scarred or fibrosed myocardium
Tissue characterization including cardiac tumors, iron overload, myocarditis, and arrhythmogenic right ventricular dysplasia
Myocardial iron and oxygen levels (beginning to become clinically utilized)
Limitations
Pacemakers are a relative contraindication
Artifacts from wires, clips, coils
Respiration can cause blurring of images at times
Lying still, sedation still necessary
Mild limitation of imaging valves and chordae, although this has been improving for many years
Many studies must be "gated" to the ECG. Certain circumstances may make recognizing the "R" wave a problem
Cannot be done "at the bedside." Equipment is large
Measuring pressures still clinically experimental. Turbulent flow can make gradient calculations unreliable

CARDIAC MAGNETIC RESONANCE PHYSICS AND CARDIAC MAGNETIC RESONANCE TECHNIQUES

The detailed physics of CMR image generation is beyond the scope of this text; however, the general concept is not. A powerful magnet, most commonly on the order of 1.5 Tesla and sometimes 3 Tesla, is used to align the spins of the hydrogen atoms in the body and a radiofrequency pulse (electromagnetic energy) combined with a special magnetic field which creates a magnetic "gradient" is used to perturb a small percentage of these molecules in a specific part of the body into a higher energy state. When the radiofrequency pulse is turned off, the hydrogen atoms from that part of the body return to their normal state, releasing energy in the process. This energy is then collected and analyzed by a complex series of mathematical computations to yield one line of data in what is called "K-space." By repeating this process many times, many lines of K-space are obtained and through a process involving "Fourier transformation," the lines of K-space are translated into lines in an image. The image itself is divided into a checkerboard [matrix] with each box called a voxel (the image is of a slice thickness and, therefore, is a 3D element). Generally, there are 128 to 256 lines of data in an image; however, this can vary from 64 to 512 lines. To allow for "noise" in the system (false data) as opposed to "signal" and to alleviate artifacts from breathing, each line can be obtained three or more times and averaged (with breathholding techniques, this is typically not necessary). To image the heart, the scanner uses the ECG (or much less commonly, pulse recording) to determine what part of the heartbeat to obtain the image.

By timing the radiofrequency pulses differently and changing their magnitude, different types of images are generated. There are many types of CMR in clinical use as shown in Figure 73.1. To define anatomy (Figs. 73.1–73.3), double inversion (DI) dark blood CMR yields high-resolution images of myocardial tissue and blood vessel walls while the blood remains signal poor. SSFP imaging also yields high-resolution images of myocardial tissue and blood vessels; however, the blood is bright and signal intense, without the use of contrast agents. Cine MR (see below) can be used to delineate morphology as well, including valve morphology (shown).

Although static noncontrast images used to be the mainstay of CMR, many other techniques have come into clinical use for morphology including contrast-enhanced imaging. A contrast, typically a gadolinium-based agent, is injected and a T_1 weighted, 3D sequence is utilized to obtain a 3D image of the cardiovascular system (Fig. 73.2). Dynamic imaging is used and follows the contrast through the cardiovascular system; each phase is a 3D image in itself. These images can then be formatted as multiple two-dimensional images or a volume-rendered object (VRT) (Fig. 73.3) from any phase of the dynamic imaging; these images can be "added" or "subtracted" from each other to delineate the salient point of the anatomy. Shaded surface displays (SSD) and maximum intensity projections (MIP) are rarely used today but played a role in CMR in past years.

CMR goes beyond anatomy, however (Figs. 73.4 and 73.5). Cine CMR (Figs. 73.1 and 73.4) produces a high signal from blood

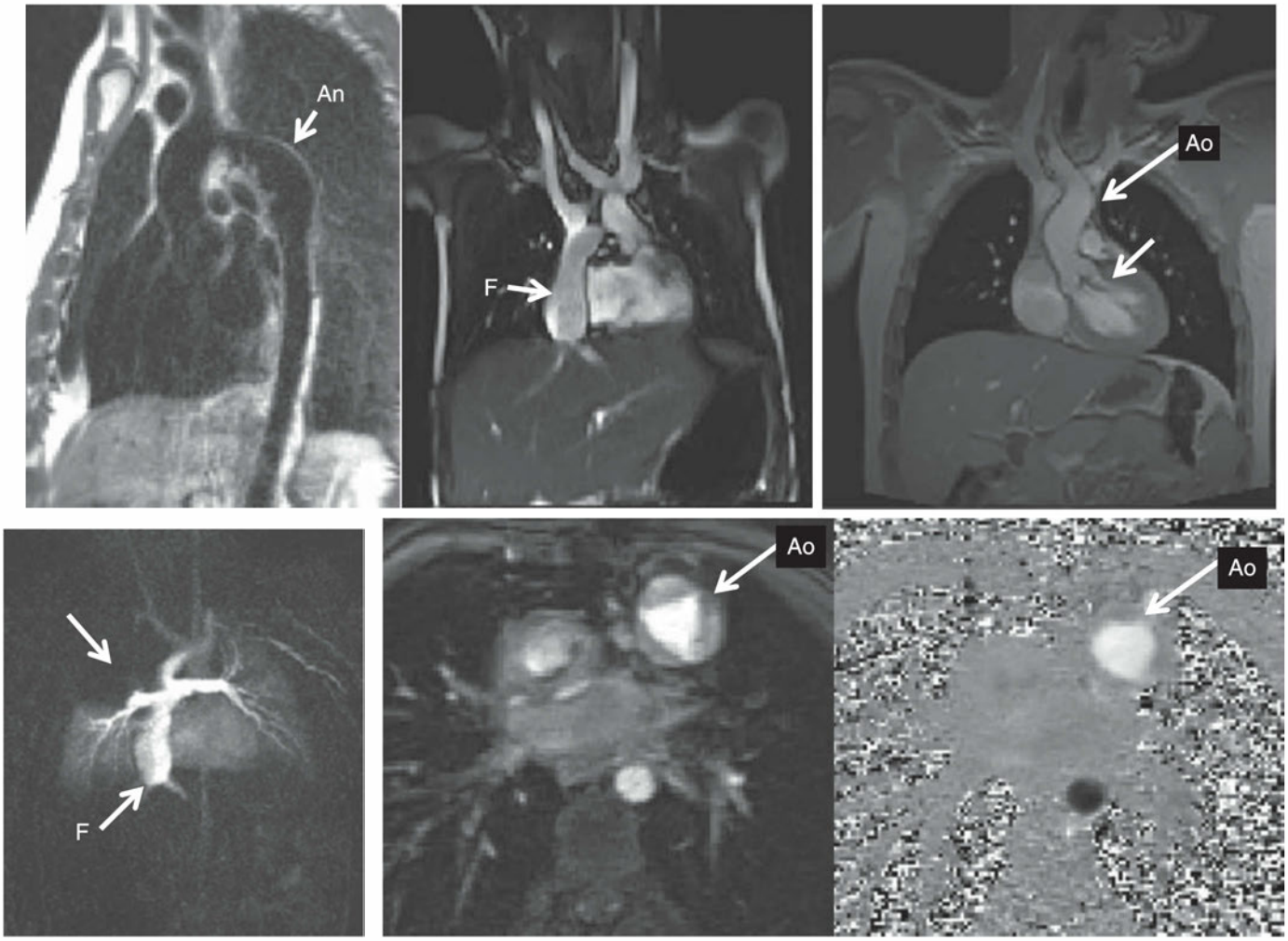


Fig. 73.1. Types of cardiac magnetic resonance in common use for anatomy. The upper left panel is a double-inversion dark blood image of a patient with a coarctation of the aorta (Ao) with residual aneurysm (An). Note that the blood is "black" and myocardial and vascular tissue give off signal. The upper middle image is a steady-state free precession (SSFP) image of a patient with pulmonary atresia with intact ventricular septum after Fontan (F); note how bright the blood is compared with the rest of the anatomy. SSFP depends upon this signal contrast for quality imaging. The upper right image is a patient with bicuspid aortic valve and mild aortic insufficiency using a spoiled gradient echo (SGE) sequence; note the turbulence underneath the aortic valve from the narrow jet. The lower left image is a three-dimensional maximum intensity projection of the Fontan baffle in a patient with hypoplastic left heart syndrome outlining the baffle, the branch pulmonary arteries and lungs; note the perfusion defect in the right upper lobe (*arrow*). The lower middle image is a magnitude image from phase-contrast velocity mapping depicting the trileaflet aortic valve in the Fontan patient mentioned in this figure; note how easily the valve leaflets are seen. The lower right image is the phase image in through plane-phase contrast velocity mapping of the aortic valve in the Fontan patient mentioned in this figure; flow, not just velocity, can be measured from this type of imaging.

and a lower amplitude signal from tissue and is usually utilized for cardiac motion, calculation of cardiac index, blood flow, and functional analysis (ventricular volumes, ejection fraction, etc.). One of the distinct advantages CMR has is that it does not rely on geometric assumptions to calculate volume, mass, and so on as other imaging modalities do. 3D echocardiography has come a long way, however, is not ubiquitous and needs specialized hardware, software, and image acquisition; CMR is nevertheless considered the "gold standard" in ventricular volume and mass measurement. Nearly all manuscripts in the literature validating 3D echocardiography compares the data with CMR; why use a surrogate, however, when a "gold standard" can be used? This is extremely important in CHD where bizarre, misshapen cardiovascular structures cannot be modeled by geometry. Further, CMR is exquisitely sensitive to turbulence and can detect even small amounts of regurgitation or stenosis. If turbulent blood flow is present, cine CMR will show a signal void in the region of turbulence, used to detect valvular regurgitation, valve stenosis, blood vessel stenosis, or baffle leaks/fenestrations (Figs. 73.1 and 73.4). Alternatively, cine CMR can obtain static images at various levels of the body, "labeling" blood as signal intense regions. This may be used, for example, to find collateral vessels off the aorta in a patient with tetralogy of Fallot and pulmonary atresia. Cine CMR can be of the spoiled gradient echo type (SGE) or of the SSFP type.

The CMR signal generated contains both the amplitude information of the signal as well as "phase" information. Phase-contrast (also known as phase-encoded) velocity mapping (Figs. 73.5 and 73.7) uses this phase information to encode velocity similar to Doppler imaging in echocardiography. The difference is that with

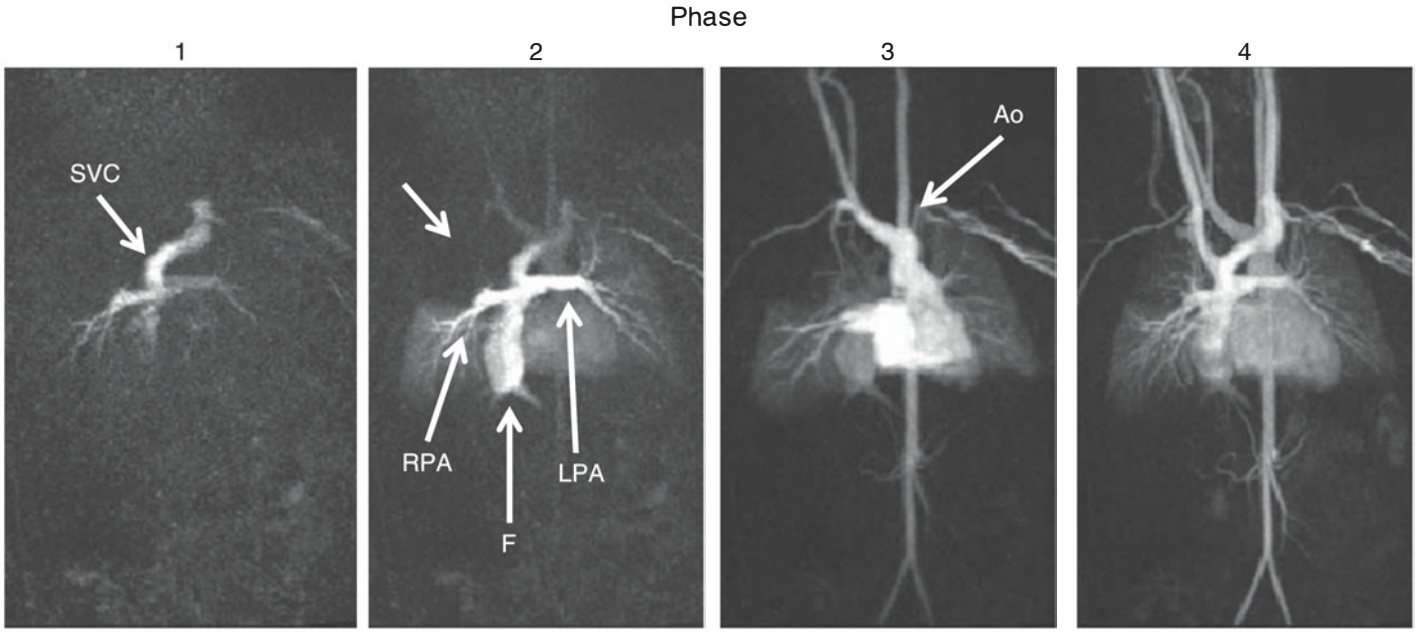


Fig. 73.2. Types of cardiac magnetic resonance in common use for anatomy—dynamic three-dimensional gadolinium imaging: The series of images, progressing from left to right, depicts the injection of gadolinium as it traverses the cardiovascular system in a patient with hypoplastic left heart syndrome after Fontan (F). Each phase is a 3D image in itself. In the left image, gadolinium is injected in the left arm and enters the superior vena cava (SVC) and the branch pulmonary arteries. In the image second to left, the gadolinium has lit up the F as well as the right (RPA) and left pulmonary arteries (LPA); note the perfusion defect in the right upper lobe (*arrow*). In the second image from the right, the left-sided phase of the injection depicts the aorta (Ao) and in the image on the right, the recirculation phase is seen with flow back into the RPA and LPA.

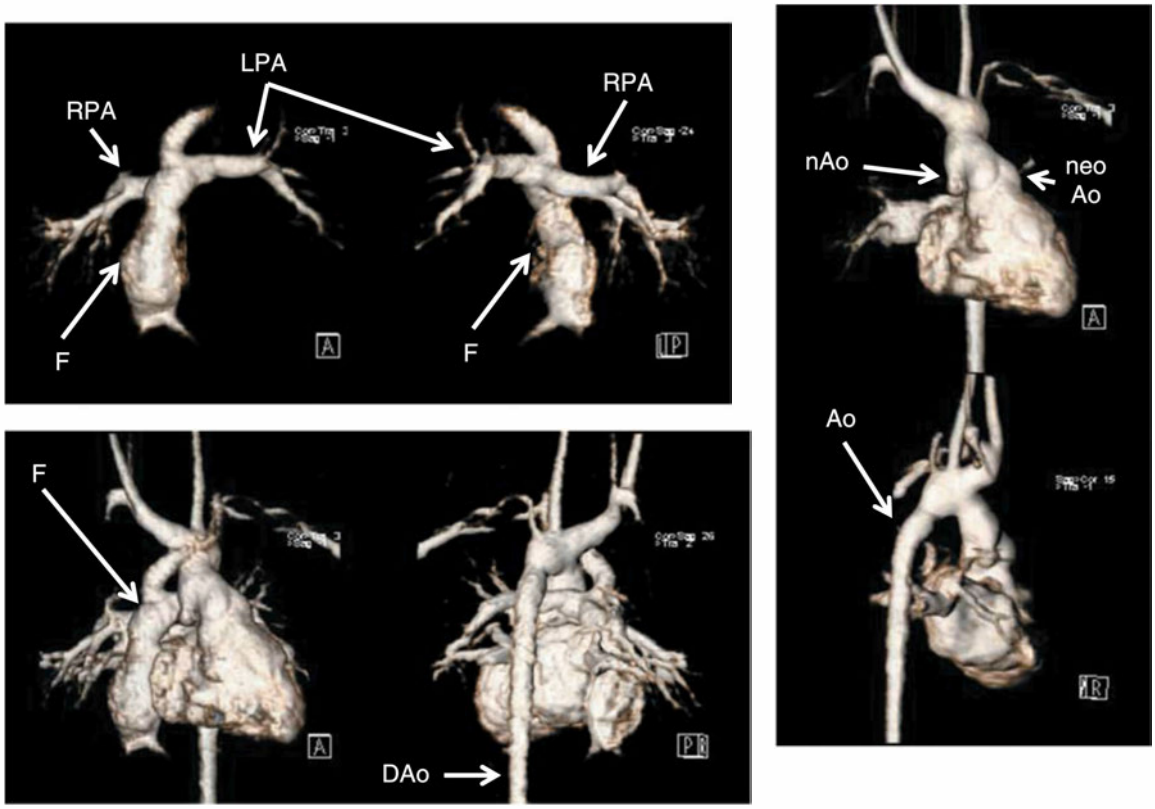


Fig. 73.3. Types of CMR in common use for anatomy—volume-rendered three-dimensional imaging: These volume-rendered images are from the patient in Figure 73.2. The advantage of dynamic 3D gadolinium imaging is that various phases can be separated out to highlight various structures. The upper left and middle panels are 3D volume renderings of the Fontan baffle (F) and right (RPA) and left pulmonary arteries (LPA) from anterior (*left*) and posterior (*middle*) views. The right upper and right lower panels are volume renderings of the ventricle and aorta (Ao) from the systemic phase of the dynamic 3D gadolinium injection; note how both the native (nAo) and neoaortas (neoAo) and the connections can be easily seen. The lower middle and lower left panels are a combination of the pulmonary and systemic phases demonstrating the entire heart intact. DAo, descending aorta.

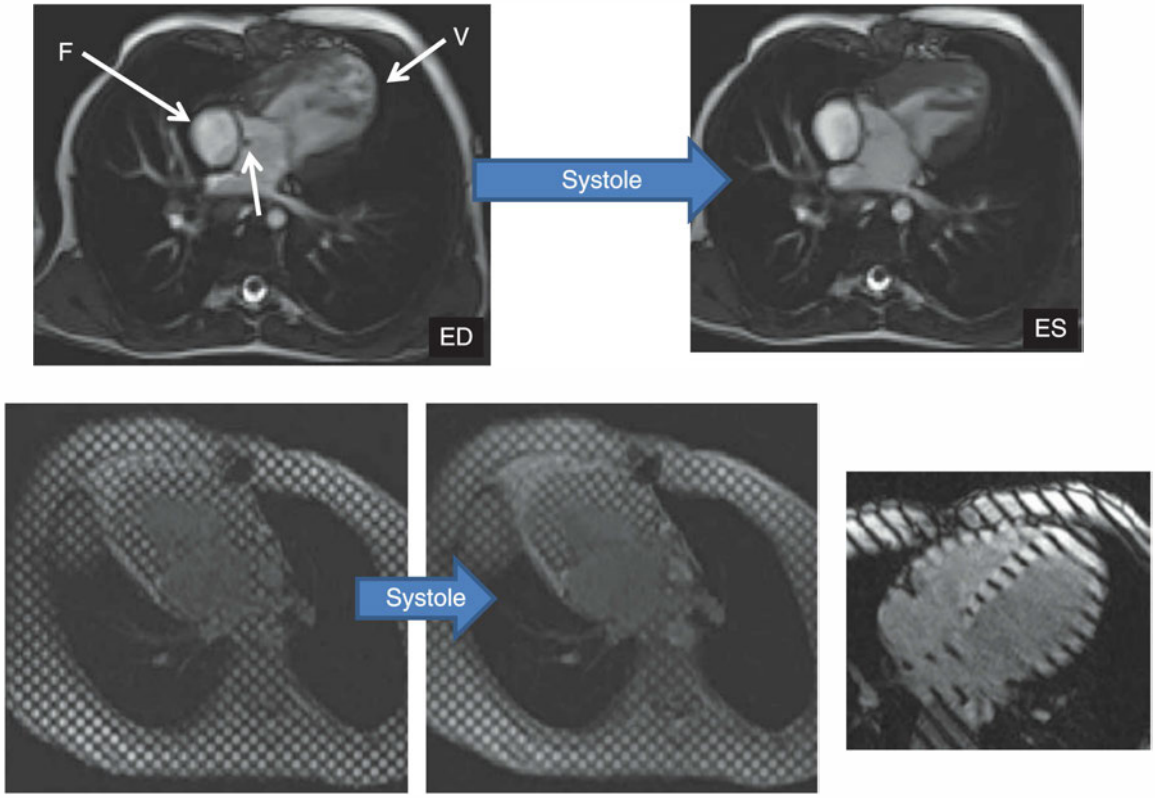


Fig. 73.4. Types of cardiac magnetic resonance in common use for physiology/function—cine CMR and myocardial tagging: The upper left and right panels are two images from the "three-chamber" cine of a patient with hypoplastic left heart syndrome after Fontan at end diastole (ED) and end systole (ES), respectively. Temporal resolution can be 20 milliseconds or less. The lower left and middle images are myocardial tagging (SPAMM—SPAtial Modulation of Magnetization) imaging of a single-ventricle patient at ED and ES; note how one can visualize the myocardial deformation of each "cube" of myocardium. Myocardial wall motion and three-dimensional strain can be measured. The right lower image is also myocardial tagging but of the one-dimensional version, used to tag thin walls such at the right ventricle in this four-chamber view; regional myocardial wall shortening fractions can be measured.

CMR, velocity can be encoded in "through plane"—that is into and out of the plane of the image—the result being that actual flow can be obtained. If a blood vessel is imaged in cross section, all the voxels that encode velocity in the vessel can be summed over the entire cross section of the vessel and integrated over the entire cardiac cycle to obtain flow (as in liters/minute, not just velocity). This can be done in nearly any vessel in the body—arterial or venous. Q_p/Q_s (Fig. 73.6), aortic to pulmonary collateral flow, regional flow to each lung, caval return can all be measured. CMR has the further advantage of performing internal checks—such as flow in the main pulmonary artery equaling the sum of the flows in the branch pulmonary arteries or the flow in the aorta equaling the sum of caval return in the absence of aortic to pulmonary collaterals; this is unique to CMR as a noninvasive imaging modality. Generally, an upper limit to the velocity to be measured is set, similar to the Nyquist limit in echocardiography, called the VENC (Velocity EN Coding). Also similar to echocardiography, velocity direction can be encoded in grayscale or color scale with "white" in one direction and "black" in another (or red and blue). Similar to Doppler echocardiography, velocity can also be encoded in the plane of the image—called "in-plane" velocity mapping. This can be encoded in either *x*- or *y*-directions of the image. The resulting three orthogonal planes that velocity can be measured in enable CMR to measure four-dimensional (4D) velocities when a "slab" is obtained, which encodes all three velocity vectors (three dimensions of velocity and one of time; Fig. 73.7). Much work is now being done in this area. Finally, phase-contrast CMR can be used to determine myocardial velocities similar to Doppler tissue imaging.

Myocardial tissue tagging (Fig. 73.4) is another CMR technique, which "magnetically labels" the walls of the myocardium and divides it into "cubes of magnetization." This allows for the calculation of regional wall strain, radial motion, and torsion. This can be of the two-dimensional tagging type, where a "grid" is laid down on the myocardium (spatial modulation of magnetization or SPAMM) or of the one-dimension type, where just a series of parallel lines are laid down. Finally, blood tagging is similar to tissue tagging except that the blood is labeled, allowing for visualization of velocity profiles as well as calculation of cardiac index (Fig. 73.8). This can be of the bolus tagging variety, where only a thin stripe is laid down on the blood vessel to label it (Bolus tagging); this has largely been supplanted by phase-encoded velocity mapping. Nevertheless, the ability to actually visualize a flow profile directly without postprocessing is still an appealing approach. Alternatively, a large stripe can be laid down on the blood to aid in shunt detection (Fig. 73.8).

Coronary imaging, as with perfusion and viability, is an important issue in CHD. Coronary issues that the imager needs to address can be divided into three broad categories: (1) congenital coronary abnormalities such

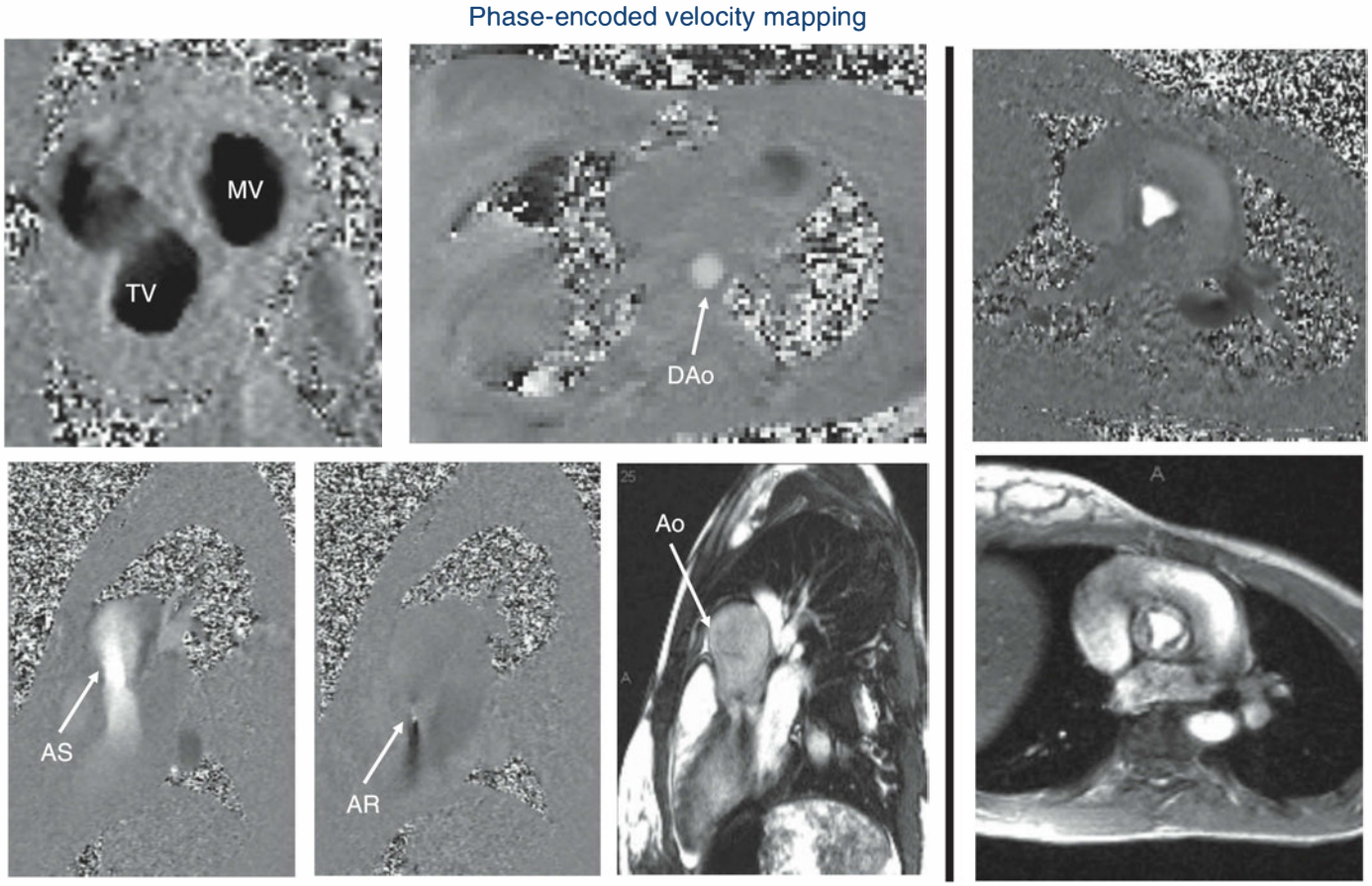


Fig. 73.5. Types of cardiac magnetic resonance in common use for physiology/function—velocity mapping: Each pixel has a velocity measurement associated with it and encodes velocity into and out of the plane of the image for the through-plane version and in the plane for the in-plane version. Direction is encoded as either increased signal (*white*) in one direction and decreased signal (*black*) in the other direction. Summation of the multiplication of pixel areas by their respective velocities yields flow in the through-plane version. Through-plane velocity mapping—the upper left image is a velocity map across the atrioventricular valves in a patient with transposition of the great arteries, whereas the upper middle panel is an axial phase-encoded velocity map of the descending aorta (DAo). The upper right and lower right panels demonstrate the velocity map and anatomic image, respectively, across a normal trileaflet aortic valve. In-plane velocity mapping—the lower left and second from left images are in-plane velocity maps and the anatomic image (*third from left*) of the left ventricular outflow tract of a patient with aortic stenosis and insufficiency. The lower left image is in systole, the second and third from left in diastole.

as anomalous left coronary artery from the pulmonary artery or anomalous coronary artery origins from the opposite sinus, (2) acquired coronary artery abnormalities such as Kawasaki's disease, and (3) iatrogenic coronary artery abnormalities such as those associated with repair of transposition of the great arteries or the Ross procedure. CMR generally utilizes 3D, fat saturated (to suppress the signal from epicardial fat), T_2 prepared (to suppress signal from myocardial muscle) sequences along with navigator techniques (a CMR technique that monitors diaphragmatic motion) to allow for coronary imaging in even small infants (Fig. 73.9).

Regional myocardial perfusion and viability are now in routine clinical use in CMR (Fig. 73.10). Utilizing gadolinium enhancement, CMR assesses regional wall perfusion by using a "first pass" injection technique. Typically, short - axis views of the ventricle are obtained and the sequence set up is such that the heart is imaged relatively motionless. Gadolinium is injected intravenously while the scanner continuously images the ventricle (up to 4 to 5 short axis slices may be imaged at once) and the gadolinium bolus is followed from right ventricular cavity to left ventricular cavity to ventricular myocardium. Defects in perfusion show up as dark portions of the myocardium while the rest of the ventricle is signal intense (Fig. 73.10). It is usually used in conjunction with a coronary vasodilator, such as adenosine, although other agents such as dipyridamole can also be used. Lung perfusion can also be assessed qualitatively using time-resolved gadolinium techniques (Fig. 73.2).

Infarcted myocardium is less of a known issue in CHD than it is in adults; however, native lesions such an anomalous left coronary artery from the pulmonary artery or operations, which scar the myocardium (e.g., repaired tetralogy of Fallot or single ventricles), may manifest myocardial infarction and scarring. Gadolinium is avidly taken up in the extracellular space by scarred myocardium and can remain in the scarred tissue for an extended period of time while it is subsequently "washed" out by coronary blood flow in perfused myocardium. Put another way, the signal intensity–time curves separate, with the infarcted myocardium gadolinium curve remaining highly signal intense after 5 minutes, whereas normal myocardium becomes much less so. CMR sequences take advantage of this property to be able to image infarcted myocardium using an inversion

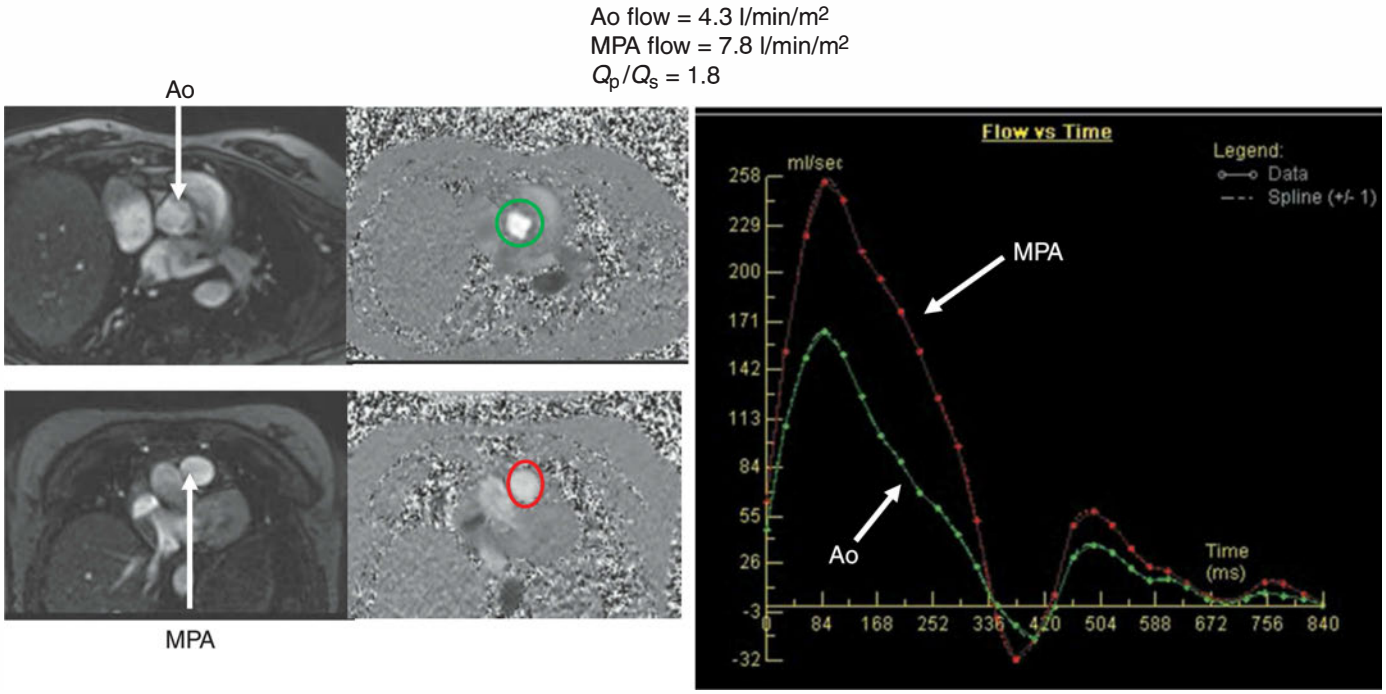


Fig. 73.6. Types of cardiac magnetic resonance in common use for physiology/function—velocity mapping analysis example—Q_p/Q_s: velocity mapping can determine complex or simple flows. For example, in a patient with an atrial or ventricular septal defect, Q_p/Q_s may be determined by placing a velocity map across the aortic (Ao) valve (*top left and top middle panels*) and the main pulmonary artery (MPA) (*lower left and lower middle panels*). The left panels are the anatomic images for reference, and the middle panels are the velocity maps with the cross section of the vessel contoured (*green* for Ao and *red* for MPA). The resultant measurements are graphed in a flow versus time curve (*right panel*). In this particular patient with partial anomalous pulmonary venous connection, Q_p/Q_s was 1.8.

pulse; this ability of CMR to detect myocardial scar is unique in noninvasive imaging. Signal intensity differences between normal and infarcted myocardium of up to 500% have been achieved. The technique has been shown to accurately delineate the presence, extent, and location of acute and chronic myocardial infarction as well as fibrous tissue (Fig. 73.10). Ventricular septal defect patches, right ventricular outflow tract patches as with repaired tetralogy of Fallot also demonstrate signal intensity with this technique. In addition, various cardiac tumors can take up gadolinium, whereas others will not and cardiac MR uses this property, along with T_1-weighted images, T_2-weighted images, and fat saturation to predict what type of tumor is present (Fig. 73.11).

Viability, myocardial edema, and tumor characterization are all examples of a third major function of CMR, which is "tissue characterization"—the ability of CMR to differentiate various forms of tissue from others and to make a diagnosis based on this and other characteristics. Other tissue characterization techniques rely on the T_2 properties of the myocardium to determine the presence of excessive water or iron. T_2 imaging, which has a long TR and TE, is sensitive to water and is used to detect edema in, for example, myocarditis (Fig. 73.10). T_2* imaging, which quantifies the decay of signal at different TEs, will be lower in patients with iron deposits in the myocardium (after all, iron is ferromagnetic) than in patients without; this is used in patients with thalassemia or sickle cell disease to monitor chelation therapy (Fig. 73.11).

There are many other specialized techniques within CMR; the two that will be explored in greater detail in this chapter are:

1. X-ray magnetic resonance (XMR)—the use of CMR in combination with cardiac catheterization. The uses range from superimposition of 3D images generated by CMR onto the fluoroscopy of the catheterization laboratory to act as a "roadmap" for the physician performing the catheterization to performing interventions in the CMR suite—and everything in-between.
2. Computational fluid dynamic modeling is the application of the Navier–Stokes equations, which is the governing equations of fluid flow in the body, to the cardiovascular system. With anatomy from CMR and blood flow measured at the inlets and outlets of the system (e.g., cavae and branch pulmonary arteries of the systemic venous pathway of Fontan patients or the main and branch pulmonary arteries in tetralogy of Fallot), a model of fluid flow can be created and tested at different flow conditions.

This chapter cannot delve into all the specialized CMR techniques although the reader should be aware they exist:

1. Exercise CMR—Performing exercise in the CMR suite itself with a CMR-compatible ergometer. Ventricular function, blood flow, and perfusion at exercise can be obtained by utilizing "real time" cine CMR and "real time" phase-contrast velocity mapping (Fig. 73.12).
2. Fetal CMR—Utilizing "real time" and ultrafast single shot CMR, fetal cardiac anatomy and ventricular function can be obtained. Newer techniques utilize "Metric Optimized Gating" (MOG) allow for measuring flow in fetal vessels (Fig. 73.12).
3. Tissue characterization: BOLD imaging, still being developed, can measure tissue oxygen levels as deoxygenated blood

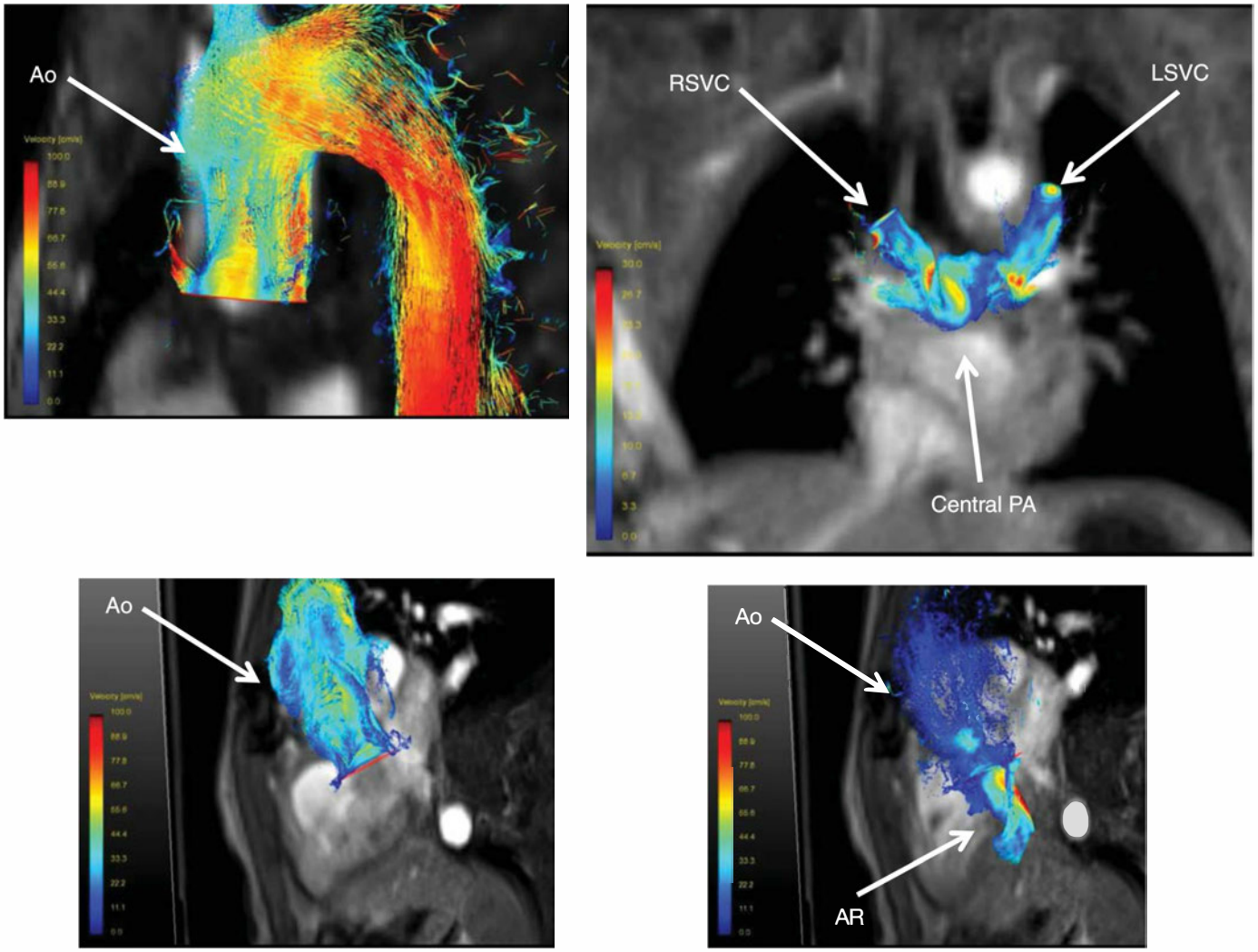


Fig. 73.7. Types of cardiac magnetic resonance in common use for physiology/function— four-dimensional (4D) velocity mapping: By obtaining a "slab" encoding all three velocity vectors, 4D flows can be visualized by streamlines. The upper left panel is 4D flow in the aorta (Ao) of a patient with transposition of the great arteries after arterial switch operation. Lower left and right panels are 4D flows of a patient with a bicuspid Ao valve and aortic insufficiency (AR) with frames in systole (*left*) and diastole (*right*), the plane which the streamlines originating from being the aortic annulus. The upper right image demonstrates 4D flows in a patient with single ventricle after bilateral, bidirectional Glenn. Central PA, central PA; LSVC, left superior vena cava; RSVC, right superior vena cava.

has different magnetic properties than oxygenated blood. T_1 mapping utilizing gadolinium can measure "diffuse" myocardial scarring.

4. Three-dimensional printing is a technique that enables creation of physical 3D models of the cardiovascular system from gadolinium images, which the surgeon or the cardiologist can actually hold in their hands (Fig. 73.12).
5. T_1 mapping: The new and emerging field of "diffuse" myocardial scarring has been developing for a few years. As opposed to viability imaging, which generally detects discreet areas of scar, T_1 mapping techniques utilizes gadolinium to detect diffuse scarring not readily apparent on the viability images.

CARDIAC MAGNETIC RESONANCE IMAGING APPROACH

The imaging approach to the patient with CHD is the same in CMR as it is in echocardiography, which was outlined previously in this chapter. However, there are various approaches that are used in the CMR evaluation of CHD, which any physician in the care of children with heart disease should be aware of. The following is a delineation of the conduct of the CMR examination and the various techniques that are performed. The reader should understand that not all techniques are run on every patient and that the study is tailored to the individual patient.

The initial images obtained are contiguous, tomographic slices in the transverse (also called "axial") plane (images are oriented anteroposterior and right–left). This allows for one complete volumetric data set to be obtained so that if the study is terminated prematurely (e.g., patient instability), multiplanar reconstruction, and 3D data manipulation can still be generated off-line and used in the analysis of the anatomy. This view depicts the following structures (preoperatively): short axes of the ascending and descending aorta, pulmonary and aortic annulus, venae cavae, azygous vein, trachea, and esophagus, the long axes of the transverse aortic arch, main and branch pulmonary arteries, pulmonary veins, and a slightly off-axis apical four-chamber view.

The next set of images obtained are determined by the region of interest.

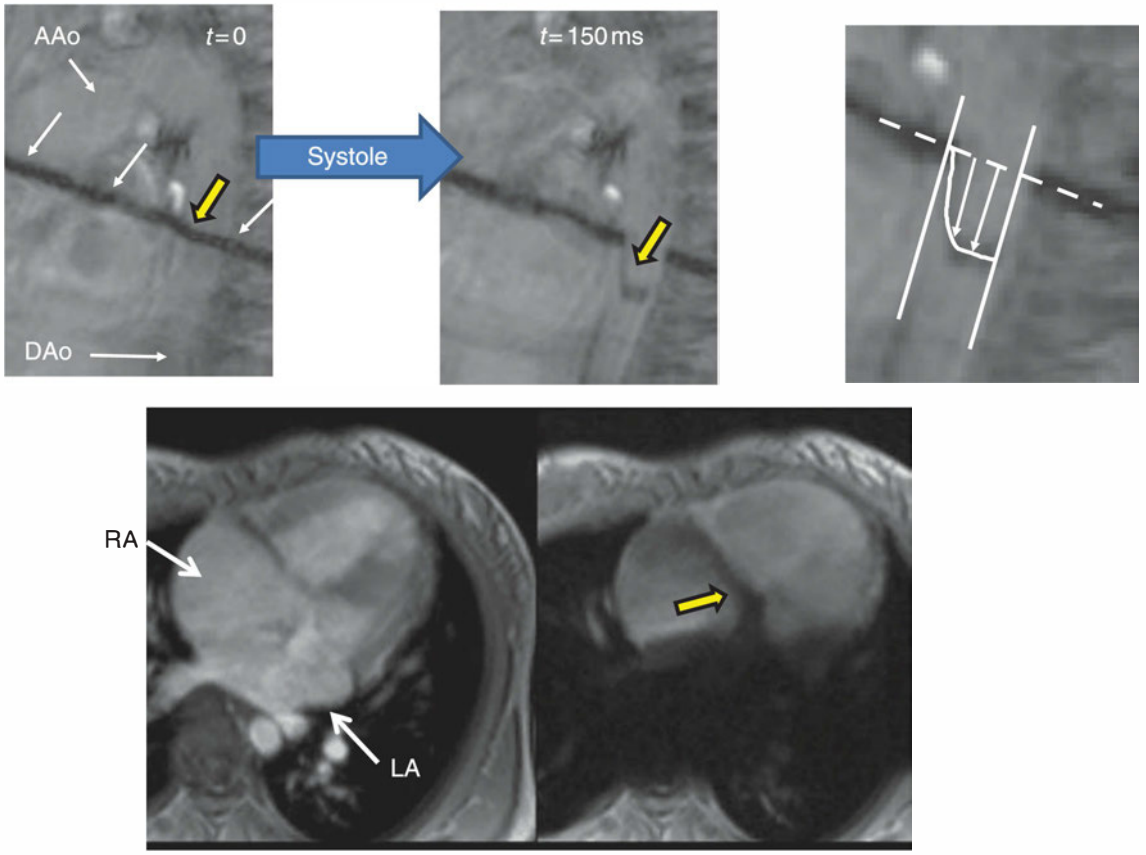


Fig. 73.8. Types of cardiac magnetic resonance in common use for physiology/function—blood tagging: Upper left and upper middle panels display blood (Bolus) tagging in the descending aorta (DAo) in the candy-cane view at end-diastole ($t = 0$) and 150 milliseconds into systole ($t = 150$ ms). White arrows represent the site of initial tagging and the yellow arrow is the tag in the DAo. Note the velocity profile of the DAo in the upper middle panel. The upper right panel is a magnification of the site of tagging in the DAo, showing how velocities at each point across the vessel diameter are measured. With the appropriate mathematical calculation, planimeterizing of the area underneath the velocity profile will yield the regional cardiac index; this technique for calculating cardiac index has been supplanted by phase-encoded velocity mapping. The lower two images demonstrate blood tagging of the left atrium (LA) and ventricle in a patient with a primum atrial septal defect. The left lower panel is the cine image without tagging the left side of the heart and the right image is one with the tag (large *black line* across the left side of the heart). The *yellow arrow* points to flow across the primum defect from LA to right atrium (RA). AAo, ascending aorta.

For example, if a coarctation of the aorta or the systemic venous pathway of a Fontan patient is being imaged, an off-axis sagittal image is used to obtain the "candy-cane" view of the aorta or the long axis of the systemic venous pathway (parallel to the path of flowing blood). If on the other hand, a double aortic arch is being imaged or the left ventricular outflow tract is assessed, a set of straight or slightly off-axis coronal images are obtained to yield the short axes of the right and left aortic arches, the long axes of the amalgamation of these structures into ascending and descending aorta, and the long axis of the left ventricular outflow tract. This can be performed using either dark or bright blood techniques.

It is the author's strong belief that physiology and function must be interpreted in light of the prevailing anatomy. Therefore, once the anatomy is sorted out, cine CMR is the next performed to determine ventricular performance (end-diastolic volume, ejection fraction as examples), valve function (e.g., aortic insufficiency or stenosis), shunts that may be present and to aid in determining stenosis of great vessels. This series of scans is also used to confirm the anatomic information obtained by static imaging (e.g., hypoplasia of the branch pulmonary arteries should show the signal void of turbulence on cine CMR). The static images are used as a localizer for cine CMR imaging (e.g., using the four-chamber view on the static images to obtain the left ventricular short axis for shortening), another reason why the static images are obtained first.

Three-dimensional gadolinium imaging is then performed to obtain a 3D volumetric set of the cardiovascular system. The author prefers to use a dynamic technique such as TWIST, which follows the peripheral injection through the cardiovascular system, creating a 3D image every 2 to 3 seconds. The reader should note that other techniques that are "static" can be performed as well utilizing (1) bolus tracking—a special sequence is used to track the bolus of contrast through the cardiovascular system in real time and when it reaches the region of interest (e.g., branch pulmonary arteries in a patient with tetralogy of Fallot), the 3D gadolinium sequence is run automatically or (2) test dose technique—a sequence is used to determine how long a small test dose of contrast takes for the contrast to reach the region of interest from the time of injection. The 3D gadolinium sequence is then run after the full dose of contrast is injected, delayed by the amount of time determined by the test dose.

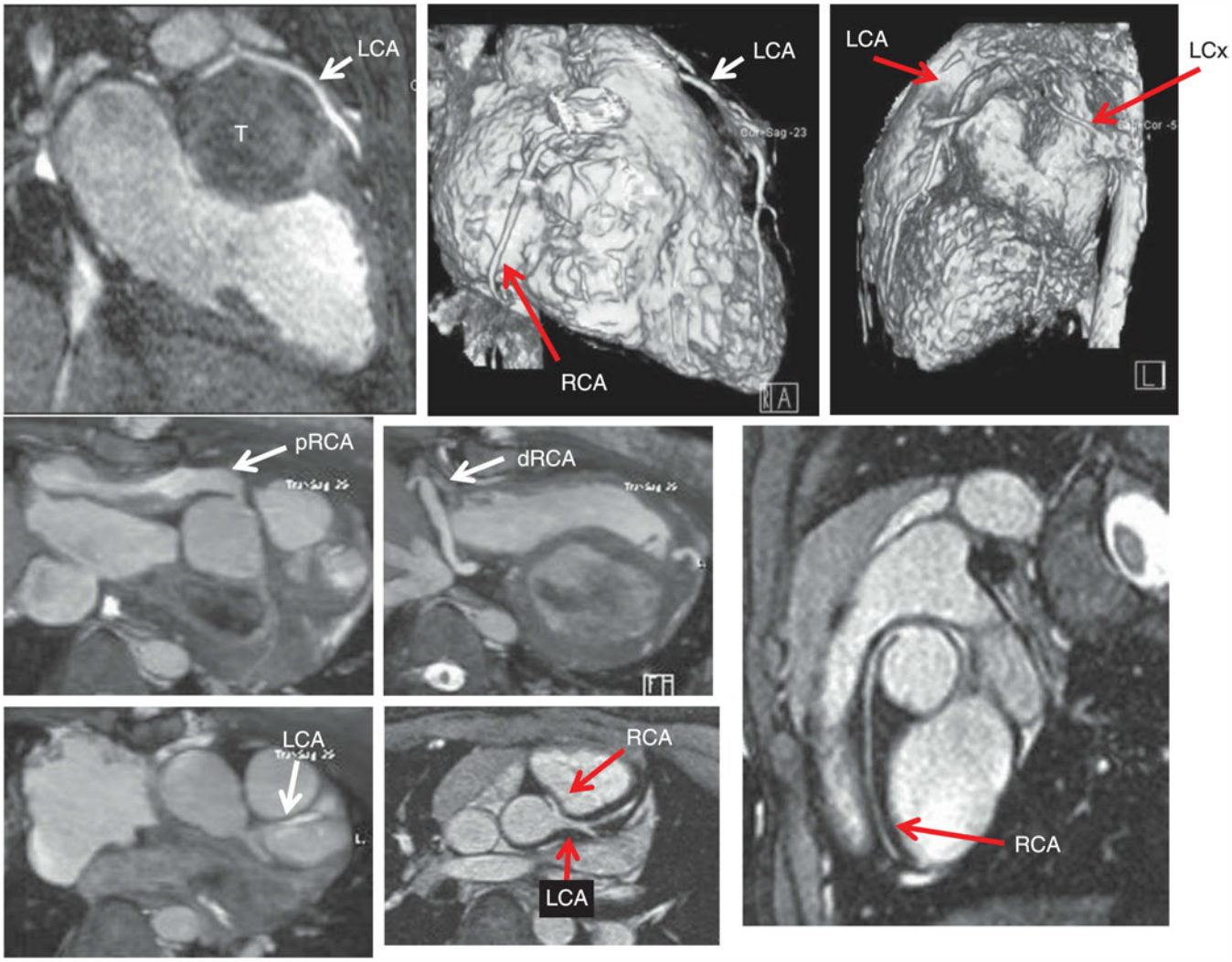


Fig. 73.9. Types of cardiac magnetic resonance (CMR) in common use—coronary imaging: CMR has come into to its own with whole heart, steady-state free precession (SSFP) imaging for coronaries that have compared favorably to catheter and CT (although resolution is less). The upper three images are of a patient with a left ventricular fibroma. The top left panel is an SSFP image of the left coronary artery (LCA)—specifically the left anterior descending coronary artery—coursing just above the tumor. The middle and right panels are 3D reconstructions demonstrating the courses of the right coronary artery (RCA), LCA and the left circumflex coronary artery (LCx); because the tumor is nearly avascular, it is not imaged on the 3D reconstruction. The middle left, second from middle and the lower left panels are taken from a patient who is after heart transplantations with large coronary aneurysms of both the right and left systems. The lower middle panel and lower right panel are from a patient with anomalous origin of the RCA from the left coronary cusp.

After the gadolinium portion of the study, phase-encoded velocity maps are obtained and these are used for flow and velocity information. The vessel to be interrogated is dependent upon the lesion, but typically, velocity maps across the aorta and main pulmonary artery are a minimum which is obtained. This serves two purposes: (1) with no shunts present, the cardiac output of the aorta should equal the cardiac output of the main pulmonary artery and (2) with a shunt present, a Q_p/Q_s may be obtained (Fig. 73.6). This author recommends obtaining right and left pulmonary artery flow when calculating a Q_p/Q_s as well since both flows should add up to flow in the main pulmonary artery. In addition, caval flows are also obtained which should equal flow in the aorta in the absence of aortic to pulmonary collaterals or a systemic to pulmonary artery shunt. These quantitative internal checks are usually performed in CMR and is one of the unique features of CMR as an imaging modality. In addition, ventricular pressure estimates and gradients can be calculated utilizing through-plane and in-plane velocity mapping as one would in Doppler echocardiography.

In a typical scenario, in a patient who is after surgery, delayed enhancement (viability) is the final portion of the study. Since velocity mapping was performed after gadolinium was injected, the requisite 10 to 15 minutes would have passed and the patient would be ready to have scar imaging performed; in this way, little time is wasted during the examination. Generally, delayed enhancement imaging is performed in a stack of contiguous short- and long-axis views of the ventricle; if a questionable region is present, other views are obtained through that region. To ensure that the questionable region is not artifact, images may be obtained with different parameters (e.g., swapping phase and frequency-encoding directions, etc.).

A number of other techniques can be performed in between these sets of sequences, in place of them, or the sequences would be performed in a different order, tailored to the individual patient's

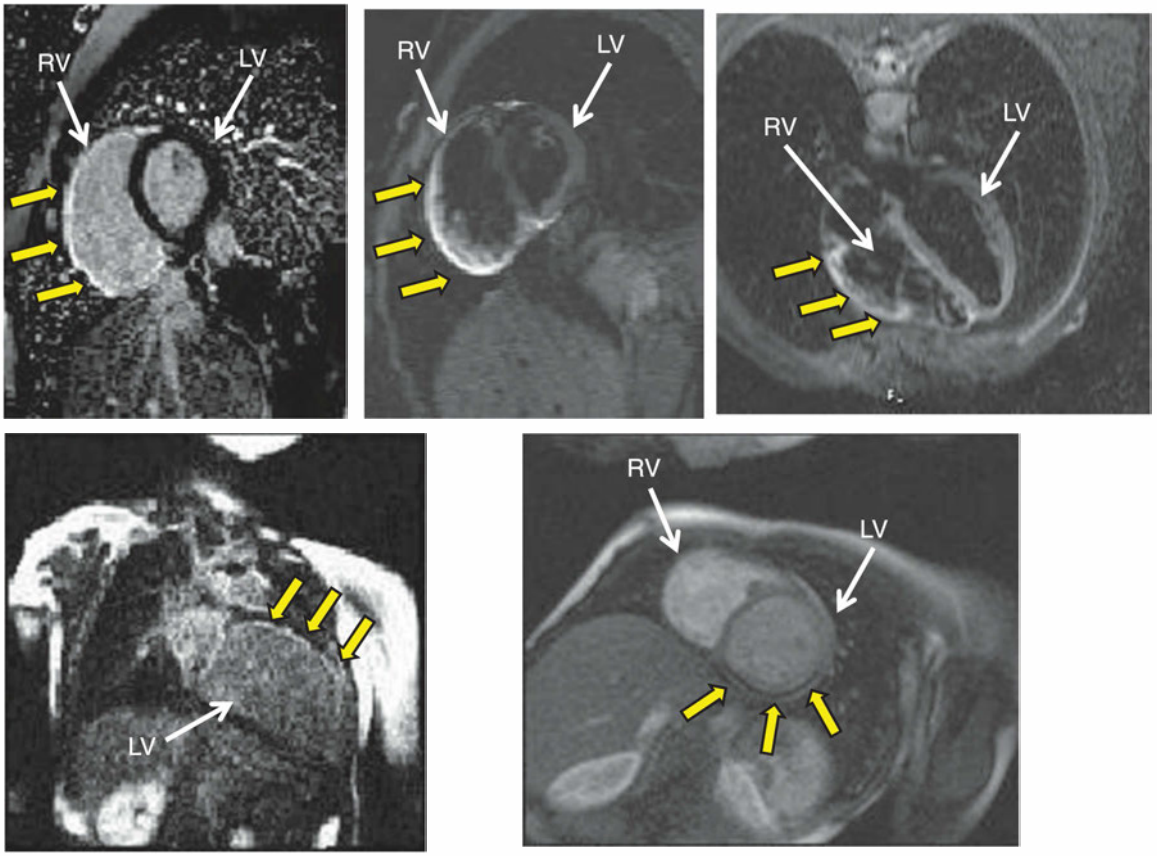


Fig. 73.10. Types of cardiac magnetic resonance in common use for tissue characterization—viability, edema imaging (T_2), and perfusion: delayed enhancement imaging, also known as viability, can detect scar/fibrous tissue; surgically implanted foreign material such as a patch; or can aid in tumor characterization. The infarcted tissue is signal intense (*white*) while normal myocardium is *black* (see text). The upper and lower left panels display a patient with right ventricular (RV) infarction in short axis (*yellow arrows*) and superior wall endocardial scarring of the left ventricle in long axis (yellow arrows), respectively. Normal myocardium should appear black as the left ventricle (LV) in the upper left image. The patient with an RV infarction also demonstrated myocardial edema (*yellow arrows*) with T_2 imaging in the upper middle (RV short axis) and upper right panels (the four-chamber view). Perfusion imaging uses gadolinium injection, and visualization of the myocardial "blush" indicates the degree of myocardial perfusion. The lower right panel is a perfusion image of a patient with transposition of the great arteries after arterial switch operation. Note the inferior and lateral wall perfusion defect of the LV (*yellow arrows*) with adenosine administration.

disease and history. Myocardial tagging or blood tagging can be done after the cine sequences. Coronary imaging can take anywhere from 7 to 10 minutes and if that was the goal, it would be performed after the initial static images and some cine imaging. If perfusion is the goal of the study, this is usually done close to the beginning of the exam but after all dark blood sequences are needed. If myocardial iron imaging (T_2^* imaging) was the goal, that would be performed first. In the case of perfusion, the following sequence of events would be used, taking 60 to 70 minutes:

- Static bright blood imaging
- Cine imaging to obtain a gestalt for myocardial shortening prior to adenosine
- Adenosine perfusion imaging with gadolinium (adenosine 140 µg/kg/minutes for 4 to 6 minutes)
- Cine imaging for function (ventricular volumes and mass)
- Velocity mapping for flows
- Resting perfusion imaging with gadolinium (15 to 20 minutes after adenosine perfusion)
- Coronary imaging
- Delayed enhancement

With this sequence of events, a full functional exam, coronary and perfusion imaging can be obtained efficiently and distinctions can be made between normal perfusion, artifact, ischemia, and infarction.

CARDIAC MAGNETIC RESONANCE: MAJOR USES FOR ANATOMY

In certain situations, CMR is so vastly superior to other imaging modalities that it is the standard of care. There are five broad categories for which CMR has found an important role (both pre- and postoperatively) in the anatomic diagnosis of CHD which, because of the varied and flexible nature of the imaging modality, are not all-encompassing but take into account the vast majority of uses. They are (1) great artery anatomy, (2) imaging extracardiac conduits and intracardiac baffles, (3) complex spatial relationships, (4) venous connections both preoperative and postoperative, and (5) general morphologic evaluation and miscellaneous, *but important* individual diseases are not included in the other categories. This also includes subsections, such as (5A) valve morphology, (5B) tissue characterization (such as characterizing right ventricular dysplasia, identifying myocardial scar tissue or cardiac tumors), and (5C) coronary imaging.

Great Artery Anatomy—Preoperative and Postoperative

Aorta

Lesions of the aorta commonly evaluated by CMR are classified as either (a) ring or

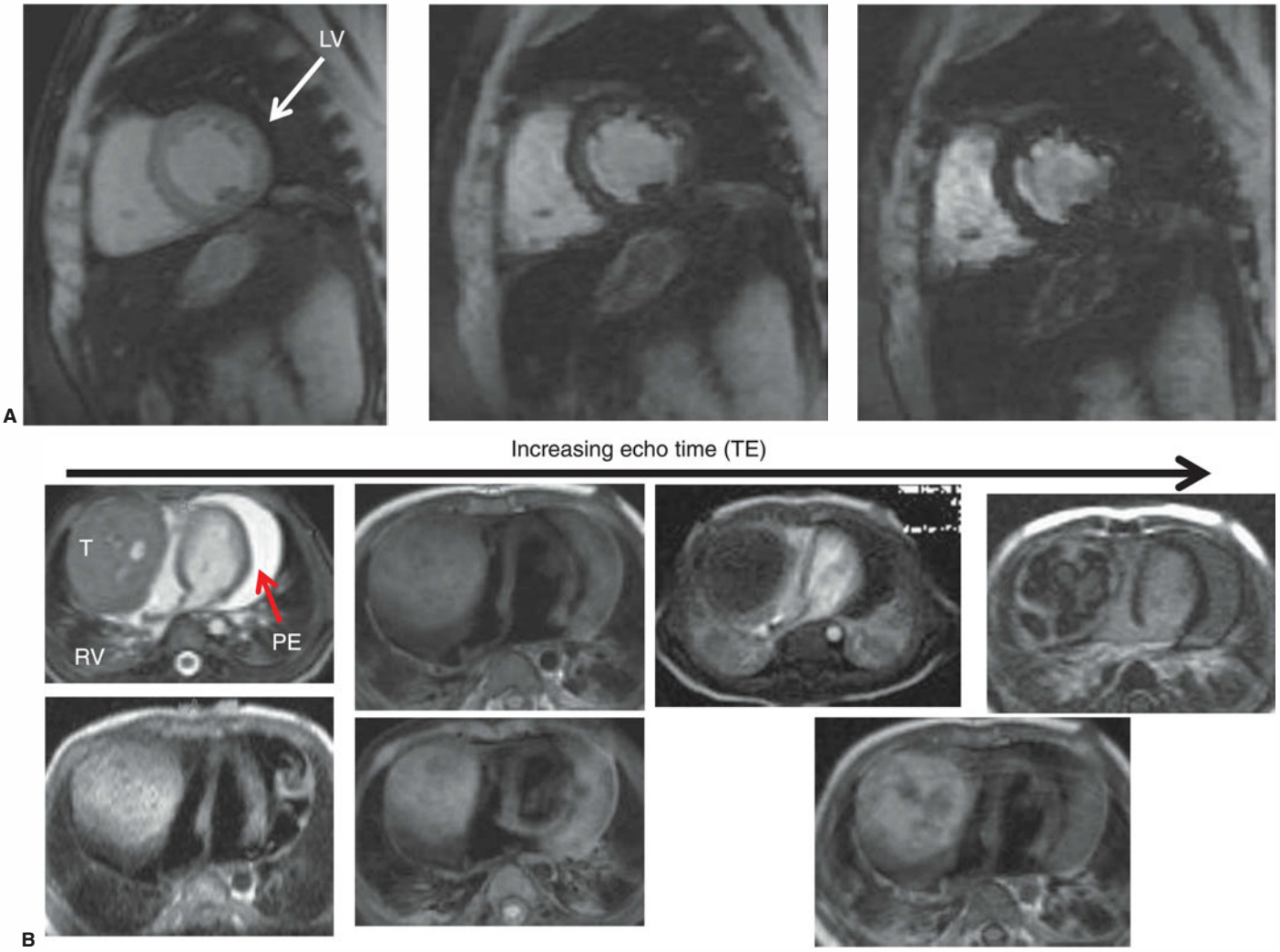


Fig. 73.11. Types of cardiac magnetic resonance in common use for tissue characterization—T_2* for myocardial iron and tumor characterization: **(A)** The T_2* approach uses the fact that iron alters a magnetic field and how much of that iron is present will determine how that magnetic field is altered. The top three panels going from left to right are T_2* images with increasing echo times (TE) from a patient with thalassemia—note how dark the myocardium and liver become and this darkness increases with increasing TE. A T_2* <20 milliseconds puts a patient at risk for decreasing ventricular performance. (**B**) For tumor characterization, there are multiple techniques used by CMR to determine a tumor type. SSFP (*middle left panel*), HASTE imaging (*lower left panel*), T_1 dark blood with and without fat saturation (second from middle left and second from lower left, respectively), perfusion (second from *middle right*), T_1 after gadolinium administration (*lower right*), and viability (*middle right*) are just a few in this patient with a right ventricle (RV) tumor (T). There is a pericardial effusion (PE) and there is obvious RV compression. The heterogeneity of the tumor's signal intensity on different types of imaging and the avascular nature of the tumor on perfusion with a rim of neovascularization suggested a mixed type of tumor with fibrous material. This patient's biopsy showed a mixed cellular fibroma.

(b) non-ring abnormalities (Figs. 73.1–73.3, 73.7, 73.13, and 73.14). A vascular ring is an aortic malformation where vascular structures (or former vascular structures) completely surround the trachea and esophagus, possibly compromising these structures (diverticulum of Kommerel). An advantage of CMR is that imaging the trachea and bronchi as it relates to the vascular ring allows the physician to examine the bronchoarterial relations and find the reason for and assess the amount of airway compression.

In the CMR evaluation of a vascular ring, the initial axial images can almost always make the diagnosis and a set of contiguous coronal images complements the axial ones as an orthogonal view (useful in assessing the diameter of the vascular structures which may ultimately affect the surgical management). Three-dimensional reconstruction is now the standard in assessing the size of the various components of the vascular ring, the geometry, and in the evaluation of tracheal or bronchial compression.

In a double aortic arch, the ascending aorta splits into two arch vessels, which cross over both bronchi and branch pulmonary arteries coalescing behind the trachea and esophagus to form the descending aorta. The aorta totally surrounds the trachea and esophagus and surgical management entails *ligation and division* of the smaller of the two arches, which is more commonly the left one. Since this is performed by thoracotomy, CMR is crucial in determining the surgical management by assessing which arch is smaller to know which hemithorax to enter.

Right aortic arch complexes may form rings, depending upon where regression in the embryonic aortic arches occurs. For example, in a right aortic arch with a retroesophageal diverticulum of Kommerel and an aberrant left subclavian artery (Figs. 73.13 and 73.14), the components

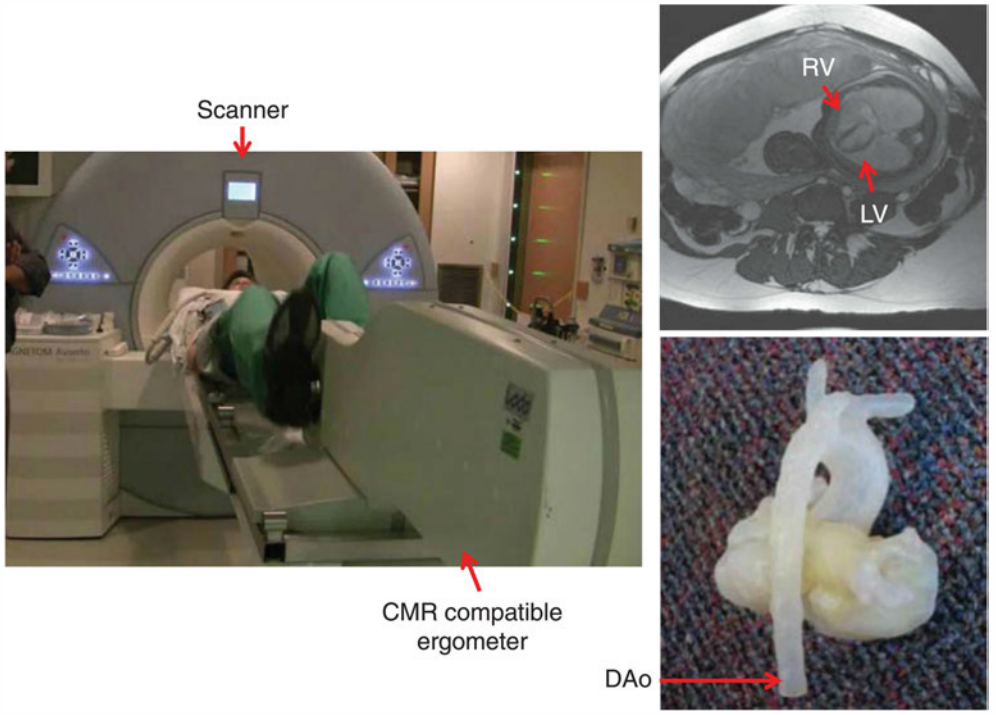


Fig. 73.12. Other types of cardiac magnetic resonance—exercise CMR, fetal CMR, and three-dimensional printing: left image is of a volunteer on a CMR compatible ergometer about to be scanned. The right upper panel is a four-chamber view of a fetus with pulmonary atresia and intact ventricular septum. The right lower panel is a 3D model fabricated from CMR images of a patient with pulmonary atresia and intact ventricular septum with a hypoplastic right ventricle from posterior; only the left-sided structures were printed for this example. DAo, descending aorta; LV, left ventricle; RV, right ventricle.

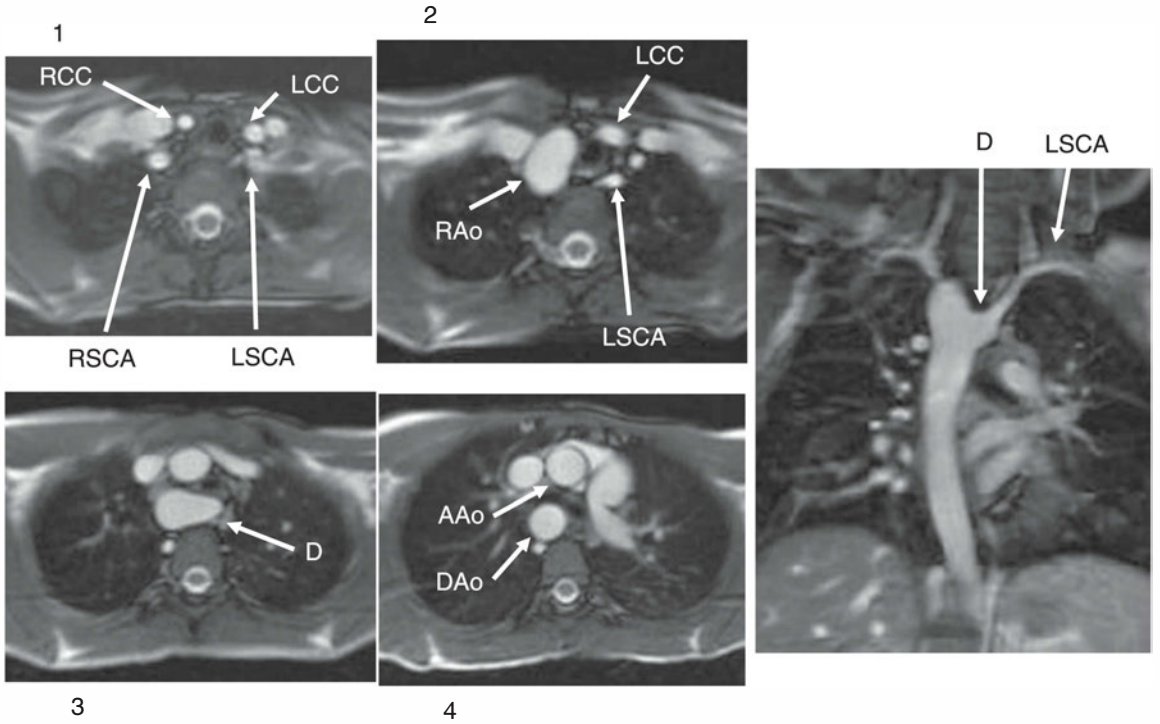


Fig. 73.13. Right aortic arch with a diverticulum of Komerrel and aberrant left subclavian artery (LSCA)—bright blood images: The upper left, upper middle, bottom left, and bottom middle are static bright blood axial images of a child with a right aortic arch (RAo) with a diverticulum of Komerrel (D) and aberrant LSCA. Images progress from superior to inferior as the numbers progress from one to four. The panel on the right is an off-axis bright blood coronal view demonstrating the D. AAo, ascending aorta; DAo, descending aorta; LCC, left common carotid artery; RCC, right common carotid artery; RSCA, right subclavian artery.



Fig. 73.14. Right aortic arch with a diverticulum of Komerrel and aberrant left subclavian artery (LSCA)—trachea and three dimension: The left upper and lower panels are images of the trachea (T) from off-axis coronal and sagittal views from the patient in Figure 73.12, respectively. The middle and right panels are two views from the 3D imaging demonstrating the right aortic arch, diverticulum of Komerrel (D), and the aberrant left subclavian artery.

of the ring are as follows: anterior and right lateral portions formed by the aorta, posterior portion from the diverticulum, and left lateral portion formed by the left ligamentum arteriosum and left pulmonary artery (LPA). The right panels of Figure 73.14 is a volume-rendered image of such a lesion.

It should be noted that echocardiography can generally not totally rule out the diagnosis of a vascular ring. A small retroesophageal ductus (ligamentum) will cause a ring whether right or left aortic arch, and this can almost never be diagnosed by echocardiography. In addition, echocardiography relies on the rule of thumb that the first branch of the aorta is contralateral to the sidedness of the arch (although technically, this branch needs to be visualized to bifurcate before this rule can be applied; even then, an early bifurcation of the carotid cannot be ruled out). There are significant exceptions to this rule that can occur such as a right aortic arch with an aberrant left innominate artery or a right aortic arch with an atretic/absent left carotid artery, and aberrant left subclavian artery both of which would have the first branch being the right carotid artery; the echocardiographer would then assume this was a left aortic arch when indeed it was a right aortic arch.

With nonring aortic arch abnormalities, as with ring abnormalities, the initial axial images can generally yield the diagnosis or at least lead one in the correct direction. Instead of a set of coronal images, a set of oblique sagittal images are obtained parallel to the path of flowing blood allowing the whole arch to be visualized in one picture. In both ring and nonring abnormalities, 3D imaging with gadolinium or noncontrast techniques (T_2 prepared, navigated SSFP) is nearly always obtained.

Of the nonring aortic arch abnormalities, assessment of the aortic arch for coarctation (Fig. 73.1, upper panel on left) is one of the most frequent referrals seen. Four extremity blood pressures and an image of the coarctation by CMR are all that are necessary for the diagnosis and referral for surgery CMR will also be used to assess for a bicuspid aortic valve and other associated abnormalities. Preoperatively, by measuring aortic flow immediately distal to the coarctation and then at the level of the diaphragm, CMR can be utilized to determine and quantitate collateral flow seen in this disease. In a normal aorta, the flow at both levels would be approximately the same or the flow in the descending aorta at the level of the diaphragm slightly lower than just distal to the isthmus; in a coarctation, because of collateral flow, aortic flow at the level of the diaphragm would be higher than that just distal to the coarctation. Postoperatively, CMR is used to assess the presence of coarctation in patients with aortic arch reconstruction or may be used a follow-up in patients who have had coarctation repair, monitoring for recurrence of coarctation or aneurysm formation. Other nonring aortic abnormalities include interruption of the aortic arch, supravalvar aortic stenosis (e.g., caused by Williams syndrome) or dilation of the ascending aorta because of

valvar aortic stenosis (Fig. 73.1), postoperative assessment of the adequacy of an aortic to pulmonary anastomosis in patients who require left ventricular outflow reconstruction, or ascending aortic aneurysm formation such as after a "wrap" procedure.

Pulmonary Artery

Initial assessment of the pulmonary arteries is performed from the axial images, followed by long-axis views of the individual branch pulmonary arteries. Pulmonary artery lesions that were imaged by CMR fall into three basic categories. Stenosis or hypoplasia of one or both branch pulmonary arteries or main pulmonary artery (e.g., pre- and postoperative tetralogy of Fallot or tricuspid atresia) is one category, in which the data are important preoperatively to help predict outcome and may affect the conduct of the surgery as pulmonary artery augmentation may be necessary. Pulmonary artery size is thought by some to be one of multiple prognosticators of surgical outcome in patients with single-ventricle lesions leading to Fontan reconstruction. CMR can characterize the geometry and size of the arteries, determine branch pulmonary artery discontinuity (ideally imaging in three orthogonal planes), the amount of collaterals present from the aorta, and if pulmonary atresia is present, to determine how far the main pulmonary artery extends to the base of the heart.

Aneurysmal dilatation of the pulmonary arteries is the second category. This occurs classically in the patient with tetralogy of Fallot with absent pulmonary valve leaflets but may also occur in patients with pulmonary artery hypertension or the poststenotic dilatation of pulmonic stenosis. CMR will yield the size and geometry of the pulmonary arteries and the amount of respiratory compromise. The last category is anomalous origin or course of the pulmonary arteries (Fig. 73.15). This category includes, for example, anomalous origin of the LPA from the right pulmonary artery (a pulmonary artery "sling") where tracheal embarrassment may occur, anomalous origin of the right pulmonary artery from the ascending aorta (hemitruncus), or when one great artery arises from the base of the heart and gives rise directly in its ascending portion to the systemic, pulmonary, and coronary circulations (truncus arteriosus). In addition, postoperative assessment of patients after arterial switch procedure and the Lecompte maneuver falls into this category. In all cases, CMR can and should be used to delineate size, geometry, and site of origin.

Extracardiac Conduits and Baffles

Echocardiography may be hampered in imaging these structures because extracardiac conduits frequently pass immediately underneath the sternum or near the lungs, and parts of the intracardiac baffle are posterior in the atria or ventricle. CMR can usually succeed where echocardiography fails.

Extracardiac conduits fall into a few categories. A right ventricular-to-pulmonary artery conduit (e.g., a Rastelli procedure), an apical left ventricular-to-pulmonary artery conduit (e.g., {I,L,L}, severe pulmonic

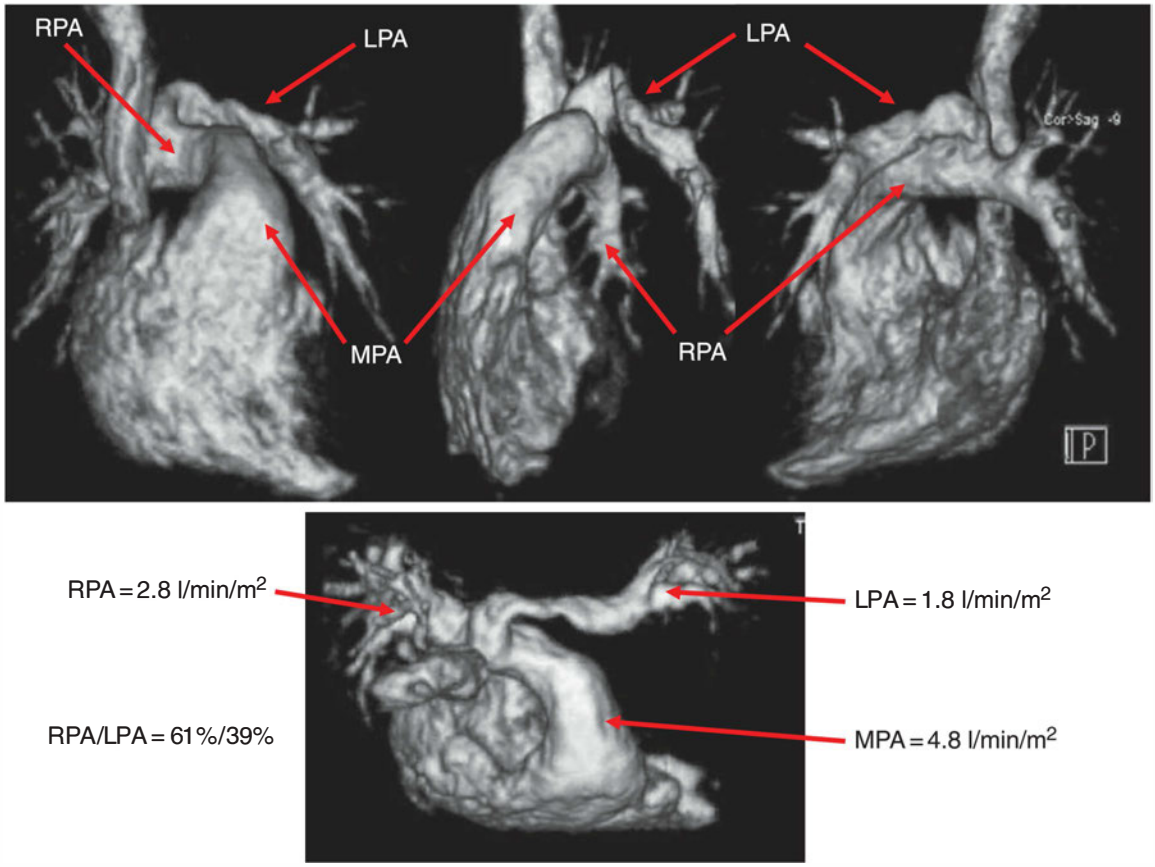


Fig. 73.15. Pulmonary artery sling: The top panels demonstrate a three-dimensional image of a pulmonary artery sling from various views of anterior (*left panel*), lateral (*middle panel*), and posterior (*right panel*) where the left pulmonary artery (LPA) arises from the right pulmonary artery (RPA). The lower panel is a transverse (axial) view. Because there was narrowing of the LPA, velocity mapping was performed across the main (MPA) and branch pulmonary arteries to determine the relative flows to both lungs. Note how close the MPA flow measurement is to the sum of the RPA and LPA flows.

stenosis, and two good size ventricles), or an apical left ventricular-to-descending aorta conduit (e.g., severe aortic stenosis) are examples of *ventriculoarterial* conduits. An example of a *venoatrial* conduit is a Baffes procedure, whereas an example of an *arterio-arterial* conduits is in the case of an aortic-to-aortic conduit in reconstructing the heart of thoracopagus conjoined twins.

Intracardiac baffles can also be categorized. *Atrial* baffles, which function to channel venous blood to arteries or the ventricles, include the Fontan reconstruction for single ventricle complexes (Figs. 73.2–73.4), an atrial baffle to direct pulmonary venous blood to the left atrium in anomalous venous connection, and the Mustard and Senning procedures (which have been performed in the past) for the transposition of the great arteries. The Rastelli procedure, performed for transposition of the great arteries with ventricular septal defect and pulmonic stenosis, is an example, which has a *ventricular* baffle placed.

After contiguous axial images are performed, which can be used to follow the conduit and baffle in short axis, double oblique-angled 2D images are usually necessary to obtain the long axis of the conduit or baffle in one image. Sometimes, this may be impossible and although the physician must be satisfied with two to three 2D images of the conduit in long axis to obtain its full extent, 3D imaging with gadolinium, or T_2-prepared SSFP imaging will nearly always enable visualization in long axis. Stenosis, regurgitation, or leaks across the baffle can be detected by cine CMR.

Complex Spatial Relationships

The orientation of various cardiovascular structures relative to each other and the rest of the body can be sorted out by CMR easier than other imaging modalities. The ability of CMR to obtain parallel, contiguous, tomographic slices, creating 3D volume-rendered displays and the use of multiplanar reconstruction gives the physician a powerful tool with which to analyze the complex geometry.

As with conduits and baffles, contiguous axial images are performed, which can be used to follow the various cardiovascular structures and, in general, to yield a first approximation of the anatomy. Afterward, double oblique-angled images (coronal angled to sagittal angled to axial) are usually necessary to further delineate the morphology and regions of interest identified on the axial images. Confirmation of certain diagnoses can be made using cine CMR.

Superoinferior ventricles with criss-cross atrioventricular relations is one example of complex spatial relationships (Fig. 73.16). In this lesion, the two ventricles are oriented superoinferiorly instead of anteroposteriorly and right–left with the ventricular septum lying parallel to the axial plane. Further, in connecting atria to ventricles, the atrioventricular valves appear to cross each other, hence, the

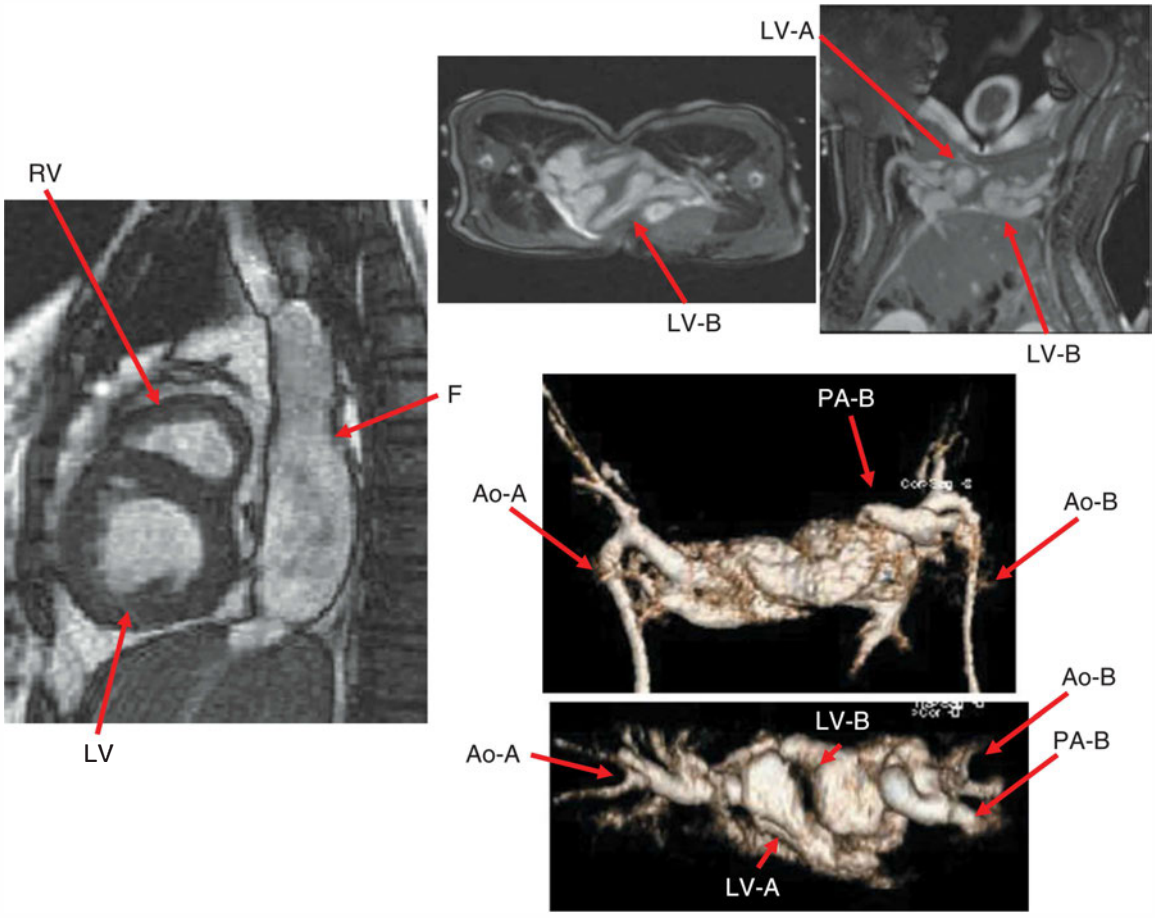


Fig. 73.16. Complex spatial relationships: The left panel is an off-axis sagittal view of a patient with superoinferior ventricles after Fontan (F). The top middle panel and top right panel are bright blood images of thoracopagus conjoined twins (designated as twin "A" and twin "B") from the axial and sagittal views (relative to the babies), respectively. The middle and lower panels are three-dimensional reconstructions from the sagittal and axial views, respectively. All structures could not be labeled as the figure would get too complex. Ao-A, aorta from twin A; Ao-B, aorta from twin B; LV-A, left ventricle from twin A; LV-B, left ventricle from twin B; PA-A, pulmonary artery from twin A; PA-A, pulmonary artery from twin B.

name criss cross. To depict the ventricular relationship, coronal or sagittal images are utilized while the criss cross of the atrioventricular valves can be shown by the standard axial images. Off-axis coronal and sagittal images can be used the delineate the criss cross in the superoinferior plane.

Thoracopagus conjoined twins is another example of a complex spatial relationship (Fig. 73.16). The twins are joined at the thorax and cardiac fusion may occur at one or multiple levels. Echocardiography may be difficult postnatally because of acoustic windows, whereas prenatal imaging may be better in this scenario. CMR is used to sort out the various cardiac structures, including the complex venous anatomy (e.g., status of the inferior venae cavae), ventricular morphology (e.g., fused central [usually left ventricular morphology] ventricle), and abdominal viscera (status of the liver). For all lesions in this category, 3D volume-rendered displays as well as multiplanar reconstruction is useful in conceptualizing the anatomy. Time-resolved 3D gadolinium imaging can be utilized to determine if vascular connections exist between the twins and if so, at which level.

Venous Connections—Preoperative and Postoperative

At times, these may be difficult to visualize by echocardiography because of their position in the chest or poor echocardiographic windows (Fig. 73.17). Many times, it may take a long time for this to be sorted out by echocardiography where 10 to 15 minutes in the scanner is all that is needed, especially with the use of gadolinium (assuming anatomy is the sole goal of the study). CMR is a useful tool in delineating these connections.

Systemic Veins

Sometimes, isolated lesions of the systemic veins may have marked physiologic consequences, such as when the right or a persistent left superior vena cava connects to the left atrium, causing cyanosis (Fig. 73.17, left panel). Other anomalies of the systemic veins may be associated with intracardiac lesions and will affect the conduct of surgery, necessitating the need to be aware of these lesions preoperatively (e.g., if a patient with hypoplastic left heart syndrome has a persistent left superior vena cava not identified prior to Fontan reconstruction, deoxygenated blood will enter the pulmonary venous pathway, mixing with the oxygenated blood and causing cyanosis). The necessity for identifying this lesion is obvious and, therefore, important to diagnose at CMR. Since most systemic veins run in a superoinferior plane, coronal images are obtained after the contiguous axial images, delineating the vein along its long axis. This allows confirmation of the connections, assessment of the size of the vessel, and identification of any areas of stenosis.

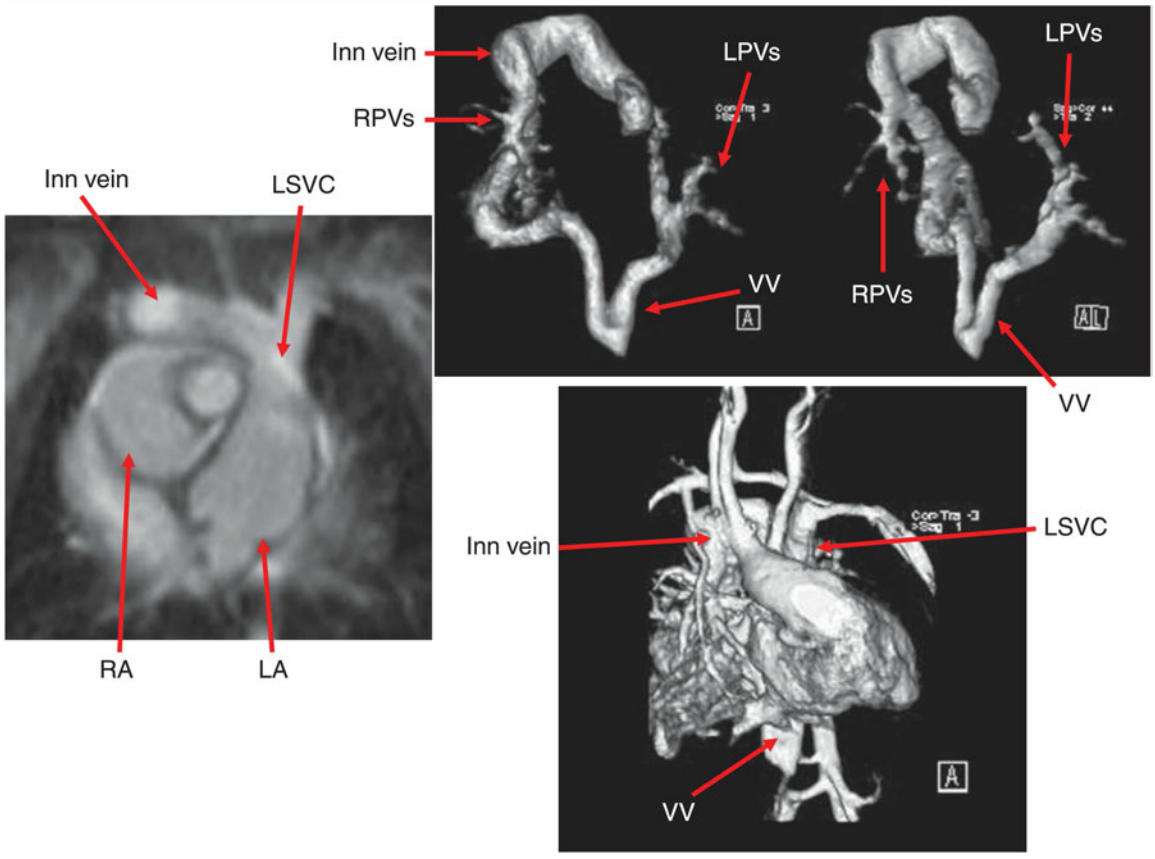


Fig. 73.17. Venous anomalies: The left panel is an off-axis coronal view of a cine image of a newborn with a left superior vena cava (LSVC) connected to the left atrium (LA) without a right superior vena cava. The other images are three-dimensional reconstructions of a patient with complex total anomalous pulmonary venous connections—the left pulmonary veins (LPVs) connect to a vertical vein (VV), which descends on the left below the diaphragm, makes a hairpin turn superiorly into the right hemithorax to ascend and accept the right pulmonary veins (RPVs), and connects to the innominate vein (Inn vein) which connects to an LSVC. The top images isolate the RPVs, LPVs, and VV in two different views; the bottom image has the whole cardiovascular tree visualized from anterior.

Pulmonary Veins

Axial images can delineate the pulmonary venous anatomy and can also follow a vertical vein along its course through the thorax or abdomen (Fig. 73.17, middle and right images). Off-axis coronal images can then be obtained to confirm the diagnosis by visualizing the pulmonary veins in long axis.

The number of anomalies of pulmonary veins is myriad and their importance are obvious and are dealt with elsewhere in this textbook. Examples include Scimitar syndrome which has an anomalous large, single right pulmonary venous connection to the right atrium, total and partial anomalous pulmonary venous connection.

General Morphology and Miscellaneous Diseases

CMR can also be used for general morphologic evaluation as well. It can be of great utility in the older child, adolescent and adult for this and because of the wide field of view, a single CMR study may serve the purpose of multiple other studies. For example, in heterotaxy syndrome, where defining the morphology and sidedness of the trachea, liver, gastrointestinal tract, and finding a spleen is important, a CMR may suffice. A chest X-ray, abdominal ultrasound, echocardiogram, abdominal CT scan, and possibly a liver–spleen scan may have to be performed otherwise.

CMR may also add another dimension to the lesion under study because of its reliance on the magnetic properties of the tissue. For example, in an intracardiac myxoma or tumor (Fig. 73.9, top left panel), the acoustic contrast (by echocardiography) may not be the same as the contrast on CMR (utilizing different pulse sequences and injecting gadolinium [which magnetically enhances vascular structures] to demarcate the extent of the tumor and its water content).

Other examples of common lesions CMR is used for, or have been diagnosed incidentally, are shown in all figures of this chapter. A pericardial effusion (Fig. 73.11) and single ventricle lesions (Figs. 73.2–73.4), for example, are fairly straightforward to recognize on CMR. Some other examples not shown include an aorto-pulmonary window, which is visualized by the turbulent jet of blood shunting from aorta to pulmonary artery in the coronal and axial views by cine CMR. Occasionally, a dark blood scan can identify this lesion as well and the shunt can be quantified using phase-encoded velocity mapping. Finally, left juxtaposition of the atrial appendages can be visualized in the coronal and axial (less well) plane of which whose findings generally include a ventricular and/or atrial septal defect, tricuspid atresia or stenosis, a hypoplastic right ventricle, pulmonic stenosis, and bilateral infundibulum.

With the advent of faster imaging sequences and improvements in hardware as well as software, capabilities of CMR for uses of generalized anatomy have expanded greatly. For example, valve morphology (category 5A referred to above) can be performed by CMR now coming close to and many times equaling echocardiography. Examples of how easily these are seen are shown in Figures 73.1 and 73.5. CMR can use en-face imaging to view this type of morphology using either SSFP or SGE sequences with a high flip angle. Even phase-encoded velocity mapping can be used to determine valve morphology.

In addition to valve morphology, tissue characterization plays an important role in CMR (category 5B referred to above). The tool has been advocated to be used to identify patients with right ventricular dysplasia by characteristics such as (1) fatty substitution of the myocardium, (2) ectasia of the RVOT, (3) dyskinetic bulges or dyskinesia of RV wall motion, (4) a dilated RV, (5) a dilated RA, and (6) fixed RV wall thinning with decreased RV wall thickening. Tumor characterization is also an important function of CMR and is based on the tissue characteristics by T_1 and T_2 weighting (with and without fat saturation), its signal intensity during and after gadolinium injection (Figs. 73.9 and 73.11), and the contraction pattern on myocardial tissue tagging. Finally, identification of myocardial scar tissue by the use of delayed enhancement 5 to 10 minutes after gadolinium injection (Figs. 73.10 and 73.11) is useful to determine ventricular performance, myocardial perfusion (see below), or as an etiology for arrhythmia as just a few examples.

Coronary imaging by CMR (Fig. 73.9) is a major emphasis in adults with heart disease; however, there is much applicability in pediatrics and CHD as well (category 5C referred to above). This is typically performed by obtaining a 3D volume data set using SSFP or (much less commonly) SGE sequences utilizing the Navigator technique, which monitors diaphragmatic motion and allows acceptance of data from only one part of the respiratory cycle (i.e., no need for breath-holding). There are the more well-known coronary anomalies found in pediatrics and CHD such as anomalous left coronary artery from the pulmonary artery, but others such as single coronary arteries, origin of the left coronary artery from the right sinus of Valsalva, or its mirror image which also need to be delineated. The status of the coronary arteries in coronary transplantation in surgeries such as the Ross procedure or the arterial switch procedure for transposition of the great arteries can also be assessed with this technique. Other CMR techniques can be used in conjunction with structural imaging of the coronaries such as delayed enhancement to evaluate for myocardial infarction (see above) or perfusion imaging (see below), once again demonstrating CMR as a "one-stop-shop" in imaging.

CARDIAC MAGNETIC RESONANCE: MAJOR USES FOR PHYSIOLOGY AND FUNCTION

Some of the uses of CMR for physiology and function are extremely clinically useful (e.g., accurate measurement of ventricular volume in a patient with borderline left ventricular size to determine whether it can support the systemic circulation) and play an important role in the management of patients (Figs. 73.4–73.8). Others are in physiology or ventricular function/fluid mechanics research or are still in the experimental or development stage clinically. And still others are on the verge of acceptance (e.g., computational fluid dynamics). Nevertheless, the physician taking care of patients with CHD should be aware of all of these capabilities as the research ones will no doubt enter clinical practice in the near future. Presented are just the most common uses in practice and in research.

The various techniques may be used in conjunction with each other. For example, to obtain the mitral regurgitant fraction in a patient with mitral insufficiency, cine CMR is used to obtain left ventricular end-diastolic volume, stroke volume, and total cardiac output. Phase-encoded velocity mapping in the aorta measures forward stroke volume, and the subtraction of this forward stroke volume from the total stroke volume measured by cine is the mitral regurgitant volume (which if divided by the total stroke volume is the mitral regurgitant fraction). Alternatively, velocity mapping across the mitral valve in diastole combined with forward flow across the aortic valve in systole allows for another method to calculate mitral insufficiency.

A feature unique to CMR is the capability of internal checks used in quantitative data, which is unique to noninvasive imaging. To use the example above, phase-encoded

velocity mapping across the mitral valve in diastole should yield the same flow volume as the mitral regurgitant volume calculated by cine imaging of the ventricle. Similarly, in patients without intracardiac shunting, velocity mapping across the main pulmonary artery should equal velocity mapping across the aorta, flow in the branch pulmonary arteries should match flow in the main pulmonary artery, and flow in the cavae should match flow in the aorta in the absence of aortic to pulmonary or veno-veno collaterals. All these strengthen the accuracy of CMR for the assessment of physiology and function.

Another unique feature of CMR is that the image can be built over multiple heartbeats and averaged (unlike echocardiography and angiography, a single CMR image can be averaged over two to many hundreds of heartbeats). Some might call this a disadvantage; however, it should be noted that using this approach, functional analysis by CMR can give a better handle on long-term performance. The image itself is an average and this average over many heartbeats is built into the image—the physician does not have to do this averaging "in his head" as in echocardiography.

Cine Cardiac Magnetic Resonance Uses

As noted in the Introduction, cine CMR (SSFP, SGE sequences) is used to image cardiac motion and blood flow (Figs. 73.1, 73.4, and 73.8). Blood is signal intense in this pulse sequence and myocardial tissue is less so and turbulence yields a signal void in the blood.

To assess ventricular shortening, a single-level short-axis view or multiple levels of the short axis of the ventricle can be obtained. This may be complemented by cine in the apical four-chamber view or the ventricular long-axis view. Regional wall motion abnormalities can be grossly visualized in this manner. Furthermore, some investigators have been using dobutamine infusions to assess the myocardium in a stressed state and determining regional wall motion abnormalities. Most software on present day scanners can give a temporal resolution of about 20 milliseconds. Newer techniques now allow for "real time" visualization of ventricular performance or "interactive scanning," where the real-time technique can be used and the user can change planes instantaneously, similar to echocardiography. ECG gating is not a requirement in these instances and, therefore, makes it very useful in patients with arrhythmias. Modifications of existing techniques use "arrhythmia rejection" algorithms to successfully image the heart in patients with arrhythmias. Clearly, a physician versed in all the nuances of CMR is needed to choose the best imaging for the particular task at hand.

Ventricular volume, mass, stroke volume, ejection fraction, and cardiac index may be accurately assessed by cine CMR. As mentioned earlier, this technique does not rely on any geometric assumptions, which are an advantage with the bizarre ventricular shapes found in CHD. Multiple contiguous cine CMR runs are performed throughout the entire ventricle at the same temporal resolution (scans usually take approximately 5 to 10 minutes) and the data then sorted by time. The results are multiple full volume data sets as low as 20 millisecond intervals. Ventricular volume at a given time (usually end-diastole and end-systole are the times of interest) is obtained by tracing the endocardial borders on all images at that time (done on computer with a cursor and mouse), planimeterizing the areas, multiplying by the slice thickness, and summing the result. Ventricular mass is obtained by tracing the epicardial borders on all images, planimeterizing the areas, multiplying by the slice thickness, summing the result, and then subtracting the ventricular volume. Stroke volume is simply the ventricular volume at end-diastole minus the ventricular volume at end-systole (usually defined by closure of the semilunar valve). Once this is known, ejection fraction is calculated in the usual manner. Cardiac index is simply the stroke volume multiplied by heart rate during the study divided by body surface area. Software packages today make this process easy, and the results may be obtained in a few minutes with minimal user interaction.

Cine CMR, as noted above, may also be used to visualize turbulence of blood flow. This is especially useful when valvar regurgitation or stenosis is present. Grading the regurgitation is similar to color Doppler echocardiographic imaging although the "volume amount" of regurgitation may be calculated using phase-encoded velocity techniques alone or in combination with cine CMR techniques as noted below. Similarly, valvar stenosis may be detected using cine CMR, and peak velocities may be measured using phase-encoded velocity techniques. Caution must be used when using the signal void, as this can be made greater or smaller by manipulation of CMR parameters (such as the echo time); this is similar to echocardiography where the Nyquist limit and color Doppler gain (to name just a few parameters) may be manipulated to change the "aliasing" in the image.

Cine CMR is useful in many disease states in CHD. Postoperative ventricular performance is certainly a common application. Assessments of ventricular performance in patients with single ventricle, tetralogy of Fallot, and transposition of the great arteries are common referrals of cine CMR. Accurate assessment of regurgitant fraction in patients with left atrioventricular valve insufficiency in a postoperative endocardial cushion defect repair is another example. Visualization of dynamic obstruction to the left ventricular outflow tract in the older patient with transposition of the great arteries after an atrial inversion operation is also a useful application.

Phase-Encoded Velocity Mapping Uses

This technique makes use of phase information obtained at CMR to encode velocity (Figs. 73.5–73.7). This may be done in images perpendicular to flow or parallel to flow (see Figs. 73.5 and 73.6). In the images perpendicular to flow (e.g., obtaining a cross-sectional area of the vessel), summing all the velocities in each pixel in a given cross section of blood vessel and multiplying by the area of each pixel will yield flow at that given period of time. Summing all phases of the cardiac cycle will yield flow during one heartbeat. Multiplying by the heart rate will yield the cardiac output. As with cine CMR, most software on present day scanners can give a temporal resolution of up to 20 milliseconds. "Real time" velocity mapping, which can yield real-time flow measurements (i.e., flow vs. time, not just velocity vs. time as in echocardiography) is currently being tested. Cardiac output can be measured to assess ventricular function in such lesions as single ventricles or transposition of the great arteries after atrial inversion. Regurgitant fractions can be measured by simply placing a velocity map across a great vessel and measuring both forward and reverse flows. This would be important, for example, in a patient after tetralogy of Fallot repair with a transannular patch.

As with echocardiography, velocities obtained at CMR are useful to obtain a noninvasive estimate of the pressure using the Bernoulli equation (see above). Similarly, to obtain, for example, the amount of atrioventricular valve regurgitation, one needs only image perpendicular to the atrioventricular valve in systole to obtain volumetric data (as described above), although their can be fraught with error at times. Other alternative methods for the assessment of atrioventricular valve regurgitation are imaging perpendicular

to the atrioventricular valve during diastole (volumetric amount of inflow—see Fig. 73.5) and perpendicular to the semilunar valve during systole (volumetric amount of outflow) and subtract outflow from inflow to obtain the amount of regurgitation. In a similar vein, one can use a combination of cine CMR techniques (measure ventricular volume at end-diastole and end-systole to obtain the total amount of blood ejected by the ventricle) and phase-encoded velocity mapping (the amount of forward flow from the ventricle) to measure the volumetric amount of regurgitation. Further, because CMR can obtain velocities perpendicular to flow, velocities in different regions of the blood vessel at a given level may be obtained and is used in fluid mechanics research of the cardiovascular system.

Flow can be measured to different organs or parts of organs. A Q_p/Q_s may be obtained in patients with an atrial or ventricular septal defect simply by placing velocity maps across the aorta and pulmonary arteries (Fig. 73.6). Internal checks are used to validate the data (i.e., flow in the branch pulmonary arteries should sum to the main pulmonary artery and flow in the cavae should sum to the flow in the aorta in the absence of aortic to pulmonary collaterals). Cerebral blood flow can be measured by velocity mapping across the jugular veins. Right and left pulmonary blood flow can be measured by placing velocity maps across the right and left pulmonary arteries, respectively, and the same information as a nuclear scan can be obtained. This is useful in patients with single ventricles, for example, or with tetralogy of Fallot.

Four-dimensional flow (Fig. 73.7) has begun to play an important role and multiple studies are being published in the literature on this.

Myocardial Tissue and Blood Tagging

A unique ability of CMR is to magnetically tag tissue or blood (Fig. 73.4). This makes use of cine CMR in combination with a special technique, which destroys all the spins in a given plane (resulting in a line of signal void). The result is "slicing up" the myocardium into "cubes of magnetization" or labeling (with a signal void from it and hence a black line on the cine image) the blood. Once the slicing into "cubes of magnetization" is performed at end-diastole, images can be obtained as much as every 20 milliseconds and the distortion of these cubes (in systole or diastole) allows for calculation of regional strain, ejection fraction, and wall motion (Fig. 73.4). Qualitatively, regional wall motion can also be assessed and this can be used in instances where there are questionable areas of contraction on cine imaging. This is similar to speckle tracking in echocardiography; however, these "cubes of magnetization" are in full 2D in the image (across the entire myocardial thickness), can "visualize" qualitatively the deformation, and can be made into a 3D strain map.

In blood tagging, however, the more useful technique is to tag prior to each image (similar to myocardial tagging, images can be obtained as much as every 20 milliseconds) and cardiac index can be calculated (although this has largely been supplanted by phase-encoded velocity mapping) and velocity profiles can be visualized (Fig. 73.8). This can also be used for shunt detection (e.g., isolation of atrial septal defects, e.g., see Fig. 73.8). As with phase-encoded velocity mapping, individual regions of the blood vessel may be isolated to evaluate for flow dynamics. It may also be useful for measuring forward flow (cardiac index) for valvular regurgitation calculations (see above).

Myocardial Perfusion

Regional myocardial perfusion is an important parameter in the assessment of the myocardium in adults, and this plays an important role in pediatrics and CHD as well (Figs. 73.10 and 73.11). Manipulation of the coronaries in diseases stated above such as transposition of the great arteries repair or the Ross procedure can lead to perfusion abnormalities. Anomalies of the coronaries such as in anomalous origin of the left coronary artery from the pulmonary artery or in hypoplastic left heart syndrome can also lead to perfusion defects. Regional wall motion abnormalities, for example, may be caused by a lack of blood supply to a certain region of the myocardium. CMR, utilizing gadolinium enhancement, has the ability to assess regional wall perfusion by using a "first pass" injection technique. Typically, short-axis views of the ventricle as described in the cine CMR section are obtained and the sequence set up is such that the heart is imaged relatively motionless in diastole. Gadolinium is injected intravenously while the CMR scanner continuously images the ventricle (up to 4 to 5 short-axis slices may be imaged at once), and the gadolinium bolus is followed from right ventricular cavity to left ventricular cavity to ventricular myocardium. Defects in perfusion show up as dark portions of the myocardium while the rest of the ventricle is signal intense. This is typically performed with and without an adenosine infusion and then followed 5 to 10 minutes later with delayed enhancement imaging.

Other Uses of Cardiac Magnetic Resonance for Ventricular Function/Physiology

Time-resolved 3D gadolinium imaging (Fig. 73.2), since temporal information is embedded in the technique, can be utilized to determine regional lung perfusion (e.g., no blood flow to the right upper lobe because of a pulmonary embolus) or to determine stenotic versus atretic vessels (e.g., continuity or discontinuity of the branch pulmonary arteries). Newer techniques such as exercise CMR is useful in patients with tetralogy of Fallot which is a disease state known to have decreased exercise performance. Functional fetal CMR may be used to determine ventricular volumes in the fetus with just recently developed techniques enabling the evaluation of fetal blood flow. XMR is being used on a research basis not only to perform interventions in the CMR suite but also to determine lung compliance and plot ventricular pressure–volume loops (see below).

SURGICAL PLANNING AND CARDIAC MAGNETIC RESONANCE

By virtue of obtaining and combining anatomy, physiology, and function, CMR can be utilized to perform surgical planning in a couple of ways—not solely by "showing" the cardiac defect to the surgeon but by actually performing "virtual" surgery and predicting outcome. In addition, with the advent of rapid prototyping and 3D printing (Fig. 73.12), a 3D "physical model" of the heart and a proposed surgery can be created; the surgeon and cardiologist can literally hold the heart of the patient in his/her hand prior to surgery and actually create a baffle or conduit to see how it looks. This has obvious utility in making efficient use of cardiopulmonary bypass and deep hypothermic circulatory arrest times as well as optimizing potential results.

Surgical Planning: Single- vs. Double-Ventricle Repair

Not infrequently, neonates are born with two ventricles where the suitability of

separating the circulations with two pumping chambers is questioned (Fig. 73.18). The left (LV) or right ventricles (RV) may be borderline in size before surgery or after baffle creation. In a patient with double outlet right ventricle, a baffle placed between the ventricular septal defect to the aorta to direct LV blood to the systemic circulation may substantially decrease the right ventricular size below the limits of tolerability. The baffle may not even be feasible if atrioventricular valve chordal attachments are in the way. By utilizing a stack of cine SSFP images of both ventricles, a "virtual" baffle can be created and the ventricular volumes, ejection fraction, and cardiac index assessed; phase-encoded velocity mapping is used as a check on these data. If the cardiac index, end-diastolic volume, or ejection fraction from a ventricle are unacceptably low with the "virtual" baffle, a double-ventricle repair may not be feasible and the patient may progress toward the single-ventricle Fontan pathway. It is true that loading conditions postoperatively may change these parameters, however; quantitative preoperative information may be a better gauge to predicting clinical outcome than the current "eyeball" method currently in use preoperatively.

Surgical Planning—Computational Fluid Dynamics and Modeling

Work performed with Ajit Yoganathan and the bioengineering department at The Georgia Institute of Technology. The Navier-Stokes equations are the fundamental equations that govern fluid flow in the cardiovascular system (Figs. 73.19 and 73.20). By obtaining the geometry of blood vessels, baffles, conduits, and so on in conjunction with 2D or 3D flows at the inlets and outlets of these structures, assessment and visualization of the flows, streamlines, and power can be obtained by applying these mathematical formulae, hence, the term "computational" in the fluid dynamic part. The combination of phase-contrast velocity mapping and static SSFP imaging by CMR allows for just such computations. This has been mostly applied to modeling of the systemic venous (Fontan) pathway of single ventricles but has found application in other diseases such as coarctation of the aorta and tetralogy of Fallot. A number of investigations in single ventricle patients after Fontan have demonstrated the degree of "power loss" in the systemic venous pathway and the effect of geometry and flows on this. In addition, streamlines can be calculated and relative contributions of caval blood to the branch pulmonary arteries can be accurately assessed. This approach not only can evaluate the present status of the circulation in vivo but can also assess various structural modifications of the cardiovascular system that the surgeon or interventional cardiologist might perform. One study, published a few years ago, assessed the effect of LPA stenosis on flow and power loss in single ventricles; this was followed by a "virtual

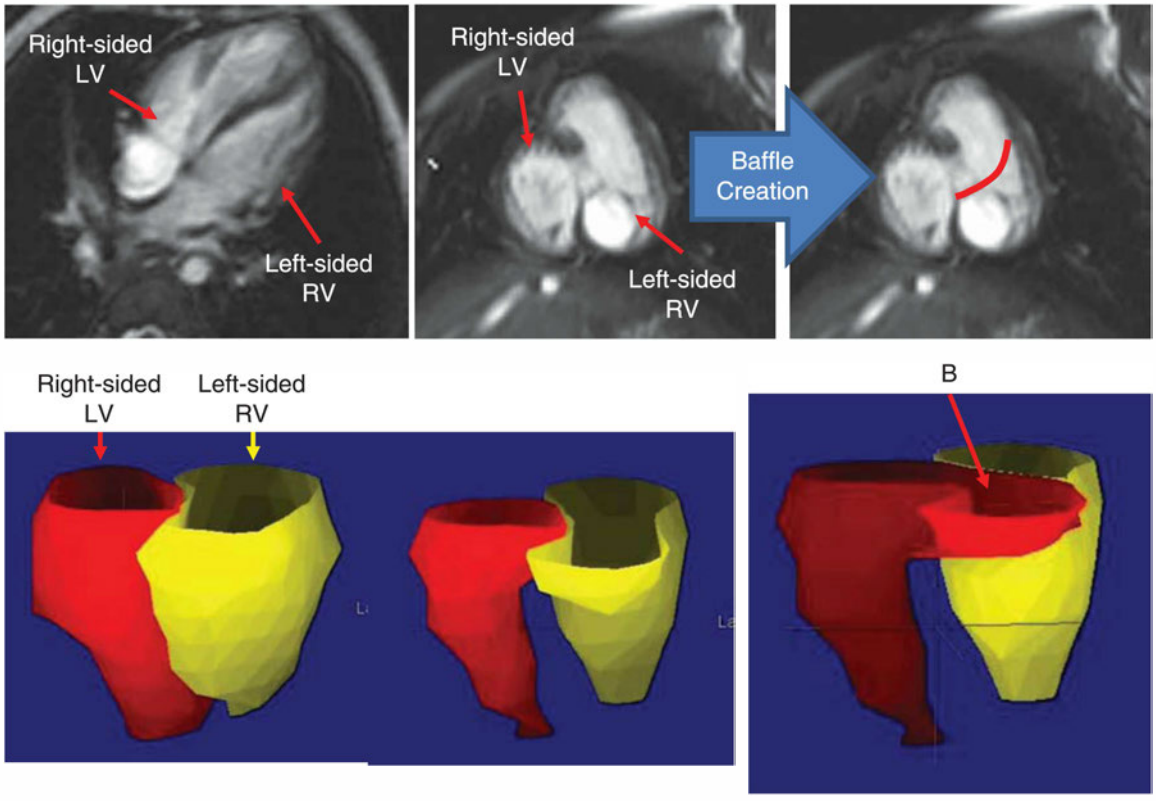


Fig. 73.18. Single- versus double-ventricle repair: In patients where a baffle needs to be created by the surgeon, a "virtual" baffle can be created from the cardiac magnetic resonance images and quantitative data can be obtained to estimate whether the ventricles are of adequate size. The patient is 3-year old with double outlet right ventricle {S,L,L} with a ventricular septal defect, pulmonic stenosis, and Ebstein's anomaly. The four-chamber view and short-axis cine images are in the upper left and upper middle panels, respectively. To repair this, the surgeon would baffle the ventricular septal defect to the aorta (*upper right* image with baffle drawn in *red*). The three-dimensional images of the ventricles before baffle creation (*lower left and middle panels*) and after baffle (B) creation (*lower right panel*) are shown. End-diastolic volumes, stroke volume, ejection fraction, and cardiac index are then calculated to determine if this approach is feasible.

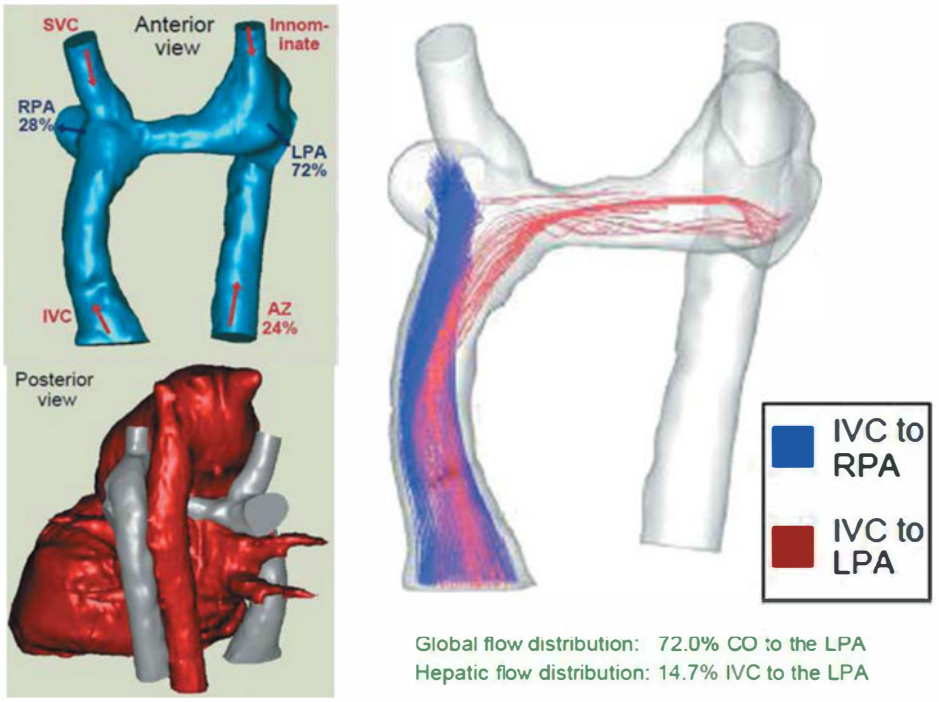


Fig. 73.19. Surgical planning—original anatomy and physiology of single ventricle after Kawashima and hepatic baffle: The patient is a single ventricle patient with interrupted inferior vena cava and azygous continuation after Kawashima operation and hepatic venous baffle with left-sided pulmonary arteriovenous malformations. In this figure, inferior vena cava (IVC) is used to signify hepatic venous blood and not IVC blood. Cardiac magnetic resonance derived anatomy of the systemic venous pathway alone (top left) and with the cardiac mass and aorta in place (lower left) is shown. The image on the right demonstrates the streamlines showing how much IVC blood gets to each one. Data are labeled and suggest why left lung arteriovenousmal formations occurred; little hepatic flow gets to that lung. Figures are created in conjunction with Dr. Ajit Yoganathan and the bioengineering department at the Georgia Institute of Technology. CO, cardiac output; innominate, truly left superior vena cava; LPA, left pulmonary artery; RPA, right pulmonary artery; SVC, superior vena cava.

angioplasty" and a reassessment of flow and power loss when the stenosis was removed. Other studies have looked at etiologies for the development of pulmonary arterial venous malformations (lack of streaming of hepatic blood to a lung) with possible alterations of the geometry to correct this (see below) while still other studies have investigated power loss in the systemic venous pathway during exercise.

With this notion, the idea that a surgeon, a cardiologist, or an engineer can create "virtual" systemic venous baffles and assess the effect of flow and power loss in various geometries has been implemented in select patients. One clinical application is the single ventricle heterotaxy patient with an interrupted inferior vena cava (IVC) with azygous continuation. A Kawashima operation in this type of patient connects the superior vena cava to the pulmonary arteries thereby diverting nearly all systemic blood flow except for hepatic venous flow to the lungs. Without hepatic venous blood, pulmonary arteriovenous malformations typically develop; however, when the hepatic veins are subsequently incorporated into the superior vena cava–pulmonary artery connection, flow phenomenon may occur which would preferentially stream hepatic flow to one lung allowing pulmonary arteriovenous malformations to occur in the contralateral lung. Figures 73.19–73.21 demonstrate one such patient with left lung pulmonary arteriovenous malformations on cardiac catheterization who has a hepatic baffle already created. It can be seen from Figure 73.19 that little hepatic blood (designated IVC blood even though this is really hepatic vein flow) streams toward the LPA. Hepatic venous baffles are virtually created on a workstation for a number of patient-specific geometric options (Fig. 73.20) and flows are then calculated to determine power loss and how much hepatic flow will get to each pulmonary artery at a given total flow to each lung. Figure 73.21 demonstrates the computational fluid dynamic modeling for one such virtual baffle (top left panels of Fig. 73.20). Figure 73.22 demonstrates preoperative planning in a patient with a Kawashima who has yet to have a hepatic baffle placed; three options with their data are presented both visually and quantitatively.

Still in another application, Whitehead et al. utilized CFD in Fontan patients to determine the etiology for decreased exercise performance. With 10 patients of various systemic venous pathway geometries and various flow splits to the branch pulmonary arteries, the cardiac output was increased to two and three times baseline and power loss and flow profiles determined. They found a "dramatic increase in a non-linear fashion" of increasing power loss in the baffle, even when normalized for various parameters. One reason was an increase in the collision of flows between superior and IVC blood. A "resistance index" that takes into account resistance to flow increased during the increasing exercise simulations to such levels that exercise limitation was almost a certainty with pressure drops across the baffle at exercise becoming nonphysiologic (up to 52 mmHg) and therefore unachievable. The work showed the importance of CFD calculations and the possibility of designing various Fontan geometries, which might minimize the exercise limitation.

X-RAY MAGNETIC RESONANCE

Traditional fluoroscopically guided cardiac catheterization in CHD is sufficient to guide catheterization procedures with good results. However, fluoroscopy requires the use of ionizing radiation, has poor soft tissue definition, and only provides 2D projections of complex 3D anatomy. As interventional procedures have become more complex, it is becoming apparent that in many instances, this imaging modality is suboptimal. A different approach to image-guided interventional procedures is to perform the procedures inside the MRI scanner with real-time magnetic resonance imaging (rtMRI) for guidance. While this strategy has promise, certain technical issues, such as the lack of MRI compatible wires and catheters and difficulties with catheter tracking, have limited the current use of this modality although it is currently an active area of research.

Multiple types of cardiac interventions have been successfully performed in the cardiac MRI suite on a research basis. Utilizing XMR to perform electrophysiologic ablations has a special appeal; beside the anatomic capabilities of CMR to be used as a roadmap to place catheters for performing the procedure, the myocardial disruption caused by radiofrequency

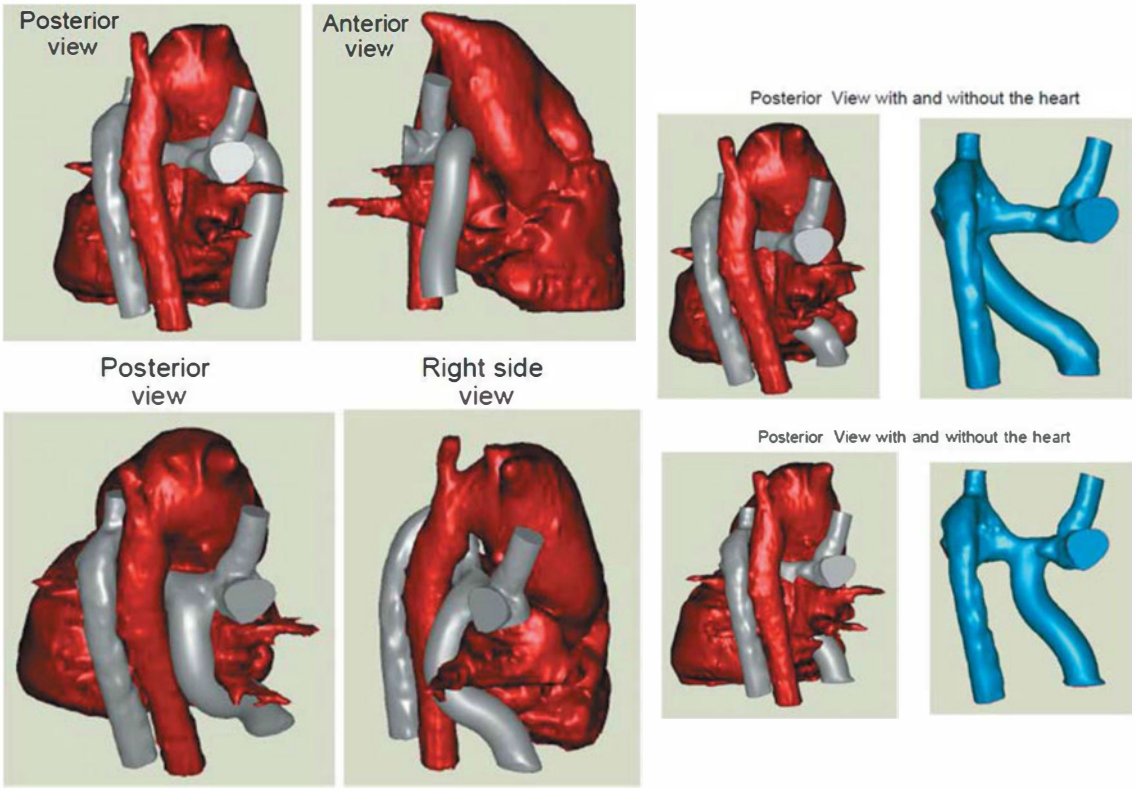


Fig. 73.20. Surgical planning—anatomic options for single ventricle after Kawashima and hepatic baffle: Multiple anatomic options were virtually created by computer for the patient in this figure and were considered of which four are shown as pairs. The top left two panels create the hepatic baffle anterior while the bottom left two panels create the hepatic baffle posterior. The right panels are all intra-atrial; the top two right panels create the hepatic baffle connection to the pulmonary artery very leftward, while the bottom two right panels create the hepatic baffle connection to the pulmonary artery in the center. Figures are created in conjunction with Dr. Ajit Yoganathan and the bioengineering department at the Georgia Institute of Technology.

energy can actually be visualized—on T_1 imaging, the myocardium will turn from a "gray" signal to a "white" signal in a matter of minutes. If a line of scar tissue needs to be created to prevent an arrhythmia, this capability can be used to ensure the line is continuous and no breaks are present that might allow an impulse to pass.

Other interventions solely in the cardiac MRI suite include such procedures as stent placement (e.g., for coarctation), balloon dilation of stenotic branch pulmonary arteries, or valves and creation of atrial septal defects via either balloon or laser (Fig. 73.23). Further, additional physiologic and diagnostic information can be obtained by using a combination of pressure measurements with invasive catheters (either placed in the catheter lab and moved to the CMR suite or placed in the CMR suite itself) and flows of ventricular volumes by CMR simultaneously or near simultaneously. For example, pressure–volume loops or pulmonary arterial impedance can be obtained in this manner; this is an on-going active area of research.

An alternative to exclusive rtMRI-guided interventions or fluoroscopy only guided interventions is to use a hybrid approach where 3D data sets from either CT, rotational angiography, or MRI are fused to the X-ray images to provide the additional information needed while still taking advantage of the good spatial and temporal resolution offered by conventional fluoroscopy (Figs. 73.24–73.26). Fusion of CT and rotational angiography images has been reported in a number of EP studies. Registration is the process of accurately overlaying one imaging technique over the other. Registration of CT and rotational angiography to X-ray images is simpler because of the similarity of the imaging methods making them amenable to automated intensity-based registration approaches. In contrast, registration of MRI images is more complex because of the inherit difference in the imaging modalities. To overcome this problem, fusion of MRI images is often carried out with external fiducial markers. Several groups have reported on fiducial marker-based registration and the utility of this modality to cardiac and vascular interventions. One group has reported on a registration method that did not require fiducial markers but used a catheter-based calibration scheme and visual matching to register MRI data for electroanatomic mapping. Accurate registration of MRI data to fused rotation angiography volumes has been reported in neurological interventions, but this modality requires significant radiation and can require a significant contrast load, which makes this modality suboptimal for interventions in children.

Fiducial marker-based registration has been shown to be accurate to high spatial resolution under certain conditions, but this method has several drawbacks. First, it requires that the MRI scan and catheterization procedure occur successively as the markers need to remain attached during both procedures. Second, skin mobility and movement of internal organs relative to the skin can make this method inaccurate. Third, the need to include the markers in the MRI images requires a larger scanned volume increasing imaging time and could result in reduced imaging resolution. Lastly, fiducial marker-based registration requires propriety software

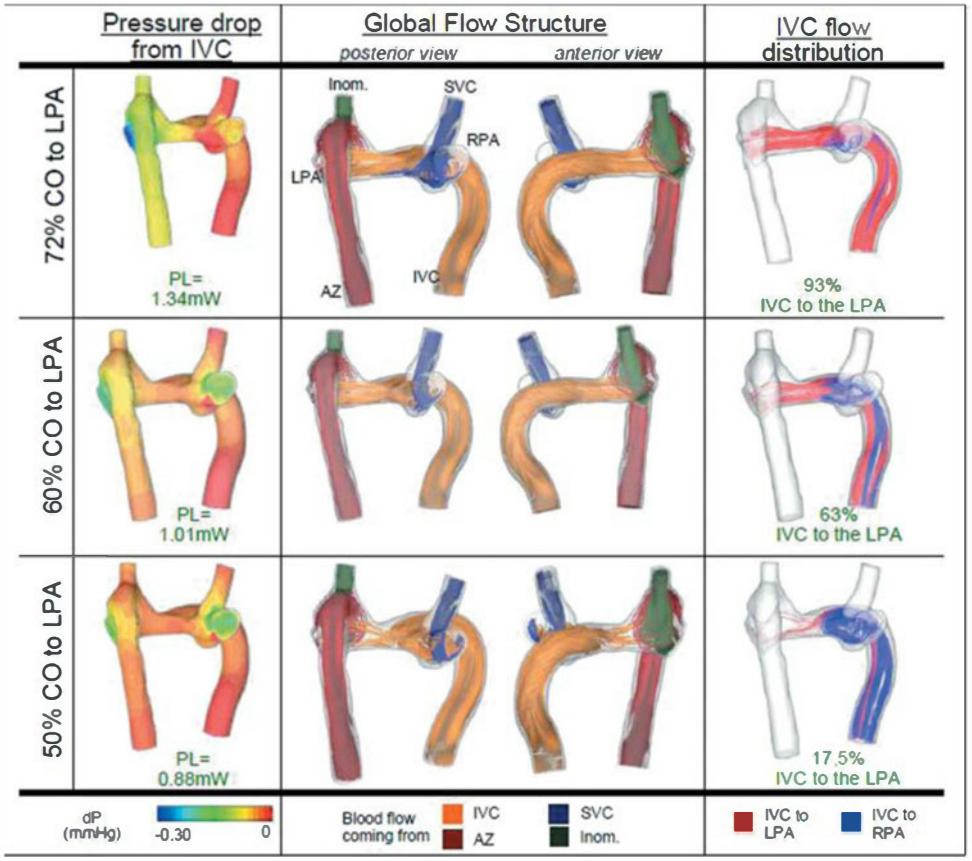
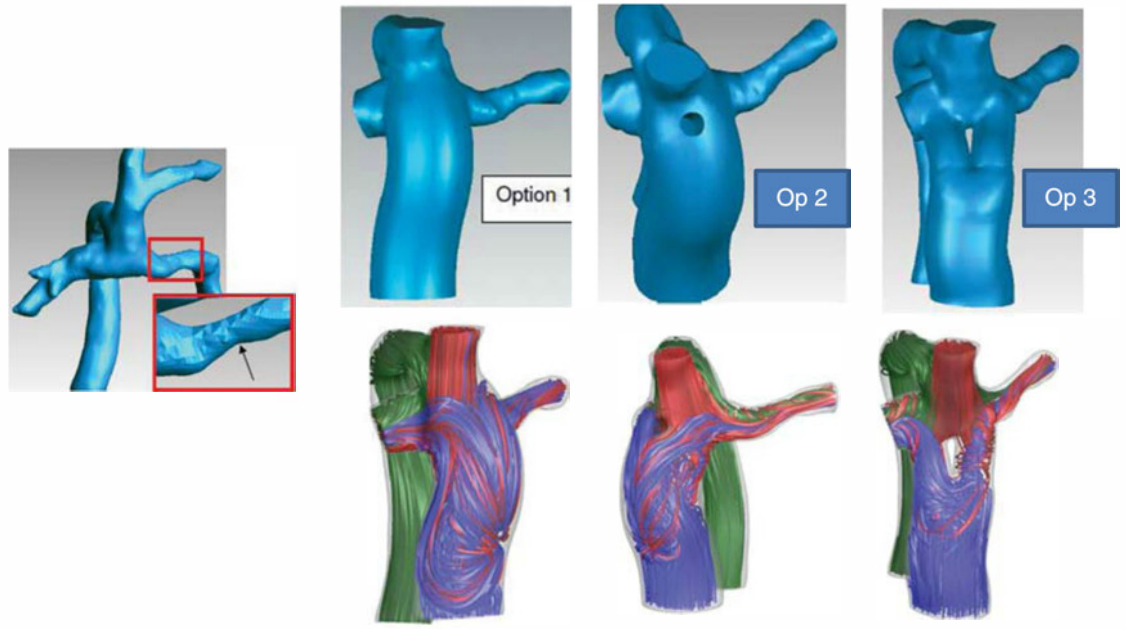


Fig. 73.21. Surgical planning—computational fluid dynamic modeling for the top left option of this figure: The three rows are different global flow distributions of blood to the left pulmonary artery (LPA); 72% on top (initial conditions from the original cardiac magnetic resonance derived data from Fig. 73.19), 60% in the middle, and 50% on the bottom. The left-most column demonstrates the calculated pressure drop (color coded), the middle column demonstrates the global flow distribution (color coded), and the right-most column demonstrates just hepatic flow to each branch pulmonary artery (color coded). Legends for the color code are on the bottom row. Figures are created in conjunction with Dr. Ajit Yoganathan and the bioengineering department at the Georgia Institute of Technology. CO, cardiac output; Innominate, truly left superior vena cava; IVC, hepatic blood flow; LPA, left pulmonary artery; PL, power loss; RPA, right pulmonary artery.



Op 1

GFD (%LPA)	34	50	60
Option 1 (%LPA)	69	76	77

Op 2

GFD (%LPA)	34	50	60
Option 2 w/Divider	20	32	45

Op 3

GFD (%LPA)	34	50	60
Ygraft	31	36	41

Fig. 73.22. Surgical planning—computation fluid dynamic modeling for a single ventricle patient after Kawashima prior to hepatic baffle creation: Original three-dimensional model from cardiac magnetic resonance is shown on the left panel. The three rightward upper panels show three of the multiple anatomic options considered, while the images below show the streamlines. Data for each option (Op) are listed at the bottom of the figure. Figures are created in conjunction with Dr. Ajit Yoganathan and the bioengineering department at the Georgia Institute of Technology. GFD, global flow distribution; LPA, left pulmonary artery; %, percent.

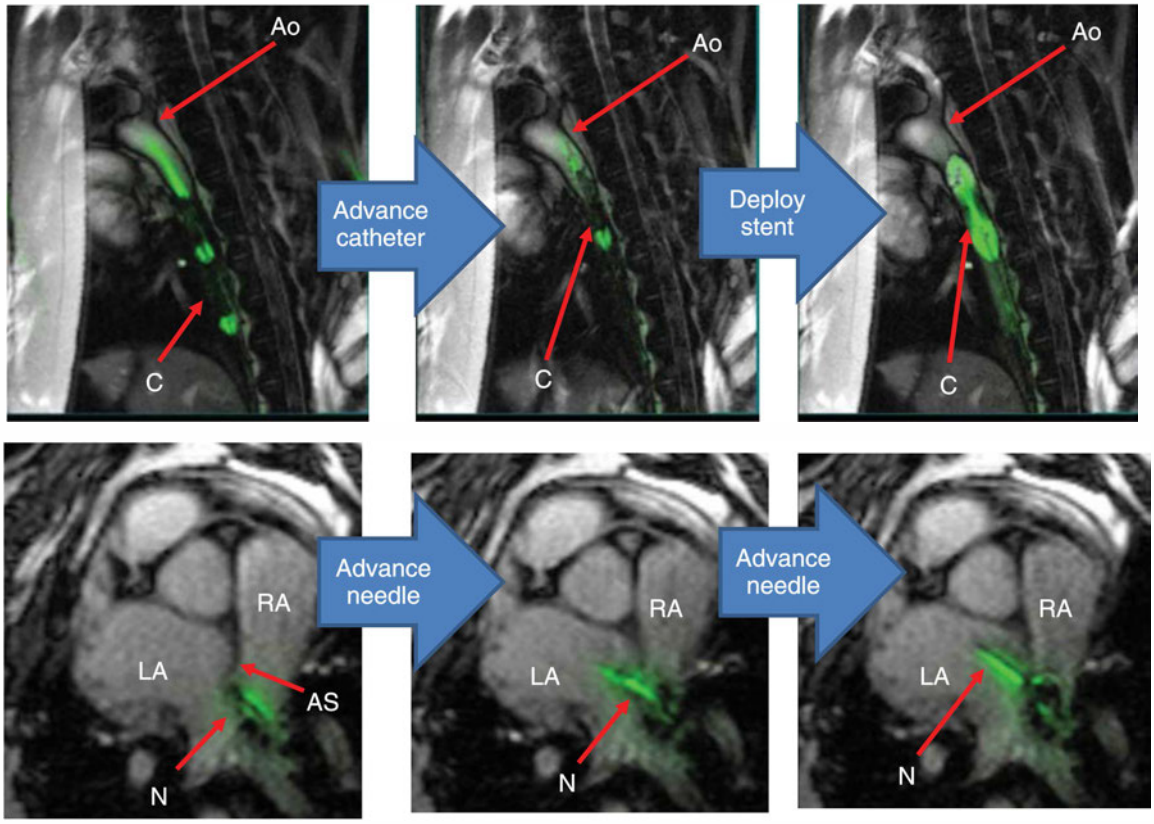


Fig. 73.23. X-ray magnetic resonance (XMR)—intervention in the CMR suite: (**A**) The top three panels are real-time CMR images of advancing a stent mounted on a catheter in the aorta and then deploying the stent; images progress chronologically from left to right. In the top left panel, the catheter (C) in green is in the distal descending aorta (Ao). In the top middle panel, the C is advanced to the isthmus and in the top right panel, the balloon is inflated and the stent deployed. (**B**) The lower three panels are real-time cardiac magnetic resonance images of advancing a needle catheter across the atrial septum to create an interatrial communication. In the lower left panel, the needle (N) in *green* is in the right atrium (RA) and is abutting the atrial septum (AS). In the lower middle panel, the N is advanced across the AS and in the lower right panel, the needle is now in the left atrium (LA). Images are courtesy of Dr. Robert Lederman of the National Institutes of Health.

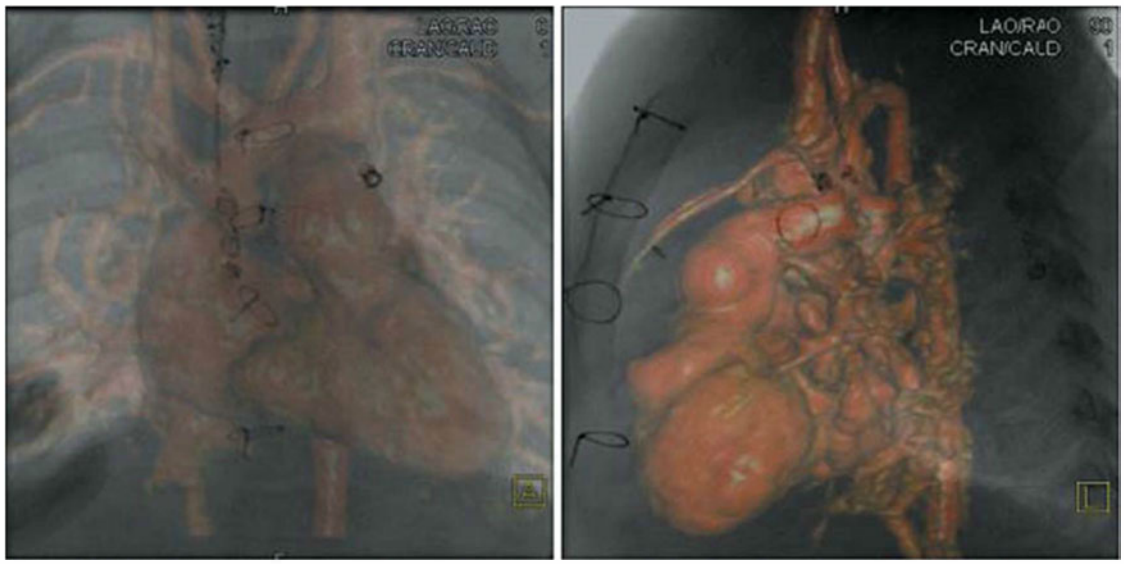


Fig. 73.24. This figure shows the anterior–posterior (AP) and lateral view as seen after the three-dimensional magnetic resonance imaging volume is fused to the fluoroscopy image in a patient with a superior cavo-pulmonary circuit and a stent in the proximal anatomic right pulmonary artery.

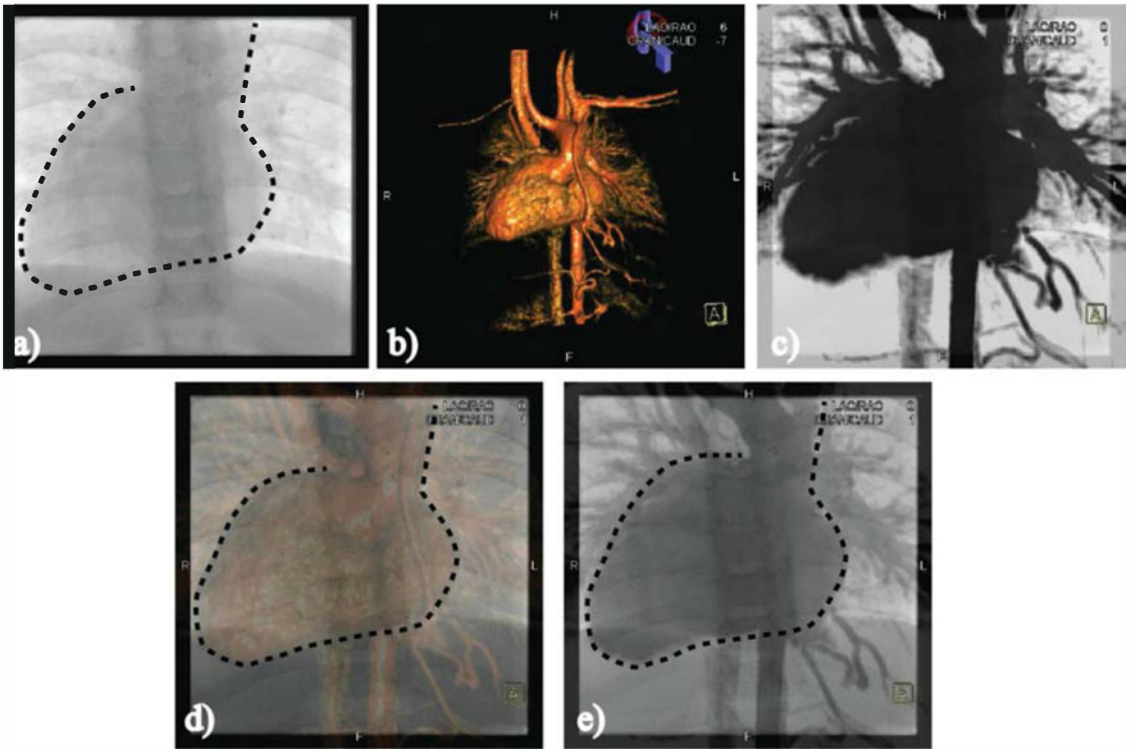


Fig. 73.25. This figure depicts X-ray magnetic resonance fusion to heart and vessel borders using volume-rendered MRI (VRT) image (**B**) and maximal intensity projection (MIP)-rendered image showing the largest cross section of the heart and great vessels (**C**). In (**A**), the *dashed line* traces the heart border and the left lateral border of the aorta on the X-ray image. (**D**) and (**E**) show the registered VRT and MIP images. All images are in the AP projection.

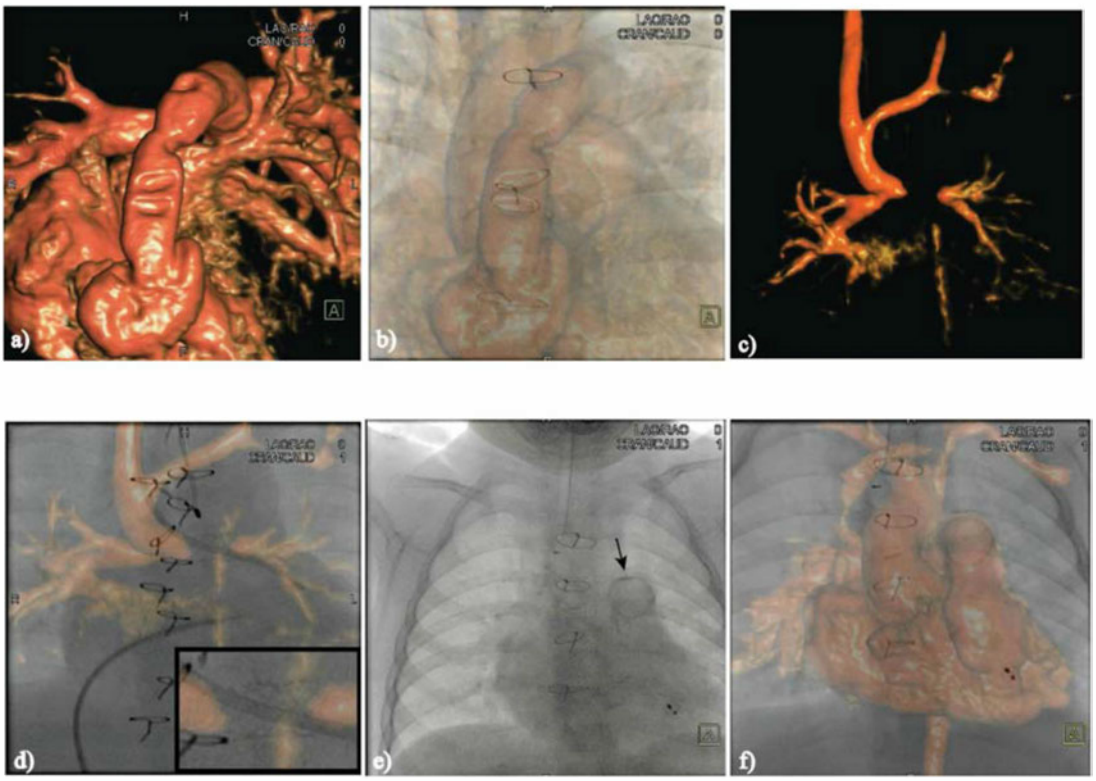


Fig. 73.26. This figure demonstrates the examples of internal marker-based registration. (**A**) and (**B**) depict XMRF to imaging artifact from sternal wires that resulted in indents on a RV-PA conduit. In (**C**) and (**D**) susceptibility artifact from a stent produced a gap in the MRI image of the LPA in a patient with a Glenn shunt. The inset in image (**D**) shows a close-up view of the stent registered between the two PA stumps. (**E**) and (**F**) depict registration to distal conduit calcification. The *arrow* in (**E**) points toward a ring of calcification that formed at the distal end of a RV-PA conduit. All images are in the AP projection.

that is not currently commercially available. A different approach is to use internal markers for the registration process (Fig. 73.26). This approach is potentially more accurate and overcomes some of the limitations of fiducial marker-based registration. Dori et al. have reported on an X-ray magnetic resonance fusion (XMRF) method to internal markers that can be performed quickly, with minimal radiation, without the need for contrast, and with high accuracy (Fig. 73.26). In addition, they have demonstrated the initial use of this modality in CHD interventions.

Internal marker-based registration offers several advantages over fiducial marker-based registration. Internal markers are inherently more stable. They allow for easy correction of patient motion and they do not interfere with contrast angiograms. Furthermore, internal marker-based registration alleviates the constraint imposed by fiducial marker-based registration, namely, the need to perform the MRI and the catheterization successively. The choice of internal marker depends on marker visualization in both modalities and the specific application. In CHD, catheterization registration to the heart and vessel borders is simple and in most cases sufficient to guide the procedure (Fig. 73.25). However, in some instances when the heart and vessel borders are not clearly visible, for example, in the presence of significant pulmonary edema or a pericardial effusion, or when there is a more distinct marker, other markers such as airways, bone, stents, devices, and calcification can be used (Fig. 73.26).

XMRF allows for roadmapping without the need for contrast load (Fig. 73.27). Furthermore, since MRI contrast angiograms provide 3D volume data of the entire circulatory system, roadmaps of an entire procedure can be created and stored as bookmarks prior to the start of the case. This offers a distinct advantage in certain cases because contrast is known to significantly affect hemodynamics. Furthermore, this has the potential to save time, reduce radiation exposure, and contrast load. An example of how XMRF was used for roadmapping is depicted in Figure 73.27. Whole volume-rendered images and images that were cut to delineate certain structures were utilized for this purpose (Fig. 73.27A–73.27C). The figure shows the heart of a patient with a small right ventricle-to-pulmonary artery conduit and LPA stenosis. A coronal cut-plane was created prior to the catheterization and used as a roadmap for navigation from the IVC to the conduit (Fig. 73.27B and 73.27E). After entering the conduit, an oblique cut-plane showing the LPA was loaded and used as a roadmap to enter the LPA (Fig. 73.27C and 73.27 F).

There are two limitations to this roadmapping modality that need to be considered. First, the spatial resolution of MRI is on the order of 1 millimeter making contrast injections necessary for roadmapping in very small vessels. Second, the presence of stiff wires or catheters can alter the local geometry of vessels rendering the registration inaccurate. Lastly, dynamic errors due to respiration and heart motion also can influence registration accuracy and need to be taken into account. Consequently, although device positioning with fused images is possible in most cases, local hand injections are needed for final confirmation of device position prior to deployment.

In addition to roadmapping, the MRI data in the catheter lab can be used for procedure planning including preliminary device sizing, camera angle selection (Fig. 73.28), and hemodynamic and

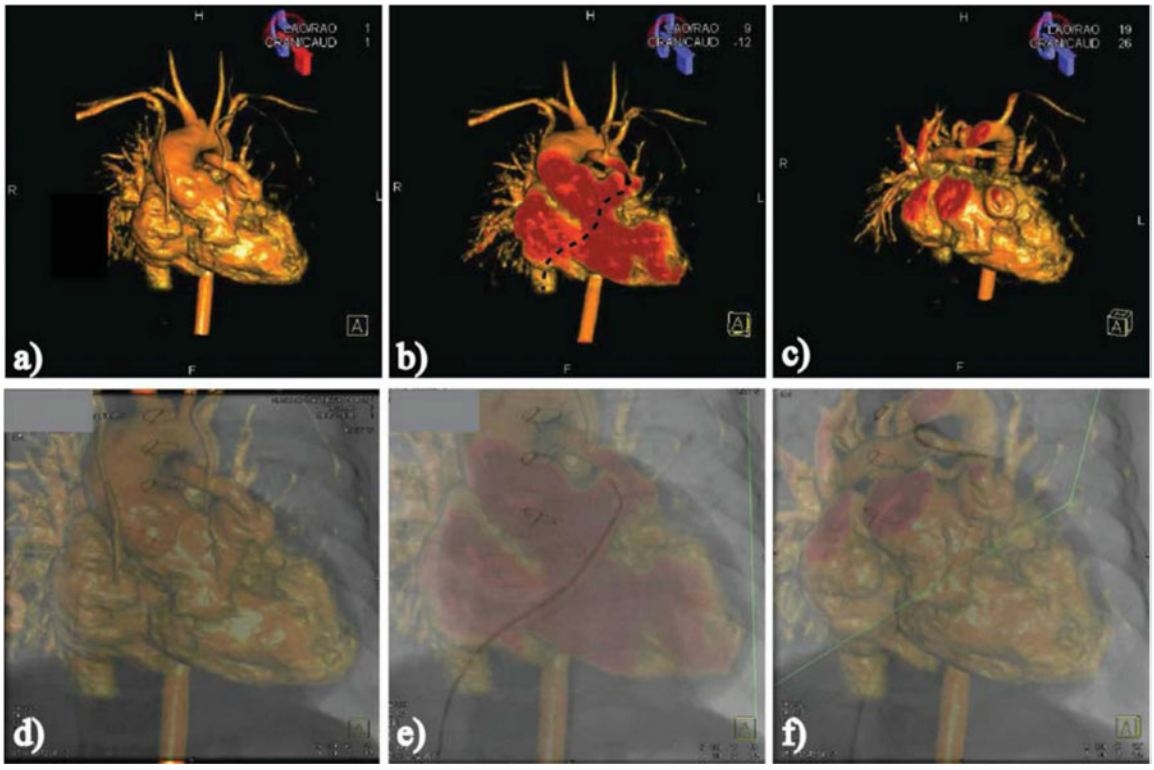


Fig. 73.27. This figure depicts the images of a patient with a small RV-PA conduit that were stored as a collection of bookmarks for roadmapping (**A–C**) and the same images during the catheterization process (**D–F**). In (**B**) the pathway from the IVC to the entrance to the conduit is shown (*dashed arrow*) on an image that was created with an AP cut plane. The corresponding catheter course is shown in (**E**). (**C**) A roadmap image that was created with an oblique cut plane exposing the PA confluence and a hypoplastic LPA. The corresponding catheter course with the tip of the catheter positioned in the distal LPA is shown in (**F**).

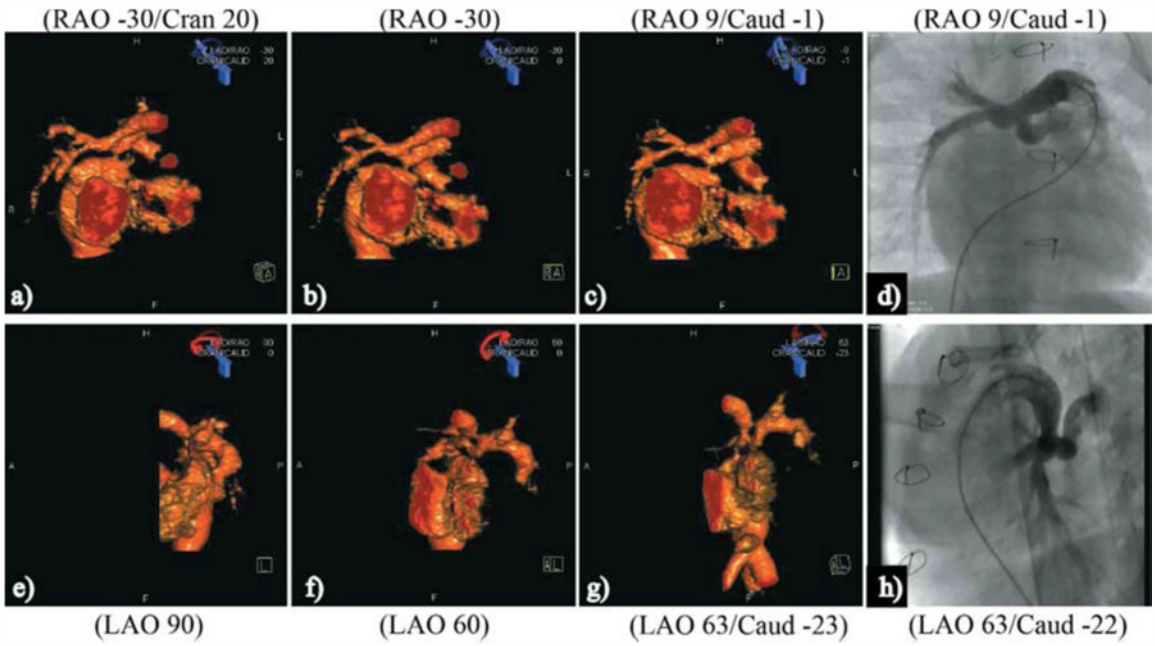


Fig. 73.28. This figure demonstrates how camera angle selection was utilized in a patient with complex pulmonary artery anatomy. In this case, the LPA originated from the proximal RPA and immediately took a 90-degree turn as it headed to the left. There was also proximal LPA hypoplasia. (**A**) to (**D**) correspond to the AP camera and (**E**) to (**H**) to the lateral camera. The anatomy is shown in the conventional camera angles (**A, B, E, and F**) and in the angle that was chosen for the contrast angiogram (**C**) and (**G**). The corresponding contrast angiograms are shown in (**D**) and (**H**).

functional evaluation. With current MRI technology, it is possible to perform initial approximate sizing of devices, such as stents. However, the actual final device sizing should be determined from measurements performed on contrast angiograms. This is a consequence of the fact that the diameter of vessels in volume-rendered data sets highly depends on thresholding levels and is consequently not reliable. Sizing from noncontrast images, such as cine or bright blood images, is more accurate but cine MRI angiograms are normally acquired over multiple heart cycles and other imaging sequences are often triggered to diastole and also averaged over multiple heartbeats.

Camera angle selection is an important utility of MRI volume data sets and XMRF (Fig. 73.28). This is especially true in CHD interventions, where complex 3D anatomy is often encountered. Complex 3D vessel anatomy visualization with fluoroscopy often requires repeated angiograms to fully delineate the anatomy. This is essentially eliminated with this new technology as the anatomy at the different angles can be visualized using the 3D MRI data prior to the procedure allowing for optimal camera angle selection, which can be entered into the C-arm control system for automatic panning to the desired camera angles. When the camera is at the desired angle, the fused image can then be used to assure that there is nothing on the fluoroscopy image that could interfere with the angiogram and to confirm that the chosen angle produced the desired result. In addition, XMRF allows for continuous visualization of the region of interest throughout the catheterization procedure. The 3D map follows the C-arm and the table such that optimal visualization of the region of interest can always be maintained. An example of how camera angle is selected to visualize a complex pulmonary artery anatomy is shown in Figure 73.28.

Clearly, 3D MRI has had a significant impact on the way in which CHD is diagnosed and managed (Figs. 73.24–73.28). Although it is customary to acquire 3D MRI data on many patients with CHD for a variety of clinical indications, the ways in which these data can be used to reduce catheterization time, radiation exposure, and contrast load are under investigation. We have recently reviewed data from our institution of 15 patients who had a purely diagnostic pre-Fontan catheterization in 2010. Seven of the patients had a cardiac catheterization following an MRI evaluation. We compared radiation exposure, contrast load, and catheterization time with the remaining eight patients who underwent a complete diagnostic pre-Fontan catheterization during the same time period without an accompanying MRI. Table 73.2 summarizes the results of a comparison between the two groups. In this small cohort of patients undergoing diagnostic pre-Fontan catheterization, we have been able to show that if the data from a precatheterization MRI are utilized correctly, this can significantly reduce radiation exposure while providing the needed diagnostic information. For this study, we chose patients undergoing a diagnostic pre-Fontan catheterization because the uniformity of this catheterization makes comparison of these patients simpler. Patients who had an interventional procedure were excluded from the study because the heterogeneity in intervention types, lengths, and complexity would make it difficult to compare these patients.

In addition to reduction in radiation, we also found that utilization of the MRI data significantly reduced catheterization time and exposure to contrast compared with subjects receiving a standard diagnostic pre-Fontan catheterization without accompanying MRI. The reduction in contrast exposure is almost certainly a direct effect of the decreased need to perform cine angiograms, as anatomic information is obtained by MRI. The reduction in radiation exposure is likely due to the combined beneficial effects of a decreased total catheterization time and a decreased number of cine angiograms performed (where a disproportionate amount of radiation exposure occurs). Although in the MRI group, the total fluoroscopy time is lower than in

Table 73.2 Comparison between XMR and control groups

	XMR group ($n = 7$)	Control group ($n = 8$)	*P*-value
Height (cm)	36.3 ± 4	36.5 ± 2.4	0.9
Weight (kg)	28.9 ± 5.9	29.7 ± 2.5	0.7
BSA (m^2)	0.57 ± 0.09	0.58 ± 0.04	0.8
Age (years)	3.2 ± 1	3.1 ± 0.9	0.9
Contrast (cm^3/kg)	0.5 ± 0.34	1.7 ± 0.24	**<0.0001**
Catheterization time (min)	34.6 ± 7.9	66.9 ± 35.6	**0.04**
Fluoroscopy time (min)	10.2 ± 3.5	10.7 ± 2.6	0.7
Fluoroscopy DAP (μGy m^2)	284.8 ± 111	368.4 ± 112.3	0.17
Total DAP (μGy m^2)	340.8 ± 139.1	593.8 ±168.9	**0.008**
Total effective biologic dose (mSv)	1.8 ± 0.9	2.8 ± 0.8	**0.048**
Number of power injections	2 (0–2)	3.5 (3–5)	**0.0009***
Number of hand injections	1 (0–3)	0 (0–3)	0.8*

the control cases, this difference did not reach statistical significance. A more significant difference in the total fluoroscopy times will likely be seen in cases where more complex cardiovascular anatomy increases the utility of XMRF and roadmapping.

Although this study looked only at one specific group of patients, it seems quite likely that extending this strategy to many others will result in similar benefits. In any circumstance, obtaining at least some of the required diagnostic information by MRI prior to catheter entry will decrease the time needed for the catheterization, with resultant decrease in contrast and radiation exposure. Furthermore, beyond the simple matter of reducing the time a patient spends under the fluoroscopy cameras, XMRF may allow for catheter manipulation to be performed at very low fluoroscopy pulse rates, further lowering radiation exposure.

It is likely that multimodality imaging techniques including MRI will significantly alter the way we perform catheterization procedures in the future. This new technology where 3D imaging data can be incorporated into live fluoroscopy images holds great promise to improve catheterization outcomes and reduce radiation exposure. Further studies into the effect of MRI and XMRF on catheterization outcomes are currently underway in our institution.

PARTING THOUGHTS

CMR has advanced considerably in the last 30 years and the next 30 years holds even more promise for additional capabilities to come on line. It is gaining widespread use but clearly, there is a long way to go. It is an extremely useful and complementary technique to other imaging modalities and has supplanted some to become the gold standard. Recognition of this fact should play a role in practicing 21st century medicine.

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EDITOR'S COMMENTS

Cardiac MRI imaging has become extremely sophisticated, and with new techniques, much additional *hemodynamic* information can be added to the *morphologic* information from MRI scanning. In addition, the ability to three-dimensionally reconstruct MRI images has provided a very useful anatomic description of cardiac anomalies, including abnormalities of the aortic arches and vascular rings. The MRI scan is now the most useful imaging modality for evaluation of these lesions, and barium swallow determinations are now most commonly used as screening modalities. The ability to assess not only great vessel anatomy but also airway and esophageal anatomy by MRI has distinct advantages in assessment in these patients. In addition, assessing abnormalities of chest wall anatomy may be useful in determining whether simple division of vascular rings or more complex vascular reconstruction or aortopexies may be necessary to completely relieve obstruction.

One of the greatest limitations to cardiac MRI is the fact that it is somewhat cumbersome in the management of very small infants, who require anesthesia to prevent motion during the scans. The limits of resolution of the scans also are important for imaging in very small children, although technology is improving rapidly in this area. Nevertheless, the unique ability of MRI to create three-dimensional images of cardiac anatomy with associated images of blood flow and ventricular function, make hemodynamic evaluations of ejection fraction and blood flow velocity, and assess regurgitant volumes, is making this modality an increasingly important tool in the assessment of congenital heart disease.

Recently, fast multislice computed tomography angiography has been added to MRI as a good modality for three-dimensional reconstruction of vascular images in children with congenital heart disease. This technique has the advantage of very rapid acquisition times, making anesthesia less important. An additional advantage is excellent resolution even in relatively small children and the ability to evaluate the airways, including the possibility of virtual bronchoscopy. A disadvantage of this technique is the relatively large radiation dose necessary to acquire the images, and this concern has significantly limited its use.

As noted by Dr. Fogel in this chapter cardiac MRI is becoming increasingly used as the primary imaging modality in congenital heart disease. The images being obtained today are of superior quality to those of only a few years ago and increasing hemodynamic and flow distribution images have been obtained, which now make echocardiographic studies and cardiac catheterization studies less and less important. One might argue that cardiac MRI will become the primary imaging modality and echocardiography will be used primarily as a surveillance tool in Cardiac Intensive Care Units or for initial screening examinations. The ability to calculate cardiac shunts and cardiac valve regurgitant volumes in addition to assessing ventricular function and flow distributions in vessels along with cardiac output and shunt calculations has made cardiac MRI virtually supplant hemodynamic cardiac catheterization except in rare cases where pulmonary vascular resistance needs to be calculated. The combined use of cardiac catheterization and cardiac MRI as has been done in our center permits the possibility to define cardiac anatomy and pulmonary flow distribution and assess collateral pulmonary blood flow with subsequent intervention to eliminate extra sources of pulmonary blood flow or shunts, and then recalculate the remaining shunt flows and distributions. In addition, cardiac MRI has become useful in preoperative planning in baffle reconstruction and complex single-ventricle repairs such as those with heterotaxy syndromes and systemic and pulmonary venous abnormalities. Defining the capabilities of, and indications for, the various imaging modalities now available to us in congenital heart surgery will be very important to prevent unnecessary duplication of imaging and to achieve the maximum possible information for surgical planning on our patients.

TLS

74 Hemodynamic Assessment and Transcatheter Therapy for Congenital Heart Disease

Matthew J. Gillespie and Jonathan J. Rome

Advances in noninvasive imaging have done away with the need for preoperative cardiac catheterization in the management of many heart lesions. Thus, cardiac catheterization is not indicated prior to operative intervention for transposition of the great arteries, hypoplastic left heart syndrome (HLHS), truncus arteriosus, complete common atrioventricular canal defect, a simple paramembranous ventricular septal defect, or tetralogy of Fallot, unless particular concerns arise. This discussion of heart catheterization in children, therefore, focuses not on which lesions require a heart catheterization for a complete evaluation but rather what information is desired when it has been decided that a heart catheterization should be undertaken.

BASIC PRINCIPLES OF HEMODYNAMIC EVALUATION

Invasive hemodynamic assessment involves calculation of flows and resistances, shunt calculations, and measurement of pressure and flow gradients.

Calculation of Flows

The simplest method of measuring flow is to use a thermodilution catheter. This method is a variation on the dye dilution method using temperature decrement to determine blood flow. However, the method is only applicable in the absence of intracardiac shunting when there is a mixing chamber between the proximal and distal ports. Since most children with congenital heart disease do have either right-to-left, left-to-right, or bidirectional shunting, thermodilution is not an appropriate method of determining cardiac output in these patients.

More often, the Fick principle is used for calculation of flow. The essence of the Fick principle is that flow can be calculated by the use of a measurable indicator, which is added or subtracted at a known rate. In practice, the indicator is oxygen.

F = unknown flow rate (ml/min)
I_1, I_2 = indicator concentrations (mg/ml) at the two ends of the flow pathway
R = rate at which indicator is added or subtracted (mg/min)

$$F\,(\text{ml/min}) = R\,(\text{mg/min})/I_2 - I_1\,(\text{mg/ml})$$

In a hemodynamic study, the unknown flow rate is systemic or pulmonary blood flow, the indicator is oxygen, and the rate of change of the indicator is the oxygen consumption. Oxygen content is determined by multiplying the hemoglobin concentration times the spectrophotometrically measured percentage of oxygen saturation of hemoglobin; dissolved oxygen must be included in the calculation when patients receive supplemental oxygen. Oxygen consumption is either measured or assumed on the basis of age and heart rate, using standardized tables. The equation for calculation of pulmonary blood flow is as follows:

Q_P = O_2 consumption/(pulmonary vein O_2 content – pulmonary artery O_2 content)

and the equation for calculation of systemic blood flow is as follows:

Q_S = O_2 consumption/(aortic O_2 content – venous O_2 content).

Effective flow describes the quantity of deoxygenated blood that flows through the pulmonary circuit, which is also equal to the volume of oxygenated blood that flows through the systemic circuit. Q_P effective equals Q_S effective and is calculated by taking the difference between the mixed systemic and pulmonary venous oxygen content and dividing that into the oxygen consumption:

Q_{eff} = O_2 consumption/(pulmonary venous O_2 content – mixed venous O_2 content)

Calculation of Shunts

Shunt flow can be thought of as the ineffective flow in the system: the left-to-right shunt is the volume of oxygenated blood flowing through the lungs and is equal to the total pulmonary flow minus the effective pulmonary flow ($Q_P - Q_{eff}$). The right-to-left shunt is the volume of deoxygenated blood flowing through the systemic circulation and is equal to the total systemic flow minus the effective systemic flow ($Q_S - Q_{eff}$).

Because of the great variation in size among pediatric patients, flows are always indexed to body surface area.

Calculation of Resistance

Resistance is the change in mean pressure divided by the flow. Thus, pulmonary vascular resistance (R_p) is the difference between the pulmonary venous and pulmonary arterial pressures divided by the total pulmonary flow.

R_p = (mean pulmonary vein pressure – mean pulmonary arterial pressure)/Q_P

and the systemic vascular resistance is the difference between the systemic arterial and right arterial pressures divided by systemic flow.

R_s = (mean arterial pressure – mean right atrial pressure)/Q_S

The resulting units, liters per minute per millimeters of mercury, are termed *Wood's units*, after the cardiologist Paul Wood. As with flows, vascular resistance is indexed to body surface area in pediatric patients.

Flow Gradients

In the cardiac catheterization laboratory, gradients across stenotic valves or vessels are generally measured as "peak-to-peak" gradients. This measurement is often inaccurately referred to as the PSEG or peak systolic ejection gradient. The peak-to-peak gradient is a different entity than the maximal instantaneous gradient that is estimated noninvasively by Doppler studies. Whether a gradient is expressed as peak-to-peak, maximal instantaneous gradient, or mean gradient, it is not a

meaningful number unless one has some idea of the amount of flow that is producing the gradient. For example, a patient with critical aortic stenosis who is in low cardiac output may have a relatively low measured gradient across a severely narrowed valve orifice.

HEMODYNAMIC EVALUATION OF PATIENTS WITH A LEFT-TO-RIGHT SHUNT LESION

Patients with left-to-right shunt lesions (ventricular septal defect, complete or partial common atrioventricular canal defect, patent ductus arteriosus [PDA]) undergo cardiac catheterization in order to address the following related questions:

1. What is the magnitude of the left-to-right shunt? Does it justify surgical repair?
2. What is the status of the pulmonary vascular bed? Is the pulmonary vascular resistance sufficiently low that a complete repair is possible?

HEMODYNAMIC EVALUATION OF PATIENTS WITH A RIGHT-TO-LEFT SHUNT LESION

In patients with right-to-left shunt lesions (critical pulmonary stenosis status postsurgical or transcatheter correction, pulmonary atresia with intact ventricular septum status postaortopulmonary shunt, Ebstein's anomaly, tetralogy of Fallot with pulmonary atresia status postright ventricular outflow reconstruction with an open ventricular septal defect, single ventricle status postfenestrated Fontan palliation), there is often some uncertainty as to the adequacy of the right heart or pulmonary vascular bed. Specifically, the question is whether one normal cardiac output can pass through the right heart with acceptable systemic venous and right ventricular pressures. If not, the presence of an atrial or ventricular level combination will often permit adequate cardiac output with acceptable hemodynamic values at the expense of oxygenation. Often, an attempt will be made to temporarily occlude the site of right-to-left shunting during the heart catheterization in order to assess the hemodynamic sequelae. Increased heart rate, increased systemic venous pressure, and decreased cardiac output with occlusion of an atrial level communication are all signs of right heart insufficiency.

HEMODYNAMIC EVALUATION OF PATIENTS WITH COMPLETE MIXING LESIONS AND FUNCTIONAL SINGLE VENTRICLE

Currently, the "ideal" surgical palliation for children with functional single ventricle lesions is the modified Fontan operation, with or without an atrial level right-to-left shunt. This type of surgical palliation requires that the pulmonary vascular resistance be low (<4 Wood's units), as the separation of the circulations is accomplished by using the functional single ventricle to pump systemic blood flow, while pulmonary blood flow occurs passively across a pressure gradient. Early in life (i.e., in the first year), the goal of management is to provide adequate but limited pulmonary blood flow. In cases of functional single ventricle with pulmonic stenosis, no intervention may be needed in the first few months of life. More often, the desired hemodynamics will be achieved via placement of a pulmonary artery band or an aortopulmonary shunt. Typically, this arterial to pulmonary flow is altered to veno pulmonary flow by 6 months of age with a bidirectional cavopulmonary anastomosis (bidirectional Glenn or "hemi-Fontan" procedure). Completion Fontan procedure (total cavopulmonary anastomosis) is usually performed by 3 years of age. Traditionally, cardiac catheterization has been performed prior to the second and third stages of surgical palliation though the need for routine diagnostic catheterization at these times has been called into question. When diagnostic catheterization is performed prior to the Fontan operation, its purpose is to assess whether "Fontan physiology" will be well tolerated in a given patient. The important questions to be evaluated at cardiac catheterization include:

1. What is the pulmonary vascular resistance? Patients with pulmonary vascular resistance >3 to 4 Wood's units are much more likely to manifest high central venous pressures and low cardiac output.
2. What is the pulmonary artery pressure? Elevated pulmonary artery pressure is not necessarily a contraindication to a modified Fontan operation provided that it is measured in the setting of elevated pulmonary blood flow. However, a mean pulmonary artery pressure greater than about 15 mmHg in the setting of normal or diminished pulmonary blood flow bodes poorly for the outcome after conversion to Fontan physiology.
3. What is the ventricular filling pressure? After a modified Fontan operation, blood flows passively across the pulmonary vascular bed. The central venous pressure will, therefore, be determined by the ventricular filling pressure and the pulmonary vascular resistance. Elevated ventricular filling pressure may be present as a result of chronic volume overload or ventricular hypertrophy, the latter case in particular is associated with poor outcome after Fontan palliation.

HEMODYNAMIC ASSESSMENT OF SEMILUNAR VALVE STENOSIS OR INSUFFICIENCY (WITHOUT SHUNT)

Pulmonary Valve

The severity of pulmonary valve stenosis is graded according to the peak-to-peak gradient across the valve (trivial, <25 mmHg; mild, 25 to 49 mmHg; moderate, 50 to 79 mmHg; severe, >80 mmHg). In general, the presence of a peak-to-peak gradient of ≥40 mmHg is an indication for treatment. Pulmonary insufficiency is difficult to assess quantitatively in the cardiac catheterization laboratory. A ventricularized pulmonary artery tracing—that is, one in which the diastolic pressure approximates that of the right ventricle—indicates severe pulmonic insufficiency.

Aortic Valve

Although cardiologists will generally grade aortic stenosis in adults according to the calculated effective valve area, in the pediatric population indications for intervention are based on catheter-measured peak-to-peak systolic pressure gradients. Because gradients across a stenotic valve will vary depending on flow and heart rate, cardiac output, left ventricular function, and mitral and aortic valve insufficiency all must be evaluated to arrive at an accurate assessment of left ventricular outflow tract obstruction. Left ventricular outflow tract obstruction may be subvalvar, valvar, or supravalvar.

Aortic valve insufficiency is assessed in a semiquantitative manner in the cardiac catheterization laboratory by angiography. A wide arterial pulse pressure not only may represent important aortic valve insufficiency but may also be seen with aggressive afterload reduction.

HEMODYNAMIC ASSESSMENT OF ATRIOVENTRICULAR VALVE STENOSIS OR INSUFFICIENCY

Tricuspid Valve

Isolated tricuspid stenosis is extremely rare; more often, tricuspid stenosis is seen in the context of right heart hypoplasia and pulmonic stenosis or atresia. Tricuspid stenosis cannot be assessed in the presence of pulmonic atresia because there will be little or no flow across the tricuspid valve in these cases. The presence of a patent foramen ovale that allows right-to-left shunting or right ventricular diastolic dysfunction (both of which are common in right heart hypoplasia) makes assessment of the degree of tricuspid stenosis problematic. Complete evaluation requires balloon occlusion of the atrial communication and simultaneous measurement of right atrial and right ventricular pressures; this may not be technically feasible particularly in neonates and small infants (e.g., the atrial communication is too large to be occluded). However, the maneuver may be quite useful in evaluating right heart competency in older children noting any resultant changes in right atrial pressure, cardiac index, and heart rate with balloon occlusion.

Angiographic assessment of tricuspid insufficiency can be difficult because the catheter in the right ventricle of necessity crosses the tricuspid valve; if the catheter stents the valve open or if some of the holes of the catheter are in the right atrium at the time the injection is made, artifactual insufficiency will result. In addition, ectopic beats will often be associated with induced atrioventricular valve insufficiency.

Mitral Valve

Mitral stenosis is generally graded in terms of calculated valve area by cardiologists treating adults and in terms of anatomy and valve gradient by cardiologists treating children. Both the mean valve gradient (arrived at by digitizing the area between the left atrial or pulmonary artery wedge pressure tracing and the left ventricular pressure tracing during the period when the mitral valve [MV] is open) and the gradient between the a-wave and the left ventricular end-diastolic pressure as well as the absolute left atrial pressure are generally considered in assessing mitral stenosis. Because gradients are meaningful only when considered in association with flow, cardiac output must be assessed to appropriately interpret the severity of obstruction. A mean left atrial pressure >25 mmHg is generally associated with moderate-to-severe reactive pulmonary hypertension.

Mitral insufficiency is assessed in a semiquantitative manner by angiography. This is more reliably accomplished than in the case of the tricuspid valve because the catheter with which the picture is taken usually does not cross the MV and the shape of the left ventricle is such that it accommodates a catheter more easily than does the right. However, entanglement of the catheter in the MV apparatus or stimulation of ventricular premature contractions can result in induced mitral insufficiency.

TRANSCATHETER THERAPY FOR CONGENITAL HEART DISEASE

Introduction

Catheter-directed treatments have assumed a major role in the care of patients with congenital cardiovascular defects. These techniques have replaced surgery as the primary mode of therapy in some instances. In many others, optimal treatment combines staged transcatheter and surgical intervention. It is, therefore, essential that the congenital heart surgeon has a working knowledge of interventional cardiology. This section introduces current catheter interventions for patients with congenital cardiovascular defects, highlighting therapies particularly useful in combination with surgery.

Catheter interventions for congenital heart defects are quite varied, but in simplistic terms can be classified into: (1) procedures to create or enlarge communications and vessels (septostomies, dilations, and stents); (2) procedures to close vessels or defects (device closures, vascular occlusions); and (3) other (thrombectomy, vascular retrievals). Another commonly performed intervention, radiofrequency ablation, is more appropriately dealt with as part of a discussion of cardiac dysrhythmias.

Creation and Enlargement of Atrial Communications

Balloon atrial septostomy described in 1966 by Rashkind and Miller for palliation of cyanosis in D-transposition of the great arteries is often considered the first intervention for congenital heart defects. Even with the use of prostaglandin E1, many infants with transposition require septostomy procedures to ameliorate compromising cyanosis. The procedure is also performed to relieve left atrial hypertension in infants with left atrioventricular valve stenosis or atresia. The basic technique has changed little since first described. A septostomy catheter is advanced through either umbilical or femoral vein into the right atrium and across the foramen ovale. The balloon is filled with fluid and the catheter pulled quickly across the foramen tearing septum primum. Balloon septostomy is often performed at the bedside under echocardiographic guidance (Fig. 74.1).

In many situations, balloon septostomy is not the best way to create an atrial communication. Balloon septostomy is ineffective in patients over 6 weeks of age because the atrial septum is too thick. In most newborns with HLHS and restrictive or intact atrial septum, balloon septostomy is not feasible or ineffective because the left atrium is too small or posterior deviation of septum primum is present. In these populations, an atrial communication is best created by other means. The intact atrial septum may be traversed either by the standard transseptal puncture technique (Brockenbrough procedure), or with the aid of a specially designed radiofrequency perforation system (Nykanen Catheter, Baylis Medical Corp., Mississauga, ON). Once the atrial septum has been crossed, a hole may be created by static balloon angioplasty, or in the case of the very thick atrial septum, endovascular stent deployment. When the procedure is performed for relief of cyanosis in patients with mixing lesions and left atrial outlet obstruction (such as the newborn with HLHS born with intact atrial septum), the goal should be the creation of an unrestrictive atrial communication. In other circumstances (i.e., augmentation of systemic blood flow in patients with right heart failure, or for left atrial decompression in patients on extracorporeal support for left heart dysfunction), smaller atrial communications suffice. In fact, when these techniques are used to palliate right heart failure (transient right ventricular dysfunction after repair of tetralogy of Fallot, or primary pulmonary hypertension, or fenestration creation for Fontan failure), the procedure results in systemic arterial desaturation; thus, the size of communication created must be carefully titrated to arterial oxygen saturation. The goal is to create a restrictive ASD to augment systemic output without excessive cyanosis. In tetralogy of Fallot or pulmonary hypertension patients, this is best accomplished by graded balloon angioplasty of the atrial septum. In Fontan patients requiring fenestration creation, the optimal strategy depends on the type of Fontan. It may include the use of standard or cutting balloon dilation (lateral

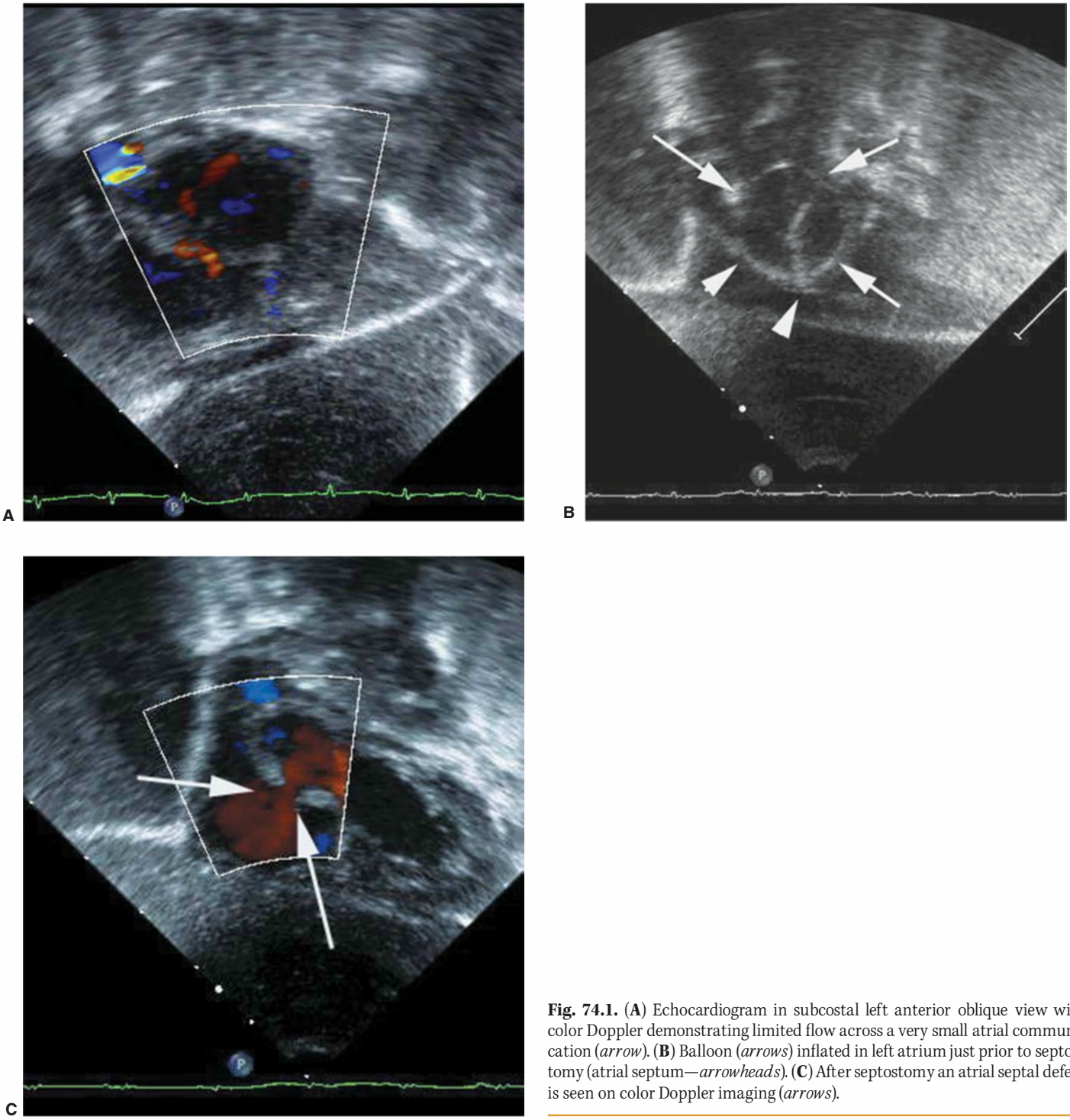


Fig. 74.1. (**A**) Echocardiogram in subcostal left anterior oblique view with color Doppler demonstrating limited flow across a very small atrial communication (*arrow*). (**B**) Balloon (*arrows*) inflated in left atrium just prior to septostomy (atrial septum—*arrowheads*). (**C**) After septostomy an atrial septal defect is seen on color Doppler imaging (*arrows*).

tunnel-type Fontan), or transseptal puncture followed by bare metal or covered stent deployment (extracardiac Fontan).

Valve Repair and Replacement Procedures

Balloon Valvuloplasty

Inflation balloon angioplasty was developed in 1974 by Gruntzig for the treatment of atherosclerotic peripheral arterial stenoses. Advances in equipment and technique have allowed the application of balloon dilation to a variety of valvar and vascular stenoses in patients of all ages.

Pulmonic Valve Stenosis and Atresia

The first congenital lesion treated with balloon dilation was pulmonic stenosis. Balloon valvuloplasty for pulmonic stenosis is usually curative and generally regarded as the treatment of choice in patients of all ages. In older children and adolescents, the procedure is straightforward. After hemodynamic evaluation, a right ventricular angiogram is performed to assess anatomy and measure the pulmonary annulus dimension. Dilation is accomplished by advancing the balloon catheter over a guide wire positioned in the distal pulmonary artery tree through the stenotic valve. The balloon size is chosen to be 120% to 140% of the annulus

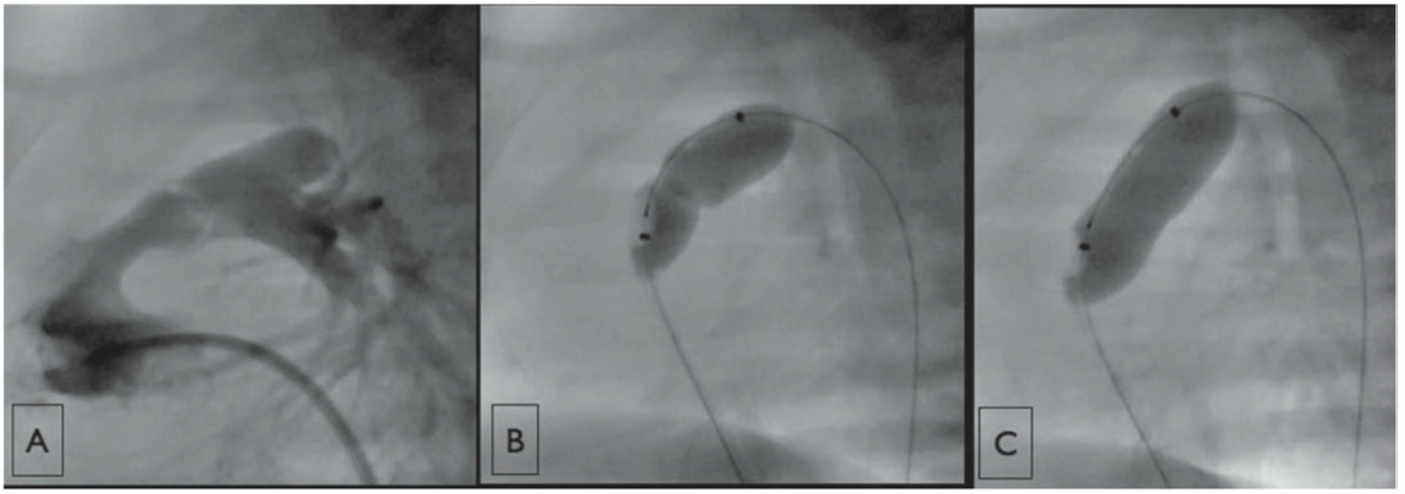


Fig. 74.2. (**A**) Lateral projection of a right ventriculogram in a newborn with critical pulmonary valve stenosis demonstrates doming of the stenotic pulmonary valve in systole. (**B**) Inflation of a balloon catheter across the stenotic pulmonary valve. A waist develops on the balloon during inflation and subsequently resolves (**C**). The guide wire crosses the patent ductus arteriosus and is positioned in the descending aorta.

diameter. As the balloon is inflated, the stenotic valve creates a waist, which then disappears. Failures may occur (1) if the pulmonary valve is thickened, nondoming, and often muscularized (so-called dysplastic valve common in Noonan's syndrome) or (2) where valvar stenosis is part of complex obstruction involving infundibulum, valve annulus, or supravalvar region.

Dilation for critical pulmonary stenosis in the newborn is successful in the majority of cases (Fig. 74.2). Moderate cyanosis is common after dilation due to atrial right to left shunting. Cyanosis diminishes as right ventricular compliance improves. Severe cyanosis after valvuloplasty should prompt further evaluation. Where inadequate relief of right ventricular outflow obstruction is present after technically adequate balloon valve dilation, right ventricular outflow patch augmentation may be required, particularly when there is hypoplasia of the pulmonary annulus.

Balloon valvuloplasty is also applicable to newborns with pulmonary atresia and intact ventricular septum. Prior to intervention, these children require diagnostic evaluation to determine that the right ventricle and particularly the infundibulum are of adequate size as well as to rule out the presence of a right ventricular dependent coronary circulation. If an infant is deemed a suitable candidate, the pulmonary valve is perforated with a radiofrequency catheter (see above) and then dilated in the standard manner (Fig. 74.3). Patients undergoing this intervention very frequently develop cyanosis after discontinuation of prostaglandin infusion as the ductus arteriosus closes. If cyanosis occurs, management is individualized. In cases where the right ventricle is deemed well developed, the patient may be maintained on prostaglandins for a time to allow improvement in right ventricular compliance. If the right heart is more significantly hypoplastic, a stable source of additional pulmonary blood flow is added usually by stenting of the ductus arteriosus (Fig. 74.4).

Aortic Stenosis

All treatments for aortic stenosis, including surgical or balloon valvuloplasty, valve replacement, or pulmonary autograft (Ross procedure), should all be considered palliative. Balloon dilation of aortic stenosis generally results in adequate gradient relief with a minimal increase in aortic regurgitation. Reported failure rates vary from 0% to 10%, and significant increases in aortic regurgitation occur in approximately 10% of patients. The duration of successful palliation varies from months to decades though most (if not all) patients will subsequently develop restenosis, progressive regurgitation, or both. Intermediate term results after balloon and surgical valvuloplasty are comparable, thus balloon valvuloplasty is generally recommended as the first treatment for patients with aortic stenosis and little regurgitation. Except in the newborn, aortic valvuloplasty is usually performed via a retrograde approach from the femoral artery. After measurement of the transvalvar gradient, aortic root injection and ventriculography are performed for valve anatomy, degree of insufficiency, and annulus measurement. The valve is dilated by advancing the angioplasty balloon over a guide wire positioned in the left ventricle (Fig. 74.5). Balloon size is chosen to be slightly less than the diameter of the aortic valve annulus. The risk of significant aortic insufficiency increases when the balloon-to-annulus ratio exceeds 110%.

Special considerations apply to critical aortic stenosis in the newborn. This lesion has traditionally been very difficult to treat with high mortality. However, improvements in stratifying patients to appropriate treatment as well as in the treatments themselves have resulted in markedly decreased mortality. The approach to such patients is primarily dependent on left ventricular function and anatomy. When the left ventricle is inadequate to support systemic circulation, patients require treatment as HLHS. Several reports have established criteria for stratification of patients to biventricular repair versus single ventricle palliation. Balloon aortic valvuloplasty is the treatment of choice where left ventricular size (including MV, ventricular volume, and aortic valve sizes) and function are deemed adequate to support systemic circulation. When the left ventricular volume and MV are adequate but the aortic valve hypoplastic, primary Ross procedure may be the optimal strategy. After successful balloon valvuloplasty, ventricular function generally recovers enough to allow discontinuation of ventilatory and intravenous inotropic support within a few days. When this cannot be accomplished, either left ventricular outflow obstruction persists or left heart size and/or function are(is) inadequate. When left ventricular outflow obstruction persists despite a technically

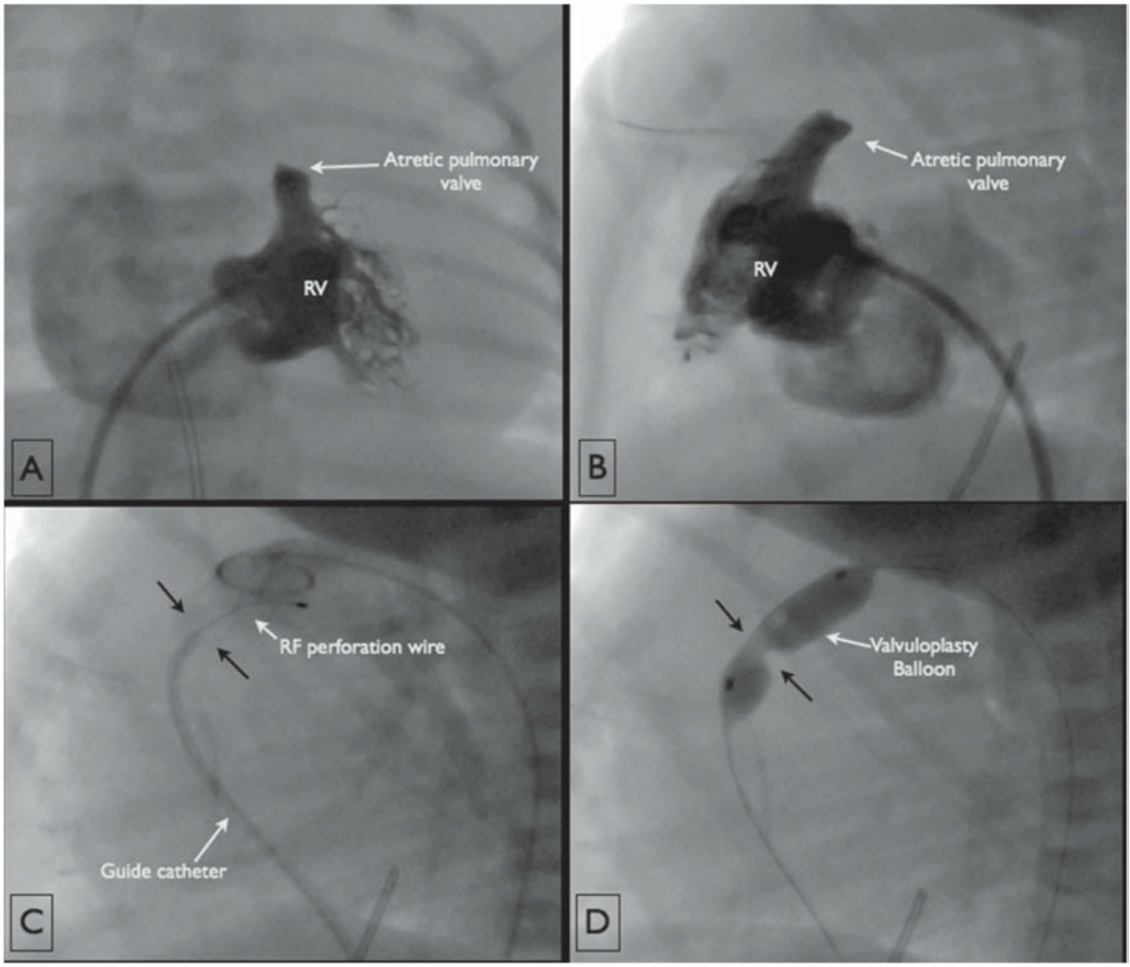


Fig. 74.3. (**A**) Anteroposterior projection of a right ventricular injection in a newborn demonstrates a tripartite right ventricle with a well-developed infundibulum and valvar pulmonary atresia. (**B**) A lateral view of the same injection. (**C**) A Nykanen catheter (arrow; Baylis Medical, ON) has been advanced through a guide catheter to the atretic valve, which has been perforated by the application of radiofrequency energy. (**D**) Subsequent balloon valvuloplasty. The arrows mark the location of the atretic valve. RF, radio frequency.

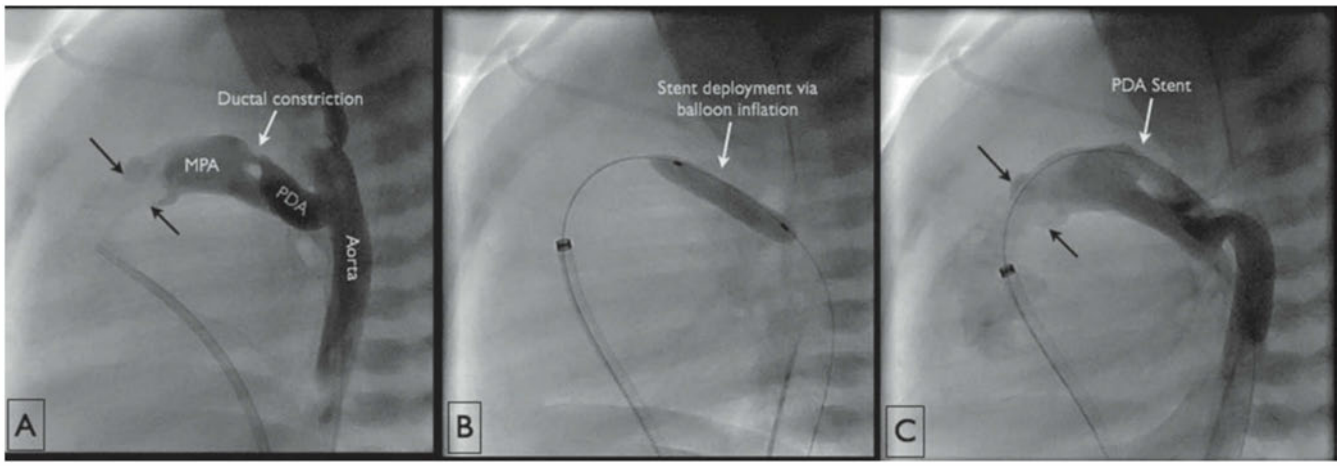


Fig. 74.4. (**A**) Lateral projection of a descending aortogram in a patient with pulmonary atresia with an intact ventricular septum following prior RF perforation and balloon valvuloplasty. There is significant constriction of the patent ductus arteriosus at the pulmonary end (*white arrow*), resulting in hypoxemia. (**B**) Deployment of a stent via balloon inflation results in unobstructed flow through the ductus (**C**) and therefore improved oxygenation. MPA, main pulmonary artery; PDA, patent ductus arteriosus.

adequate balloon procedure, it is usually the result of annular hypoplasia and may be successfully treated by autograft root replacement. In the latter circumstance, an alternative surgical palliative strategy is the only approach likely to lead to long-term survival. This strategy may be a Norwood operation committing the patient to subsequent single ventricle palliation. Alternatively, a so-called hybrid procedure (see below) may be undertaken. After the hybrid approach, subsequent anatomic

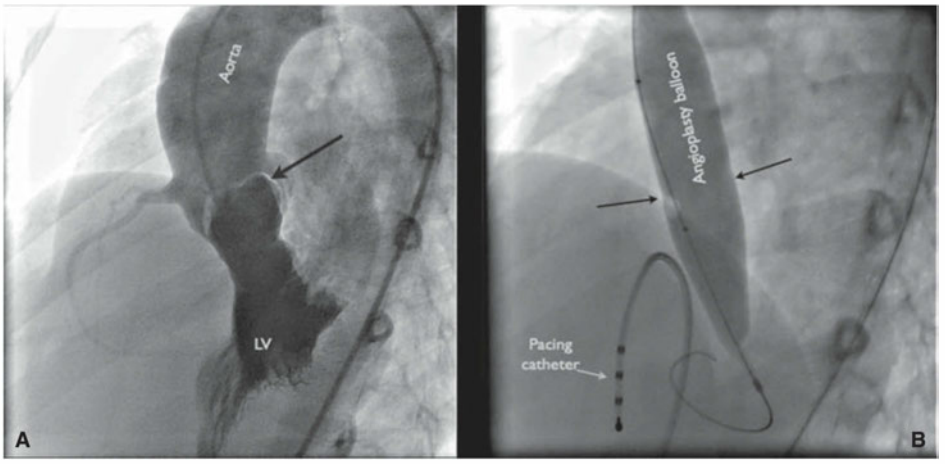


Fig. 74.5. (**A**) A left ventriculogram in a patient with aortic stenosis demonstrates systolic doming of the aortic valve leaflets. (**B**) A valvuloplasty balloon advanced over a guidewire through the aortic annulus is inflated across the stenotic valve. A pacing catheter is placed in the right ventricle to permit rapid ventricular pacing, used to transiently reduce cardiac output and therefore facilitate stable balloon positioning. LV, left ventricular.

and physiologic evaluation is performed to determine whether the patient is best served with a single or biventricular approach.

Other Valvular Lesions

Balloon dilation has been applied to mitral, tricuspid, and bioprosthetic stenoses. In contrast to the excellent outcome after balloon dilation for rheumatic mitral stenosis, results after dilation in infants and children with congenital mitral stenosis are mixed and restenosis common. Nonetheless, balloon valvuloplasty is an appropriate first-line treatment in certain anatomic subtypes of congenital mitral stenosis because these lesions respond poorly to surgical valvuloplasty. Dilation may be reasonably attempted in patients with either "typical" congenital mitral stenosis or multiple orifice MV. Neither parachute MV nor supravalvar mitral ring should be dilated; in the former because success has never been demonstrated and in the latter because surgical therapy is clearly superior. While results after dilation of bioprosthetic valves have by and large been disappointing, percutaneous valve therapy has emerged as an effective treatment.

Catheter-Based Valve Repair and Replacement

Over the last decade, advances in catheter-based technology have resulted in new minimally invasive therapeutic options for pulmonary and aortic valve replacement, and for MV repair. These advances have transformed the treatment of valvular and structural heart disease for both pediatric and adult patients.

Pulmonary Valve Replacement

The era of percutaneous heart valve replacement began in the earnest following the first published report of percutaneous pulmonary valve replacement (PPVR) procedure using the Melody Valve (Medtronic, Inc., Minneapolis, MN) in September 2000. Since that time, there have been approximately 3,000 procedures performed worldwide using either the Melody or the SAPIEN (Edwards Lifesciences LLC, Irvine, CA) device. Both devices consist of balloon-expandable stents, with a valve mechanism sewn into the center (Fig. 74.6). At present, due to size limitations, PPVR is mostly limited to postoperative patients with preexisting RV–PA conduits and/or bioprosthetic valves. Alternative devices and strategies for PPVR in patients with dilated RV outflow tracts are in development and will likely play a major role in the management of postoperative pulmonary insufficiency in the years to come.

Figure 74.7 shows an example of a typical PPVR procedure using the Melody valve in a patient with a stenotic and insufficient RV–PA conduit. Through femoral or jugular venous access, conduit stenosis is usually alleviated first by placement of a bare metal stent, with subsequent implantation of the Melody valve through a 22F delivery system. Figure 74.8 shows an example of Melody valve implantation into a dysfunctional bioprosthetic valve.

The early and intermediate results of PPVR have been reported, with high success rates, low morbidity and mortality, and acceptable reintervention rates. However, procedural complications, including coronary artery compression and RV–PA conduit rupture, can be life threatening, and stent fractures following implantation are common, necessitating ongoing medical follow-up.

Transcatheter Aortic Valve Implantation

Transcatheter aortic valve implantation (TAVI) was first performed in 2002; since that time it has evolved into a routine procedure for symptomatic aortic stenosis performed in specialized centers around the world. First introduced as an alternative to medical treatment for elderly patients deemed too high-risk for standard surgical valve replacement, TAVI has subsequently been proven to be equivalent to surgery in respect to mortality rates in randomized prospective clinical trial in a standard-risk population (PARTNER and PARTNER 2). Currently, two TAVI devices are commercially available: the SAPIEN valve (Edwards Lifesciences LLC, Irvine, CA) and

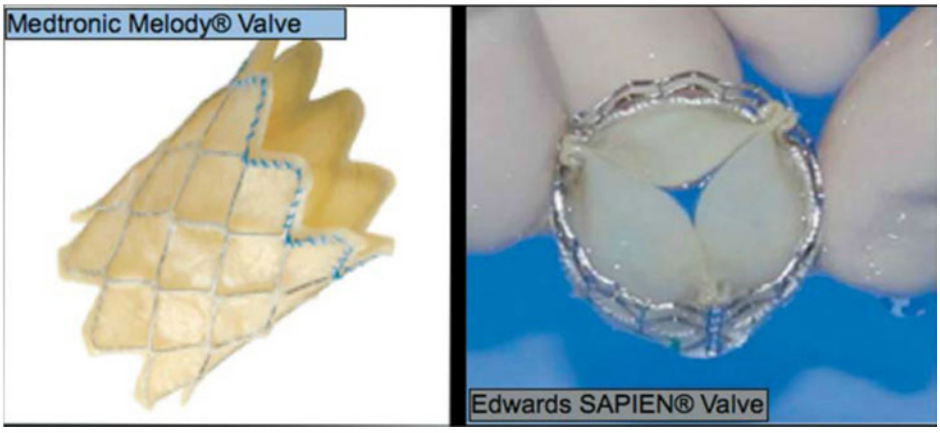


Fig. 74.6. Two devices are currently in use for percutaneous pulmonary valve replacement: The Melody Valve (Medtronic, Inc.) and SAPIEN Valve (Edwards Lifesciences, LLC).

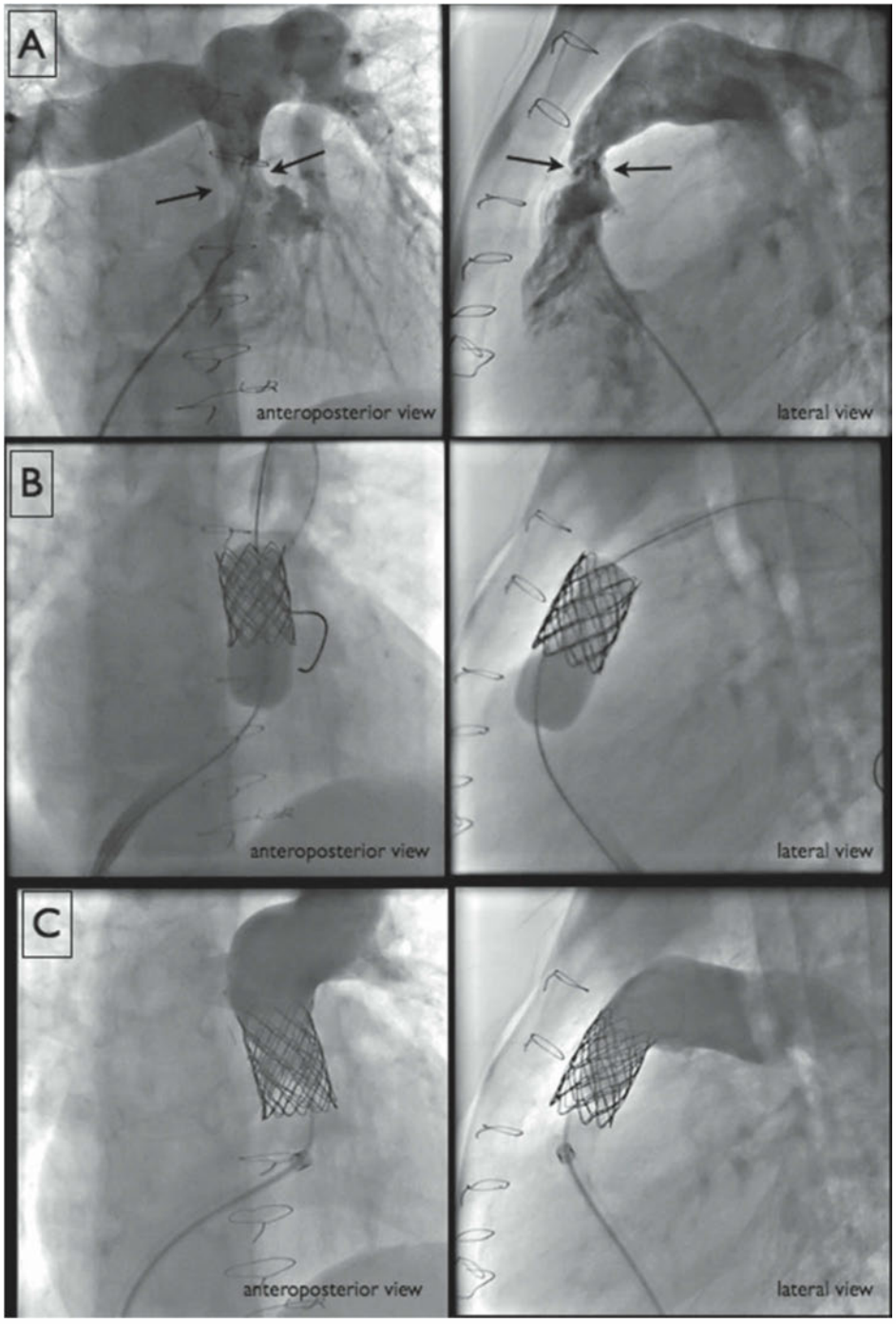


Fig. 74.7. Percutaneous pulmonary valve replacement using the Melody valve in a RV–PA conduit. (**A**) Anteroposterior and lateral views of an angiogram showing severe RV–PA conduit narrowing (*arrows*) and severe pulmonary insufficiency. (**B**) A bare metal stent was deployed to alleviate the stenosis, and then the Melody valve was implanted via balloon inflation. (**C**) Follow-up angiogram in the main pulmonary artery shows no pulmonary insufficiency, and a dramatic increase in the diameter of the conduit.

the CoreValve (Medtronic, Inc., Minneapolis, MN) (Fig. 74.9).

The role of TAVI in younger patients and in those with congenital heart disease is still evolving with the experience thus far limited to small case series using off-label devices. Next-generation devices are on the horizon, which may expand the indications for TAVI into younger, healthier individuals, including those with congenital defects.

Mitral Valve Repair Technology

Improved understanding of the mechanisms of MV dysfunction, combined with advances in catheter-based technologies, has resulted in the rapid development of percutaneous mitral valve repair (PMVR) techniques over the last decade. All of these new technologies are designed to mimic well-established surgical MV repair principles. These include therapies aimed at the MV leaflets, the MV annulus, and at the submitral apparatus (chordae). The most successful PMVR device to date is the MitraClip device, which mimics the surgical Alfieri technique by bringing the anterior and posterior leaflets of the MV together, creating a "double-orifice" MV. This approach re-establishes leaflet coaptation, thereby reducing mitral regurgitation. The MitraClip has been shown to be noninferior to surgery in randomized clinical trial (EVEREST and EVEREST2). Other technologies, including percutaneous annuloplasty devices and artificial chordae, are in clinical or preclinical testing.

Despite the many promising catheter-based MV repair technologies under development, one-stage percutaneous MV replacement remains elusive. However, surgical (transapical or transatrial) and transcatheter MV replacement via valve-in-valve (VIV) and valve-in-ring (VIR) (Fig. 74.10) procedures using the SAPIEN and Melody valves have been published in both preclinical and clinical reports. This early experience underscores the potential of repurposing "failing" surgical hardware (bioprosthetic valves and annuloplasty rings) as "landing zones" for subsequent minimally invasive valve replacement.

Balloon Angioplasty and Endovascular Stenting

Balloon dilation has been used for over a decade to treat native and postoperative aortic, pulmonary artery, and venous obstructions. Balloon-expandable endovascular stents are used to treat pulmonary artery stenoses, venous obstructions, and aortic coarctation. In balloon angioplasty, the vessel is dilated to a diameter significantly greater than the expected final size. Relief of obstruction is generally due to intimal and medial disruption at the site of stenosis. Vascular integrity after successful angioplasty is dependent on an intact adventitia. As a general rule, dilation should not be performed within 6 weeks to 2 months after surgical dissection of the region.

Aortic Arch Obstructions

Balloon angioplasty for native coarctation and postsurgical recoarctation has similar results with residual gradients

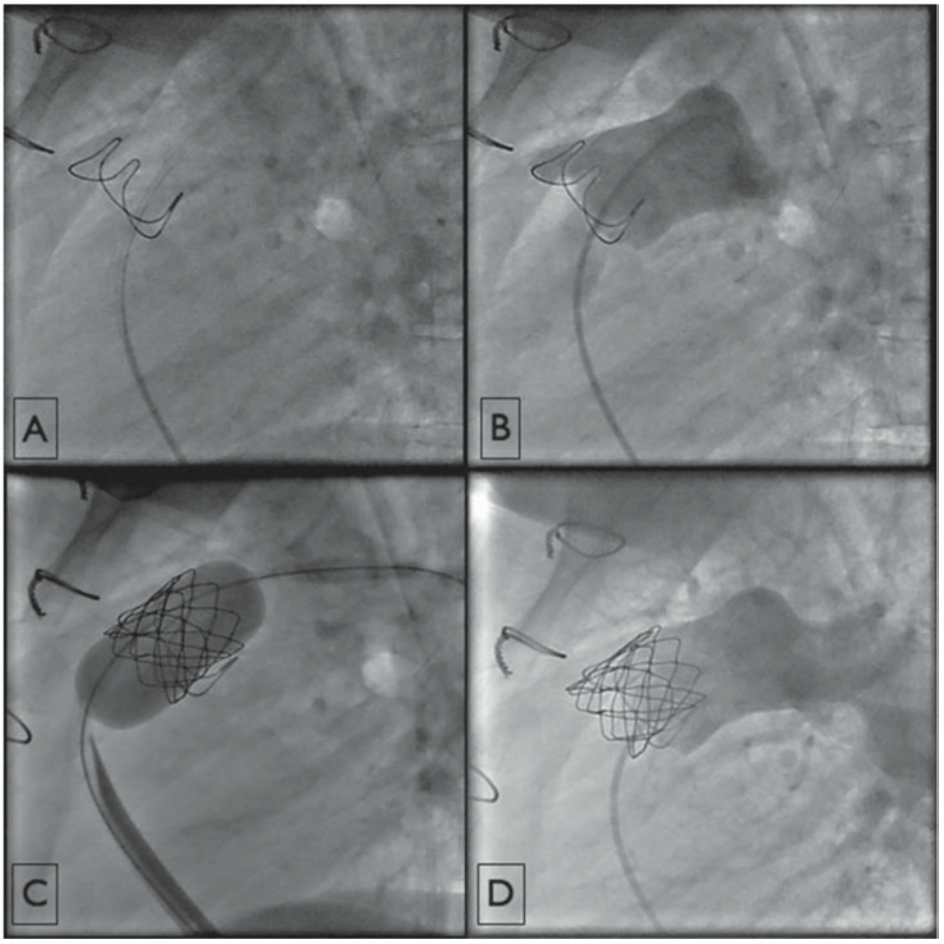


Fig. 74.8. (**A**) A lateral view of a bioprosthetic valve wire "cage" in the pulmonary position. (**B**) Pulmonary arteriogram shows significant pulmonary valve insufficiency. (**C**) Deployment of the Melody valve into the bioprosthetic valve cage via balloon inflation. (**D**) Follow-up pulmonary arteriogram shows no pulmonary insufficiency.

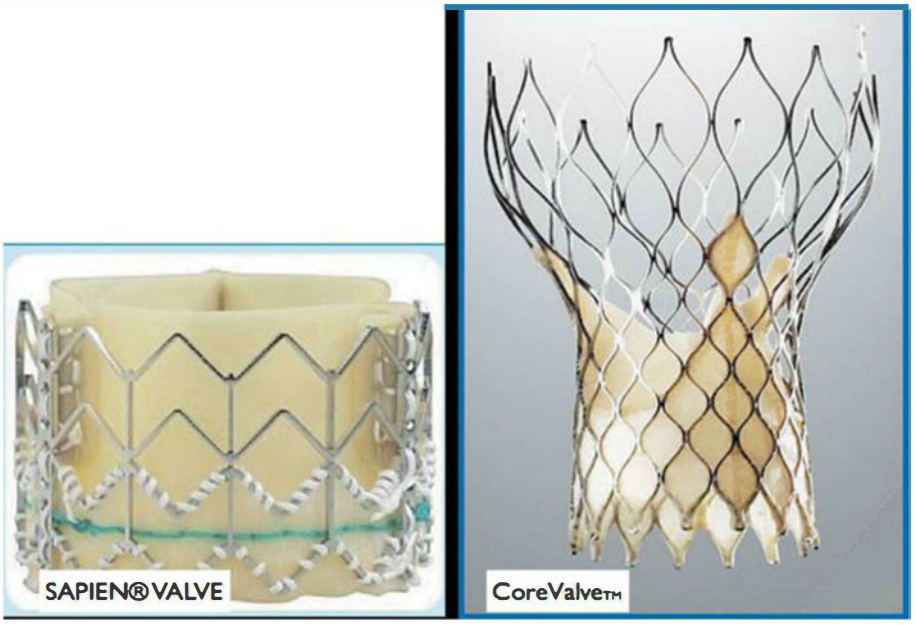


Fig. 74.9. Current devices used for transcatheter aortic valve implantation. On the left is the Edwards SAPIEN valve, and on the right is the Medtronic CoreValve.

of <20 mmHg in 80% to 90% of cases. Restenosis is more common after dilation of native coarctation, largely due to a particularly high rate of restenosis when dilation is performed in young infants. Balloon angioplasty is the treatment of choice for recoarctation of the aorta after previous surgical repair. Given the high restenosis rate after dilation, current data suggest that surgery is the optimal treatment for native coarctation in the very young infant. For native coarctation in older infants and children, dilation is an acceptable alternative to surgery. Endovascular stenting for coarctation is now a widely used form of therapy for both native and recurrent coarctation in older children and adults. The early and intermediate-term results of this form of therapy have been documented in several series with very high success rates. In most reported series complication rates are low but not insignificant; aneurysm formation may occur even late after intervention and necessitates follow-up with scheduled CT or MRI arch imaging. Aortic wall complications are more prevalent in patients with aortopathies (such as William and Turner syndromes) and older adults. Covered stent angioplasty is generally the recommended catheter-based treatment approach for such patients and results in acceptable short- and intermediate-term outcomes. Given the increased surgical risk in many such patients, transcatheter treatment is often the optimal strategy.

Results of dilation for recoarctation after Norwood procedures for HLHS are comparable to those in simple recoarctation though risks tend to be higher.

Branch Pulmonary Artery Stenosis

Balloon angioplasty for pulmonary artery stenosis is successful in 80% of instances, when success is defined as an increase in vessel diameter of more than 50%. In patients with complex or multiple pulmonary artery narrowings, angioplasty procedures may be lengthy and challenging. Wires are positioned in the largest distal branches and dilations performed with short balloons to minimize risk of aneurysm formation. Potential vascular complications include perforation, aneurysm formation, dissection, or thrombosis. Mortality rates for the procedure may be as high as 0.5% to 1%. Patients require careful follow-up, as restenosis after successful dilation is not uncommon. Endovascular stenting has proven to be a very effective treatment for branch pulmonary artery obstruction. Stents are deployed on

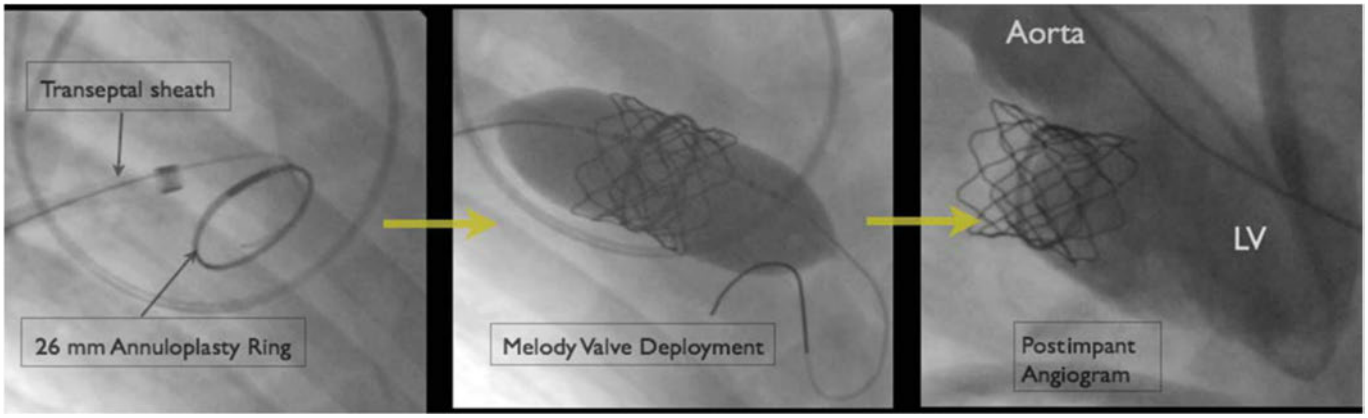


Fig. 74.10. This figure shows percutaneous, transvenous mitral valve replacement using the Melody valve in a previously placed annuloplasty ring in an Ovine model. The valve is well seated within the ring, and there is no mitral regurgitation, and no left ventricular outflow tract obstruction. LV, left ventricular.

angioplasty balloons advanced through long sheaths to the site of the stenosis (Fig. 74.11). Disadvantages of endovascular stenting include the need for redilation to accommodate growth, restenosis from neointima formation (most significant in small diameter stents), and prothrombotic potential of the foreign body (particularly in low flow vessels). In general, stents are most effective in the treatment of proximal branch pulmonary artery obstructions where it is rare for angioplasty alone to completely relieve vascular obstruction.

Branch pulmonary artery narrowings may occur in isolation. More commonly, these lesions occur in association with other defects, either congenitally (tetralogy of Fallot—particularly with pulmonary atresia, truncus arteriosus) or as a result of prior surgery (pulmonary artery banding, shunt procedures, and arterial switch operations). Treatment of these complex lesions often requires a combination of catheter and surgical intervention. Dilation in the early postoperative period and surgery early after dilation may both be risky. Thus, it is essential that balloon angioplasty and surgery be planned and staged to minimize such risks. Catheter intervention should nonetheless be considered for the occasional patient who develops signs of right heart failure immediately postoperatively due to previously unrecognized or inadequately treated pulmonary artery distortion. Endovascular stenting is the preferred transcatheter treatment in these patients as the risk of vascular disruption is lower than with angioplasty.

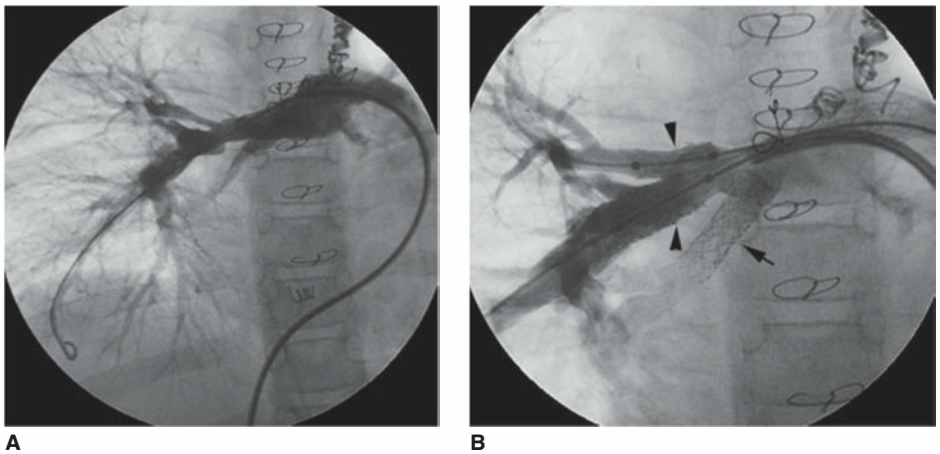


Fig. 74.11. (**A**) Right pulmonary arteriogram in patient with multiple complex branch pulmonary artery stenoses in patient with tetralogy of Fallot, pulmonary atresia, and multiple aortopulmonary collaterals after unifocalization. (**B**) Repeat angiogram after complex stenting procedure with placement of stents in tandem (*arrow*) as well as so-called "kissing" (*arrowhead*) orientations, demonstrated significant improvement.

Complex branch pulmonary artery obstruction due to multiple stenoses in series and parallel remain among the most difficult problems in the care of children with congenital heart defects. Current therapy includes standard balloon angioplasty, cutting balloon angioplasty, and the use of stents. While stents usually result in the most complete resolution of stenoses, their use in more distal vessels is limited because of the risk of "jailing" multiple branching segmental arteries effectively reducing the arborization of an already compromised pulmonary artery tree. Cutting balloon angioplasty is more effective than the standard static balloon dilation, however, probably carries an increased risk of vessel wall injury.

Systemic and Pulmonary Veins

Balloon angioplasty or stent placement for systemic venous obstructions may be very effective and has been particularly useful in patients with systemic venous pathway obstruction after atrial switch operations and cavopulmonary connections. Treatment of pulmonary vein stenoses by angioplasty has generally been disappointing. Though dilation often results in initial angiographic and hemodynamic improvement, restenosis is rapid and inevitable. Endovascular stenting of pulmonary vein stenosis has been somewhat more successful but only in select patient groups. The duration of success depends on the underlying disorder. Thus, some patients with discrete obstruction of large normal pulmonary veins after previous procedures (usually older individuals after lung transplantation or repair of partially anomalous pulmonary venous return with anastomotic narrowing, or stenosis after radiofrequency ablation for atrial flutter) will have lasting improvement with

stenting. However, infants with pulmonary vein stenosis invariably develop restenosis either from neointimal proliferation within the stent or progressive vascular disease in the more peripheral pulmonary veins. There has been interest in the use of adjuvant therapy with systemic antimetabolites or drug-eluting stents in this disease, but the efficacy of these methods has yet to be demonstrated.

Conduits and Shunts

Endovascular stents can be very useful for relief of obstruction in surgical conduits and shunts. Progressive obstruction in conduit reconstruction of the right ventricular outflow tract is to be expected as infants grow. However, in many cases the process is accelerated due to external compression or shrinkage of homograft conduit from host inflammatory response. In these instances, the conduit can often be stented back to near its original diameter prolonging the life of the prosthesis. In younger patients, this technique finds its most useful application for the treatment of cyanosis due to conduit stenosis after the Sano modification of the Norwood operation (Fig. 74.12). In the older patient population, it is in use of the percutaneous valve for conduit failure as described previously.

The wide availability of coronary stents mounted on very low profile balloons allows treatment of surgical shunt obstruction or occlusion even in very small children (Fig. 74.13).

Closure Procedures

A variety of techniques have been developed for closure of vascular connections and intracardiac defects. The versatility of these techniques has allowed their use in a wide range of congenital lesions.

Embolizations

A variety of catheter techniques are available for vascular embolization in patients with congenital cardiovascular defects. These include metal coils, synthetic particles, and self-expanding controlled delivery nitinol plugs (Amplatzer Vascular Plug). After careful angiographic definition of vessel size and anatomy, the delivery catheter is advanced into the structure to be embolized. The type of delivery catheter varies from small microcatheters for delivery of particles or microcoils to large guide catheters or sheaths for deployment of vascular plugs. Under angiographic guidance the embolization device or material is then placed. The most common complication of transcatheter embolization is misplacement of the embolization material. While errant coils or plugs can usually be removed by transcatheter retrieval, this is not possible in the case of particles. Other less common complications include vascular injury at catheter entry site, hemolysis from partially occlusive devices, and endovascular infection.

Embolization is definitive therapy for some lesions. The technique has been widely

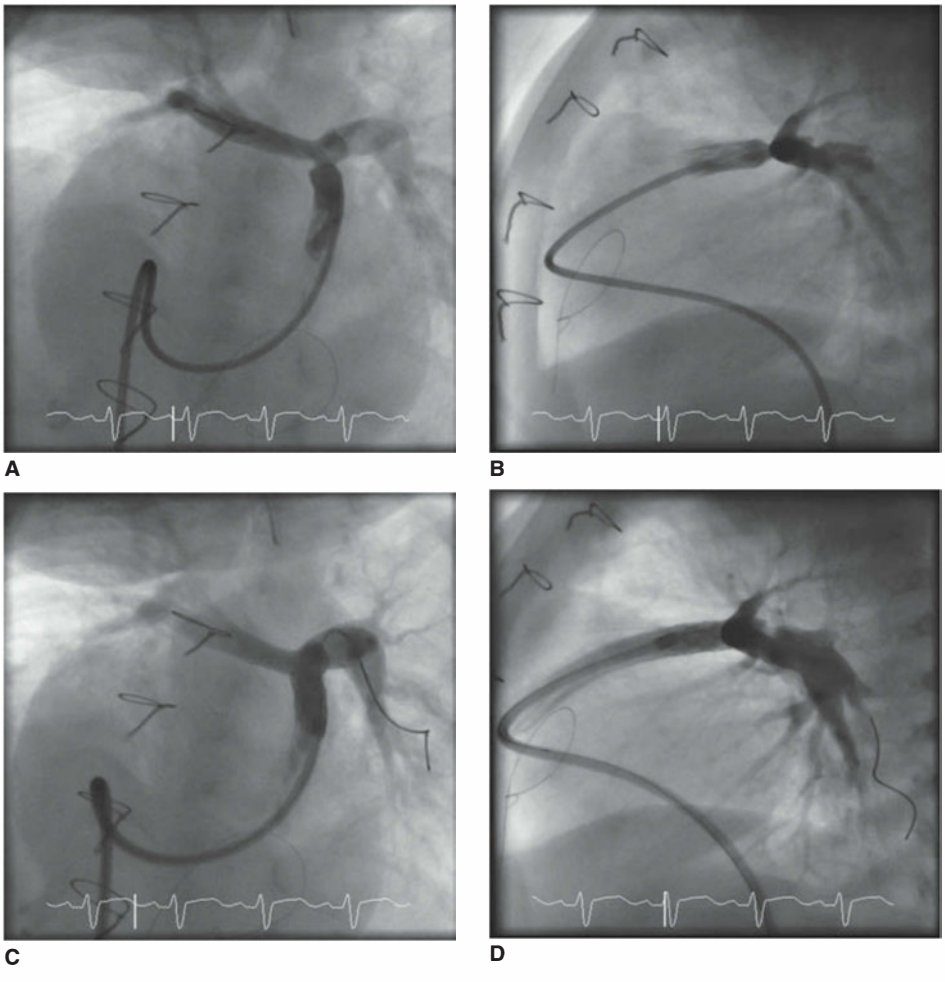


Fig. 74.12. Anteroposterior (**A**) and lateral (**B**) projections of right ventricular angiogram in a patient who has obstruction at the insertion of a right ventricle to pulmonary artery conduit at its distal insertion after Norwood with Sano modification. Repeat angiogram in this patient after stenting of the distal conduit shows relief of obstruction in anteroposterior (**C**) and lateral (**D**) projections.

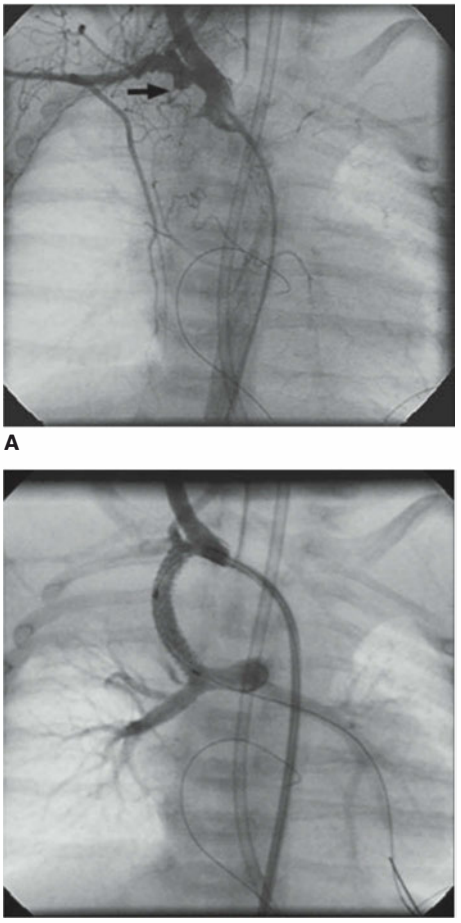


Fig. 74.13. (**A**) Retrograde pigtail catheter injection in innominate artery demonstrating completely occluded right modified Blalock–Taussig shunt (*arrow*). (**B**) Restored patency of shunt after stenting.

used for the treatment of persistent PDA (see below) and has proven efficacious in the treatment of coronary cameral fistulae. In addition, transcatheter embolization is often performed as part of staged intervention in patients with complex defects. Examples include for redundant aortopulmonary collateral arteries in tetralogy of Fallot with pulmonary atresia and previously placed surgical shunts in patients with multiple sources of pulmonary blood flow. Patients with single ventricle heart disease often undergo embolization procedures. This may be for persistent left superior venae cavae or other decompressing veins in patients undergoing bidirectional Glenn or Fontan procedures, or for the treatment of systemic to pulmonary collateral arteries. The indications for and efficacy of this procedure in the latter circumstance remain poorly defined. All patients with single ventricle heart disease develop systemic to pulmonary collateral artery formation. In some patients, these collaterals may result in a substantial volume load, accounting for over 50% of total pulmonary blood flow. Embolization seems to result in acute decrease in this flow but preliminary evidence suggests that the effect may not be durable. Further work is underway to clarify this issue.

A small percent of patients with a variety of congenital heart defects including tetralogy of Fallot, transposition of the great arteries, and single ventricle defects are plagued by recurrent hemoptysis often years after surgical therapy. Catheter embolization is the main treatment modality in this situation. Bronchoscopy may be useful in localizing the side and lobe where hemorrhage originates. Selective digital subtraction angiography is performed to identify potential sources of bleeding. Though it is exceedingly rare to visualize extravasation of contrast (active hemorrhage), candidate vessels are identified by their characteristic appearance. These are usually bronchial arteries. The vessels are occluded with particle emboli. Typically, multiple feeder vessels must be embolized (Fig. 74.14).

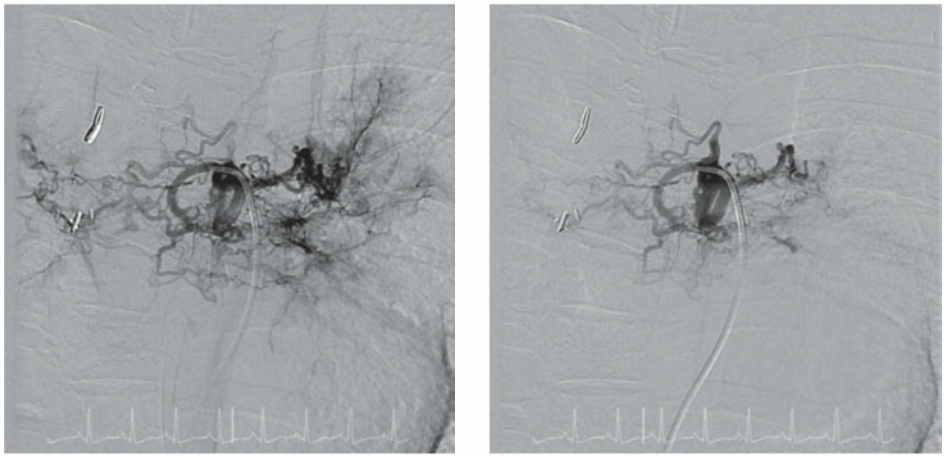


Fig. 74.14. Digital subtraction angiography performed in bronchial artery in a patient with hemoptysis after Fontan. (**A**) Prior to embolization therapy, there is an extensive network of vessels arising from the bronchial artery with pathologic opacification of pulmonary parenchyma on both sides. (**B**) After particle embolization of feeders to both right and left, the feeder vessels appear truncated and there is markedly reduced opacification of pulmonary parenchyma.

Device Closures

Devices designed for closure of defects including PDA, atrial septal defects, and ventricular septal defects have been developed and tested for almost 40 years. Many such devices are in routine clinical use and several others are in clinical trials. The majority of transcatheter device procedures are performed for closure of PDA or atrial communications: either typical secundum atrial septal defects or patent foramen ovale in patients with prior stroke. Device closure of PDA is well established in young children and adults, and recent advances have made it possible in infants as small as 2 kg (Fig. 74.15).

The most widely used device for closure of atrial defects is the Amplatzer Septal Occluder. This device has been used in tens of thousands of cases worldwide. It is composed of a woven frame of superelastic nitinol in which have been sewn three disks of woven Dacron (Fig. 74.16). The device is attached via a microscrew to a delivery cable. The assembly is advanced through a delivery sheath, which has been positioned through the defect to be closed. It is then deployed such that it straddles the defect (Fig. 74.17). The Helex Septal Occluder is currently the only other available device approved in the United States for ASD closure (Fig. 74.18). This device is composed of a Goretex patch sewn onto a single helical strand of nitinol. When advanced out of its delivery catheter, the device forms two disks—one on each side of the atrial septum (Fig. 74.18). This device is easily repositioned and retrieved but is not suitable for large defects. The Amplatzer PFO Occluder and the Gore HELEX device have Humanitarian Device Exemptions for closure of patent foramen ovale in patients who have had strokes and failed anticoagulant therapy.

The majority of secundum atrial septal defects can be successfully closed by catheter-delivered devices. Typically, the

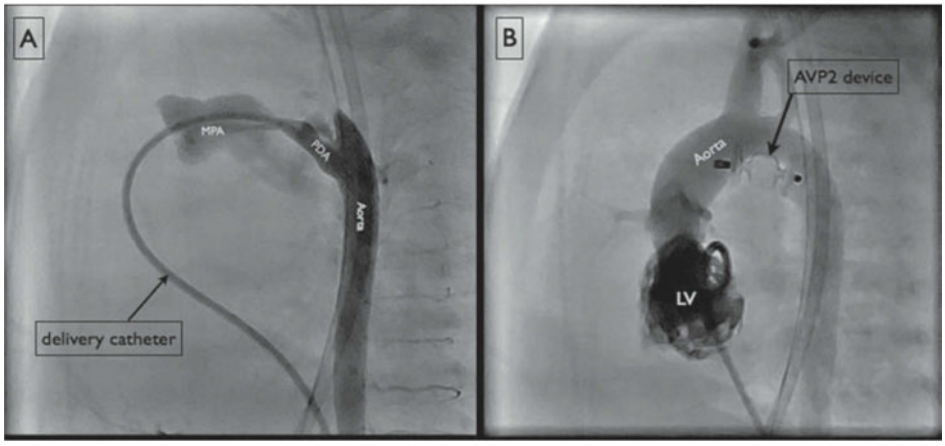


Fig. 74.15. (**A**) Lateral view of a descending aortogram in a premature infant with a hemodynamically significant patent ductus arteriosus. (**B**) Left ventriculogram following PDA closure with an Amplatzer Vascular Plug 2. There is no residual flow through the ductus arteriosus, and the aortic arch is unobstructed.

procedure is performed with combined echocardiographic (transesophageal or intracardiac) and fluoroscopic guidance. Though less data have been accumulated for transcatheter closure of ventricular septal defects, results are encouraging for certain lesions. Because of impingement on atrioventricular or semilunar valves, the devices are not suited for closure of atrioventricular canal or malalignment defects. Transcatheter devices, such as the Amplatzer muscular VSD Occluder, have been used successfully for closure of muscular and postoperative peripatch septal defects (Fig. 74.19). In smaller children, intraoperative closure via a sheath placed periventricularly has been successfully used to close difficult defects. The Amplatzer Membranous VSD Occluder is in clinical trial for closure of membranous VSD.

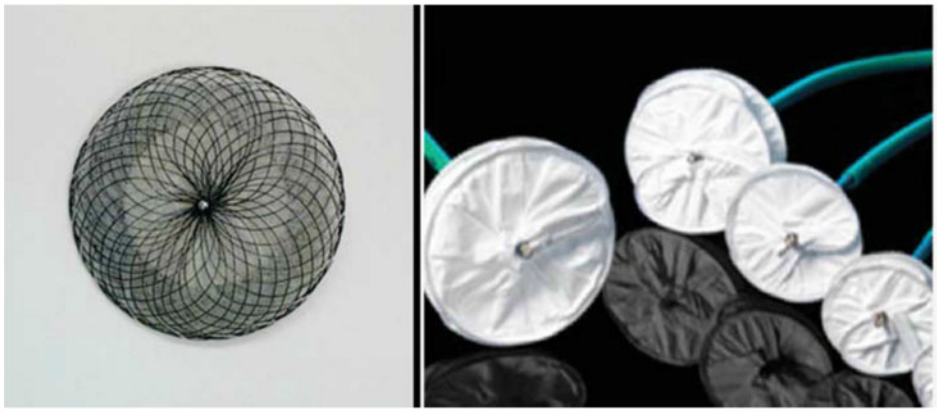


Fig. 74.16. The Amplatzer Septal Occluder device (St. Jude Medical) and the Helex® device (Gore, Inc.). Both are FDA approved for closure of atrial septal defects.

Retrievals

Several specialized catheter systems have been designed for intravascular retrievals. These devices include snares, baskets, and grabber catheters. Most can be advanced through small introducers and are therefore applicable even in small infants. With the exception of large noncollapsible

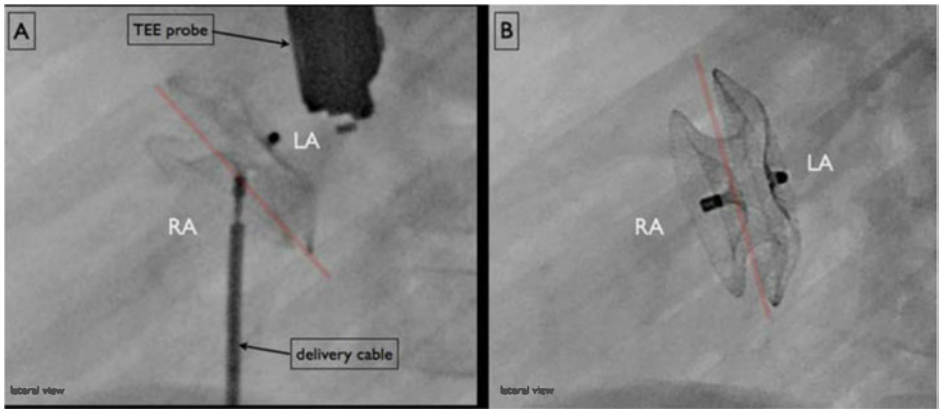


Fig. 74.17. (**A**) Lateral view of Amplatzer device implantation for atrial septal defect closure. In this picture, the device is opened within the ASD, but still attached to the delivery cable. The plane of the atrial septum is shown in *red*. (**B**) The device reorients slightly following release from the delivery cable. TEE, transesophageal echocardiogram.

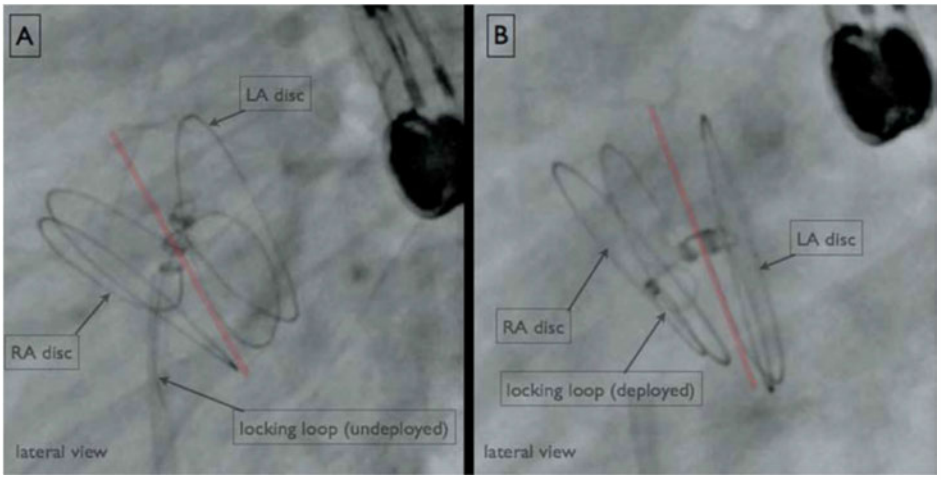


Fig. 74.18. (**A**) Lateral view of HELEX device implantation for closure of an atrial septal defect. The device is positioned well across the atrial defect, still attached to the delivery system. The plane of the atrial septum is shown in red. (**B**) HELEX device fully deployed and released in stable position within the atrial septum.

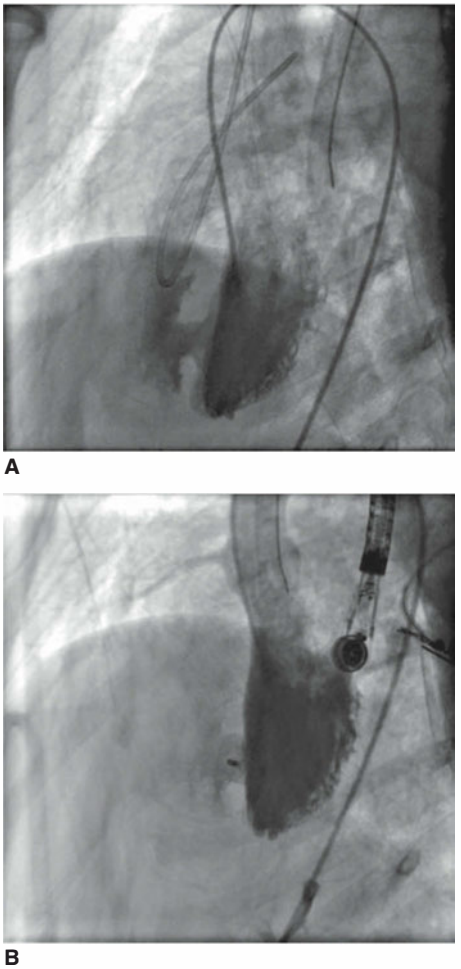


Fig. 74.19. (**A**) Retrograde pigtail injection into left ventricle demonstrates a large muscular ventricular septal defect. (**B**) After deployment of and Amplatzer Muscular ventricular septal defect device, the defect is effectively closed with trivial residual shunt through the Dacron of the device.

endovascular devices (expanded stents), most errant catheters, and intravascular foreign materials can be successfully removed without the need for surgery. In children undergoing heart surgery, this approach is particularly useful for removal of fractured, retained lines.

Combined Surgical and Catheter Interventions

The last decade has witnessed a proliferation of procedures combining surgical and transcatheter techniques. The most commonly performed of these "hybrid" procedures include per-ventricular closure of ventricular septal defects and what has been referred to as the "hybrid stage I": bilateral pulmonary artery banding combined with ductal stenting for palliation of HLHS or its variants. This alternative approach for HLHS avoids newborn cardiopulmonary bypass. There have been no concurrent controlled trials comparing outcome of hybrid with standard surgical palliation for HLHS. Contemporary results among centers suggest equipoise. There is great variability in approach; at some centers only one form of treatment is performed, but more commonly both are offered to patients. However, the reasons for stratifying patients to one treatment versus the other vary greatly. Choice of procedure may depend on concomitant risk factors deemed to increase the risk of cardiopulmonary bypass such as low birth weight or associated anomalies. Parental preference may also play a role. While there are as many different approaches to surgical stratification of HLHS patients as there are centers, it should be emphasized that there are clear anatomic risk factors for hybrid palliation. The most important is anatomy predisposing to coronary ischemia after ductal stent placement. Patients at risk are those with mitral or aortic atresia in whom there is anatomic substrate for retrograde coarctation of the aorta. Such patients should be treated with conventional Norwood operations.

The optimal environment for hybrid palliation is a specifically designed hybrid operating/catheterization suite but the procedure can be accomplished in a traditional catheterization laboratory. Bilateral pulmonary bands are placed by median sternotomy usually by using a cut ring of Gore-Tex shunt. Next, a purse string is placed into the most proximal portion of the main pulmonary artery through which a sheath is placed. Angiography is performed assessing the adequacy of the pulmonary bands as well as to evaluate ductal anatomy for stent placement. A guide wire is then advanced to the descending aorta and ductal stenting performed. After follow-up angiography, the wire and sheath are removed (Fig. 74.20). Prior to hospital discharge, the patient is brought back to the catheterization laboratory for balloon atrial septostomy.

Patients who have undergone hybrid palliation for HLHS require particularly close monitoring to screen for arch obstruction (either retrograde or anterograde) as well as the development of restrictive transatrial flow. They are typically referred for the subsequent stage palliation the so-called comprehensive stage II, at 3 to 4 months of age. This operation incorporates both the Norwood and the bidirectional Glenn procedures.

Fetal Intervention

A series of principles both medical and ethical must guide fetal intervention. The fundamental medical principles include the following: First, the condition in question occurs as a consequence of a relatively simple primary abnormality arising late in embryogenesis (or after). Second, this primary abnormality leads to subsequent changes that are either lethal or result in major morbidity. Third, that prenatal intervention is feasible at a stage where the above process can be interrupted. The guiding ethical framework includes (1) the proposed fetal intervention, which is reliably expected on the basis of animal studies to be life saving or prevent serious irreversible disease. (2) Among the alternatives, the intervention is designed to involve the least risk of death and morbidity to the fetus. (3) The risk of death or injury to the pregnant woman is very low. The application of these principles to cardiac malformations has been imperfect because investigators have failed in attempts to generate animal models for the relevant malformations.

Balloon dilation of aortic stenosis was the first fetal cardiac catheter intervention attempted, over two decades ago. The procedure was clearly groundbreaking and a technical tour de force, but unsuccessful in altering the clinical outcome. Approximately a decade later, other investigators rekindled interest in this method in dramatic manner. Approaching the same lesion at a substantially earlier stage

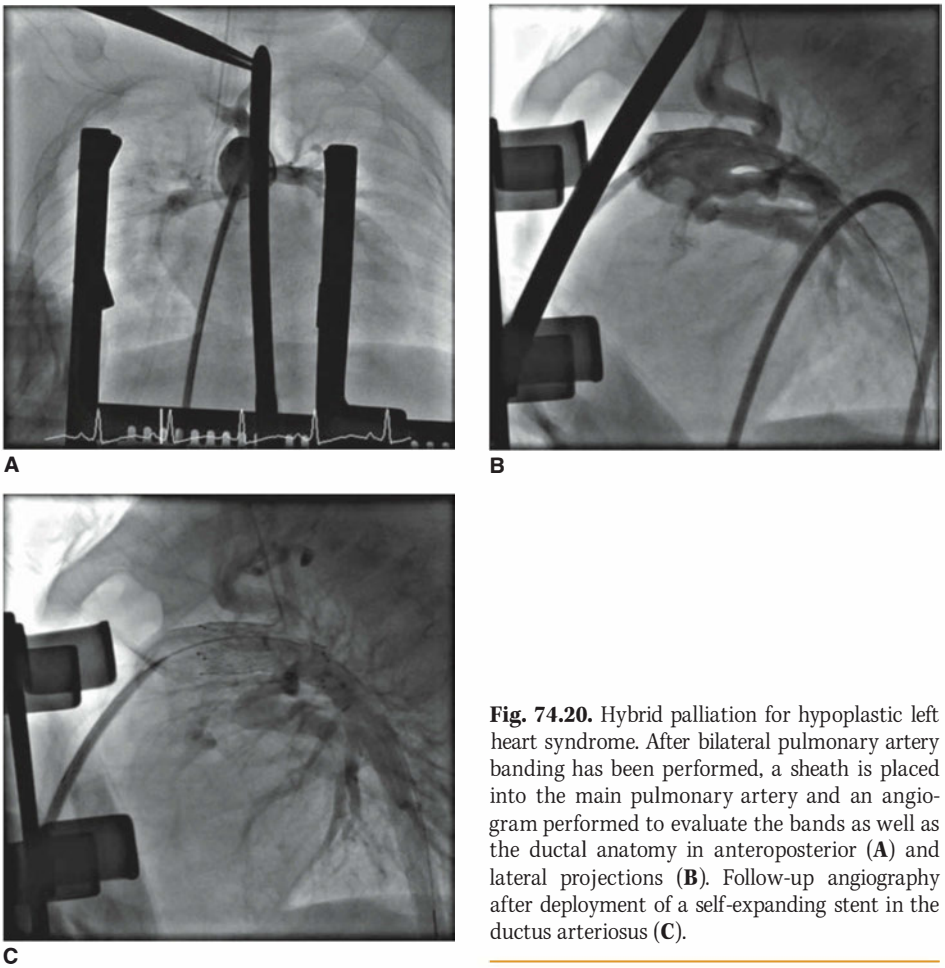


Fig. 74.20. Hybrid palliation for hypoplastic left heart syndrome. After bilateral pulmonary artery banding has been performed, a sheath is placed into the main pulmonary artery and an angiogram performed to evaluate the bands as well as the ductal anatomy in anteroposterior (**A**) and lateral projections (**B**). Follow-up angiography after deployment of a self-expanding stent in the ductus arteriosus (**C**).

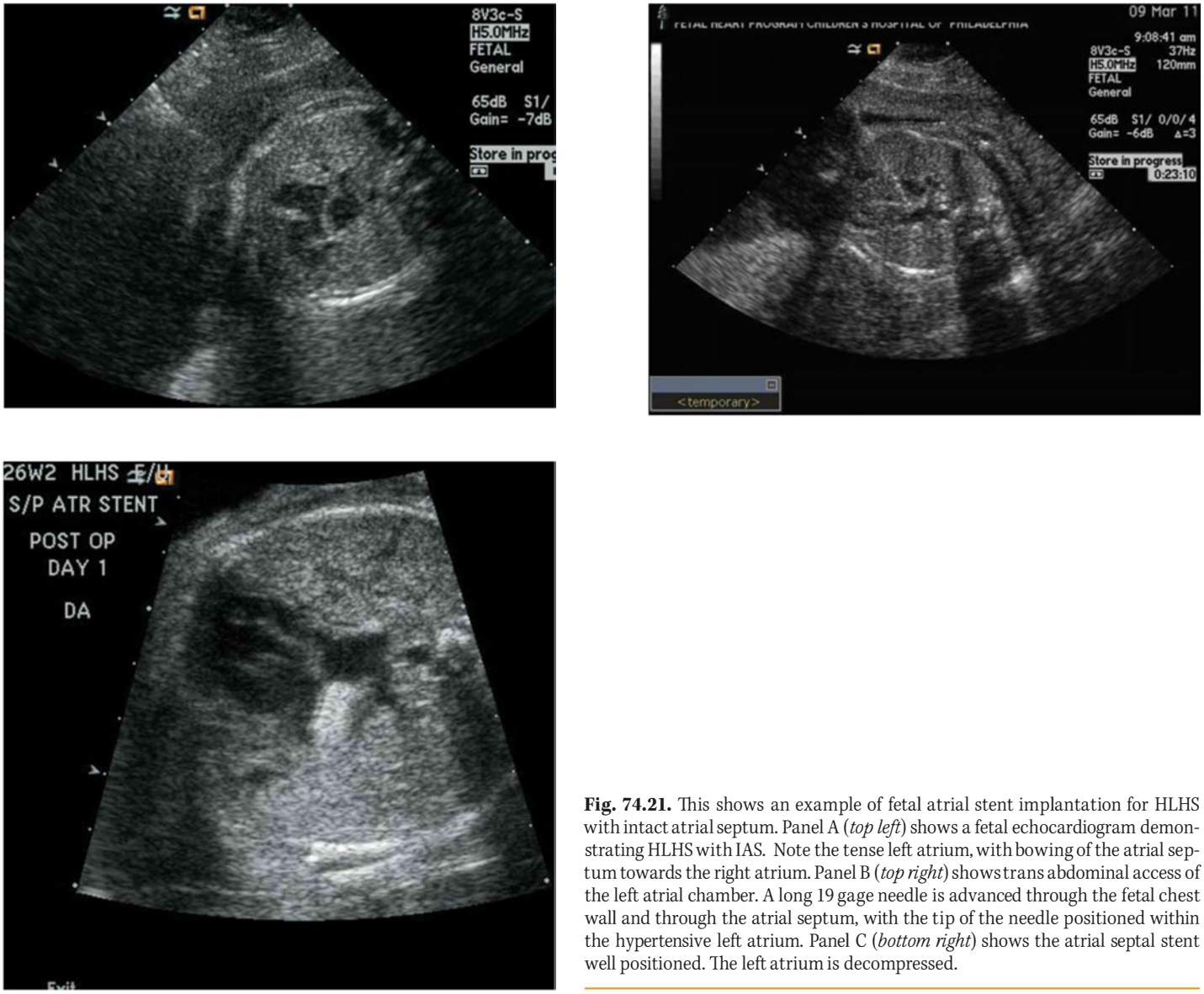


Fig. 74.21. This shows an example of fetal atrial stent implantation for HLHS with intact atrial septum. Panel A (*top left*) shows a fetal echocardiogram demonstrating HLHS with IAS. Note the tense left atrium, with bowing of the atrial septum towards the right atrium. Panel B (*top right*) shows trans abdominal access of the left atrial chamber. A long 19 gage needle is advanced through the fetal chest wall and through the atrial septum, with the tip of the needle positioned within the hypertensive left atrium. Panel C (*bottom right*) shows the atrial septal stent well positioned. The left atrium is decompressed.

in gestation, not only was the procedure technically successful, but also in some cases it appeared that therapeutic success was achieved by preventing the development of HLHS. It should be emphasized that only a small fraction of HLHS has even the potential to be averted in this manner, as most cases of HLHS are not due to fetal aortic stenosis. Nonetheless, over the years a substantial experience with fetal intervention has accrued for a small number of conditions. The majority of procedures have been for dilation of fetal aortic stenosis to prevent HLHS. As most infants born with HLHS now survive, the goal in this lesion is primarily to decrease morbidity rather than avert mortality. Another condition for which fetal intervention has been performed is HLHS and intact atrial septum. In patients born with this combination of abnormalities, in utero left atrial outlet obstruction results in pathologic alteration of the pulmonary vasculature. Neonatal mortality is markedly increased compared with other cases of HLHS. Fetal intervention is performed to create an atrial septal communication with the goal of preventing pulmonary vascular disease and decreasing mortality (Fig. 74.21).

SUMMARY AND CONCLUSIONS

Transcatheter therapies are now available for many cardiovascular lesions. The patient with complex lesions may require several operations and interventional catheterizations. In these cases, outcome is optimal when surgeon and cardiologist, both educated in the range of therapeutic options, plan treatment strategies collaboratively.

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EDITOR'S COMMENTS

The advent of transcatheter approaches for the treatment of many cardiovascular lesions in children has resulted in the need for close cooperation between surgeons and interventional cardiologists in planning operative therapies. Because the results with balloon dilation of the aortic valve in critical aortic stenosis in newborns are similar to those of open surgical valvotomy, there is little added benefit to a surgical approach in these patients. It should be emphasized that the results are no better with transcatheter approaches, and there remains significant mortality in these newborns. A certain proportion of patients will have significant residual aortic valve stenosis or insufficiency and ultimately require additional intervention. Because of the success of balloon dilation of aortic stenosis in young patients, however, it is rare to require pulmonary autograft valve replacement or other surgical approaches to the aortic valve in infancy. Most patients can be at least satisfactorily palliated with balloon dilation, and then if significant insufficiency or recurrent stenosis not amenable to additional balloon dilation occurs, pulmonary autograft aortic valve replacement can be undertaken in early childhood. As noted by the authors of this chapter, the long-term results of even pulmonary autograft valve replacement are unknown, and therefore all of these procedures should be considered palliative at the present time.

Hemodynamic evaluation at cardiac catheterization is important for determining operability in many patients with complex congenital heart disease with significant shunts. A shunt calculation in many of these patients of 2:1 or greater is an indication for surgical repair. However, there is evidence that the presence of even modest shunts should prompt operative or transcatheter correction because the long-term effects on pulmonary vascular resistance of even a modest shunt are still variable. Therefore, in most situations a left-to-right shunt ratio of 1.5:1 or greater should be considered an indication for operative or transcatheter intervention, and even minor ductus arteriosus patency should be considered an indication for surgical or coil closure because subacute bacterial endocarditis is still a risk factor in these patients. Even ASD with a modest left-to-right shunt should not be considered a completely benign lesion because some of these patients will develop pulmonary vascular changes even with a relatively small defect. It appears that there are some patients who have a particular predisposition to vascular disease, and even a modest left-to-right shunt may exacerbate the development of progressive pulmonary vascular obstructive disease in these patients. In addition, as patients increase in size and age, shunts are not fixed; therefore, a significant shunt may be present in a patient with an ASD who at one isolated determination may have a relatively modest left-to-right shunt. As growth occurs, the shunt can increase significantly. With the relatively low risk of intervention in the current era, in my opinion, even modest shunts should be addressed.

The usefulness of occluder devices for ASD is established. It is of interest that the use of atrial septal occluder devices has not been associated with a significant incidence of pericardial effusion, which is the cause of the greatest morbidity after surgical closure of ASD. The reasons for this difference are not apparent. With surgical closure of ASD, no prosthetic material or mechanical struts are implanted, and the operative risk is extremely low. However, an incidence of pericardial effusion that requires treatment in approximately 5% to 20% of patients is noted in ASD closure, and the reason for this high occurrence of pericardial effusion is unknown. Transcatheter devices have resulted in high closure rates with a low incidence of recurrence or infection and have become the procedure of choice for small to moderate-sized secundum ASDs.

The larger Amplatzer devices that have been used for larger-sized secundum atrial septal defects are quite large in the atrium. Some of the transcatheter devices have resulted in significant complications. There have been several reports of infection, thromboembolism, and erosion of the devices into the base of the sinus of the aortic valve or the atrial wall, resulting in rupture of sinus of Valsalva into the atrium. It appears that the incidence of these complications is low; however, the larger devices seem to occupy much more space in the heart, and therefore longer follow-up will definitely need to be obtained before these devices are proved to have similar safety and efficacy as the smaller devices for small to moderate-sized atrial septal defects.

Although results with dilation of the native infant coarctation are suboptimal and certainly no better than for surgical therapy, the long-term results remain to be determined. Balloon dilation of recurrent coarctation has become accepted standard therapy in most patients. The incidence of aneurysm formation and other long-term complications requires longer follow-up. It is certainly possible to argue that in an older patient with a normal-sized aorta, primary stenting of coarctation may result in relief of gradient and improvement in hypertension with no greater morbidity or mortality than surgical intervention. Stenting in younger patients, however, may be suboptimal because growth will frequently require additional dilation of the stent to prevent recurrent stenosis.

Catheter intervention for residual defects after surgical intervention is increasingly utilized. Patients with multiple ventricular septal defects are particularly amenable to this type of approach. Although most apical defects and perimembranous defects can be directly addressed with standard surgical

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EDITOR'S COMMENTS (*continued*)

techniques, muscular ventricular septal defects (especially anterior defects) are often obscured from right ventricular or transatrial approaches. In patients with multiple defects, a primary surgical approach to the defects in the apical and perimembranous regions and then subsequent catheter closure if necessary of residual muscular defects is reasonable. Because most multiple ventricular septal defects are generally amenable to surgical and transcatheter closure, we have taken the approach that patients with multiple defects in general should not undergo pulmonary artery banding. Avoidance of banding may prevent hypertrophy of ventricular muscle, which can obscure margins of muscular defects and make later repair more difficult. Cooperation and communication between the catheterizing interventional cardiologists and surgeons can result in optimal outcomes in these patients.

An area of increasing interest is that of "hybrid" treatment options for various forms of congenital heart disease. In the hybrid approaches, transcatheter and surgical procedures are done simultaneously. There has been an increasing experience with the perventricular approach to muscular VSD closure in the operating room, with excellent short-term results. The ability to access the VSD directly through the ventricular wall results in a very straight catheter course and ease of implantation of these devices. In addition, there has been increasing interest in the use of hybrid therapies for first-stage palliation in hypoplastic left heart syndrome. In these approaches, the ductus arteriosus is stented over its entire length and bands are surgically placed on the pulmonary arteries.

As noted by the authors of this chapter, the use of hybrid procedures for hypoplastic left heart syndrome has been variable. There are anatomic contraindications to the use of this therapy and unfortunately a significant number of patients with aortic atresia will have a substrate that is predisposed to retrograde coarctation of the aorta. In most centers, hybrid therapy is used selectively for patients who are considered at particularly high risk for surgical Stage I Norwood operation. As in many of the interventional techniques where stents are implanted in vessels, the stents are difficult to remove at the time of surgical intervention and often residual stent material must be left in place during surgical reconstruction, making reoperation somewhat more complex. Thus, in any given patient it is particularly important that the long-term needs for staged surgical reconstructions be recognized and the plans for surgical and interventional procedures coordinated between the surgeons and interventionalists to insure that the long-term treatment plan is optimal.

The availability of stents for maintaining ductal patency has also led to greater interventional use as an alternative to aortopulmonary shunts in the newborn. Patients with pulmonary atresia and intact ventricular septum may undergo a radiofrequency perforation of the valve plate with balloon dilation and placement of a ductal stent to avoid the need for surgery. Nevertheless, the long-term outcomes for patients with these strategies have not been determined, and in most cases the ductal stenting needs to be performed in a somewhat restrictive ductus to allow the stent to anchor adequately. There have been cases of stent migration and stenosis, and the hemodynamic characteristics of a central stent may be suboptimal in terms of protection from pulmonary vascular resistance over the short term. Nevertheless, increased experience is being gained with the use of these interventional techniques.

There has not been a direct comparison of surgical versus catheter approaches for pulmonary atresia with intact ventricular septum with long-term outcomes. However, the intermediate stage results of catheter-based therapy seem to be quite good.

Although they are not yet readily available in the United States, the Amplatzer perimembranous VSD occluder devices are being used more extensively in Europe and Asia. Relatively large series of VSD closures with these devices are being reported and new devices are being studied. Although the closure rates appear to be quite good, there is an incidence of late development of complete heart block that needs to be addressed with additional long-term follow up and device modifications. Nevertheless, many VSDs could have device closure with appropriate patient selection.

As transcatheter interventions become more sophisticated, the communication between interventional cardiologists and surgeons becomes even more critical. In many cases, transcatheter interventions can be undertaken in conjunction with surgical therapy to occlude shunts that may be difficult to access intraoperatively, to occlude additional sources of pulmonary blood flow in patients who are undergoing the Fontan procedure, or to occlude decompressing veins after staged palliation for single ventricle. In some cases, intraoperative transcatheter interventions may be useful for stenting pulmonary arteries (e.g., in patients with compression of the left pulmonary artery after staged reconstruction for Hypoplastic Left Heart Syndrome), which can significantly improve flow to the lungs after the bidirectional Glenn or hemi-Fontan procedure or completion of Fontan procedures. In many cases, such as patients with discontinuous pulmonary arteries and a ductal source of pulmonary blood flow to one lung, early, complete operative repair can be undertaken but with a very high risk of stenosis of the pulmonary arteries because of involution of ductal tissue after neonatal life. These patients should be referred for transcatheter dilation early and scheduling of these catheter interventional procedures should be considered part of the operative intervention.

It should be noted that diagnostic catheterization has particular value in patients with residual lesions after surgical intervention because many of these lesions cannot be directly identified by echocardiography. Postoperatively, echocardiographic windows may be inadequate, and therefore catheterization may be necessary to identify the exact anatomy of the residual lesion.

In our center, as in many others, patients who are not following a predicted postoperative course are referred for diagnostic catheterization if the echocardiographic information does not fit clearly with the patient's clinical progression. We will very aggressively take patients for diagnostic and interventional catheter procedures postoperatively to eliminate any potential residual defects that could influence the patient's

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EDITOR'S COMMENTS (*continued*)

hemodynamic course. It is not uncommon for residual VSDs or other significant shunts to be underestimated by postoperative echocardiography, and the risk of catheterization is low enough that it appears optimal to actually measure the hemodynamic situation to obtain the best overall outcome.

One of the areas of most active interest in interventional cardiology is the use of stent mounted valves. As mentioned by the authors of this chapter, this technology is rapidly increasing, and catheter-based valve implantation is being increasingly used in adult populations. The relative benefit of catheter-based valve implantation in children remains to be determined. The durability of these valves is not yet known since most have been placed in older patients who have contraindications to surgical intervention. It is well known that tissue valves deteriorate rapidly in very young patients and therefore it is difficult to know how useful this technology will be as primary therapy. However, implantation of catheter-based valves in the pulmonary outflow tract in conduits has clearly been effective, and on the right side of the heart these valves seem to last a reasonable period of time which may be comparable to homograft or other tissue valves. Nevertheless, it is inevitable that these valves will fail and require additional intervention. Placement of stent-mounted valves in the aortic position in children, while certainly feasible, is probably not the optimal therapy due to the poor durability of tissue valves in children at the present time. Nevertheless, there will certainly be instances, with careful patient selection where these technologies will be extended into the pediatric age ranges.

As noted in the previous chapter, MRI has supplanted diagnostic catheterization and to some extent echocardiography for many congenital heart lesions and much hemodynamic information can be obtained with MRI. Thus, the incidence of hemodynamic cardiac catheterization is significantly decreasing and interventional catheterization procedures are becoming increasingly prominent worldwide. Nevertheless, there are certain circumstances where hemodynamic catheterization is optimal, such as to evaluate postoperative repairs that are not responding as expected and in patients where filling pressures and vascular resistances need to be calculated. In addition, if the anatomic and hemodynamic information can be obtained at catheterization, the use of diagnostic catheterization permits the opportunity for interventional treatment of identified lesions without requiring an additional procedure (which would be true if MRI were used at the primary diagnostic modality). Thus, selection of the optimal procedure in each individual patient becomes an important consideration.

Finally, as emphasized by the authors, surgical outcomes in complex congenital heart disease with often multiple operative interventions are optimal when both surgeons and cardiologist construct treatment strategies in a collaborative and prospective manner.

TLS

75 Palliative Operations for Congenital Heart Disease

Hyde M. Russell and Carl L. Backer

With continuing advances over the years in neonatal cardiopulmonary bypass (CPB) and surgical techniques, the chapter on palliative procedures in all textbooks on congenital heart surgery has grown shorter and shorter. There seem to be progressively fewer indications for palliative operations, although there is a select, small group of patients for whom (at least with our current understanding of cardiovascular physiology) palliation apparently always will be required. Countering this trend, there has been a recent resurgence in the use of certain palliative techniques performed in conjunction with transcatheter procedures: the "hybrid" approach. The history of cardiac surgery has been one of an evolution from operations that were solely palliative to ones in which there is complete correction in the neonatal period with one procedure. Recognition of the complications associated with palliative operations and the realization that neonates and infants do not necessarily fare worse than older children during open-heart surgery have provided the impetus for primary corrective surgery at an earlier age. In this process, many previously commonly performed palliative procedures are now considered completely obsolete and are no longer indicated. It is important, though, for the practicing congenital heart surgeon to understand these palliative procedures because there are many patients who have had these operations in the distant past who require later surgical intervention. The two primary palliative procedures still frequently used are the aortopulmonary shunts (Table 75.1) for patients with diminished pulmonary blood flow and cyanosis and pulmonary artery banding for certain patients with excessive pulmonary blood flow and congestive heart failure. The pulmonary artery band has recently undergone a rebirth in its use as part of the hybrid approach to the treatment of infants with hypoplastic left heart syndrome. The importance of the initial palliative procedure cannot be overemphasized. A poorly performed aortopulmonary shunt that destroys a pulmonary artery may prohibit the child from having a completion Fontan correction. Hence, although these are older and seemingly less important procedures, they must be performed well to ensure a smooth eventual corrective procedure.

AORTOPULMONARY SHUNTS

Classic Blalock–Taussig Shunt

The Blalock–Taussig shunt (Table 75.2) was the first aortopulmonary shunt; it was first performed in 1944 by Alfred Blalock of the Johns Hopkins University Medical Center. The classic Blalock–Taussig shunt is a direct end-to-side anastomosis of the transected subclavian artery to the pulmonary artery. Legend has it that Helen Taussig, the pediatric cardiologist at Johns Hopkins University, observed that the condition of infants and children with severe forms of tetralogy of Fallot worsened after their patent ductus arteriosus closed. She traveled to Boston and asked Robert Gross if he could create an artificial ductus in these children. He supposedly told her that he was in the business of closing the ductus, not creating new ones. She then met with Blalock, the chairman of the department of surgery at her institution. Blalock had successfully created a left subclavian artery-to-pulmonary artery anastomosis 6 years earlier while developing a canine model of pulmonary hypertension at Vanderbilt University. Taussig suggested to Blalock that he will apply his experimental procedure to the many patients that she saw in her clinic who had cyanosis from insufficient pulmonary blood flow. The first operation was performed on November 29, 1944, on a 15-month-old girl with the diagnosis of tetralogy of Fallot and severe pulmonary stenosis. After that first successful case, hundreds of cyanotic children went to Baltimore for "the operation." Within the next 2 years, more than 500 patients had undergone the Blalock–Taussig shunt. During this time, there was an early mortality of 16%, and 6% of the patients were considered inoperable. This procedure was very commonly used until the introduction of the modified Blalock–Taussig shunt, which uses an interposition graft of Gore-Tex, avoiding sacrifice of the subclavian artery. The Hopkins group recently reported a six decade experience with 2,000 Blalock–Taussig shunts.

The anastomosis for the classic Blalock–Taussig shunt is classically constructed on the side opposite the aortic arch. When there is a left aortic arch, there is typically a right innominate artery, and using the subclavian artery on this side provides a gentle curve down to the pulmonary artery. With a right aortic arch and mirror-image branching, the left innominate artery provides a similar gentle curve to the pulmonary artery. In contrast, the other subclavian artery for each arch would require an angulation of the artery of nearly 180 degrees to effect the anastomosis. Figure 75.1 illustrates the anatomy for exposure of the subclavian and pulmonary arteries through a right thoracotomy (see inset). The branches of the right subclavian artery are ligated and divided along with the distal subclavian artery, and the subclavian artery is pulled through the loop formed by the right recurrent laryngeal nerve. The carotid artery is freed to provide more mobility. The end of the subclavian artery is spatulated, so the anastomosis is 1.5 to 2.0 times larger than the subclavian artery per se. The anastomosis is constructed to the proximal pulmonary artery, with care taken to avoid the complication of performing the anastomosis to the right upper lobe branch. The subclavian artery will then be in a groove just posterior to the superior vena cava (SVC) and the phrenic nerve (Fig. 75.2).

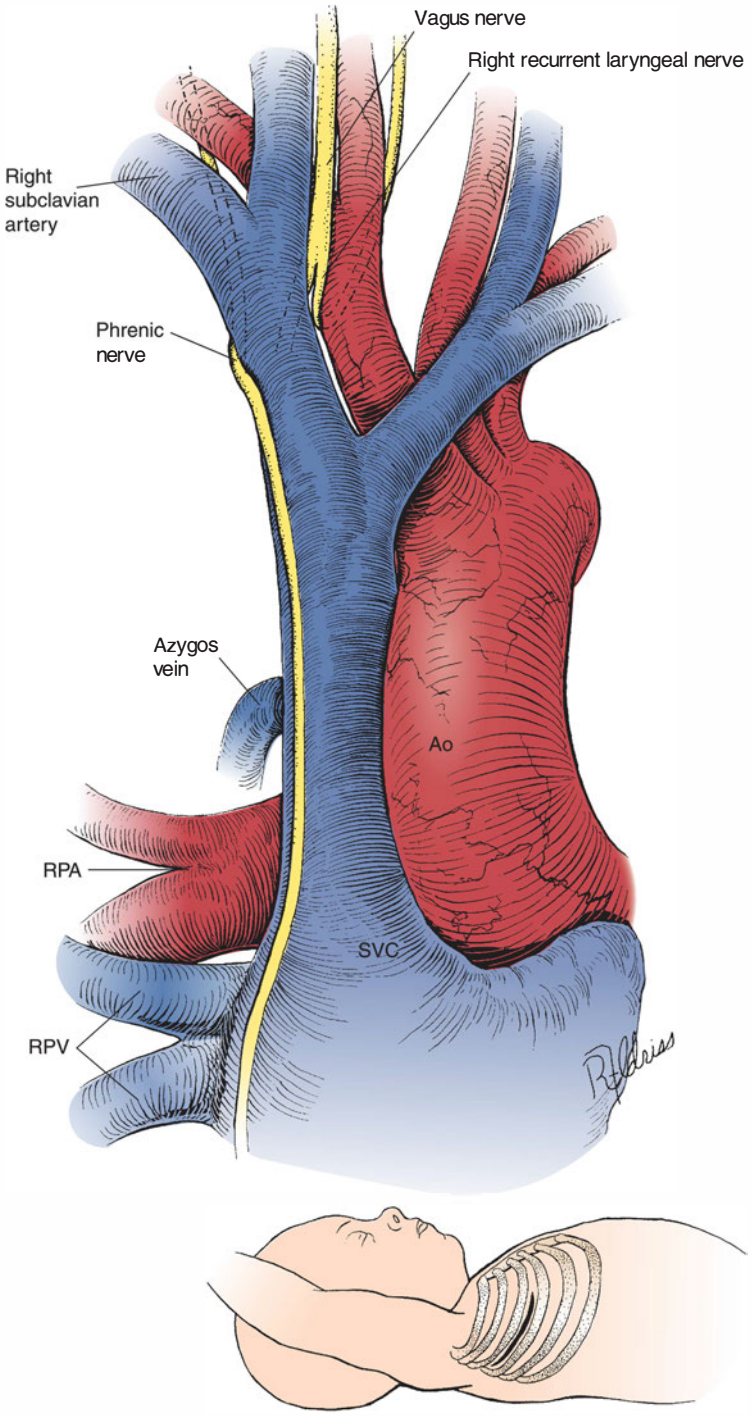
Table 75.1 Aortopulmonary Shunts

Blalock–Taussig shunt
Modified Blalock–Taussig shunt
Waterston/Cooley shunt
Potts shunt

Table 75.2 Blalock–Taussig Shunt

Date/surgeon	1944/Alfred Blalock
Technique	Direct anastomosis, subclavian artery to pulmonary artery
Most common indications	Tetralogy of Fallot
	Pulmonary atresia
	Tricuspid atresia (right-sided obstructive lesions)
Advantages	Precise amount of pulmonary blood flow
	Grows with the patient
Disadvantage	Sacrifices subclavian artery

The classic Blalock–Taussig shunt does not require prosthetic material and provides a precise amount of pulmonary blood flow limited by the orifice of the subclavian artery. In addition, the shunt grows with the patient, providing more pulmonary blood flow as the child grows. However, the Blalock–Taussig shunt sacrifices the subclavian artery, which in a small number of cases can result in hand or arm ischemia. In addition, the affected arm is usually shorter than the contralateral arm, is always somewhat cool to the touch, and will not have a palpable pulse. Finally, even with mobilization of the carotid artery and division of the inferior pulmonary ligament, the subclavian artery may still be so short as to cause the pulmonary artery to be "pulled" up and kinked. Takedown of the classic Blalock–Taussig shunt at the time of complete correction through a median sternotomy involves dissection posterior to the SVC. The artery can then be encircled and double ligation performed (Fig. 75.3).



Modified Blalock–Taussig Shunt

The use of a polytetrafluoroethylene (PTFE) tube for an aortopulmonary shunt was first reported by Gazzaniga and associates in 1976. Three infants with pulmonary atresia underwent aorta-to-pulmonary artery shunt using an interposed 4-mm PTFE tube. DeLeval coined the term modified Blalock–Taussig shunt when he reported on 99 patients operated on between 1975 and 1979 having a prosthesis of Dacron (13) or PTFE (86) interposed between the subclavian and pulmonary arteries (Table 75.3). Shunt failure rate was 6%, and mortality was 8%. The advantages of the modified Blalock–Taussig shunt,

Fig. 75.1. The anatomy as seen through the right thorax in preparation for a classic or modified Blalock–Taussig shunt. (Inset) Right thoracotomy. SVC Ao, aorta; RPA, right pulmonary artery; RPV, right pulmonary vein; SVC, superior vena cava.

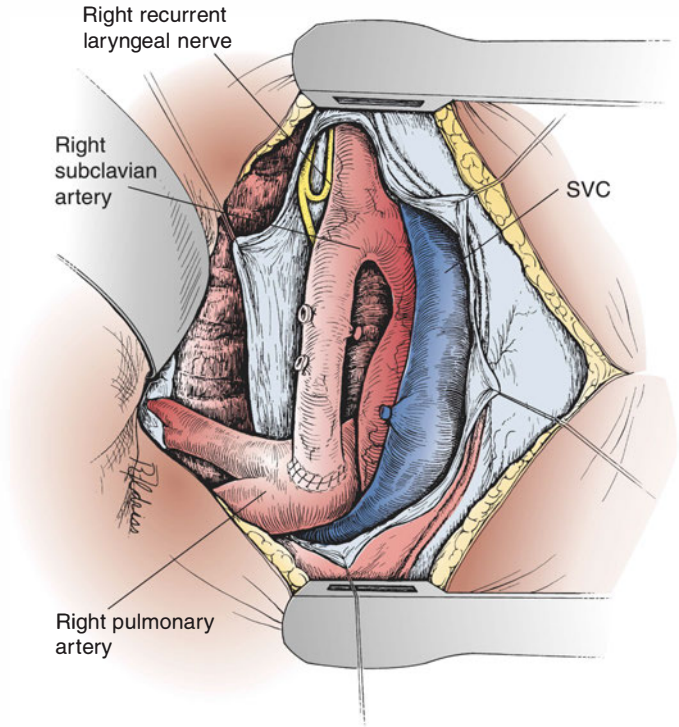


Fig. 75.2. The classic Blalock–Taussig shunt. SVC, superior vena cava.

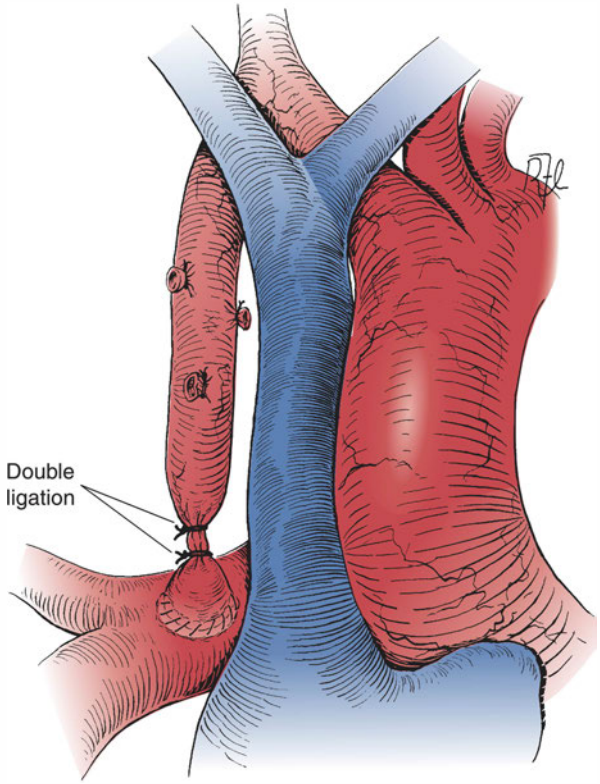


Fig. 75.3. Takedown of a classic Blalock–Taussig shunt with double ligation.

which has now become the shunt of choice at most congenital heart surgery centers, include (1) preservation of the circulation to the affected arm, (2) regulation of the shunt flow by the size of the systemic (subclavian or innominate) artery, (3) high early patency rate with minimal tissue ingrowth of even a small-diameter expanded PTFE arterial prosthesis (Gore-Tex; W.L. Gore & Associates, Inc., Flagstaff, AZ), and (4) guarantee of adequate shunt length. One disadvantage of the modified Blalock–Taussig shunt is the occasional excessive leaking of serous fluid through the interstices of the fabric of the PTFE. This may result in excessive and prolonged chest tube drainage, localized seroma formation around the graft, or both. This complication occurs in 3% to 5% of patients.

The modified Blalock–Taussig shunt can be performed through a right or left thoracotomy or a median sternotomy. In the last 10 years, we have exclusively used a median sternotomy approach. This facilitates the use of CPB support if the child has significant oxygen desaturation during the procedure. This, of course, is much easier with a median sternotomy approach. CPB should always be readily available for such an instance when a shunt is performed. There are several other advantages to the median sternotomy approach. A patent ductus arteriosus can be ligated as soon as the shunt is opened or when the patient is placed on CPB. The lungs are not compressed by the exposure through a thoracotomy (which can affect the O_2 saturations). If a thoracotomy approach is used, the side selected depends on the subclavian and pulmonary artery anatomy, the presence and location of a ductus arteriosus, and the great-vessel relationship. The disadvantages (relatively minor) of the median sternotomy are the adhesions with reoperation. With a thoracotomy approach the latissimus dorsi muscle is divided, the serratus anterior is mobilized and spared. The thorax is entered through the fourth interspace. The lung is retracted anteriorly and inferiorly (Fig. 75.4). The mediastinal pleura is opened posterior to the SVC and phrenic nerve. The azygos vein is doubly ligated and divided. The subclavian artery is encircled with a vessel loop, with care being taken to avoid the right recurrent laryngeal nerve, which passes around the distal innominate artery at the takeoff of the subclavian and common carotid arteries. The right pulmonary artery is dissected free, with care being taken to identify the right upper lobe branch and main pulmonary artery continuation to the right lower and middle lobes. The right upper lobe and the distal right pulmonary artery branches are encircled with vessel loops, which can then be occluded with small Rommel tourniquets. Alternatively, a small vascular clamp can be placed on the pulmonary artery. The patient is administered 1 mg/kg of heparin intravenously.

The size of the PTFE graft selected is based on the size of the patient. We currently

Table 75.3 Modified Blalock–Taussig Shunt

Date/surgeon	1975/Marc DeLeval
Technique	Interposition of polytetrafluoroethylene graft between subclavian artery and pulmonary artery
Most common indications	Right-sided obstructive lesions (tetralogy of Fallot, pulmonary atresia, tricuspid atresia)
Advantages	Flow limited by subclavian orifice
	Does not sacrifice subclavian artery
Disadvantage	Seroma formation

use only the heparin-bonded "stretch" PTFE. We use a 3.0-mm shunt for neonates <2.0 kg, a 3.5-mm shunt for neonates 2.0 to 4.0 kg, a 4.0-mm shunt for infants >4 kg, and only rarely a 5-mm shunt in an infant over 5 kg. Nearly all neonates having a shunt procedure will have a bidirectional cavopulmonary anastomosis or a complete repair by 6 months of age; so long-term patency is not an issue. The PTFE is cut to size before the clamps are placed; the clamps distort the relative distance between the subclavian and pulmonary arteries. The graft is usually cut to give it a gentle curve, which allows for some patient somatic growth. It should not kink or distort the pulmonary artery, which can easily happen if the length is not accurate. The vessel loop is used to pull the subclavian artery, and a segment of the vessel is then occluded with a small Castaneda clamp. The PTFE graft is beveled as illustrated in Fig. 75.4A, and an arteriotomy is created in the inferior aspect of the subclavian artery. The PTFE graft is anastomosed to the opening in the subclavian artery with 7-0 polypropylene suture and a parachute technique. The clamp on the subclavian artery is left in place until the pulmonary artery anastomosis is completed. Repositioning the clamp to the

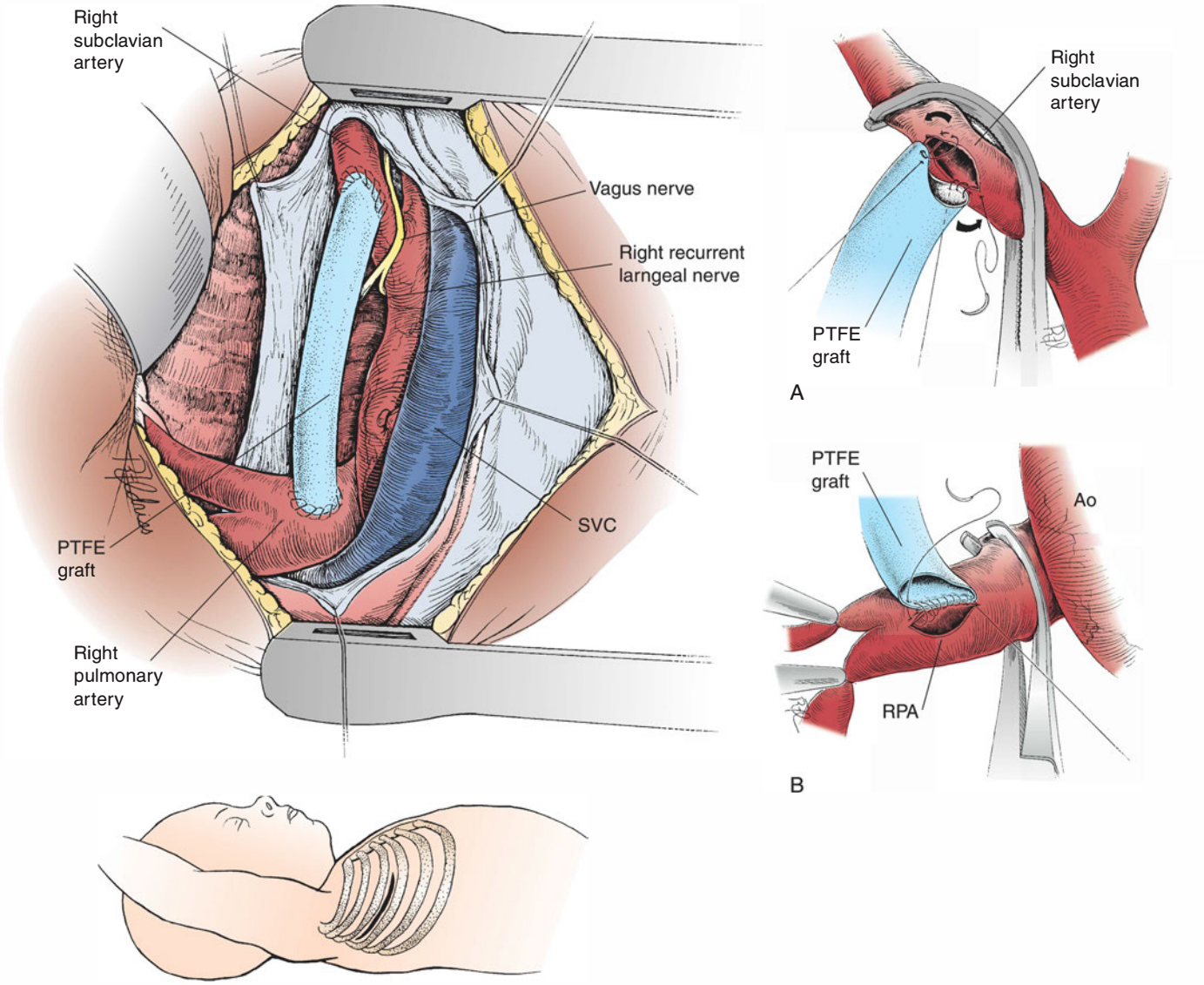


Fig. 75.4. The modified Blalock–Taussig shunt using a polytetrafluoroethylene graft. (Inset) Right thoracotomy. (**A**) Right subclavian artery anastomosis. (**B**) Right pulmonary artery anastomosis. Ao, aorta; RPA, right pulmonary artery; SVC, superior vena cava.

PTFE graft may increase the risk of blood stasis and shunt thrombosis. The proximal pulmonary artery is controlled with another small Castaneda clamp. The Rommel tourniquets are snugged on the right upper and right distal pulmonary arteries. A longitudinal arteriotomy is created in the superior aspect of the right pulmonary artery. With a single 7-0 polypropylene suture, the PTFE graft is anastomosed to the pulmonary artery (Fig. 75.4B). The distal Rommel tourniquets are released first, then the clamps on the pulmonary and the subclavian arteries.

There should be a nearly instantaneous rise of approximately 10% to 15% in the patient's oxygen saturation as monitored by pulse oximetry. A thrill should be palpable in the shunt and in the distal pulmonary artery. It is important to maintain adequate systemic arterial pressure before, during, and after the operation to prevent early shunt thrombosis. This may require the use of inotropic support (dopamine, dobutamine). Bleeding is controlled with absorbable gelatin sponge (Gelfoam) soaked with topical thrombin. The heparin is not routinely reversed with protamine unless there is excessive bleeding from the suture lines. The graft should lie in a groove posterior to the SVC, where it is accessible at the time of takedown for intracardiac repair through a median sternotomy approach. The chest is closed in layers with a single chest tube directed posteriorly and superiorly. We routinely sedate and ventilate the child for the first 24 hours postoperatively.

Williams and colleagues recently reviewed 2,000 shunt procedures performed at the Johns Hopkins hospital since Alfred Blalock's first operation. Of these patients, 70% carried a diagnosis of tetralogy of Fallot as a result of the large number of cases done in the 1940s and 1950s before the advent of complete intracardiac repair. Operative mortality ranged from 15% in the first decade of use (1940s) to 8% to 10% in the modern era. Complications of stroke, bleeding, and infection are all <5% in the current era. Over the six decades, the review encompassed its use decreased, the operative mortality declined, and there was a shift in application to single ventricle patients.

Petrucci reviewed outcomes of all shunt procedures recorded in the Society of Thoracic Surgeons Congenital Heart surgery database during the modern era (2002 to 2009). This study included 1,273 patients. The overall mortality rate was 7.2% and the complication rate (need for mechanical circulatory support or unplanned reoperation) was 13%. Low weight (<3 kg), functionally univentricular hearts, or a diagnosis of pulmonary atresia with intact ventricular septum were all risk factors for death. Notably, one-third of deaths occurred within the first 24 hours of operation.

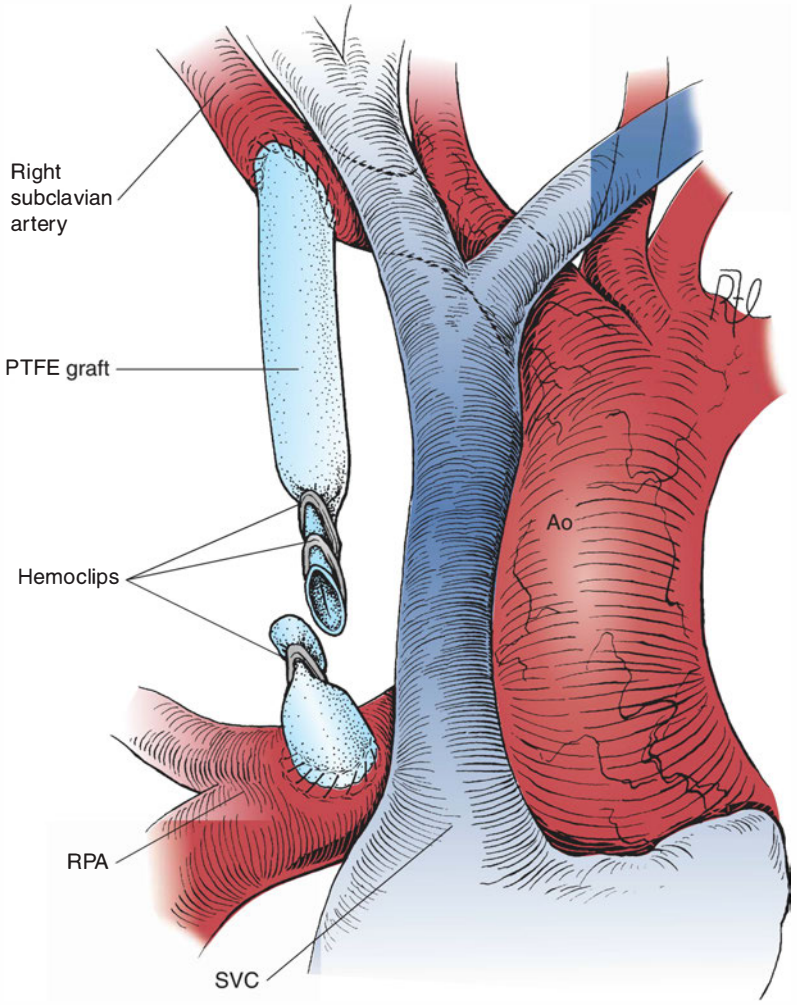


Fig. 75.5. Takedown of a modified Blalock–Taussig shunt with the hemoclip and division technique. Ao, aorta; PTFE, polytetrafluoroethylene; RPA, right pulmonary artery; SVC, superior vena cava.

The takedown at the time of intracardiac repair through a median sternotomy is quite easily performed, particularly if the shunt was placed on the right side (Fig. 75.5). The shunt can be identified by dissecting the medial aspect of the SVC posteriorly. Locating a shunt on the left side is more difficult, but it can be identified either by dissecting along the left pulmonary artery, along the aorta to the shunt, or by entering the pleural space and approaching the shunt laterally. The dissection will first encounter a thick fibrous "peel," which typically forms around the PTFE graft. After the plane between the "peel" and the PTFE graft is entered, the dissection proceeds relatively quickly and easily and enough graft length can be achieved for double hemoclip application proximally and single hemoclip distally. The graft should be divided between the hemoclips to prevent distortion of either the subclavian artery or the pulmonary artery as the patient grows and the distance between these two vessels naturally increases. No attempt is made to remove the proximal portion of the graft. In some patients, the distal graft will require removal to facilitate performing a bidirectional Glenn anastomosis. However, if the operation involves repair at the site of the main pulmonary artery, the distal graft can be left in place and typically does not cause a residual peripheral pulmonary artery stenosis.

As mentioned earlier, our preference is to perform the modified Blalock–Taussig shunt through a median sternotomy approach. This is particularly useful for the child who is having uncontrollable episodes of oxygen desaturation despite sedation, paralysis and full ventilation, and administration of phenylephrine (Neo-Synephrine). CPB with a single atrial cannula and cooling to 32°C will allow the Blalock–Taussig shunt to be performed in a safe and unhurried

fashion, with the heart beating in normal sinus rhythm throughout the procedure. We and others have adopted the median sternotomy as the standard approach for a modified Blalock–Taussig shunt, believing that the safety factors outweigh the adhesions at reoperation.

One other consideration at the time of Blalock–Taussig shunt construction is the question of whether or not to ligate the patent ductus arteriosus. Our practice until recently has been to ligate the patent ductus arteriosus either when CPB is started or when the clamps for the shunt are released (no CPB). A recent review by Zahorec and colleagues noted a higher early hospital mortality (9.7% vs. 0%) when the arterial duct was surgically closed versus being left open ($n = 62$).

One other trend in the management strategy of neonates requiring a source of pulmonary blood flow is the use of transcatheter ductal stenting. McMullan and colleagues recently reported 11 neonates who underwent percutaneous transcatheter placement of an endovascular stent to maintain patency of the arterial duct as an alternative to modified Blalock–Taussig shunt. When compared with 43 patients who had a modified Blalock–Taussig shunt during the same time period, they found that percutaneous stenting had greater freedom from reintervention and fewer procedural complications. These promising results merit consideration as worldwide experience with this new technique expands. Continued work with this alternative is necessary prior to widespread adoption of this new transcatheter approach.

Waterston/Cooley Shunt

In 1962, Waterston (Table 75.4) first reported an aortopulmonary shunt that was an anastomosis between the posterior ascending aorta and the anterior right pulmonary artery. This procedure was performed through a right thoracotomy with the anastomosis posterior to the SVC. Denton Cooley reported the same technical shunt but it was performed anterior to the SVC. After mobilizing the proximal and distal right pulmonary artery, the distal pulmonary artery branches can be controlled with small Rommel tourniquets. Proximally, a Castaneda clamp is used to occlude both a portion of the ascending aorta and the right pulmonary artery. The aorta is rotated slightly anteriorly with a forceps so that a posterior rather than a lateral portion of its wall is exteriorized by the clamp for the anastomosis. As illustrated in Figure 75.6, matching incisions are made in the posterior aorta and anterior pulmonary artery. These incisions are between 3 and 4 mm in length, depending on the size of the patient. An anastomosis is then created with running polypropylene suture as illustrated in Figure 75.7. The tourniquets are released and the clamp is removed, and the oxygen saturation should increase appropriately.

Table 75.4 Waterston/Cooley

Date/surgeon	1962/David Waterston 1966/Denton Cooley
Technique	Waterston: right thoracotomy, anastomosis of ascending aorta to right pulmonary artery posterior to superior vena cava
	Cooley: right thoracotomy, anastomosis anterior to superior vena cava
Most common indications	Right-sided obstructive lesions
Advantages	Technically "easier" than modified Blalock–Taussig shunt
	No prosthetic material
	Preserves subclavian artery
Disadvantages	Right pulmonary artery distortion
	Possibility of excessive or inadequate shunt flow

A major problem with the Waterston shunt is that it is not as controlled a shunt as either the classic or the modified Blalock–Taussig shunts. If the incisions in the aorta and pulmonary artery are too long and the anastomosis is too large, there will be excessive pulmonary blood flow with a risk of pulmonary vascular disease. If the incisions are too short and the opening is too small, there will be inadequate pulmonary blood flow. A second problem with the Waterston anastomosis is that as the patient grows and the aorta rotates, the anastomosis tends to put traction on the right pulmonary artery and actually to kink and distort the right pulmonary artery. This may cause preferential flow to one lung. For this reason, almost all patients who have takedown of a Waterston shunt will require a major reconstruction of the right pulmonary artery.

The Waterston shunt (Fig. 75.8) is taken down at the time of intracardiac repair

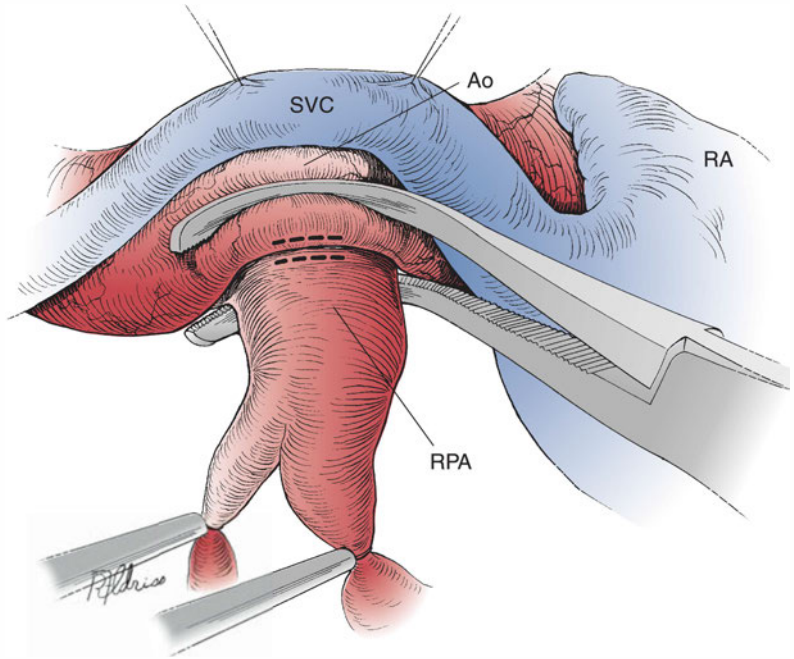


Fig. 75.6. Preparation for a Waterston anastomosis. Ao, aorta; RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.

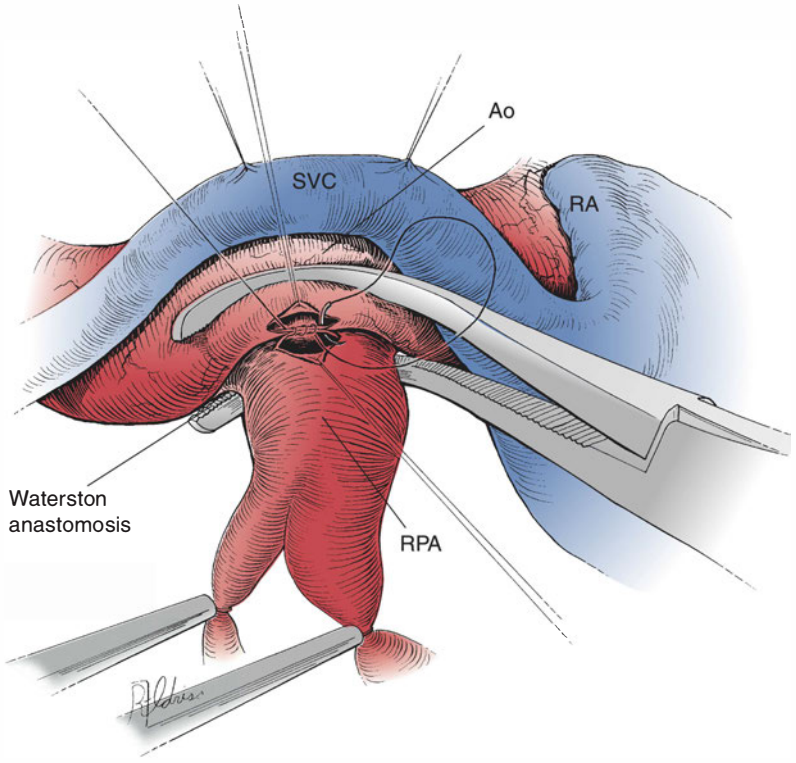


Fig. 75.7. Construction of a Waterston anastomosis. Ao, aorta; RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.

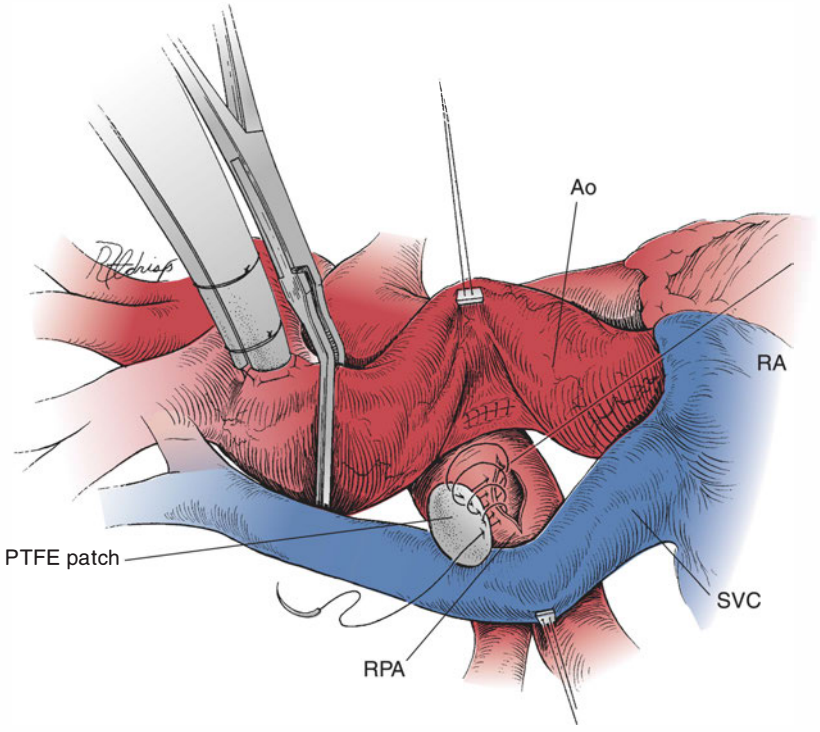


Fig. 75.8. Takedown of a Waterston anastomosis with cardiopulmonary bypass and a polytetrafluoroethylene patch. Ao, aorta; RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.

of the primary cardiac lesion through a median sternotomy. It is important to dissect out the shunt area and pulmonary arteries before CPB to occlude the shunt on initiation of CPB to prevent pulmonary runoff with inadequate systemic perfusion. Alternatively, the right and left pulmonary arteries can be snared or clamped. The repair is done with the patient on CPB with the aorta cross-clamped distal to the shunt. After cardioplegic solution has been administered (occluding the shunt or pulmonary arteries to prevent cardioplegic solution runoff), the aorta is separated from the right pulmonary artery with an incision made along the original anastomotic line. Typically, the aorta in these patients is large, and the opening in the aorta can be closed primarily with running suture. The cross-clamp can then be temporarily removed and a PTFE or pericardial patch used to patch open the area of the right pulmonary artery where focal stenosis is usually created by the shunt. Another approach to the Waterston shunt at the time of takedown (originally described by Cooley) is to open the aorta anteriorly after the cross-clamp has been applied and inspect the opening from within the aorta itself. The aorta can also be completely divided to provide better exposure of the right pulmonary artery. The Waterston shunt has fallen out of favor because of the previously mentioned disadvantages and has been replaced in most centers by the modified Blalock–Taussig shunt. However, there are still a number of patients (now constantly declining) who have had this shunt who will require eventual takedown as described.

Potts Shunt

The Potts shunt is described here for almost solely historical purposes. The Potts shunt (Table 75.5) is an anastomosis between the descending thoracic aorta and the left pulmonary artery performed through a left lateral thoracotomy incision; it was first reported by Willis J. Potts from Children's Memorial Hospital in Chicago. The first operation was performed on September 13, 1946. The child was 21 months of age and weighed 8.3 kg. She had been cyanotic since 3 months after birth and had multiple hypercyanotic spells. She was intensely cyanotic and clubbed. She had eventual intracardiac repair and lived to be 64 years old. The Potts shunt was performed using a special clamp developed by Potts that only partially occluded the descending thoracic aorta. The proximal and distal left pulmonary artery was occluded. The anastomosis was performed between parallel 4-mm incisions made in the descending thoracic aorta and the posterior left branch pulmonary artery. The completed shunt is shown in Figure 75.9.

The Potts anastomosis was widely used at Children's Memorial Hospital in the late 1940s and 1950s. Between 1946 and 1967, 659 such shunts were performed. However,

Table 75.5 Potts Shunt

Date/surgeon	1946/Willis J. Potts
Technique	Anastomosis of left pulmonary artery to descending thoracic aorta
Most common indications	Right-sided obstructive lesions
Advantages	"Easier" anastomosis than classic Blalock–Taussig shunt
	No prosthetic material
	Preserves subclavian artery
Disadvantages	Left pulmonary artery aneurysm formation
	Risk of stroke with shunt takedown
	Excessive pulmonary blood flow, pulmonary hypertension
	Cannot be performed with right aortic arch

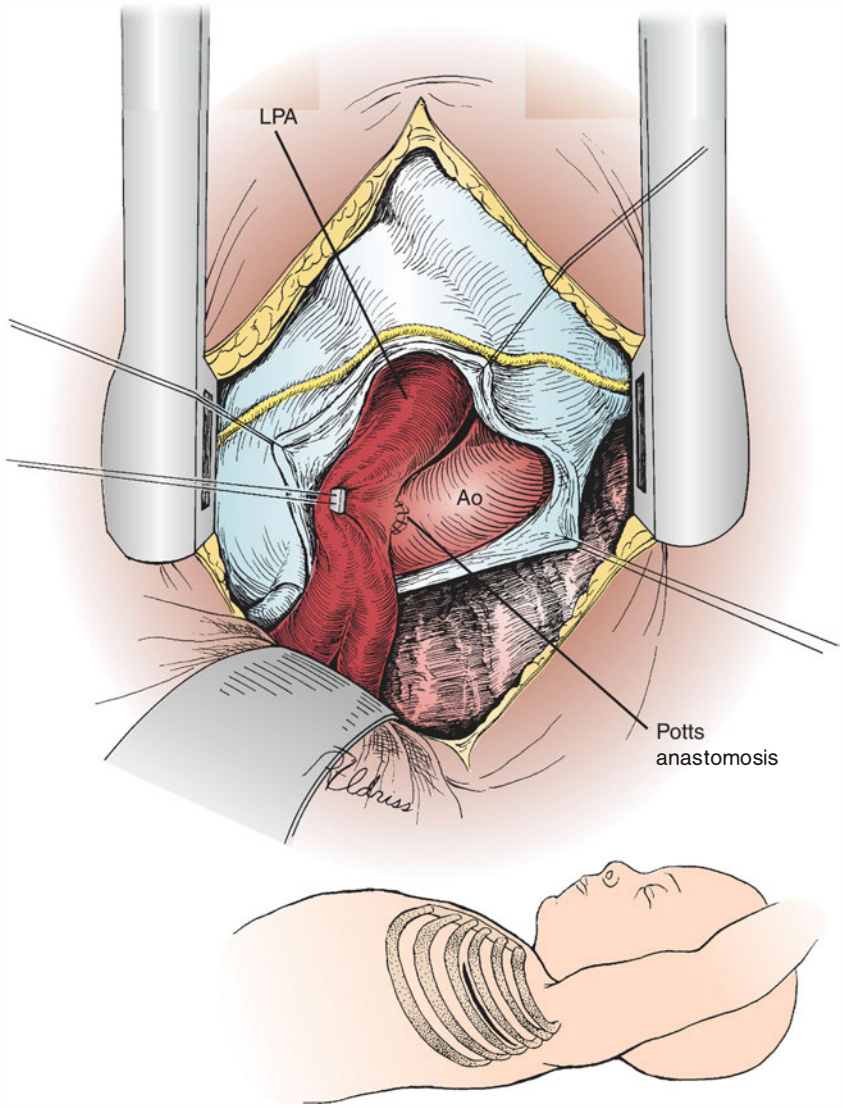


Fig. 75.9. Potts anastomosis. (Inset) Left thoracotomy. Ao, aorta; LPA, left pulmonary artery.

there were several serious complications that developed from the Potts shunt. Many children developed large aneurysms of the left pulmonary artery. Another complication was that the Potts shunt was frequently either too small and the patient remained cyanotic, or it was too big and caused congestive heart failure. The final and most significant problem with the Potts shunt was the difficulty in taking down the shunt at the time of complete correction of the intracardiac lesion. Initial attempts at simply ligating the shunt resulted in uncontrollable hemorrhage in the operating room or in the immediate postoperative period. The preferred technique now is to use deep hypothermia and circulatory arrest, with adequate precautions taken to avoid air embolism to the cerebral circulation with the descending aorta opened. After CPB is initiated through a median sternotomy with aortic or femoral arterial cannulation, the aortopulmonary anastomosis is digitally occluded with a finger from outside the pulmonary artery to limit the left-to-right shunt and improve the efficiency of cooling (Fig. 75.10). The head vessels are encircled with vessel loops. The aorta is cross-clamped, and the heart is arrested with cardioplegic perfusion. The head vessels are snared, and the patient is placed on circulatory arrest. Only then can the left pulmonary artery be safely opened anteriorly (Fig. 75.11). Under circulatory arrest, the communication between the pulmonary artery and the aorta can be visualized and closed with a PTFE patch. The circulation can then be resumed, taking standard precautions to prevent air embolus to the cerebral circulation, venting air out of the patch occluding the Potts shunt before the knot is tied.

Other complications during Potts shunt takedown can occur because of the potential for aneurysm formation of the left pulmonary artery and the difficulty with gaining access to a very posterior anastomosis from the anterior mediastinum. Because of the complications of left pulmonary artery aneurysm, excessive pulmonary blood flow and resultant pulmonary hypotension, and the difficulties in taking down the Potts shunt, this procedure is essentially no longer performed at congenital heart surgery centers. This has been true for many years, and there are very few patients alive with a Potts shunt intact.

PULMONARY ARTERY BANDING

Pulmonary artery banding (Table 75.6) was first suggested by Muller and Dammann in 1952 for children with a large left-to-right shunt or single ventricle. For many years, pulmonary artery banding was the preferred initial palliation for any small child with a large left-to-right shunt and increased pulmonary blood flow, that is, ventricular septal defect, atrioventricular

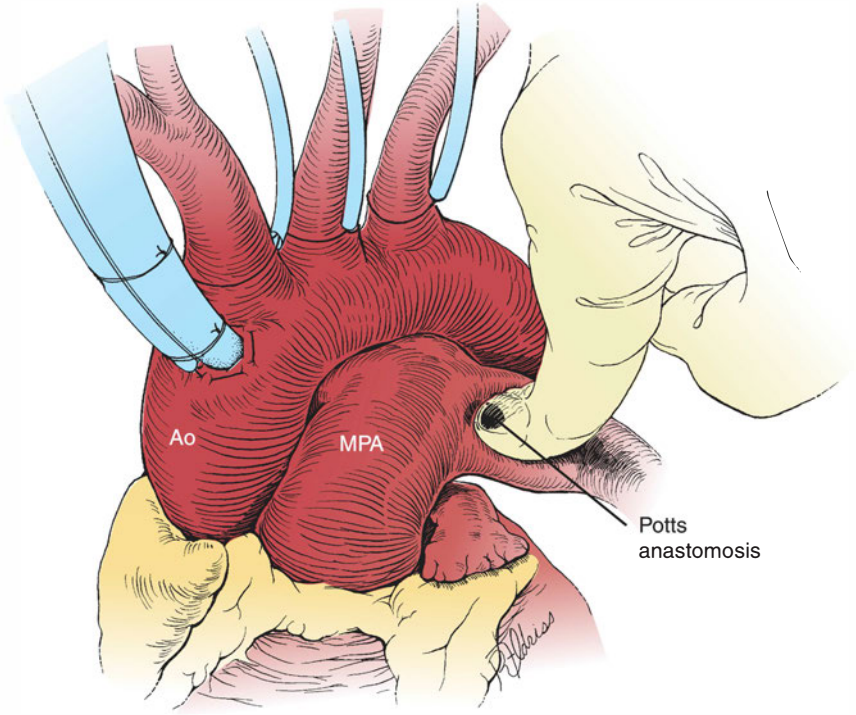


Fig. 75.10. Takedown of a Potts anastomosis with digital occlusion of the Potts anastomosis during cooling and ascending aortic cannulation. Ao, aorta; MPA, main pulmonary artery.

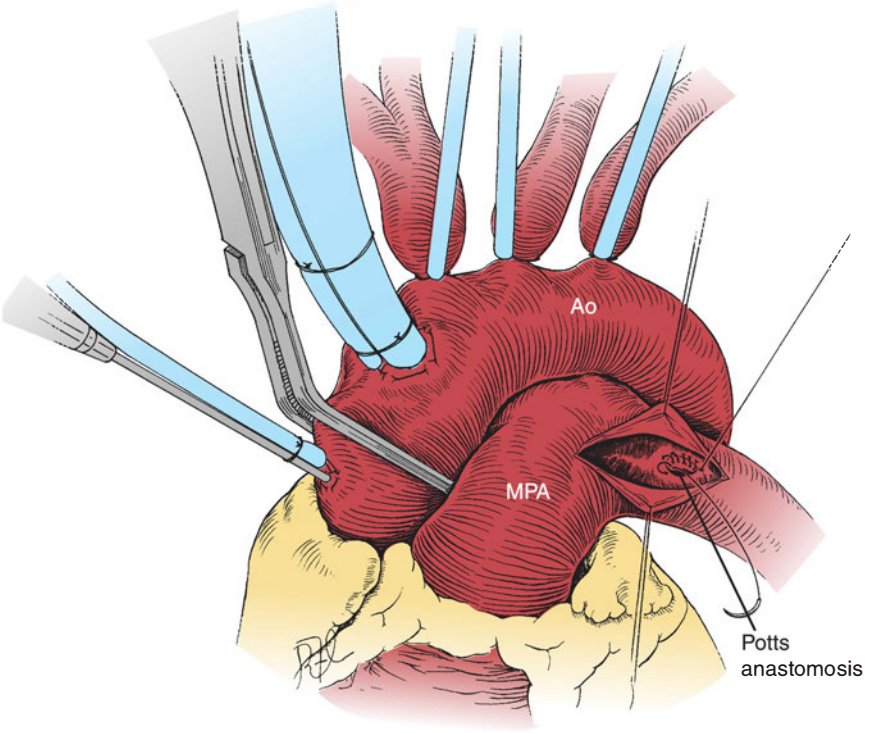


Fig. 75.11. Takedown of a Potts anastomosis with placement of a polytetrafluoroethylene patch in the left pulmonary artery under circulatory arrest. Ao, aorta; MPA, main pulmonary artery.

canal, or common arterial trunk. However, as improvements in neonatal CPB and surgical techniques have taken place, pulmonary artery banding has fallen out of favor for almost all lesions with the exception of a very few precise defects, which include (1) Swiss-cheese muscular ventricular septal defects, (2) multiple ventricular septal defects with coarctation, and (3) single ventricle (i.e., tricuspid atresia type IIc) with increased pulmonary blood flow in anticipation of an eventual Fontan procedure; (4) to prepare the left ventricle of a patient with transposition of the great arteries for the arterial switch procedure either (a) after presentation after 6 to 8 weeks of age or (b) after prior atrial repair; and (5) to prepare the left ventricle of a patient with congenitally corrected transposition of the great arteries in preparation for a "double switch."

There has recently been a resurgence of interest in the use of pulmonary artery banding for infants with hypoplastic left heart syndrome. There are two strategies that have incorporated the use of pulmonary artery banding. One new approach is the "hybrid Norwood" using bilateral pulmonary artery banding and a catheter-delivered stent in the ductus as the first stage of the classic three-stage approach. The other use for pulmonary artery bands is as part of a strategy for patients slated for orthotopic cardiac transplantation. Bilateral pulmonary artery bands are placed with a stent in the ductus to avoid prostaglandin E1 administration while awaiting a donor heart. Another "twist" for pulmonary artery banding that I will review is the modification using an "intraluminal" pulmonary artery band.

Pulmonary artery banding in a child with normally related great vessels can be performed either through a left lateral thoracotomy or a median sternotomy incision. We prefer the median sternotomy approach. Many of the same advantages noted for a modified Blalock–Taussig shunt apply to use of a median sternotomy for pulmonary artery banding. The left thoracotomy is used with simultaneous coarctation repair. The bands we use are either Teflon-impregnated Dacron or a strip of PTFE. The pericardium is opened anterior to the phrenic nerve through a lateral left thoracotomy incision (Fig. 75.12). Stay sutures are placed to hold the pericardium open. The left atrial appendage generally sits just at the site where the band is to be applied and can be retracted with a stay suture. Encircling a dilated, thin-walled pulmonary artery can possibly lead to inadvertent entering of the pulmonary artery and thus needs to be done with great care. The safest way to encircle the pulmonary artery is by the subtraction technique. This is illustrated in Figure 75.12A. The band is first placed around both the aorta and the main pulmonary artery proximally (Fig. 75.12B). This initial maneuver also avoids the complication of encircling only the left pulmonary artery. A plane is then developed between the aorta and the pulmonary artery with a combination of sharp dissection and electrocautery. A right-angled clamp is then passed around the aorta

Table 75.6 Pulmonary Artery Band

Date/surgeon	1952/William Muller
Technique	Band sequentially tightened around pulmonary artery
Most common indications	Excessive pulmonary blood flow not amenable to primary repair (Swiss-cheese ventricular septal defect, tricuspid atresia type IIc)
	Prepares left ventricle for arterial switch
Advantage	Performed without cardiopulmonary bypass
Disadvantages	Causes pulmonary artery distortion; may cause ventricular hypertrophy
	Band can migrate and occlude right pulmonary artery

(not the pulmonary artery) and the free end of the band is grasped (Fig. 75.12C). This is then pulled around the aorta and (by subtraction technique) encircles the pulmonary artery. The band is then sequentially tightened by placing interrupted sutures in the band, as illustrated in Figure 75.13. Alternatively, hemoclips can be serially applied to tighten the band. A catheter is placed in the distal pulmonary artery to monitor the distal pulmonary artery pressure in comparison with the aortic pressure. The length of the band can be predicted initially by the formula developed by George Trusler: 20-mm +

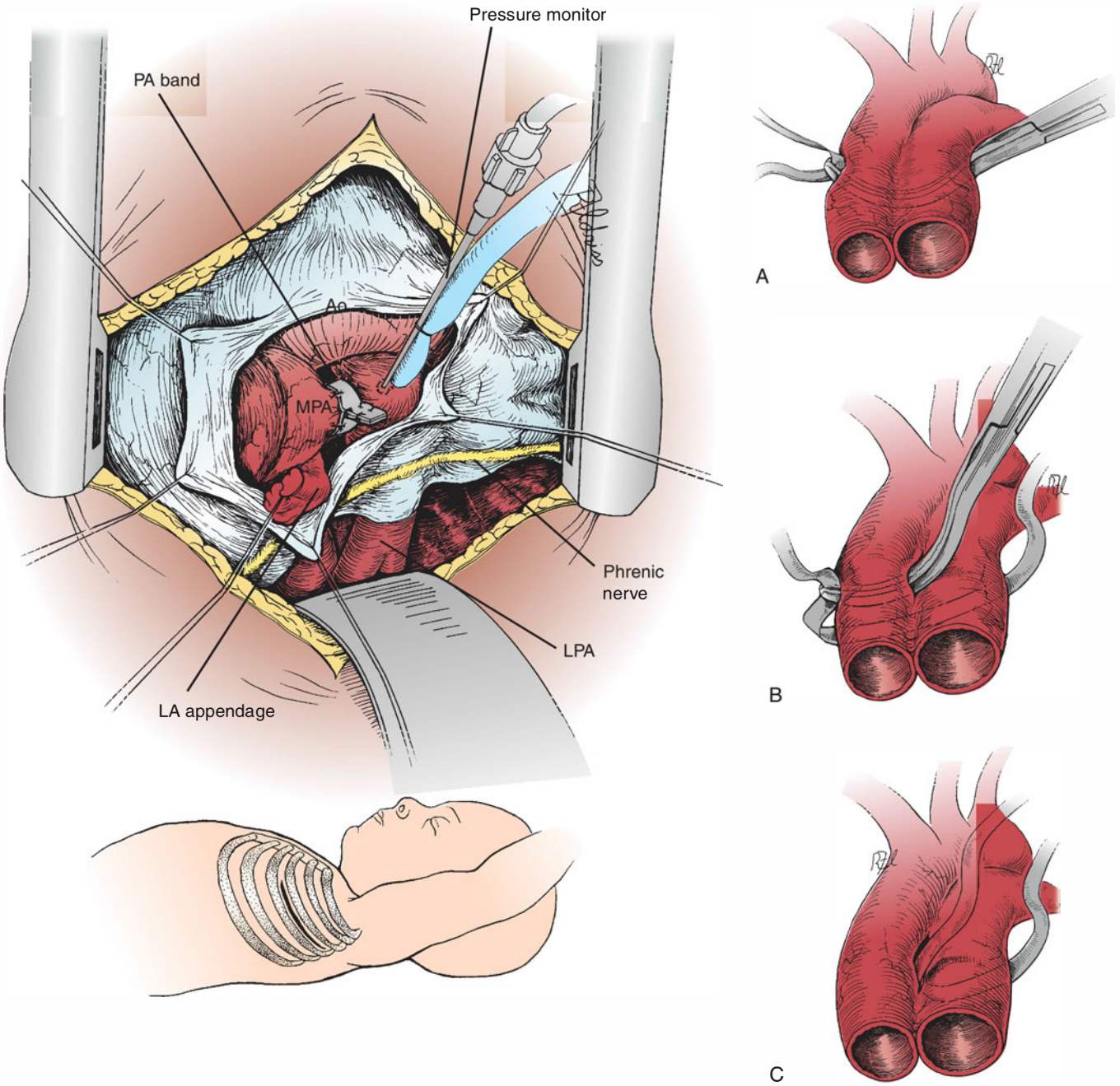


Fig. 75.12. Placement of pulmonary artery band. (Inset) Left thoracotomy. (**A**) Encircling the aortopulmonary trunk. (**B**) Encircling the aorta. (**C**) Final location of the pulmonary artery band. Ao, aorta; LA, left atria; LPA, left pulmonary artery; MPA, main pulmonary artery; PA, pulmonary artery.



Fig. 75.13. Details of sequential tightening of the pulmonary artery band.

1-mm/kg body weight. A narrower band is required for infants with a functionally univentricular heart. We have based final band tightness on the distal pulmonary artery pressure and O_2 saturation.

Placement and tightening of the band in general result in an elevation of the aortic systolic blood pressure by 10 to 20 mmHg. The distal main pulmonary artery systolic pressure should be reduced to <50% of the measured aortic systolic blood pressure for the patient who is to undergo a two-ventricle repair. For a patient who is going to eventually undergo a Fontan procedure, the lowest possible distal main pulmonary artery pressure that can be achieved with acceptable oxygen saturations is desired. Oxygen saturations for a patient who is going to undergo biventricular repair can be left at 95%. For the patient who eventually is going to undergo a Fontan operation, the oxygen saturation should preferably drop to between 80% and 85%. It should be kept in mind that as the child grows, the band will automatically become "tighter" and further reduce the distal pulmonary artery pressure. After the band has been tightened to the desired degree, it is fixed to the proximal pulmonary artery with several interrupted sutures to prevent distal migration of the band and encroachment on the right pulmonary artery. A possible complication of band placement is the aforementioned encroachment, which tends to pinch off the right pulmonary artery while allowing excessive blood flow to the left pulmonary artery. This results in severe proximal right pulmonary artery stenosis and left pulmonary artery hypertension. After the band has been secured in place, the pericardium is irrigated with saline, so there is less chance of intrapericardial adhesions at the time of intracardiac repair. The pericardium is approximated with several interrupted polypropylene sutures, with care being taken to avoid injury to the phrenic nerve. The chest is closed in the usual fashion with a single pleural drainage tube.

A modification that may be useful in complex patients is the adjustable pulmonary artery band. There are many different types of adjustable pulmonary artery bands reported. Some are constructed with standard suture materials, others have reported mechanical devices capable of precise transcutaneous calibration.

Pulmonary artery band takedown is performed at the time of intracardiac repair through a median sternotomy. The patient is placed on CPB. Generally, the intracardiac repair is performed first, and the pulmonary artery reconstruction can be done while the patient is being warmed. All portions of the band should be removed because even a small portion of Dacron band left posteriorly can create scarring, which can cause late pulmonary artery stenosis. Removing the band completely, however, is not always adequate for providing pulmonary blood flow without pulmonary stenosis because the pulmonary artery wall does not necessarily rebound open. The area of banding usually must be either patched anteriorly or excised. The patch technique is illustrated in Figure 75.14. A patch of either pericardium or PTFE is used to provide an opening for the pulmonary blood to flow over the posterior scarred area and into the right and left pulmonary arteries without hemodynamic obstruction to flow. This sometimes requires the use of a "pantaloon"

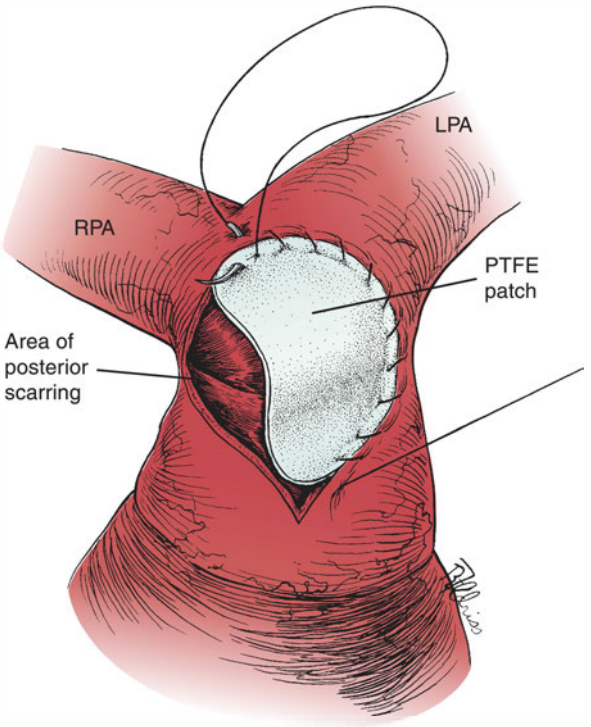


Fig. 75.14. Takedown of a pulmonary artery band with anterior polytetrafluoroethylene patch placement. LPA, left pulmonary artery; RPA, right pulmonary artery.

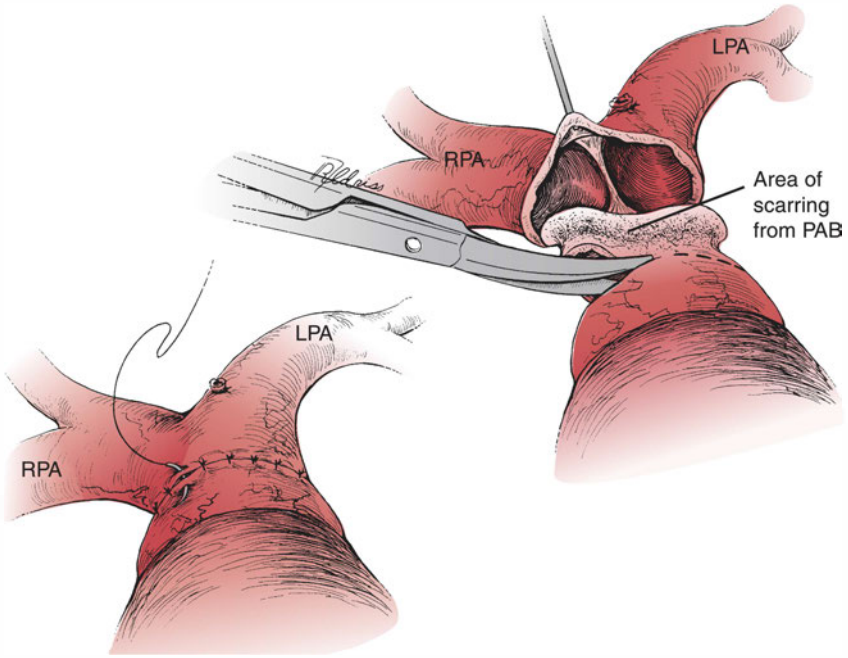


Fig. 75.15. Takedown of a pulmonary artery band with pulmonary artery transection, removal of the band site, and end-to-end reconstruction with interrupted absorbable suture. LPA, left pulmonary artery; PAB, pulmonary artery band; RPA, right pulmonary artery.

patch with extension into the right and left anterior sinuses of Valsalva. The other technique of pulmonary artery band takedown is illustrated in Figure 75.15. This involves resecting the area where the band was positioned and then doing an end-to-end anastomosis between the two remaining pulmonary arterial segments with interrupted, absorbable, fine monofilament suture. The distal right and left pulmonary arteries must be completely mobilized and the ligamentum arteriosum ligated and divided to provide a tension-free anastomosis. Although most surgeons have used a patch anteriorly, this usually still results in a mild main pulmonary artery stenosis and residual murmur. The use of transection of the site of the band and direct end-to-end anastomosis in most cases results in no gradient and no residual murmur. If the band is only in place for a few weeks, pulmonary artery reconstruction may not be necessary.

Another method of pulmonary artery band placement is the "intraluminal" technique (Fig. 75.16). This technique is used only in patients who require CPB for other, simultaneous procedures. The technique utilizes a Gore-Tex patch with a calibrated precut hole in the center that is sutured as a patch in the main pulmonary artery. It results in a consistent and significant reduction in pulmonary artery pressure and flow. It essentially eliminates the problem of band "slipping" with resultant pinching of the right pulmonary artery. One of the advantages of this "band" is that it can be dilated with transcatheter techniques if the patient should become progressively cyanotic with growth. Pulmonary artery band takedown is performed by incising the pulmonary artery and resecting the Gore-Tex patch and then performing an end-to-end anastomosis. Many of the patients, however, who have the intraluminal band go on to a bidirectional Glenn (Fontan strategy), in which case the pulmonary artery is transected at the site of the patch.

MISCELLANEOUS PALLIATIVE PROCEDURES

Miscellaneous palliative procedures include atrial septectomy (Table 75.7), either the surgical Blalock–Hanlon atrial septectomy or the transcatheter Rashkind balloon septostomy, and palliative Glenn and Mustard operations. The Blalock–Hanlon atrial septectomy was first performed in 1950 and was done through a right thoracotomy. A portion of the right and left atria is occluded with a single clamp. The atria are then opened within the confines dictated by the clamp with two parallel incisions, one on either side of the septum, and a portion of intra-atrial septum is grasped, pulled up, and excised from within the clamp. The clamp is then repositioned so that the atrial septum falls back into the atrial cavity and the clamp is only holding the cut edges of the atrium, and the atrial suture line is closed. This procedure is generally no longer performed, and most patients now who require an atrial septal defect undergo Rashkind balloon septostomy. The Rashkind septostomy is typically performed in an infant with transposition of the great arteries and intact ventricular septum. A balloon-tipped catheter is passed up the femoral vein across the patent foramen ovale and into the left atrium. The balloon is initially inflated in the left atrium and then rapidly pulled across the septum into the right atrium, tearing the atrial septum. For patients with a thick atrial septum that is recalcitrant to Rashkind balloon septostomy, open atrial septectomy with CPB is the safest procedure. The atrial septectomy can be performed either during a short period of aortic cross-clamping or with induced electrical fibrillation of the heart. The entire atrial septum within the fossa ovalis can be excised, with the surgeon taking care to avoid injury to the atrioventricular node or cutting outside the heart. Rashkind balloon septostomy in the cardiac catheterization laboratory now is the preferred approach for most cases.

Some patients with complex cyanotic congenital heart disease and suboptimal anatomy for complete correction undergo a Glenn operation or a Mustard procedure for palliation of their cyanotic heart disease. These patients will have an elevation in their oxygen saturations after these palliative procedures, but are not completely corrected.

CONCLUSION

In summary, the primary palliative operations for congenital heart disease are aortopulmonary shunts and the pulmonary artery band. We prefer a median sternotomy approach for both techniques. The aortopulmonary shunt of choice is the modified Blalock–Taussig shunt. Transcatheter ductal stenting may decrease the number of patients having a modified Blalock–Taussig shunt. Pulmonary artery banding is selected for very few patients now, but is still indicated for patients with Swiss-cheese muscular ventricular septal defects and infants with single ventricle and increased pulmonary blood flow, who are candidates for the Fontan procedure. There has been a

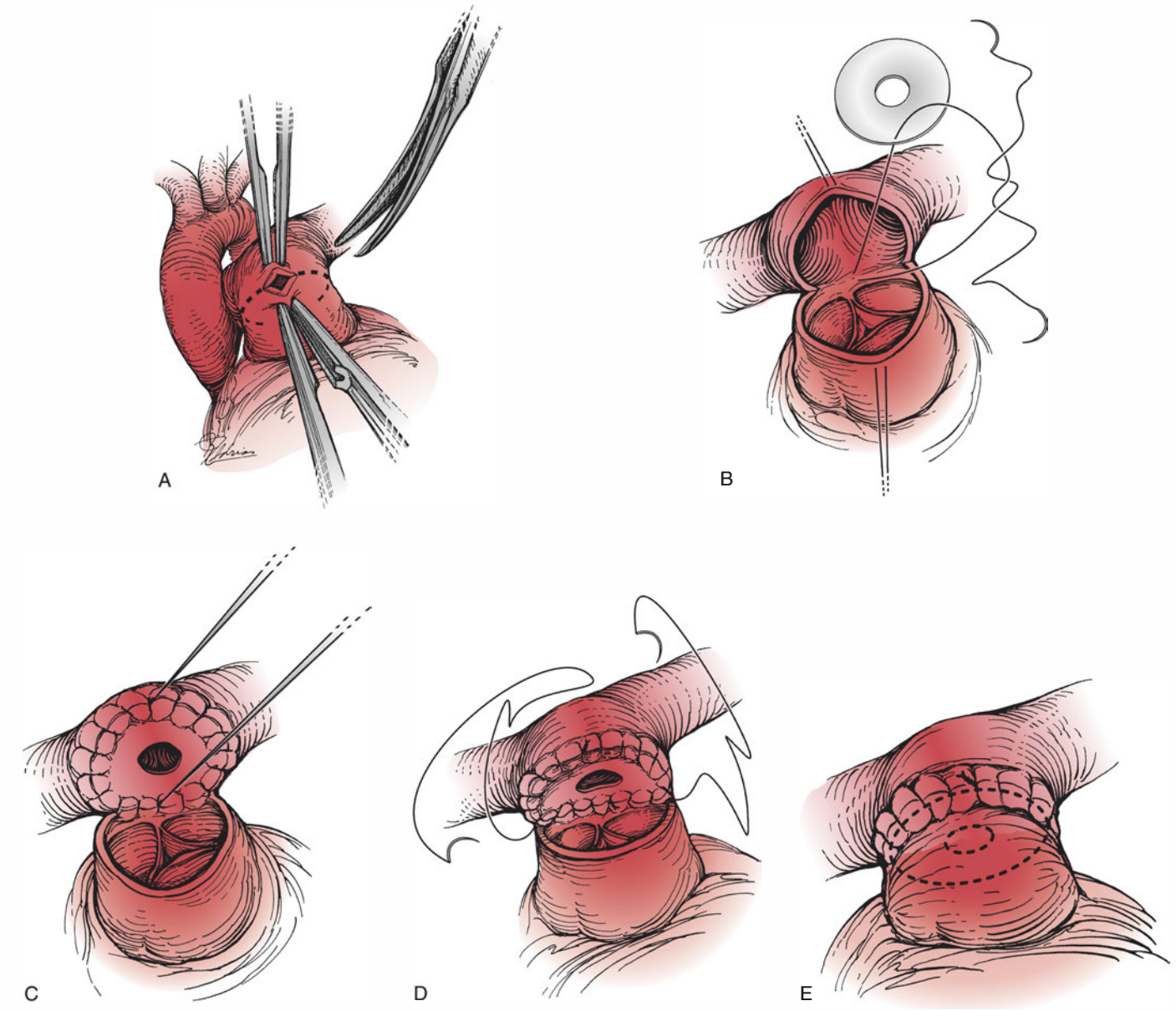


Fig. 75.16. Technique of "intraluminal" pulmonary artery banding. (**A**) The main pulmonary artery is opened circumferentially approximately 5 mm above the pulmonary valve commissure. (**B**) The incision is extended 180 to 270 degrees circumferentially around the pulmonary artery. (**C**) A Gore-Tex patch with a calibrated hole punched in the middle (2.7 mm, 3.6 mm, or 4.0 mm) is sutured in place with running continuous suture. (**D**) The pulmonary artery is reanastomosed. (**E**) The completed intraluminal pulmonary artery band.

Table 75.7 Atrial Septectomy

Technique	Right thoracotomy; Blalock–Hanlon atrial septectomy: 1950
	Rashkind balloon: 1966
	Open septectomy on cardiopulmonary bypass
Most common indications	Restrictive atrial septal defect in transposition of the great arteries; restrictive atrial septal defect with single ventricle

resurgence in the use of pulmonary artery banding for infants with hypoplastic left heart syndrome either as a replacement for the Norwood procedure or as part of a heart transplant strategy. Surgeons need to be aware of the techniques used to create a Waterston or Potts anastomosis and the possible complications of these shunts to care for those patients who have had these procedures in the past.

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EDITOR'S COMMENTS

As the authors have noted, palliative procedures have become less important for patients with congenital heart disease because early primary repair has become the standard approach to treatment. A small number of patients, however, still require palliative shunts or pulmonary artery banding. Most typically, shunting is required as a component of more complex reconstructions, such as the Norwood operation for hypoplastic left heart syndrome. Isolated shunting is still required for certain patients with single-ventricle physiology with limited pulmonary artery blood flow, who require immediate intervention in infancy as staged palliation to later Fontan procedures. Similarly, pulmonary artery banding may be required in certain patients with excessive pulmonary artery flow in infancy, who are also candidates for later single-ventricle repairs. Early limitation of pulmonary blood flow to prevent elevation of pulmonary vascular resistance is very important.

The standard Blalock–Taussig shunt takes advantage of growth of the subclavian artery, which may improve the pulmonary artery blood flow into early infancy and childhood. However, the sacrifice of a subclavian artery, with limitation of use of that artery for monitoring purposes and reduction in limb growth, has limited the usefulness of the standard Blalock anastomosis. In most centers, PTFE-modified Blalock–Taussig shunts, which do not require the sacrifice of a subclavian vessel, are preferentially used. We have generally used smaller PTFE grafts of 3.5 to 4.0 mm in most neonates and infants because in most cases additional surgical intervention, either for complete repair or additional palliation, will occur before the child is 6 months of age, and therefore long-term patency is not necessary.

We have chosen sternotomy as the primary approach for almost all palliative operations, including the systemic-to-pulmonary shunt and pulmonary artery banding. For shunting procedures, the side of the aortic arch is not of concern if a median sternotomy approach is used, and, in addition, the shunt can be placed to the main pulmonary artery if necessary in very small infants with this technique. Access to the patent ductus for ligation to remove competitive flow is always possible through the sternotomy approach. As noted by the authors of this chapter, it is controversial whether ligation of patent ductus arteriosus should be routinely performed at the time of placement of an aortopulmonary shunt. There are conflicting data in the literature regarding this practice. In most centers, ductus arteriosus ligation is performed after placement of any aortopulmonary shunt primarily to insure that shunt flow is adequate; however, it is not uncommon with ligation of the ductus arteriosus for oxygen saturation to dramatically drop and yet over time the saturations tend to recover. The ductus arteriosus tends to be quite large in most of these patients, who are often on prostaglandin therapy, and therefore the aortopulmonary shunt flow, while adequate, can be relatively less than the flow through an open patent ductus arteriosus. Thus, there may be transient changes in pulmonary blood flow that affect pulmonary resistance and oxygen saturation which subsequently resolve. Maintenance of a patent ductus arteriosus after aortopulmonary shunting can create a situation where competitive flow occurs in the pulmonary arteries and could lead to higher incidence of shunt thrombosis. Computer modeling techniques have suggested that leaving the patent ductus arteriosus is suboptimal while other clinical studies as suggested by the authors have indicated that leaving the ductus open can lower early postoperative mortality. The question remains an open one. An additional advantage of a sternotomy for aortopulmonary shunting is the possibility of creating a more central shunt from the main aorta to the pulmonary artery if the innominate vessel is small or there is an aberrant right subclavian artery making the first branch from the ascending aorta the right carotid artery. The use of the carotid artery for the proximal anastomosis of the modified Blalock–Taussig

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EDITOR'S COMMENTS (*continued*)

shunt is possible; however, often there is limitation to flow by the size of the vessel, and therefore placement more centrally on the ascending aorta may be an advantage. An additional advantage of the sternotomy approach is the fact that thoracotomies in children may be associated with late development of scoliosis, and in cyanotic patients significant aortopulmonary collateral vessels may develop from the chest wall into the lung, which can be problematic at reoperations or if potential cardiopulmonary replacement is necessary as final palliation.

Left modified Blalock–Taussig shunts are discouraged because of the difficulty in isolating the shunt at reoperation; often the left phrenic nerve is intimately adherent to the shunt, making left phrenic nerve injury more common with dissection to ligate the shunt at reoperation. We, therefore, prefer right modified Blalock-Taussig shunts through a sternotomy approach if at all possible. Adhesions are generally not a major issue at reoperation. Division of right modified Blalock–Taussig shunts can be accomplished easily at the time of a modified Fontan or Glenn procedure, whereas division of left-sided modified Blalock–Taussig shunts can still be associated with pulmonary artery distortion at the shunt insertion site, which can be complex and difficult to repair, requiring pulmonary arterioplasty, often out into the hilum of the left lung. Waterston shunts are not a preferred approach for palliation. There may still be isolated circumstances where a Waterston-type approach may be used in patients with tetralogy of Fallot and pulmonary atresia with very diminutive central pulmonary arteries, as advocated by Roger Mee. In these cases, the central pulmonary confluence can be directly anastomosed to the back of the aorta in a Waterston-type fashion to encourage pulmonary arterial growth. In most cases, these procedures are performed with cardiopulmonary bypass and even a brief period of circulatory arrest to allow for the best possible alignment of the pulmonary bifurcation to the back of the ascending aorta and to prevent kinking during growth.

For pulmonary artery banding, the variations in location of the great vessels allow approach to the pulmonary arteries safely through a sternotomy incision in all cases, and we therefore reserve the left thoracotomy approach for pulmonary artery banding to selected patients where additional surgery on the ductus arteriosus or coarctation is necessary.

The authors described the use of the Blalock–Hanlon atrial septectomy, which is a closed technique for removing the atrial septum. As they stated, the Rashkind balloon septostomy or atrial septal stenting has virtually replaced the Blalock–Hanlon operation. In patients in whom the atrial septum is particularly thick or in cases in which the capacity is lacking to perform a Rashkind balloon septostomy or atrial septal defect stenting in the catheterization laboratory or when restrictive atrial septal flow remains and operative intervention is necessary, it is preferable to use open techniques to widely excise the atrial septum. In patients with bilateral superior venae cavae and restrictive atrial septal defect, it may be necessary to cut back the roof of the coronary sinus into the left atrium to achieve a wide opening of the atrial septal wall and avoid recurrent obstruction even after open septectomy.

Although palliative operations for congenital heart disease are relatively uncommonly performed, there may be a resurgence of interest in these techniques, as noted by the authors, for certain kinds of neonatal conditions, including hypoplastic left heart syndrome. The recognition that neonates are particularly sensitive to neurologic injury has led to interest in avoiding neonatal operations requiring cardiopulmonary bypass if possible. Delaying intervention into early infancy (6 to 8 weeks) may potentially decrease the risk of neurologic injury, although data have not yet been accumulated to define whether the additional risks of abnormal physiology may outweigh the potential benefits in neurologic outcome. Early primary repair has become the standard approach for lesions where it can be accomplished in the newborn or infant period, and therefore this "hybrid" approach represents a relatively radical change in the approach to newborns with congenital heart disease. However, if data continue to accumulate suggesting that there is a specific neurodevelopmental advantage in avoiding early operation, then palliative procedures, either operative or in the catheterization laboratory, may take on a greater role.

There has been much interest in ductal stenting as an alternative to surgical aortopulmonary shunt creation. Ductal stenting provides a stable source of pulmonary blood flow; however, there continue to be issues with the ability to create a stable shunt size with the use of ductal stenting techniques and the potential for stent migration and for stent ingrowth and obstruction if the stent does not completely extend across the entire length of ductal tissue. Hybrid approaches using intraoperative pulmonary banding and ductal stenting for hypoplastic left heart syndrome are gaining increasing interest, although the results are not superior to standard surgical approaches. In the past, banding of the pulmonary arteries had been used for patients with truncus arteriosus; however, the difficulties in placing bands on small pulmonary arteries accurately and the potential for occlusion of the artery or significant distortion, making repair more difficult, have led to abandonment of this technique. It is, therefore, interesting that it is being resurrected for patients with hypoplastic left heart syndrome as a potential way to avoid early surgical morbidities. There have even been attempts to create endovascular pulmonary artery bands with calibrated openings in a endovascular plug that can be delivered in the catheterization laboratory. As these techniques evolve, the relative risks and benefits of these more modern palliative approaches and hybrid approaches remain to be evaluated.

Although the authors describe pulmonary artery banding as being useful for patients with Swiss-cheese muscular ventricular septal defects and complex ventricular septal defect (VSD) with coarctation, in most cases primary repair of multiple VSDs has been accomplished with good results. Therefore, pulmonary artery banding is reserved for patients with multiple ventricular septal defects in whom direct operative intervention is not considered possible, such as true "Swiss-cheese" defects or isolated ventricular "noncompaction" in which little true septum is present. The problem with pulmonary artery banding in these patients is the development of significant right ventricular hypertrophy, which may obscure

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the margins of the VSDs at the time of operative intervention. In addition, after banding, angiography may suggest that most muscular ventricular septal defects have become small or closed, whereas when ventricular hypertrophy regresses after pulmonary artery debanding, residual VSD lesions may be significant. We believe that generally it is best to address these VSDs early and that in most cases, direct closure can be accomplished. With the advent of transcatheter interventions, small residual or moderate residual defects can be addressed in the catheterization laboratory, and, on occasion, device delivery in the operating room via a transventricular puncture may aid in complete closure of VSDs that are difficult to access via the right atrium.

Complex VSDs with coarctation can often be addressed by primary coarctation repair and VSD closure at a single operation. Although results with coarctation repair and pulmonary artery banding for significant ventricular septal defects are excellent with later repair of the defect and debanding, the progression of techniques of infant repair allows one-stage repair of both defects in most circumstances.

Pulmonary artery banding is now used most commonly in patients with single ventricle with increased pulmonary blood flow to prepare the patient for an eventual Fontan procedure and as a secondary procedure to prepare the left ventricle in patients with late presentation of transposition of the great arteries or for right ventricular failure after the Mustard operation in anticipation of a takedown and an arterial switch procedure. Remotely adjustable banding devices are being developed for this purpose. The authors stated that simple removal of pulmonary artery banding is rarely sufficient to allow relief of pulmonary outflow obstruction. We and others have noted that if second operations are undertaken early, then pulmonary artery debanding can often be accomplished with complete relief of residual obstruction. As the trend toward early intervention for complete repair continues, it may be possible to use very short terms of pulmonary artery banding in most patients with single-ventricle physiology with takedown of the band and performance of a hemi-Fontan or bidirectional Glenn procedure by 3 to 6 months of age. A similar scenario can be used for most patients with multiple VSDs, as noted by the authors. We have not elected to use pulmonary artery banding for atrioventricular canal defects because the use of a band results in significant right ventricular hypertrophy, which may complicate the actual repair of the intracardiac defect. Even in very small patients, primary repair of atrioventricular canal defects has been associated with excellent results; therefore, banding is very rarely indicated.

Knowledge of palliative approaches to congenital heart disease is still essential for congenital heart surgeons to permit the appropriate staging of patients to complete repair. In addition, as noted by the authors, knowledge of the range of palliative operations is particularly important so that successful takedown can be accomplished at time of complete repair.

TLS

76 Anomalies of Systemic Venous Drainage

Sanjiv K. Gandhi and Ralph D. Siewers

INTRODUCTION

This chapter discusses the abnormalities of position and connection of the major systemic venous channels that drain into the heart. Although an understanding of the morphogenesis of the venous system is important, a classification of venous anomalies that is helpful to the surgeon is most appropriately based on anatomic considerations. Thus, this chapter is organized on an anatomic basis, and the anomalies of the venous system within each anatomic segment are addressed. Few of these venous anomalies are intrinsically pathologic to the circulation. However, many, if not most, present important surgical considerations during the palliation or correction of congenital cardiac conditions. Diagnosis of these abnormal systemic venous connections can usually be made echocardiographically, although angiography, computed tomography, and magnetic resonance imaging serve as adjunctive and very useful modalities.

Where appropriate, the relative frequency of presentation and incidence of the major venous anomalies is provided. This incidence is based on the Cardiology Patient Database at Children's Hospital of Pittsburgh (CHP), which numbers >34,200 patients entered from 1955 to 1979. The information is based on a diligent coding of all cardiovascular anomalies in this patient database. Further information from the Children's Hospital of Pittsburgh Heart Museum database is also used when appropriate to illustrate certain relationships between classes of congenital heart defects and anomalies of the systemic venous circulation.

ANOMALIES OF THE SUPERIOR CAVAL VEINS

Bilateral Superior Caval Veins Draining to the Systemic Venous Atrium

The superior caval drainage embryologically begins by means of paired cardinal veins (Fig. 76.1). After development of the left brachiocephalic (innominate) vein at week 7 of gestation, the left superior vena cava (SVC) usually involutes and becomes the ligament of Marshall. The most common anomaly of the superior caval circulation is persistence of the left SVC. This is readily recognized during cardiac catheterization or echocardiography and is frequently seen with the absence of the left innominate vein. Small bridging veins between the right and left SVCs may be present, thus joining the superior cavae. The presence of a left SVC may cause the right SVC to be somewhat smaller than usual, but the relative size of either the right or left vein is variable. In the usual circumstance, with rare exception, if there are no lateralization abnormalities, the left SVC drains into the coronary sinus and thus into the systemic or morphologic right atrium. It produces no physiologic abnormalities in its isolated condition and only becomes important when it is associated with other cardiac anomalies that require surgical correction or during cardiac transplantation.

The presence of bilateral SVCs, with the left caval vein entering the coronary sinus, is by far the most common major venous anomaly in the CHP database. It was found in 0.8% of all the patients and in >4% of the specimens in the heart museum. Seventeen percent of these patients had ventricular septal defects, and 10% to 15% had coarctation of the aorta, tetralogy of Fallot, or atrioventricular septal defects. A slightly smaller percentage had atrial septal defects, double-outlet right ventricle, transposition of the great vessels, and various forms of single-ventricle anatomy.

The left SVC originates at the junction of the left jugular and left subclavian veins. It is located anterior to the aortic arch and left pulmonary artery. Before entering the pericardial space, it accepts the hemiazygos vein, then runs inferiorly and medially into the posterior atrioventricular groove and joins the coronary sinus passing between the left pulmonary veins and the left atrial appendage (Fig. 76.2).

When the right atrium must be opened during open cardiac surgical procedures, provision must be made for management of both right and left SVC drainage. Short periods of occlusion of one, either the right or the left SVC, are tolerated nicely, but long procedures usually require cannulation of not only the right superior vein but also the left. It is believed that venous pressures proximally in the left SVC system that are <30 mmHg when the left SVC is occluded are well tolerated during most surgical procedures.

There are several options for managing drainage of the left SVC. This may be managed by cannulation through the coronary sinus from within the right atrium or directly with surgical cannulation of the left SVC using techniques similar to those on the right side. In very small infants, the use of circulatory arrest is another potential option. Bilateral SVCs, when joined by a bridging vein reminiscent of the left innominate vein, allow for the usual right SVC cannulation and simple (temporary) occlusion of the left SVC. The presence or absence of a left innominate vein is readily determined early during the operation when a midsternal approach is taken.

The augmented blood flow through the coronary sinus enlarges the coronary sinus, and this becomes an important echocardiographic manifestation of the presence of a left SVC that drains via the coronary sinus. Knowledge that a left SVC is present in the face of a coronary sinus orifice that appears normal is a tip that the left SVC may drain directly into the left atrium or that the coronary sinus may be "unroofed." An enlarged or dilated coronary sinus is an important consideration during the surgical repair of specific heart defects, the most important one perhaps being the repair of an atrioventricular septal defect. In this abnormality, which was present in 15% of our patients with a left SVC draining into the coronary sinus, the large and dilated coronary sinus changes somewhat the anatomic relationships during the surgical repair. In some centers, the placement of the septal

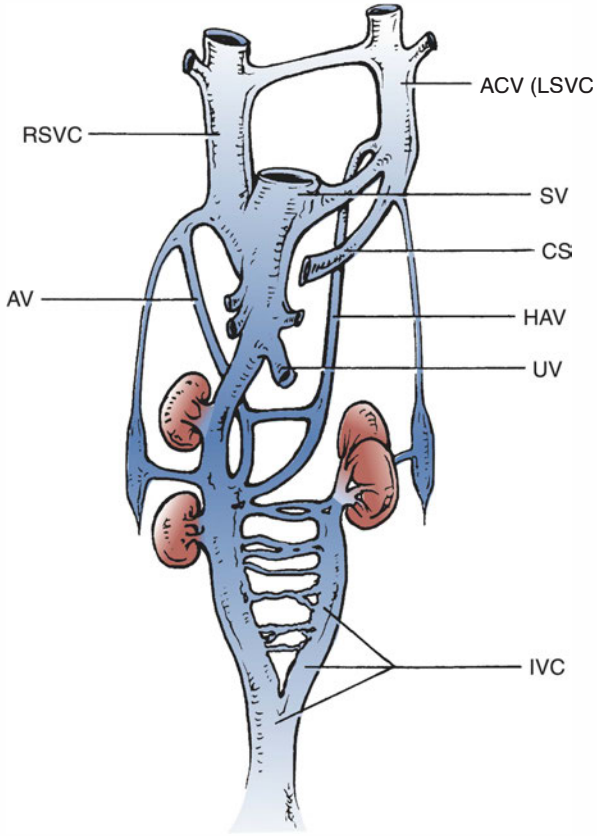


Fig. 76.1. Composite schematic of the venous embryology. ACV, anterior cardinal vein; AV, azygos vein; CS, coronary sinus; HAV, hemiazygos vein; IVC, inferior vena cava; LSVC, left superior vena cava; RSVC, right superior vena cava; SV, sinus venosus; UV, umbilical vein.

patch, because of concern over conduction system injury, will leave the coronary sinus orifice on the pulmonary venous side of the reconstructed atrial septum. This, of course, would not be acceptable with a left SVC draining via the coronary sinus, resulting in a significant right-to-left shunt, unless the SVC could be moved or perhaps closed if bridging veins to the right SVC were present.

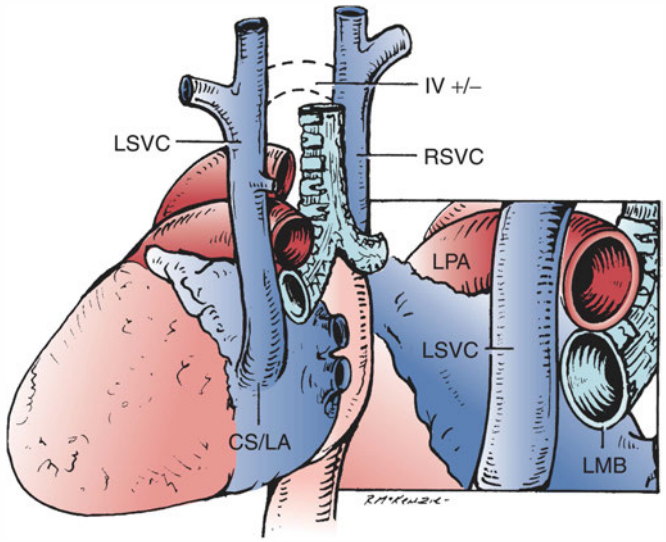


Fig. 76.2. Bilateral superior caval veins. CS, coronary sinus; IV, innominate vein; LA, left atrium; LMB, left main bronchus; LPA, left pulmonary artery; LSVC, left superior vena cava; RSVC, right superior vena cava.

Another rare problem can then be encountered with a left SVC entering the coronary sinus in patients who have elevations of right atrial pressure and pulmonary hypertension in the presence of a dilated coronary sinus, which can impinge on the wall of the left atrium and create what appears echocardiographically and functionally to be a cor triatriatum with limitation of free flow from the pulmonary venous confluence across the mitral valve. The coronary sinus, when decompressed on bypass with the heart under cardioplegic arrest, eliminates the ridge seen on echocardiography, and the defect can be quite obscure.

Although persistence of the left SVC, which drains through an intact coronary sinus into the right atrium, is not intrinsically a pathologic state, a defect in the common wall between the coronary sinus and the left atrium will produce a variable degree of interatrial communication and often must be surgically repaired. These defects in the partition may be small and relatively minor and become of greater significance only in association with complex disease. However, almost complete absence of this partition causes mixing and bidirectional shunting at the atrial level with the resultant systemic desaturation. It is difficult to diagnose various forms of unroofing of the coronary sinus when in association with a persistent left SVC. However, during the surgical repair of interatrial septal defects and other lesions in association with the persistence of the left SVC, the integrity of the coronary sinus with its drainage into the right atrium should be ascertained. Surgical options may include ligation of the SVC if a left innominate vein is present, and then a repair of the coronary sinus interatrial communication in the usual way, or simply a patching of the coronary sinus/left atrial defect to maintain left superior caval drainage into the right atrium through the coronary sinus. This condition is discussed in detail in a later chapter.

Isolated Left Superior Caval Vein

The presence of a left SVC may rarely be associated with the absence of the right SVC. When this occurs, there is a right brachiocephalic (innominate) vein that joins the left SVC. We found 15 cases (0.05%) in the CHP database and 5 additional cases in the Heart Museum. The most commonly associated anomalies were ventricular

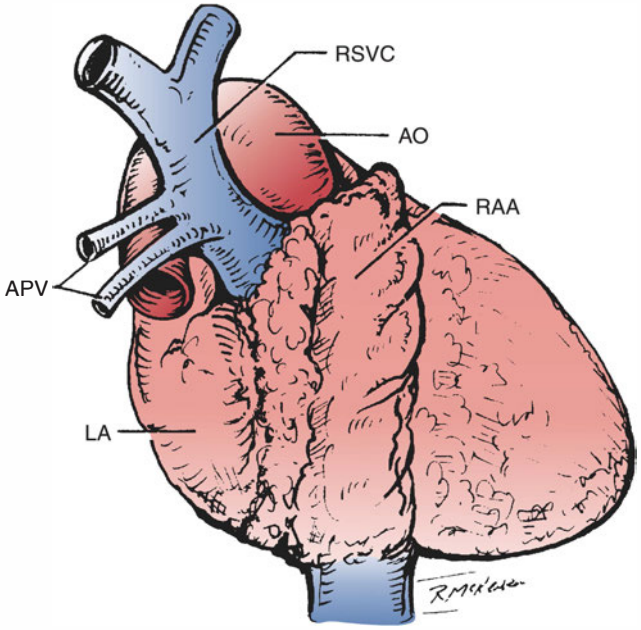


Fig. 76.4. Right superior caval vein connected to the left atrium, with partial anomalously draining pulmonary veins. AO, ascending aorta; APV, anomalous pulmonary veins; LA, left atrium; RAA, right atrial appendage; RSVC, right superior vena cava.

These patients often escape a diagnosis in infancy, but cyanosis usually worsens and a diagnosis will likely be established. There are open and closed techniques for the connection of the right superior cava to the right atrial appendage, removing it from its posteriorly located position with its attachment to the left atrium. The association of partial anomalous pulmonary venous return into the abnormally located right SVC must be recognized. If this is an isolated situation draining a small portion of the right lung, the anomalous pulmonary veins may be left alone. If a large portion of the right pulmonary venous return is connected to the anomalously draining right SVC, the portion of the right SVC containing the anomalous pulmonary veins should be left attached to the left atrium and only the more proximal part of the right SVC attached to the right atrium.

There have been several cases in which the right SVC has been noted to drain into both atria. This is conceptually identical to the superior type of sinus venosus interatrial defect, in which the right SVC "overrides" the atrial septum and offers drainage into both the right and left atria. Correction of this anomaly is usually achieved with enlargement, if necessary, of the interatrial opening (the sinus venosus defect) and baffling of the anomalously connected right-side pulmonary vein or veins to the left of the atrial septum. This sinus venosus defect repair functionally corrects the SVC drainage solely into the right atrium.

Aneurysmal Dilation of the Superior Caval System

Aneurysmal dilation of portions of the superior caval system may occur. These malformations are usually localized and may involve the SVC on either the right or left side. They have also been found in the left innominate vein and in the right and left internal jugular veins. There is usually no need for surgical involvement or correction unless they produce symptoms because of mass or displacement. Thrombus formation and emboli are theoretical concerns but have not been recognized. We have resected one such aneurysm, believed to be a congenital malformation, of the internal jugular vein in a child who had symptoms of fullness and respiratory effort on assuming the supine position. The symptoms were thought to be caused by displacement of the trachea because of the mass effect of the large internal jugular vein and were relieved by simple resection of the hugely dilated vein.

Retroaortic Innominate Vein

Occasionally, the surgeon will encounter what appears to be an absence of the left innominate vein. However, on opening the pericardium, a large vein may be found passing across the upper pericardial space, connecting the left internal jugular and subclavian veins to the right SVC. This vein passes dorsal to the ascending aorta, and its course parallels the right pulmonary artery. It is not thought to be associated with any specific cardiac abnormality; however, most patients with a retroaortic innominate vein have a congenital cardiac malformation. It has been noted with a left SVC, but this association is uncommon. There are no specific surgical connotations associated with this interesting finding other than an awareness of its occurrence so that it may be protected during dissection and not be confused with other structures.

ANOMALIES OF THE INFERIOR CAVAL VEIN

Anomalies of the inferior vena cava (IVC) are less common than anomalies of the SVC. The Cardiology Patient Database at CHP indicates an incidence of 0.3%. Slightly >1% of the hearts in the museum collection were associated with IVC anomalies. Other than lateralization abnormalities, atrioventricular septal defect was the only cardiac malformation with an apparent association with abnormalities of the IVC. Fully 15% of the patients with interruption of the IVC in our database have this anomaly.

Absence of the Hepatic Segment of the Inferior Caval Vein

Clearly, the most noteworthy form of IVC anomaly is the absence of the hepatic segment of the IVC with azygos or hemiazygos continuation of the lower portion of the IVC to the superior caval system. This malformation, referred to as an interrupted IVC, may occur in situs solitus or in the context of a lateralization abnormality.

Azygos Continuation to the Right Superior Caval Vein

In the absence of lateralization abnormalities, IVC blood return to the heart is through an azygos vein continuation to the right SVC from the infrahepatically interrupted IVC (Fig. 76.5). Rarely, the continuation is via the left (hemi) azygos system to a left SVC, even without lateralization abnormalities. It is the most common inferior caval anomaly, and, aside from surgical consideration during the repair of other cardiac anomalies, it is of no major consequence. The hepatic veins usually connect singly through a short suprahepatic confluence resembling a normal IVC to the right atrium. On rare occasions, there may be several separate hepatic vein connections to the atrium, and in the absence of heterotaxy they connect to the right atrium.

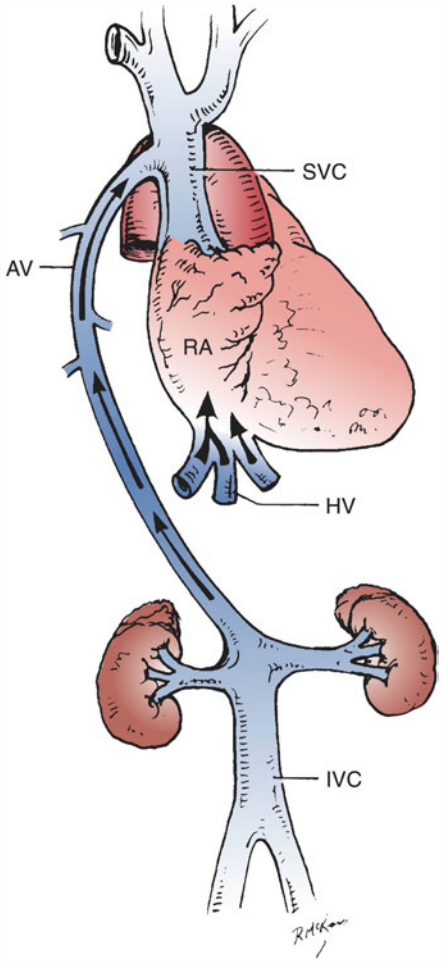


Fig. 76.5. Absence of the hepatic segment of the inferior vena cava (inferior vena cava interruption with azygos continuation). AV, azygos vein; HV, hepatic veins; RA, right atrium; SVC, superior vena cava.

Closure of the azygos vein, for instance, during a right subclavian-to-pulmonary artery shunt should not be performed when the IVC is congenitally interrupted. The tip-off to the surgeon, unless it is known preoperatively, would be an unusually large azygos vein. During the creation of a bidirectional Glenn shunt, the azygos vein is ordinarily ligated. With IVC continuation via the azygos vein, however, the shunt will direct all of the systemic venous blood, save the hepatic venous return, into the pulmonary artery circuit. This has been called the "Kawashima operation," and it is intrinsically attractive as a form of palliation for some patients with a functionally univentricular heart. However, it is occasionally associated with the development or enlargement of intrahepatic venous communications between the hepatic venous and systemic venous circuits, and it may provide a progressive decompression of the systemic venous flow away from the pulmonary circuit. Additionally, the development of arteriovenous pulmonary connections (fistulae) is much more common when the pulmonary circuit is deprived of hepatic venous blood flow. Most patients with a bidirectional SVC-to-pulmonary artery connection in the presence of azygos continuation of an interrupted IVC will eventually require baffling of some or all of the hepatic veins to the pulmonary artery—a completion of the modified Fontan operation.

Interruption of the Inferior Caval Vein in Visceral Heterotaxy

Fifty percent (37 patients) of the CHP database patients with IVC interruption had lateralization abnormalities. All but one were left isomerism with the IVC continued into a left SVC by means of the hemiazygos vein. The surgical considerations are the same as those that apply to the right-side continuation with the azygos vein. Exceptions depend on how the left SVC connects with the atrial mass, either directly or by means of the coronary sinus.

In patients with right isomerism, interruption of the IVC is extremely rare, and the azygos vein anatomy is of much less concern. However, there is a tendency for the hepatic veins to connect directly to the atrial mass, either on the right or the left side. This direct connection of the hepatic veins is very common, of course, in left isomerism because of the higher frequency of IVC interruption in this cardiac defect. The hepatic vein anatomy is much more variable in right isomerism and must be delineated carefully in anticipation of procedures designed to separate the pulmonary and systemic venous drainage and during cardiac transplantation.

Left Atrial and Biatrial Connection of the Inferior Caval Vein

The IVC may connect directly to the left atrium, with or without the presence of an interatrial communication. The inferior form of the sinus venosus interatrial defect provides the situation in which the IVC "overrides" the atrial septum and streaming from the IVC delivers a large amount of inferior caval blood into the left atrium. This anomaly is invariably associated with some degree of partial anomalous pulmonary venous return from the right lung. Depending on the relative degree of "override," the patient may be hypoxic, but shunt studies will miss the diagnosis unless done from the IVC.

Surgical correction of this anomaly involves correction of the interatrial communication with baffling of the IVC to the right side of the atrial septum. In circumstances in which there is a direct connection without interatrial communication, the inferior caval blood flow may be directed to the systemic venous atrium either by reconnection of the IVC or by means of an interatrial baffle construction. This anomaly with an intact atrial septum is extraordinarily unusual.

Other Abnormalities of the Inferior Caval Vein

Duplication of the IVC below the renal veins is not infrequent, but it has no relation to congenital abnormalities of the heart. Its presence or the presence of a left infrarenal IVC that crosses sharply to the right at a subhepatic level may cause the cardiologist some problems during catheterization. The surgeon may experience difficulty during the unusual circumstance of passing large cardiopulmonary bypass venous cannulae from the femoral approach in circumstances when bypass is necessary before the chest is opened or when transthoracic IVC cannulation is not feasible.

Congenital membranous obstruction of the IVC as it enters the right atrium has also been described, and, similar to Budd–Chiari syndrome, may present with liver dysfunction. Resection of such membranes can be performed using cardiopulmonary bypass.

Rarely, the IVC may be completely absent, and the venous return from the lower body is through small paravertebral venous channels that communicate through the azygos systems.

PULMONARY VEIN CONNECTION TO THE SYSTEMIC VEINS

Anomalous connection of a portion or all of the pulmonary veins to either the superior or inferior caval systems may occur. Other than enlargement from increased flow, the systemic vein anatomy and connections are usually normal in this circumstance. The subject is addressed in a later chapter.

TOTAL ANOMALOUS SYSTEMIC VENOUS CONNECTION

There are reports of patients with absence of both the right superior and infrahepatic caval veins with azygos continuation to a left SVC and then to the left atrium. Surgical considerations pertain to separation of the pulmonary venous and systemic venous drainage.

It is postulated that most of these patients have some form of lateralization abnormality.

ANOMALIES OF THE CORONARY SINUS

The coronary sinus receives venous blood from the cardiac veins and is usually the major venous pathway of the coronary circulation. A minor group of cardiac veins (venae cordis minimae) provides a variable degree of additional drainage directly into the cardiac chambers, predominantly the right atrium. The anterior cardiac vein drains directly into the right atrium, and the recognizable thebesian foramina provide outlets for either the minor cardiac veins or the sinusoidal systems of the heart with their drainage into the right atrium and, to some extent, into the right ventricle. Because of these alternative pathways, long-standing (slow onset) obstruction of the coronary sinus or absence of the coronary sinus, most frequently seen in right isomerism, is well tolerated without impairment of the coronary circulation.

Whereas the phenomenon of "unroofing" (partial or complete absence of the common wall between the left atrium and the coronary sinus) of the coronary sinus is found more commonly in patients with a left SVC draining via the coronary sinus, it may also occur in situations of a solitary (normal) right SVC and, in either circumstance, provides an interatrial communication by means of the coronary sinus (coronary sinus atrial septal defect).

The coronary sinus may be entirely absent in an otherwise normal heart, although this is quite unusual. More commonly, the coronary sinus is absent in the lateralization abnormalities, most frequently with right isomerism. In addition, the coronary sinus may be small and underdeveloped with the predominance of the venous drainage of the coronary system through the minor venous channels directly into the right atrium and right ventricle.

Although stenosis or hypoplasia and agenesis of the coronary sinus are well tolerated as a developmental anomaly, acute closure or narrowing of the structure with a sudden rise in cardiac venous pressure is associated with myocardial edema and may be implicated in myocardial dysfunction. It has, however, been difficult to document whether the lateral tunnel approach to the Fontan procedure, which leaves the coronary sinus draining into the lower pressure atrium, carries any functional benefit. Narrowing or obstruction of the coronary sinus should be guarded against, and preferably one should avoid leaving the coronary sinus in an acutely increased venous pressure situation such as the classic right atrium-to-pulmonary artery connection with the Fontan procedure.

Coronary sinus diverticula and aneurysms have also been described. They are often associated with accessory atrioventricular pathways and consequently with arrhythmias.

The most important anomaly of the coronary sinus is related to its enlargement because of flow increases caused by a left SVC connection or by an anomalous pulmonary venous connection. The latter is addressed in a later chapter as one form of anomalous pulmonary venous return. The dilated coronary sinus should alert the surgeon to these possibilities.

ANOMALIES OF THE VALVES OF THE VENOUS SINUS

A consideration of the morphogenesis of the valves of the sinus venosus is fundamental to the understanding of atrial anatomy and the systemic and pulmonary venous connections to the atrial mass. Involution of the right venous valve leaves remnants we recognize as the thebesian and eustachian valves, guarding the coronary sinus and the IVC, respectively. Failure of various degrees of the involution process may leave harmless remnants we recognize as a Chiari network in the right atrium or may produce a fully developed membrane that partitions the systemic atrium and may severely restrict the fetal development of the right heart structures because of its consequences for venous flow patterns in the right atrium.

Persistence of the right venous valve of the sinus venosus to form a variously fenestrated right atrial partition (cor triatriatum dexter) divides the atrium into a venous portion, which receives the systemic veins and coronary sinus and communicates with the left atrium through a patent foramen ovale or atrial septal defect, and a trabeculated portion, which communicates with the right atrioventricular valve (Fig. 76.6). Seriously restrictive partitions are usually associated with and are considered one cause of tricuspid stenosis or hypoplasia, right ventricular hypoplasia, or pulmonary stenosis or atresia.

An interesting and more benign and treatable form of persistence of the right valve of the sinus venosus is the occasional windsock-shaped fibrous structure that attaches in a parachute-like manner to the former attachment points of the right venous valve along the terminal crest and the eustachian and thebesian valves. This windsock structure may, during ventricular diastole, be filled with blood and travel through the tricuspid valve into the right ventricle and even comes out of the right ventricular outflow tract and through the pulmonary valve. It may be confused with an abnormality of the tricuspid valve, a possible vegetation of a structure of the

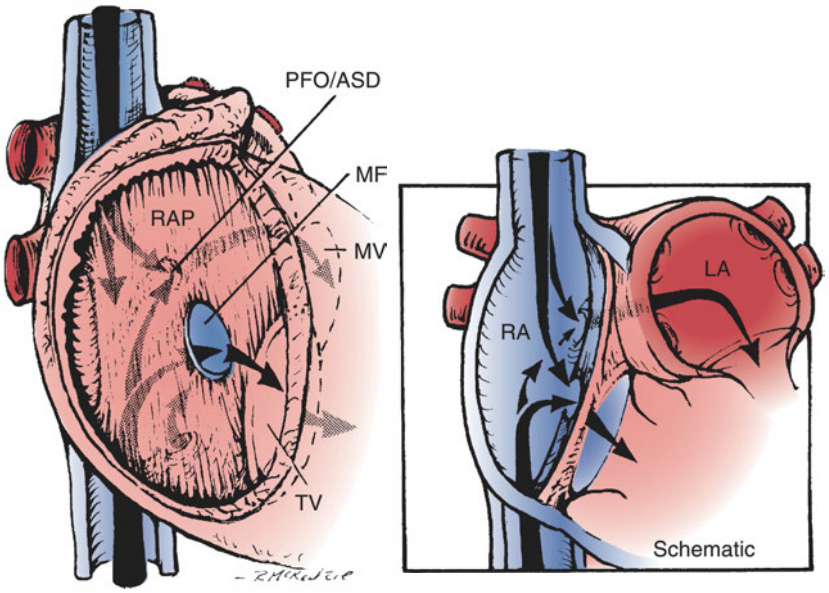


Fig. 76.6. Persistence of the right atrial venous valve (cor triatriatum dexter). ASD, atrial septal defect; LA, left atrium; MF, membrane fenestration; MV, mitral valve; PFO, patent foramen ovale; RA, right atrium; RAP, right atrial partition (persistent venous valve); TV, tricuspid valve.

right side of the heart, or a pedunculated right atrial tumor. We have resected three of these interesting lesions, each of which occurred without associated cardiac abnormalities other than patent foramen ovale. They provide interesting echocardiographic findings for the cardiologist and may produce long-term problems with trauma to the tricuspid or pulmonary valves. It has been speculated that they may cause arrhythmias because of their movement throughout the right heart system in addition to progressive interference with systemic venous return and may be associated with thrombus formation. These structures should be removed when identified.

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EDITOR'S COMMENTS

Anomalies of systemic venous return to the heart are often ignored in texts on congenital heart abnormalities. Although the majority of systemic venous return abnormalities are of no particular physiologic consequence, an understanding of venous anatomy and development and the variations of venous connections to the heart is critically important for congenital cardiovascular surgery in which cavopulmonary anastomoses and separation of pulmonary and systemic venous return are required, as in the Fontan operation.

As noted in the chapter, the most common venous anomaly is a left SVC that drains through the coronary sinus into the systemic venous atrium. This venous anomaly, although physiologically unimportant, can have significant consequences if unrecognized at the time of cardiac operation. It is possible to snare the left SVC temporarily, especially with the use of vacuum-arrested venous drainage, simplifying cannulation during cardiac repair. In complex anastomoses, we have used circulatory arrest to avoid having to separately cannulate the LSVC. Although the presence of a left SVC entering the coronary sinus was thought to complicate cardiac transplantation, in fact, the vein can be left in situ and cardiac transplantation performed in the usual manner with only minor modification, as described in Chapter 99 on Pediatric Cardiac Transplantation. One problem we have seen on occasion with a left SVC entering the coronary sinus in patients who have elevations of right atrial pressure and pulmonary hypertension is the presence of a dilated coronary sinus, which can impinge on the wall of the left atrium and create what appears echocardiographically and functionally to be a cor triatriatum with limitation of free flow from the pulmonary venous confluence across the mitral valve. The mitral valve may also be relatively small in these patients. This abnormality has on occasion resulted in exploration of the left atrium at the time of cardiac repair without finding any abnormality of the left atrium. The coronary sinus, when decompressed on bypass with the heart under cardioplegic arrest, eliminates the ridge seen on echocardiography, and the defect can be quite obscure.

As is apparent from the summary presented in this chapter, knowledge of the possible venous connections to the heart and recognition that the venous drainage must be identified at the time of atrial repairs and cavopulmonary connections are important for the management of children with congenital heart disease. Minor unrecognized systemic venous connections after cavopulmonary anastomoses can create decompression of the SVC and result in significant arterial desaturation. Thus, in patients with cavopulmonary anastomoses in whom significant desaturation is noted postoperatively, a search in the catheterization laboratory for other systemic venous pathways to the atrium should be undertaken and coil embolization performed to eliminate these decompressing channels. In spite of the theoretical need to ligate the azygos vein in patients who undergo SVC-PA anastomosis or a hemi-Fontan procedure for single ventricle, we have found in many patients that the azygos vein can be left open without producing significant desaturation despite relative elevation of superior vena caval pressures. Thus, it would appear that development of significant systemic venous collaterals to the left or right atrium in patients with cavopulmonary anastomoses is likely related to elevated resistance to pulmonary blood flow.

The authors noted that there are often abnormalities of coronary sinus drainage in patients with lateralization abnormalities. On occasion, the absence of the coronary sinus can be associated with direct drainage of the major cardiac veins into the right atrium from the epicardial surface. In these situations, care must be taken not to divide these veins when performing a right atriotomy for access to the intracardiac lesions. Often these patients have associated left juxtaposition of the atrial appendages, which makes the right atrial wall smaller than normal and limits access to the right side of the heart.

Perhaps the major surgical issue with anomalies of systemic venous return is the situation of a left superior vena cava that enters the left atrium directly. In these cases, it may be difficult to reconnect the left superior vena cava to the right atrial chamber. As outlined in the chapter, various techniques have been described including detachment of the left superior vena cava and anastomosis to the right superior vena cava, recreating

(*continued*)

EDITOR'S COMMENTS (*continued*)

an innominate vein. Often, however, the size of the great arteries and space limitations in the superior mediastinum prevent there being enough length for this reconstruction to be accomplished without tension. Other approaches have involved detaching the left superior vena cava and reattaching it to the dome of the right atrium or right atrial appendage if there is no compression in the pulmonary artery or aorta. In some cases, it may be possible to create an intra-atrial baffle that redirects the flow from the superior vena cava as it enters the roof of the left atrium over to the right side of the heart either with the use of prosthetic patch material or by imbricating left atrial tissue to create a conduit. In situations where there is left juxtaposition of the atrial appendages, we have found it useful to divide the left superior vena cava and then anastomose it to the appendage of the right atrium, which is now on the left side, adjacent to the left superior vena cava. In situations where the left superior vena cava cannot reach the right-sided structures adequately, there have been reports of ligation of the left superior vena cava without serious sequelae, although at least transient left facial and arm edema is often present and chylothorax may be a problem. In situations of single ventricle or if the pulmonary artery pressure is expected to be low, another alternative is to perform a left bidirectional Glenn shunt, which allows the left superior vena caval flow to go directly to the pulmonary arteries.

It is particularly important to understand the systemic venous anomalies when dealing with patients with lateralization abnormalities (heterotaxy syndromes). In patients with interrupted inferior vena cava and azygos or hemiazygos continuation, creation of the Kawashima-type connection between the vena cava and pulmonary artery can result in maldistribution of hepatic blood flow such that arterio venous malformations in the lung can be prevalent. The hepatic veins generally have to be redirected into the pulmonary vascular bed; however, if directed on the side opposite the Kawashima connection, most of the hepatic flow maybe pushed to the ipsilateral lung. In this situation, patients may return with progressive cyanosis despite an otherwise good functional Fontan result. There has been increasing interest in the use of computational fluid dynamic modeling with MRI to redesign the Fontan pathways to maximize flow distribution in these patients. In some cases, it may be advisable to connect the hepatic veins to the retrocardiac inferior vena cava or azygos vein such that the large return to the pulmonary arteries is distributed equally to both lungs. In other cases, use of a Y-graft may distribute hepatic flow into the right and left pulmonary arteries to optimize flow distribution.

While the modification of the Fontan operation with the use of an extracardiac conduit leaves the coronary sinus in a low-pressure system and therefore should not result in significant elevation of coronary sinus pressure, there have been situations where lateral tunnel connections have left the coronary sinus in the systemic venous aspect of the repair. This has resulted in some cases in progressive dilation of the coronary sinus with potential decompression into epicardial coronary veins, which ultimately can find their way back to the left-sided atrium and result in cyanosis. Thus, in most cases the coronary sinus should be carefully left in the low-pressure side of the Fontan circulation to avoid these potential problems.

TLS

77 Patent Ductus Arteriosus

William M. DeCampli

DEFINITION

The ductus arteriosus is a vascular communication between the systemic and pulmonary vasculature, usually between the isthmus of the aortic arch and the origin of the left pulmonary artery, which forms a vital part of the fetal vascular anatomy. Normally, the ductus arteriosus arises from persistence of the distal portion of the embryonic sixth aortic arch.

FETAL CIRCULATION

In the fetus, the lungs are not ventilated and pulmonary resistance is high. The majority of relatively desaturated venous blood from the superior vena cava returning to the right ventricle passes through the ductus arteriosus into the descending aorta, whence some of it exits the fetal body through the umbilical artery to be oxygenated by the placenta. The ductus arteriosus is normally strategically positioned so that this desaturated blood does not travel to organs with the greatest oxygen metabolism (the brain and myocardium). After birth, pulmonary gas exchange commences and the circulation is "in series." With the ductus arteriosus no longer essential, spontaneous closure normally occurs in hours to a few days after birth.

MECHANISMS OF SPONTANEOUS DUCTAL CLOSURE

Ductal closure occurs in two stages. The first is functional closure, occurring within the first 6 to 15 hours after birth. This stage results from medial smooth muscle constriction, promoted by four principal factors: (1) increase in arterial oxygen tension, (2) decrease in circulating prostaglandin E2 (PGE2), (3) decrease in mean ductal blood pressure, and (4) decline in the density of PGE2 receptors in the ductal wall. Part of the oxygen dependence is intrinsic to the specialized smooth muscle cells. The second stage (anatomic closure) is usually completed in a few days to weeks after birth and results from (1) subendothelial deposition of extracellular matrix, (2) resorption of the internal elastic lamina and elastin fibers in the media, and (3) migration of undifferentiated smooth muscle cells into the subendothelial space. The ductus closes from the pulmonary end toward the aortic end, often leaving a "ductal ampulla" seen on echocardiography. The resulting nonpatent ductus is then called the *ligamentum arteriosum*. The *patent ductus arteriosus* (PDA) is defined as persistent patency of the fetal ductus arteriosus beyond its normal time of spontaneous closure.

RISK FACTORS FOR PATENT DUCTUS ARTERIOSUS

Any pathology that alters the molecular mechanisms of normal ductal closure may result in PDA. By far, the most frequently encountered risk factor for PDA is *prematurity*. The incidence of PDA increases with lower birth weight and estimated gestational age (EGA), reaching 77% at EGA 28 weeks. Circulating PGE2 activity is greater in the preterm infant than in the full-term infant, promoting persistent ductal patency, presumably because PGE2 is completely metabolized with first pass through the mature lung but incompletely in the premature lung. The risk for PDA is further increased with infant respiratory distress syndrome (RDS). Ninety percent of babies with RDS and $<$32 weeks EGA have PDA.

PATHOPHYSIOLOGY, CLINICAL MANIFESTATIONS, AND DIAGNOSIS

Premature Neonates

In premature neonates, the normal decrease in pulmonary vascular resistance with time leads to a left-to-right shunt, which results in pulmonary vascular overcirculation and an excess volume load on the left ventricle. If the shunt is large, clinically significant congestive heart failure can develop, leading to compromised pulmonary function and systemic cardiac output. Findings include pulmonary congestion, tachypnea, ventilator dependence, peripheral edema, and poor organ and tissue perfusion.

Lung immaturity confounds the relationship between PDA and these clinical manifestations. Pulmonary dysfunction may be due to RDS and bronchopulmonary dysfunction (BPD) and difficult to separate from the clinical effects of pulmonary overcirculation. Necrotizing enterocolitis may be related to microbial effects rather than to PDA. Discriminating the underlying causes has proven to be very difficult. Until an evidence-based guide to clinical impact is developed, McNamara and Sehgal have proposed a combined clinical and echocardiographic staging system for determining the "importance" of the hemodynamically significant PDA in the premature neonate, described in Table 77.1.

Full-Term Infants and Children

As in premature newborns, the principal pathophysiology of PDA is a left-to-right shunt leading to congestive heart failure. With large shunt, infants manifest failure to thrive, upper respiratory infections, and fatigue with exertion (including feeding). Physical examination shows a continuous "machinery murmur" in the left second intercostal space, bounding pulses, an overactive precordium, wide pulse pressure, and enlarged liver. Chest film shows increased vascular markings, interstitial edema, and enlarged cardiac silhouette. Echocardiography demonstrates the ductal and aortic arch anatomy, direction of flow, and evidence of chamber dilatation.

With a large shunt present for a sufficiently long time (but as little as 6 months), pulmonary vasculopathy may develop. Patients will have symptoms consistent with pulmonary hypertension (PAH). They may exhibit differential cyanosis due to

Table 77.1 Representative "Staging System" for Determining the Magnitude of the Hemodynamically Significant Ductus Arteriosus Based on Clinical and Echocardiographic Criteria

Clinical	Echocardiography
C1 Asymptomatic	**E1** No evidence of ductal flow on two-dimensional or Doppler interrogation
C2 *Mild* Hypoxemia (OI <6) Occasional (<6) episodes of oxygen desaturation, bradycardia, or apnea Need for respiratory support (nCPAP) or mechanical ventilation (MAP <8) Feeding intolerance (>20% gastric aspirates) Radiologic evidence of increased pulmonary vascularity	**E2** *Small* PDA Transductal diameter <1.5 mm Restrictive continuous transductal flow (DA V_{max} >2.0 m/s) No signs of left heart volume loading (e.g., mitral regurgitant jet >2.0 m/s or LA:Ao ratio >1.5:1) No signs of left heart pressure loading (e.g., E/A ratio >1.0 or IVRT >50) Normal end-organ (e.g., superior mesenteric, middle cerebral) arterial diastolic flow
C3 *Moderate* Hypoxemia (OI 7–14) Frequent (hourly) episodes of oxygen desaturation, bradycardia, or apnea Increasing ventilation requirements (MAP 9–12) Inability to feed due to marked abdominal distension or emesis Oliguria with mildly increased creatinine Systemic hypotension requiring a single-inotropic agent Radiological evidence of cardiomegaly or pulmonary edema Mild metabolic acidosis (pH 7.1–7.25 and/or base deficit 7–12.0)	**E3** *Moderate* HSDA Transductal diameter 1.5–3.0 mm Unrestrictive pulsatile transductal flow (DA V_{max} <2.0 m/s) Mild-to-moderate left heart volume loading (e.g., LA:Ao ratio 1.5:2.1) Mild-to-moderate left heart pressure loading (e.g., E/A ratio >1.0 or IVRT 50–60) Decreased or absent diastolic flow in superior mesenteric artery, middle cerebral artery, or renal artery
C4 *Severe* Hypoxemia (OI >15) High ventilation requirements (MAP >12) or need for high-frequency ventilation Profound or recurrent pulmonary hemorrhage "NEC-like" abdominal distension with tenderness or erythema Acute renal failure Hemodynamic instability requiring >1 inotropic agent Moderate-to-severe metabolic acidosis (pH <7.1) or base deficit >−12.0	**E4** *Large* HSDA Transductal diameter >3.0 mm Unrestrictive pulsatile transductal flow Severe left heart volume loading (e.g., LA:Ao ratio >2.1, mitral regurgitant jet >2.0 m/s) Severe left heart pressure loading (e.g., E/A ratio >1.5 or IVRT >60) Reversal of end-diastolic flow in superior mesenteric artery, middle cerebral artery, or renal artery

DA V_{max}, ductus arteriosus peak velocity; E/A, early passive to late atrial contractile phase of transmittal filling ratio; HSDA, hemodynamically significant ductus arteriosus; IVRT, isovolumic relaxation time; LA:Ao ratio, left atrium to aortic ratio; MAP, mean airway pressure; nCPAP, nasal continuous positive airway pressure; NEC, necrotizing enterocolitis; OI, oxygenation index.

Modified from McNamara and Sehgal.

flow reversal in the PDA. Echocardiography will confirm bidirectional or right-to-left flow in the PDA. With progressive vascular injury, PAH becomes irreversible, leading to Eisenmenger's syndrome. Cardiac catheterization is indicated for any patient with PDA with clinical or echocardiographic suspicion of PAH.

PDA with a small left-to-right shunt may be asymptomatic and produce no chamber enlargement. Even smaller PDAs may lack an audible murmur (so-called "silent ductus") and are found incidentally on echocardiography or contrast studies.

In Adults

Adults with small, restrictive PDA may be asymptomatic and show no other pathological findings. Adults with anatomically large PDA may show signs of congestive heart failure or they may present with symptoms and signs of PAH or Eisenmenger's syndrome, as described above. Rarely, an adult may present with infective endocarditis, ductal aneurysm, aortic or pulmonary aneurysm, or aortic dissection. Chest film and echocardiography may demonstrate calcification of the ductus.

NATURAL HISTORY

Premature Neonates

The natural history in this population is confounded by the coexisting morbidities of prematurity. No study clearly establishes PDA as a cause of death in this population. Spontaneous ductal closure continues to occur at least up to 1 year of age. In a study of 122 infants with birth weight <1,000 g, closure occurred within 3 days in 25, within 8 days in 42, and before hospital discharge in 46. Among infants with birth weight >1,000 g, the ductus closed without intervention in 67% by age 7 days and in 94% prior to discharge, at a median age of 7 days.

Children and Adults

The spontaneous closure rate in children >1-year old is about 0.6% per year. Among patients with significant sized PDA, the overall mortality in older patients with PDA at 0.49% per year (for age 2 to 19 years) and 1.8% per year (for age >20 years), 30% are dying from congestive heart failure. Patients with large, untreated PDAs that progress to severe PAH have survival curve typical for that of Eisenmenger's syndrome. The risk of bacterial endocarditis is very low in the antibiotic era but not zero, even with a "silent ductus."

TREATMENT

Historical Notes

The first successful PDA ligation was performed on a 7-year-old girl by Robert

Edward Gross (1905 to 1988) on August 26, 1938. The history, along with some legend, is detailed by Alexi-Meskishvili and Böttcher. In 1966, Werner Porstmann (1921 to 1982) was the first to close a PDA by catheter technique. In 1976, Heymann, Rudolph, and Silverman first reported the use of indomethacin to close PDA in premature infants.

Indications for Treatment

Premature Neonates

The staging system proposed by McNamara and Sehgal (Table 77.1) is a reasonable guide to therapy—a "C3E4" patient would certainly be recommended for PDA closure in most neonatal centers. Most commonly, such a baby has (1) a large PDA with a large shunt, (2) refractory or worsening ventilator dependence and/or poor tissue perfusion in the absence of some other dominating and reversible cause, and (3) has failed a trial of COX inhibitors. Babies with stage C4 are a very high-risk group and the decision to undergo surgical ligation should be based on careful discussions with the neonatologists and parents. PDA management in babies with stage less severe than C3E3 is subject to the most debate.

The surgeon should understand the basis for uncertainty in the indications for PDA closure in this population. The primary reason is that, despite at least 75 randomized-controlled trials of interventions to close PDA in premature infants, there is scant evidence that either prophylactic or therapeutic PDA closure, either by medical or by surgical means, improves any clinical outcome measure. Taken individually, each of these randomized-controlled studies had weaknesses—most were small studies (<100 subjects), many had "rate of ductal closure" as their primary outcome measure and cross-over rates were frequently high. Only 17 of the 75 trials were conducted after the advent, in 1989, of exogenous surfactant therapy. Benitz, in 2010, attempted a meta-analytic study of these trials. Such an approach mathematically narrows the confidence intervals of odds ratios for the chosen outcomes, but it widens the underlying variability of the covariates. The Cochrane Reviews apply more stringent criteria to selection of studies for meta-analysis, eliminating studies with significant methodological deficiencies. The reviewers found only one eligible study pertaining to *prophylactic* surgical ligation of PDA, which showed that ligation decreased the incidence of NEC but had no long-term effect on mortality or BPD incidence. Furthermore, the reviewers found only one eligible study pertaining to surgical ligation for the *treatment* of PDA, concluding that this study contained insufficient data to evaluate the effectiveness of the intervention. Thus, as of year 2011, there are no studies, properly designed and containing sufficient data that evaluate surgical ligation for the treatment of PDA in premature infants.

Full-term infants: In full-term infants >6 to 12 months old, occlusion of an audible PDA with left-to-right flow is reasonable. PDA occlusion is also indicated at any age when the PDA is moderate to large and produces congestive heart failure. Infants and children with PDA undergoing other cardiac or thoracic operations should undergo simultaneous PDA ligation.

Children and Adults

Beyond infancy, probably the spontaneous ductal closure is low. Audible PDA with left-to-right flow should be occluded. Recommendations for PDA closure in adults are further described in the *American College of Cardiology/American Heart Association 2008 Guidelines for Adults with Congenital Heart Disease* (ACC/AHA).

Contraindications to Treatment

PDA should not be occluded in patients with PAH associated with significant pulmonary vascular obstructive disease (PVOD). At cardiac catheterization, if pulmonary vascular resistance index is greater than eight Woods units per meter squared and does not decrease with oxygen, nitric oxide, or other pulmonary vasodilators, then PDA closure is contraindicated. Indications for PDA closure in the presence of some reactivity of PAH are controversial.

In premature neonates, bidirectional flow in a PDA is not a contraindication to ligation. PDA should not be ligated as an isolated procedure in the presence of "ductal dependent circulation." The treatment of an inaudible PDA is not contraindicated but is controversial.

Medical Treatment

COX inhibitors close PDA in premature neonates in about 70% of cases, a significantly lower success rate than the closure rate of surgical ligation. Complications are reported for all COX inhibitors, including ibuprofen, in this population. COX inhibitors are not effective in closing PDA beyond a few weeks of age.

Catheter-Based Treatment

Catheter-based occlusion of PDA is an acceptable method of treatment in some cases. The choice of device is often made on the basis of the classification of Krichenko et al. Gianturco or Flipper coils are generally deployed by the femoral artery approach and used for types D and E, or ductus <2 mm diameter. The Amplatzer vascular plug IV is deployed usually via the femoral vein and is applicable for types C, D, and E. Finally, the Amplatzer II occluder device (ADO) is usually deployed via the femoral vein and applicable for type A. In patients <10 kg, this device may obstruct the aorta. In large diameter or short PDA, the incidence of residual flow has been shown to be less with the ADO than with coils.

Complications of catheter-based PDA occlusion include femoral artery pseudoaneurysm, limb ischemia, device embolization, left pulmonary artery stenosis, endarteritis, radiation exposure risks, dye-induced renal dysfunction, residual ductal flow, device-related hemolysis, descending aortic obstruction, hemorrhage, need for transfusion, and failure to complete the procedure (Fig. 77.1). The precise incidences of these complications are not well known due to the paucity of published data from catheterization registries, multi-institutional studies, or Cochrane reviews. The incidence of device embolization is approximately 2% and that of residual flow is 1.4% to 7%. Hemolysis typically occurs with incomplete ductal closure, though it can also occur with complete occlusion. The incidence of complications during and after catheter interventions of all types has been estimated to be nearly 20%.

Experience with catheter-based PDA occlusion has extended to smaller infants and even premature neonates. Long-term outcomes were not determined in these series.

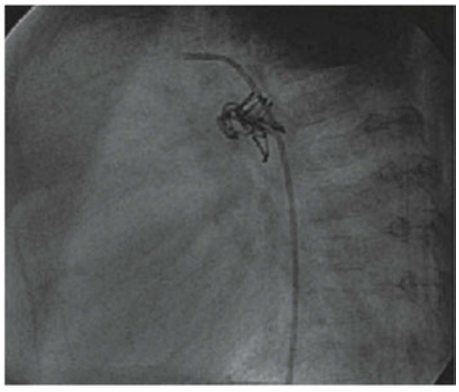


Fig. 77.1. Gianturco coils occluding the descending aorta after attempted coil occlusion of patent ductus arteriosus. The Doppler gradient was 40 mmHg. Through a lateral thoracotomy, proximal and distal aortic control was achieved, a short aortotomy made, and the coils removed. The PDA was doubly ligated.

Catheter-based closure of PDA is effective in adults and preferred in adults with calcified PDA or with significant other medical problems. Relative contraindications include the presence of ductal aneurysm or active ductal/pulmonary arterial endarteritis. Case reports of the use of covered aortic stents have recently appeared. This approach may be appropriate for the large or aneurismal ductus.

In infants and children, catheter-based PDA occlusion has been compared with open surgical ligation. Total complication rates are similar, but coil occlusion is less costly than *open* surgical ligation. In a single-institution matched cohort study by Dutta et al. comparing video-assisted thoracoscopic ligation (VATS) and coil occlusion, however, the total costs were not significantly different. The mean length of stay was 1.6 ± 0.2 days for VATS. With "same day discharge" following VATS PDA ligation, the cost difference has shifted in favor of VATS ligation. In 2011, Chen et al. published a study of 294 patients undergoing either Amplatzer device closure or VATS ligation. The complication rate was 1.5% in the VATS group and 10.2% in the catheter-based device closure group. The cost of VATS therapy was only *one-third* that of catheter-based closure. Thus, the current evidence shows that, for many patients with PDA, the catheter-based and VATS approaches are comparable in safety, effectiveness, and expediency, and VATS appears to be less costly.

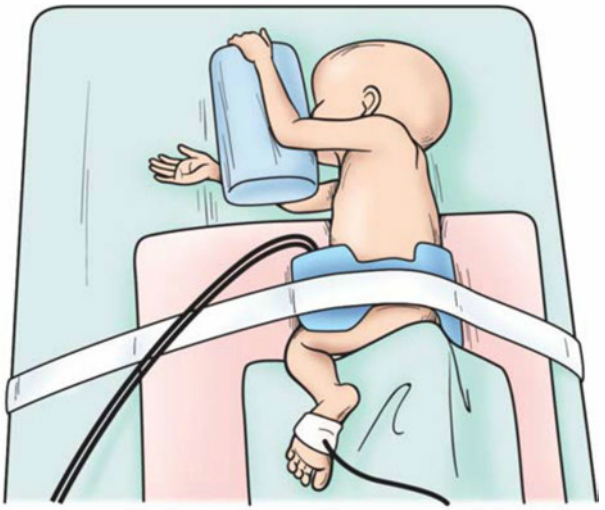


Fig. 77.2. Positioning of premature neonate for PDA ligation.

Surgical Treatment

Open Technique

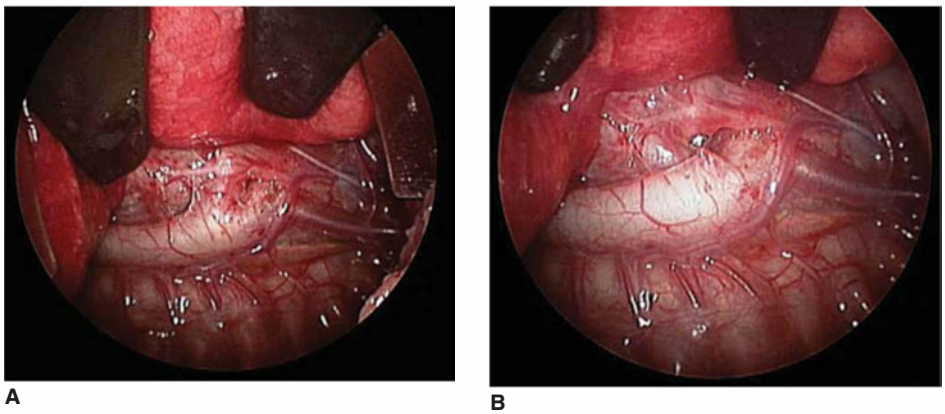
Premature neonates: Open ligation is the standard technique for premature neonates and can be performed in neonates as small as 350 g. These babies are often unstable. The risks of transport can be obviated by performing the procedure in the neonatal intensive care unit. Intraoperative and postoperative systolic ventricular dysfunction, hypotension, and decreased cerebral blood flow have been reported. These effects underscore the importance of expediency, minimal retraction on the lungs, avoidance of visceral pleural air leak, and meticulous hemostasis during the operation, and compulsive care postoperatively.

The situs of the ductus should be ascertained from the echocardiogram report. The patient is placed in the lateral decubitus position, prepped, and draped (Fig. 77.2). Adherent drapes should be avoided. A lateral thoracotomy incision, with minimal curvature, is made. Ideally, one can spare both the latissimus dorsi and serratus anterior muscles. The pleural cavity is entered through the fourth interspace, which is frequently identified as the first somewhat wider interspace just under the scapula. Even a slight injury to the lung surface during pleural opening may result in pneumothorax. A rib retractor is placed. Small rubberized malleable retractors, controlled by the first assistant, are used to retract the lung lobes.

Using low-power "needle point" cautery, a short pleural flap is mobilized, then planes just cephalad and just caudad to the ductus are developed with a Jacobson clamp, not cautery (Fig. 77.3A and 77.3B). Small cotton swabs can be used to facilitate dissection or blot minor bleeding should it be present. The clip applicator should be tested on the drape, as scissoring can tear the duct. Depending on ductal size, one "medium" or "medium-large" hemoclip is applied to the aortic end of the ductus (Fig. 77.3B). If the ductus is sufficiently long, a second clip can be applied, being careful not to overlap the first clip, by mild retraction of the descending aorta.

If the ductus tears during mobilization or ligation, one should use a cotton swab to apply direct pressure to the site, maintaining flow to the pulmonary artery and aorta, until blood is available. After sufficient time, removal of the cotton swab usually does not result in immediate bleeding. At that point, hemoclips can generally be applied to achieve hemostasis and ductal closure. Alternatively, one can obtain proximal and distal control of the aorta with clamps or tourniquets before ligating the duct with prolene suture or clips.

If hemostasis is good, the lung retractors are immediately released to allow full ventilation. The blood pressure may



A B

Fig. 77.3. (**A**) Juxtaductal pleural mobilization in a preterm infant undergoing PDA ligation. Note the position of malleable, rubber-coated hand-held lung retractors. Note that the vagus nerve near the recurrent nerve has been gently mobilized anteriorly to allow space for clip application. (**B**) Single clip placed across the PDA. Note the position of the vagus and recurrent nerves and the degree of hemostasis.

transiently decrease after ligation due to the acute increase in left ventricular afterload, but it usually recovers within minutes. With large PDA, the diastolic blood pressure should rise after this transient effect. Often, with moderate-sized PDA, one does not see a significant change in blood pressure acutely. A pleural drain should be avoided if (1) the lung surface was not violated during the procedure and (2) there is no residual bleeding or evidence of lymphatic leak. If necessary, a 5-F pigtail catheter is sufficient for a drain. Any hemodynamic or respiratory instability immediately following the procedure should prompt immediate auscultation and transillumination of the chest by a neonatologist, as pneumothorax is the most common cause. This complication can be treated with needle aspiration or percutaneous placement of a drain. A post-procedure chest film is always obtained.

Full-term infants and children: The open surgical procedure is similar to that of premature neonates, with some modifications. After cephalad and caudad dissection, a right angle clamp is used to bluntly dissect under the ductus and is then used to pull through one or two silk or Tevdek ties. The ductus can then be doubly ligated with two ties, or one tie and a hemoclip (Fig. 77.4).

If the ductus tears during dissection or ligation, bleeding should be immediately controlled with a cotton tip or sponge stick while blood is made available. Proximal and distal control of the ductus should be obtained with clamps, and the ductus divided and the ends securely oversewn, as described below.

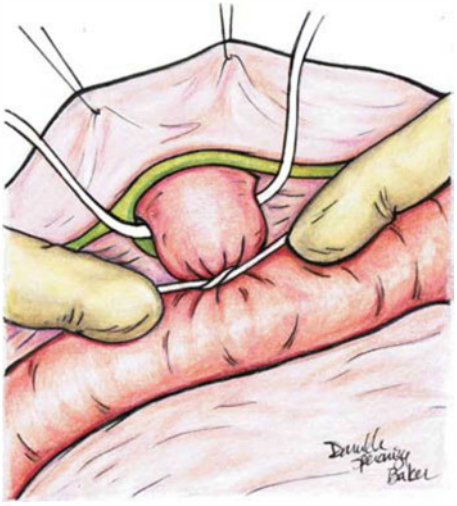


Fig. 77.4. Open ligation of PDA. Note two ties that have been passed under the ductus. The first one is being tied with traction of the other.

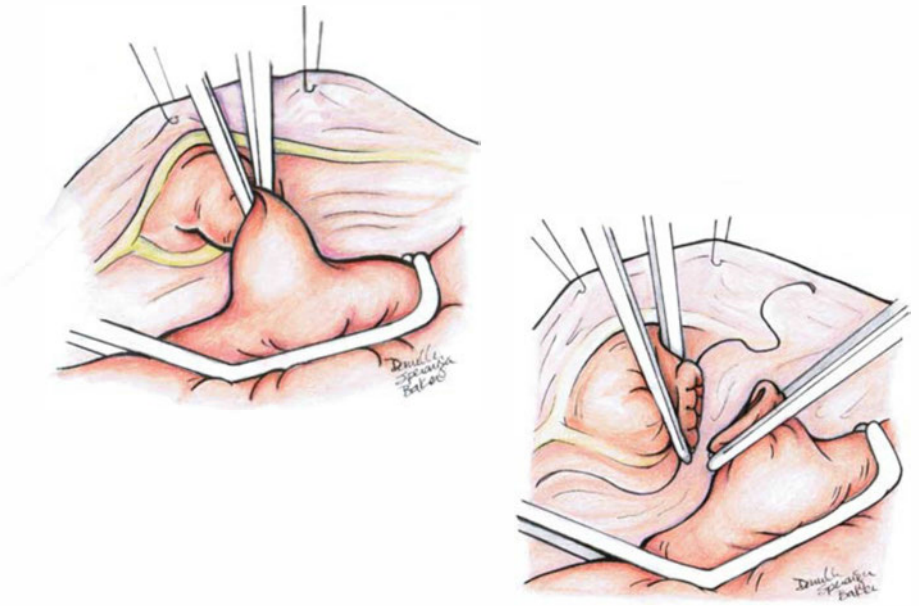


Fig. 77.5. Division and oversewing of large, short PDA. A partially occluding clamp is placed across the ductal origin, then a straight Potts clamp across the mobilized ductus, with care taken to visualize the vagus and recurrent nerves as the clamp is placed. The ductus is divided and each end oversewn. Friable edges can be reinforced with pledgetted mattress sutures if necessary.

A very large and/or short ductus should be divided. After mobilization, a partially occluding clamp is placed across the ductal origin. Alternatively, proximal and distal clamps may be applied on the aorta. Then, a straight, toothed Potts clamp is applied across the pulmonary artery end of the ductus. The ductus is divided and each end oversewn with doubly run prolene suture (Fig. 77.5).

Adults: The ductus is frequently thin-walled and calcified, typically on its aortic end, in adults. Evidence of calcification should be sought by echocardiography or computerized tomography. If the ductus has some length and is not calcified, the standard approach described above can be taken. If the ductus is heavily calcified or is very short, the transpulmonary approach, using a median sternotomy and cardiopulmonary bypass, should be taken. Under moderate hypothermia and reduced flow, the pulmonary artery is opened and the ductal orifice is closed with a polytetrafluoroethylene patch. Fogarty balloon occlusion of the orifice may facilitate patch placement. Circulatory arrest should not be required.

VATS Ligation With and Without Robotic Assistance

The technique of VATS PDA ligation is described by Laborde, Burke, DeCampli, and Nezafati et al.

Premature neonates: Burke performed VATS PDA ligation in a series of premature neonates <800 g weight. He used a 2.7-mm 30-degree angle thoracoscope. Hines et al. performed VATS PDA ligation in 100 preterm infants with a weight range of 420 to 1,500 g.

Full-term infants, children, and adults: VATS PDA ligation can be performed in young infants as well as adults. This approach is suitable for small to moderate-sized ductus. "Giant ductus" or "AP window-type" ductuses are best repaired with an open technique.

The preoperative echocardiogram should be inspected to note the position of the ductus. This will minimize the need for dissection to search for the ductus. Selective lung ventilation is preferred. In infants, a cuffed single-lumen tube can be advanced into the right mainstem bronchus. In older children and adults, a double lumen tube, or a single-lumen tube with a left mainstem bronchial blocker can be used. Single-lung ventilation should be instituted well ahead of the incision time to allow time for lung collapse. Instruments should be available for full thoracotomy, in case conversion is necessary. A unit of blood should be available. The procedure is described in Figures 77.6 and 77.7. At the completion of the ligation, hemostasis is checked. A transesophageal echocardiogram is obtained prior to completion of the procedure so that residual flow can be secured with an additional clip if needed. The instruments are withdrawn and an 8-F pigtail catheter

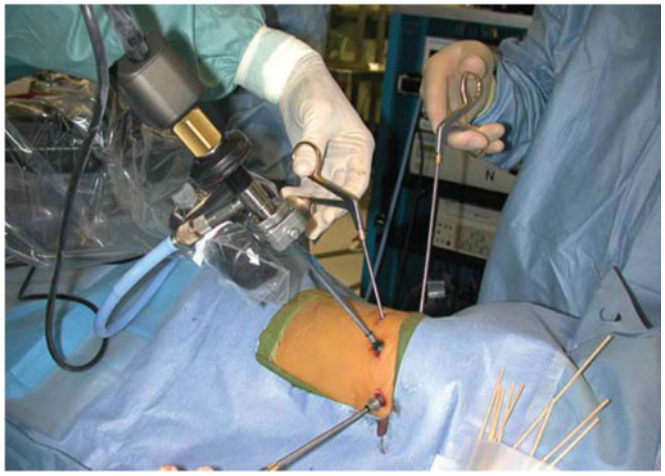


Fig. 77.6. Videoscopic (VATS) PDA ligation, here with robot assistance. Port incisions are made at the third interspace at the anterior axillary line (#1), fourth interspace at the mid-axillary line (#2), fifth interspace just anterior to the scapular tip (#3), and sixth interspace posterior to the scapula (#4). In this procedure, the lung retractor, grasper, camera (here robot manipulated using voice control), and cautery are placed through ports #1, 2, 3, and 4, respectively. The surgeon should be flexible and may move the relative port locations of the instruments during the procedure. With the lung retractor fixed on a bar, the procedure can be performed by a single surgeon.

is placed. The lung is re-expanded under camera guidance. The wounds are closed in layers with absorbable suture. If there is no evidence of pneumothorax or air leak, the drain is removed in the recovery room. The patient is discharged home a few hours later.

If bleeding is encountered during the VATS procedure, and especially if ductal tear is suspected, the procedure should be converted to open. Generally, three or four of the port incisions can be joined together and a thoracotomy performed. A sponge stick can be used through one port to control bleeding while this is done.

Robotic-assisted VATS: DeCampli, at The Children's Hospital of Philadelphia, used AESOP (ComputerMotion, Inc., Santa Barbara, CA), a voice-controlled robot arm, to manipulate the thoracoscope, as described in Figures 77.6 and 77.7. Suematsu et al., at Boston Children's Hospital, used the Da Vinci System (Intuitive Surgical, Sunnyvale, CA) to perform nine PDA clip ligation procedures. The grasper was controlled by the left robotic arm and the cautery tip by the right robotic arm. All patients did well. These cases comprised an intriguing proof of concept that with future improvements in technology may someday allow for superior surgical precision.

COMPLICATIONS OF SURGICAL THERAPY

Premature Neonates

Complications of surgical PDA ligation include pneumothorax, chylothorax, bleeding, aortic obstruction, pulmonary artery obstruction, residual flow, vocal cord dysfunction, wound infection. Treatments of some of these complications are discussed in the prior section. Perhaps the most common, and potentially serious complication is injury to the recurrent laryngeal nerve, resulting in unilateral vocal cord paresis (VCP). In series published since 2008, the incidence in premature infants ranges from 4.8% to 67% (24% to 67% in infants $<$1,000 g at the time of closure). Clement et al. showed that, as compared with matched controls without VCP following PDA ligation, infants with VCP require longer duration of tube feeding (relative risk 9.0; $P = 0.003$), supplemental oxygen, ventilator support, and hospital stay. Benjamin et al. showed that infants with VCP are more likely to develop BPD (82% vs. 39%; $P = 0.002$), reactive airway disease (86% vs. 33%), and need for gastrostomy tube feeding (63% vs. 6%). Truong et al. examined the rate of recovery of VCP in patients undergoing cardiac surgery. They found that among 80 patients who were followed up for a minimum of 3 months after injury, 35% recovered function at median time of 6.6 months and 65% of patients had persistent VCP at median follow-up of 16.4 months. Forty-five percent of patients demonstrated evidence of aspiration and 27% of patients required intervention for feeding difficulties.

Clearly, VCP represents a frequent and serious complication of PDA ligation. The risk increases with lower weight. The risk does not seem to depend on method of ligation (clip vs. ligature). The complication may explain the observed increased incidence of BPD after surgical ligation and may be largely responsible for the failure of studies to show a clear benefit of PDA ligation in the overall outcomes of premature neonates.

Roclawski et al. showed that among 38 patients following PDA ligation, the incidence of scoliosis was 55%, compared with 16% in a matched control. In those with scoliosis, the curve was thoracic in 76% and left-curved in 57%. The magnitude ranged from 10 to 42 degrees.

Some authors have suggested an increased risk of retinopathy of prematurity and neurosensory impairment at 18 months of age in premature infants undergoing PDA ligation. The principal study, however, was observational, the control group was not matched, and covariates were not accounted for in the analyses.

Full-Term Infants and Children

In older infants and children after PDA ligation, reported incidences of residual flow range from 0% to 8%, chylothorax range from 0.2% to 4%, and VCP of 0% to 12%. Aortic obstruction or left pulmonary artery obstruction, presumably due to impingement or misplacement of the ligature or clip, has been reported but occurs rarely.

Adults

Jatene et al. reported on 34 adult patients (aged 18 to 53 years) undergoing open PDA ligation between 1997 and 2008. Only one patient required cardiopulmonary bypass. Eight patients had ductal calcification, and 12 patients had a prior attempt at catheter device closure. Twenty-five patients had ligation and division, eight patients had ligation alone, and one required an aortic patch. Complications included dysphonia (14.7%), hemorrhage (8.8%), residual flow (5.9%), and pneumothorax (5.9%). Three of the patients with dysphonia had documented VCP (one permanent). The authors felt that a risk factor for VCP was a ductus diameter $>$10 mm.

CONCLUSIONS

Moderate-to-large PDA can cause congestive heart failure and, later, PVOD. Any PDA presents a persistent risk of endocarditis or, rarely, aneurysm. The vast majority of PDAs should, therefore, be closed.

In children, options for closure include catheter-based device placement and surgical ligation. Catheter-based and VATS approaches are comparable in safety, effectiveness, and expediency, but VATS may be less costly.

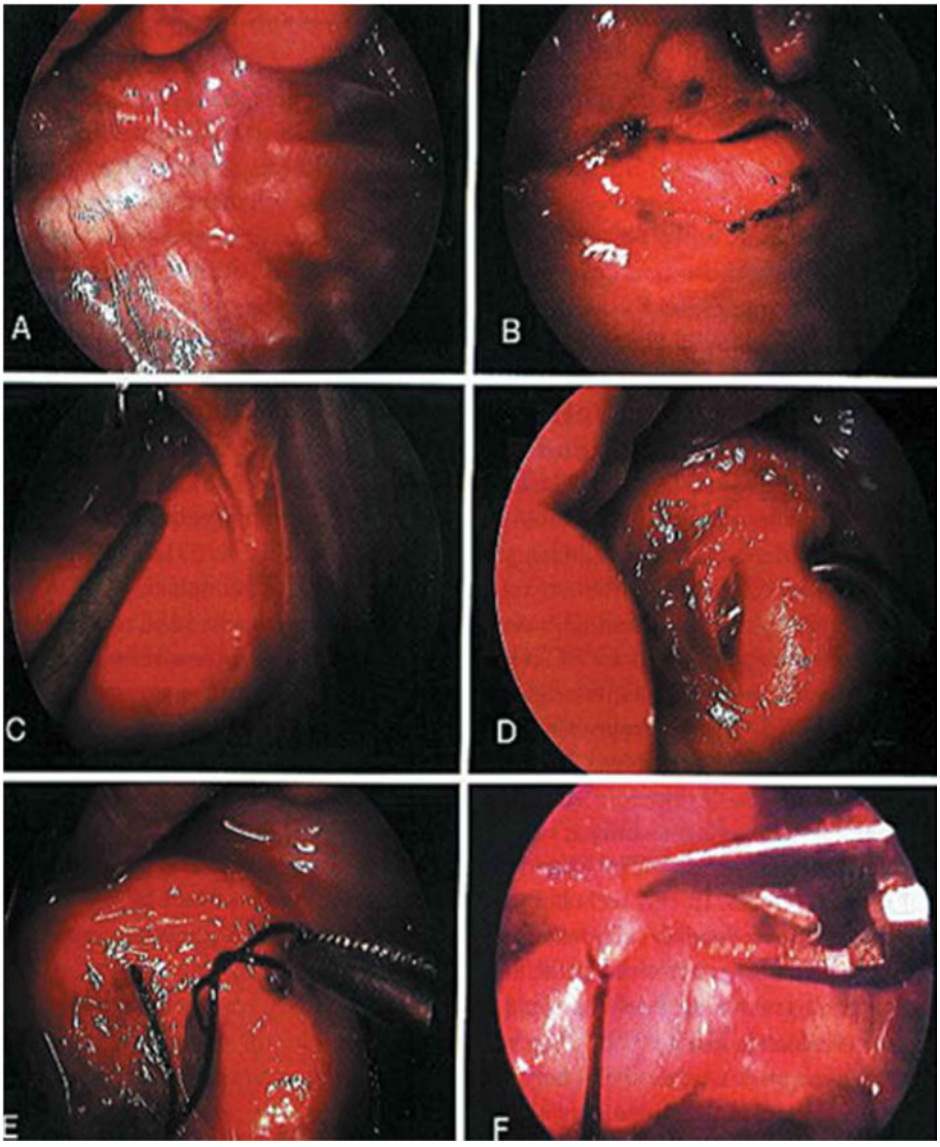


Fig. 77.7. VATS PDA ligation. (**A**) With appropriate lung retraction, the juxtaductal segment of aorta is exposed. (**B**) The pleura over this segment is opened with cautery, superiorly onto the left subclavian artery. Minor bleeding should be suctioned through any one of the ports and controlled with cautery or with temporary q-tip pressure. With repositioning of the lung retractor, the ductus will come into view. (**C**) Using cautery and long-armed q-tips, the ductus is exposed. The recurrent nerve should be visualized. The use of blunt dissection with q-tips reduces the chance of recurrent nerve injury. (**D**) An adequate length of ductus, away from the recurrent nerve, is exposed, then a right-angle clamp is used to mobilize behind the ductus in a cephalad to caudad direction. A suitable forceps should be used under the caudad aspect of the ductus to provide countertraction as the right angle comes through. It is important to keep the right angle heading parallel to the aorta so it does not head more deeply under the ductus. A cotton tip is used to tease the recurrent nerve away from this area as the right angle comes through. One should be able to visualize the recurrent nerve travelling anterior and deep to the clamp jaws. (**E**) A 2-0 or 0-silk tie is passed around the ductus and tied with intracorporeal technique on the pulmonary end of the ductus. (**F**) A manual hemoclip applier is advanced through port # 2. The use of an "endo-clip applier," especially any ratcheted device, is avoided. The clip is applied to the aortic end of the ductus, away from the recurrent nerve. The angle of approach here is important, so the clip does not angle toward the recurrent nerve.

The effect of PDA ligation, and its comparison with COX inhibitor therapy, on outcomes of very premature infants has not been studied adequately. In infants with signs attributable to congestive heart failure, surgical ligation is a reasonable therapeutic measure as it will completely eliminate the cause of congestive heart failure due to the left to right shunt. VCP is a serious complication of surgical PDA ligation in premature infants, and its avoidance should be compulsively addressed by every surgeon performing the procedure.

PDA closure in adults should generally be performed with catheter-based devices, especially in the presence of calcification. Large, short, calcified or infected ducts that cannot be closed with a device should be closed with a patch through the pulmonary artery using cardiopulmonary bypass.

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EDITOR'S COMMENTS

Because the ductus arteriosus is a normal fetal structure representing the left sixth aortic arch, persistent patency of the ductus arteriosus is one of the most common congenital heart defects. Ductal patency seems to be more common in girls, and persistent ductal patency occurs in approximately 1 in 2,000 term births. In premature infants, ductal patency is common. By 1 year of age, only about 1% of ducts remain open, and after 1 year of age, <1% per year spontaneously close. Thus, persistent patency of the ductus arteriosus at 6 months to 1 year of age is generally considered an indication for ductal closure. Certainly, ducts that are large, producing significant left-to-right shunts with congestive heart failure symptoms should be closed promptly.

As discussed in this chapter, the approach to ductal closure remains controversial. Although the incidence of endarteritis associated with ductus arteriosus is low, the occurrence of this problem even in clinically silent ducts (Balzer D, Spray TL, Cantor CE, Strauss AW. Endarteritis associated with a clinically silent patent ductus arteriosus. Am Heart J 1993;125:1192.) suggests that ductal closure should be effected in all patients including those with a clinically silent duct.

Coil occlusion techniques are being used increasingly for ductal closure and have the advantage of avoiding chest incisions and direct dissection of the ductus with attendant risks of recurrent nerve injury or bleeding; however, these techniques are limited somewhat by vascular access in very small children, and in very large ducts, coils may embolize or be difficult to place without protruding into the pulmonary artery or aorta. In addition, an inadequately closed ductus with a metallic foreign body and the attendant potential risk of late endarteritis remain a concern with this technique. If, however, complete ductal closure can be effected with coil occlusion, there appears to be no disadvantage to this approach. Certainly, adults with calcified ducts have a significant enough risk from operative intervention that transcatheter occlusion should be considered primary treatment if at all possible. In older patients, the incisions for thoracoscopy closure or routine thoracotomy closure are relatively large and the morbidity higher, and therefore, transcatheter closure should be considered.

Surgical closure of the ductus arteriosus with clips or simple ligatures has been associated with recanalization rates as high as 20% to 25% in some series (Sorenson KE, Kristensen BO, Hansen OK. Frequency of occurrence of residual ductal flow after surgical ligation by color flow mapping. J Am Coll Cardiol 1991;67:653.). Therefore, despite documented complete closure after thoracoscopic ductal ligation with clips, recanalization must be considered a relative risk. Certainly, the gold standard for ductal occlusion with the lowest incidence of recanalization is ductal division, although the technical aspects of ductal division require larger incisions and far more complete control of the vessel before division. Longer follow-up on the use of transcatheter occluder devices and clip ligation with the thoracoscopic technique should address the issue of late recanalization and residual patency before these techniques completely supplant open division, especially for the significant-sized ductus arteriosus.

Ligation of the ductus arteriosus through a median sternotomy incision is relatively simple, although it is very important to identify the origin of the duct just beyond the take-off of the right pulmonary artery and to adequately dissect the groove between the ductus and the left pulmonary artery before the duct is mobilized for ligation.

As nicely described by Dr. DeCampli, there has been increasing recognition that studies have not clearly identified a benefit of ductal ligation in premature infants with respiratory insufficiency. The limitations of these studies have been well described; however, there has been a relative decrease in the referral of premature infants for duct ligation based on this accumulating body of data. It is interesting, however, that some studies have suggested a lower incidence of necrotizing enterocolitis after duct ligation, which maybe of particular benefit to these infants. It is unclear why the incidence of vocal cord paralysis is so high in patients after PDA ligation. Sweeping the recurrent laryngeal nerve away from the ductus is relatively straight forward in these patients, although clip ligation often leaves the tip of the clip obscure as it is being applied, and it potentially could impinge upon the recurrent laryngeal nerve as it passes behind the ductus and out of the visual field. Surgeons have been increasingly cognizant of this concern and I suspect that the incidence of vocal cord paralysis is somewhat diminished in more recent experience, especially with better visualization techniques as described in this chapter.

The issue of ductal recanalization is a significant one. The use of metallic clips, especially double clip ligation, seems to decrease the risk of this complication, but simple ligation and even simple clip application have been associated with recanalization rates that are not insignificant. In addition, residual or recurrent ductal patency has been shown in catheter-based techniques in a frequency that is not dissimilar to that recorded in surgical series.

TLS

78 Vascular Rings, Slings, and Other Arch Anomalies

Christopher E. Mascio and Erle H. Austin

HISTORICAL ASPECTS

Recognition of anomalies of the aortic arch and pulmonary artery began in 1737 when Hommel first described a double aortic arch. Fifty-seven years later, Bayford discovered an aberrant retroesophageal subclavian artery in a patient with a history of dysphagia. He called the anomaly a "lusus naturae," or "prank of nature," and coined the term "dysphagia lusoria" to describe the symptoms. The first surgical correction of a double aortic arch was performed by Gross in 1945 on a 1-year-old boy with chronic wheezing. The pulmonary arterial sling anomaly was described by Glaevecke and Doehle in 1897 and was first repaired by arterial division with reimplantation by Potts in 1954.

EMBRYOLOGY

By 5 weeks of fetal development, the primordial heart tubes have fused and six aortic (branchial) arches have formed between the ventral roots and dorsal aortae (Fig. 78.1). Migration and involution of the arches result in the complex system of the higher mammal. During normal development, persistence of the left fourth aortic arch forms the arch of the aorta between the left common carotid and left subclavian arteries. The right fourth aortic arch forms the proximal right subclavian artery. Involution of the distal right aorta results in an unpaired single aortic arch. Table 78.1 shows the fate of the remaining arches.

Failure of involution or migration of specific segments of the aortic arches results in anomalies that may form partial or complete vascular rings about the trachea and esophagus. The most common of these are diagrammed in Fig. 78.2.

Pulmonary artery development arises from two separate vascular sources: (1) the lung buds deriving their blood supply from the splanchnic plexus and (2) the proximal left and right sixth aortic arches (Fig. 78.3). Figure 78.4 demonstrates how abnormal migration of the separate blood supplies can result in dislocation of the left pulmonary artery posterior to the trachea, resulting in the pulmonary artery sling anomaly with its associated tracheal compression.

CLASSIFICATION AND INCIDENCE

Aortic arch anomalies producing tracheoesophageal constriction account for 1% to 2% of all congenital heart defects. A simple and commonly used system of classifying these vascular anomalies is given in Table 78.2. The relative prevalence of these lesions in surgical series is also indicated.

Group I: Complete Vascular Rings

Anatomy

A double aortic arch (type IA) occurs when the right dorsal aorta fails to involute (Fig. 78.2), resulting in persistence of the right and left fourth aortic arches. The right aortic arch is most commonly dominant. The descending aorta is usually in its normal position on the left, as is the ligamentum arteriosum (or patent ductus arteriosus). Generally, each subclavian and common carotid artery arises independently from its respective aortic arch. The innominate vessels do not develop. Twenty percent of cases have other cardiac anomalies, with ventricular septal defect and tetralogy of Fallot being most common.

Type IB vascular rings (right aortic arch with retroesophageal ligamentum arteriosum) account for 45% of complete vascular rings. The majority of these defects have left descending aortas. A right aortic arch with retroesophageal left subclavian artery and ligamentum results from abnormal involution of the left fourth arch. A remnant or stump of the left fourth arch (Kommerell's diverticulum) may persist. The left subclavian artery arises aberrantly behind the esophagus from the right-sided arch or from Kommerell's diverticulum. Near this point, the left-sided ligamentum emanates and joins the left pulmonary artery to create a complete ring about the trachea and esophagus. Another variant of right aortic arch with left descending aorta is called the circumflex aorta. This variant is characterized as having a hypoplastic retroesophageal arch and a coarctation (Fig. 78.9A and 78.9B).

A right aortic arch may occur with mirror image branching with the left innominate artery arising anteriorly and dividing into the left carotid and subclavian arteries. If the ligamentum arteriosum occurs between the left innominate artery and the left pulmonary artery, as is commonly seen in association with tetralogy of Fallot, there is no vascular ring. Therefore, these patients are usually asymptomatic. However, when the ligamentum arises from the aorta behind the esophagus and connects with the left pulmonary artery, a compressive vascular ring is formed.

A rare variant among vascular rings is the cervical aortic arch, in which case the arch exists in the neck above the clavicle, sometimes as high as the C2 vertebral body. There are two main subtypes. The first and the larger group are type IB anomalies with a right-sided arch, an anomalous left subclavian artery, and a retroesophageal ligamentum arteriosum. The second group has a left-sided arch with a normal branching pattern and thus does not form a complete ring. These anomalies are thought to result from a failure of the normal descent of the aortic arch from its cephalic location at 3 weeks to its intrathoracic location at 7 weeks of gestation.

Clinical Presentation

The clinical presentation of vascular rings can vary from mild dyspnea or dysphagia to respiratory distress and apnea. Signs and symptoms often depend on the type of anomaly.

A double aortic arch produces the earliest and most severe symptoms as a result of the presence of a tight ring around

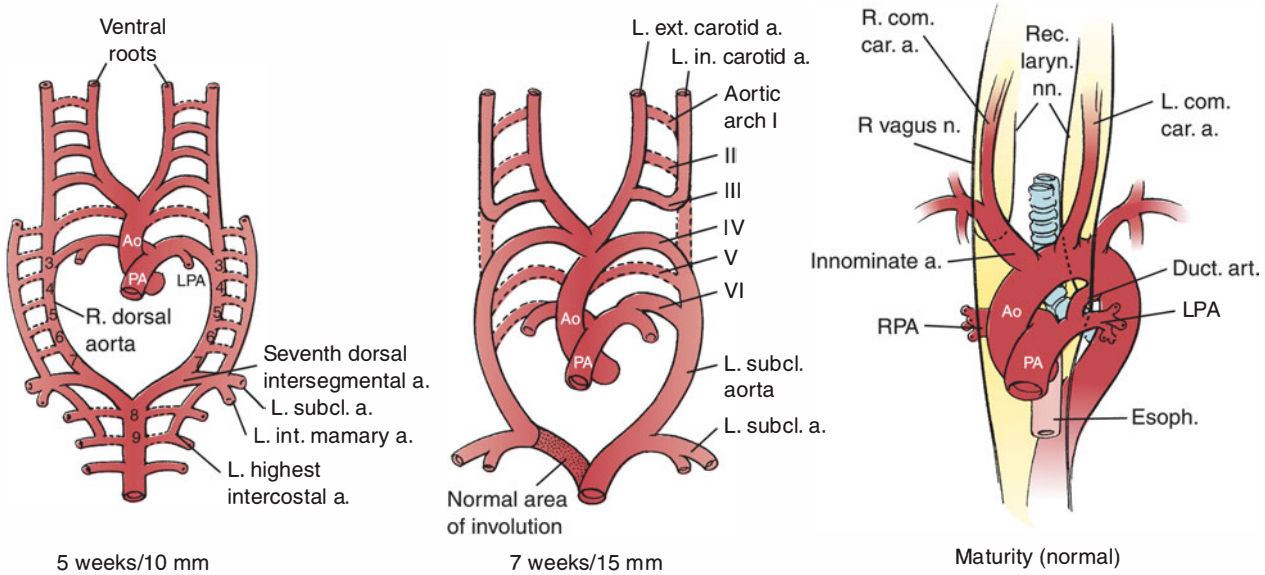


Fig. 78.1. Normal aortic arch development. The normal aortic arch arises from the persistence of the left fourth embryologic arch. Note that the recurrent laryngeal nerve recurs about the derivative of the sixth embryologic arch on the left, that is, the ductus arteriosus, and the derivative of the fourth embryologic arch on the right, that is, the right subclavian artery. This occurs because of the normal involution of the right sixth embryologic arch. a., artery; Ao, aorta; car., carotid; com., common; Duct. art., ductus arteriosus; Esoph., esophagus; ext., external; int., internal; L., left; LPA, left pulmonary artery; n., nerve; PA, pulmonary artery; R., right; Rec. laryn. nn., recurrent laryngeal nerves; RPA, right pulmonary artery; subcl., subclavian.

Table 78.1 Outcome of the Embryonic Aortic Arches

Embryonic vessel	Outcome
1. Truncus arteriosus	Proximal ascending aorta and pulmonary root
2. Aortic sac	Distal ascending aorta, brachiocephalic artery, and arch up to the origin of the left common carotid artery
3. First arch	Portions of the maxillary artery
4. Second arch	Portions of the stapedial artery
5. Third arch	Common carotid artery and proximal internal carotid artery
6. Fourth arch	
Right	Proximal right subclavian artery
Left	Aortic arch segment between the left common carotid and left subclavian arteries
7. Fifth arch	No known derivations
8. Sixth arch	
Right	Proximal part becomes proximal segment of the right pulmonary artery; distal portion involutes
Left	Proximal part becomes proximal segment of the left pulmonary artery; distal portion becomes ductus arteriosus
9. Right dorsal aorta	Cranial portion becomes right subclavian artery distal to contribution from right fourth arch; distal portion involutes
10. Left dorsal aorta	Aortic arch distal to left subclavian artery
11. Right seventh intersegmental artery	Distal right subclavian artery
12. Left seventh intersegmental artery	Left subclavian artery

the trachea and esophagus. Seventy-five percent of affected patients develop symptoms by 1 year of age, often within the first month of life and rarely after 6 months of age. Stridor, a nonproductive cough, or a hoarse cry may be noted soon after birth. The stridor generally worsens with feedings, especially solid foods. The cough is characterized as a "seal bark" or "brassy cough." Respiratory distress can lead to choking, cyanosis, and apnea. Vomiting may precede or follow choking. Despite the esophageal compression, most infants with double aortic arch tolerate liquids well and appear well fed.

When tracheoesophageal compression results from a right aortic arch and a retroesophageal ligament, symptoms are similar in nature but are usually less severe and present later in infancy or in early childhood. These rings are generally less stenotic and become notable as the aorta grows.

Those with a right arch and left descending aorta (circumflex aorta) may present with murmur, upper extremity blood pressure differential, exertional dyspnea, and/or chest pain.

Patients with cervical arches are usually asymptomatic but may present with a pulsatile mass in the neck or the

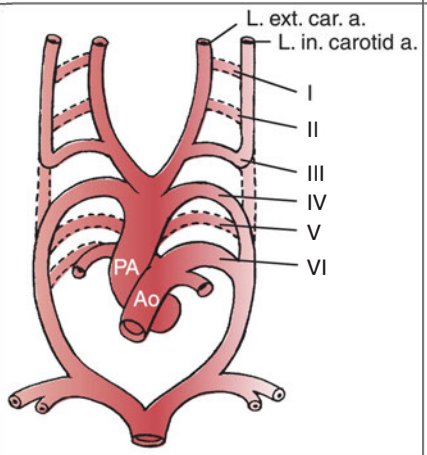
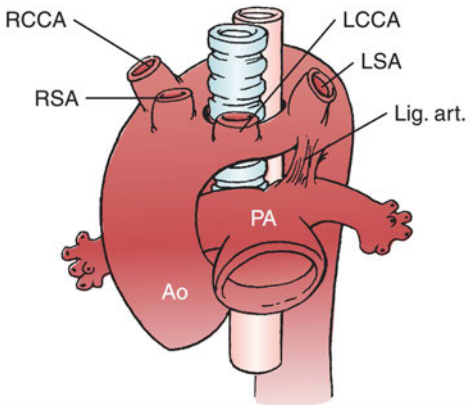
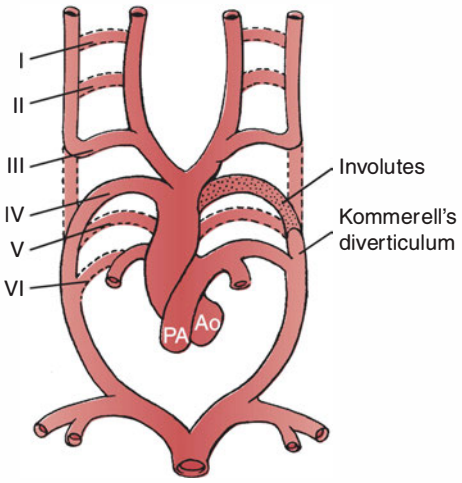
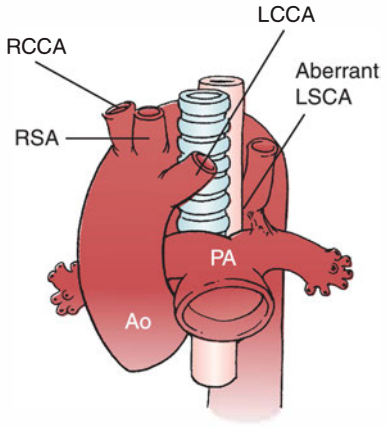
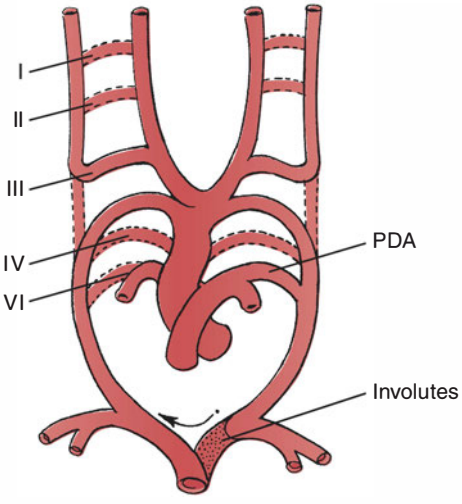
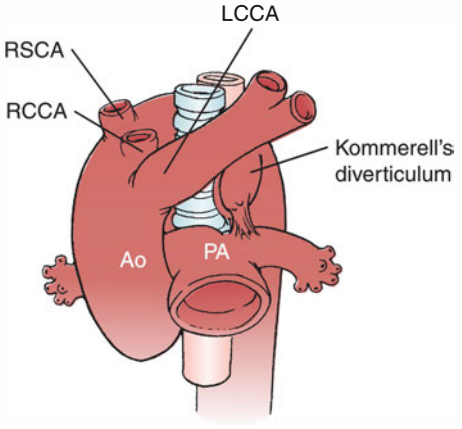
Anomaly	7-wk/15-mm embryo	Maturity
Double aortic arch Right arch dominant Left arch dominant Arches equal		
Right aortic arch with retroesophageal left and subclavian artery and left ligamentum arteriosum		
Right aortic arch with mirror image branching and retroesophageal left ligamentum arteriosum		

Fig. 78.2. Vascular rings most frequently requiring surgical intervention and their embryologic development. Kommerell's diverticulum is a remnant of the involuted left fourth aortic arch, which may be the origin of the ductus (ligamentum), the left subclavian artery, or both. a., artery; Ao, aorta; L. ext. car. a., left external carotid artery; L. int. carotid. a., left internal carotid artery; LCCA, left common carotid artery; Lig. art., ligamentum arteriosum; LSA, LSCA, left subclavian artery; PA, pulmonary artery; PDA, patent ductus arteriosus; RCCA, right common carotid artery; RSA, RSCA, right subclavian artery; roman numerals refer to embryologic aortic arches.

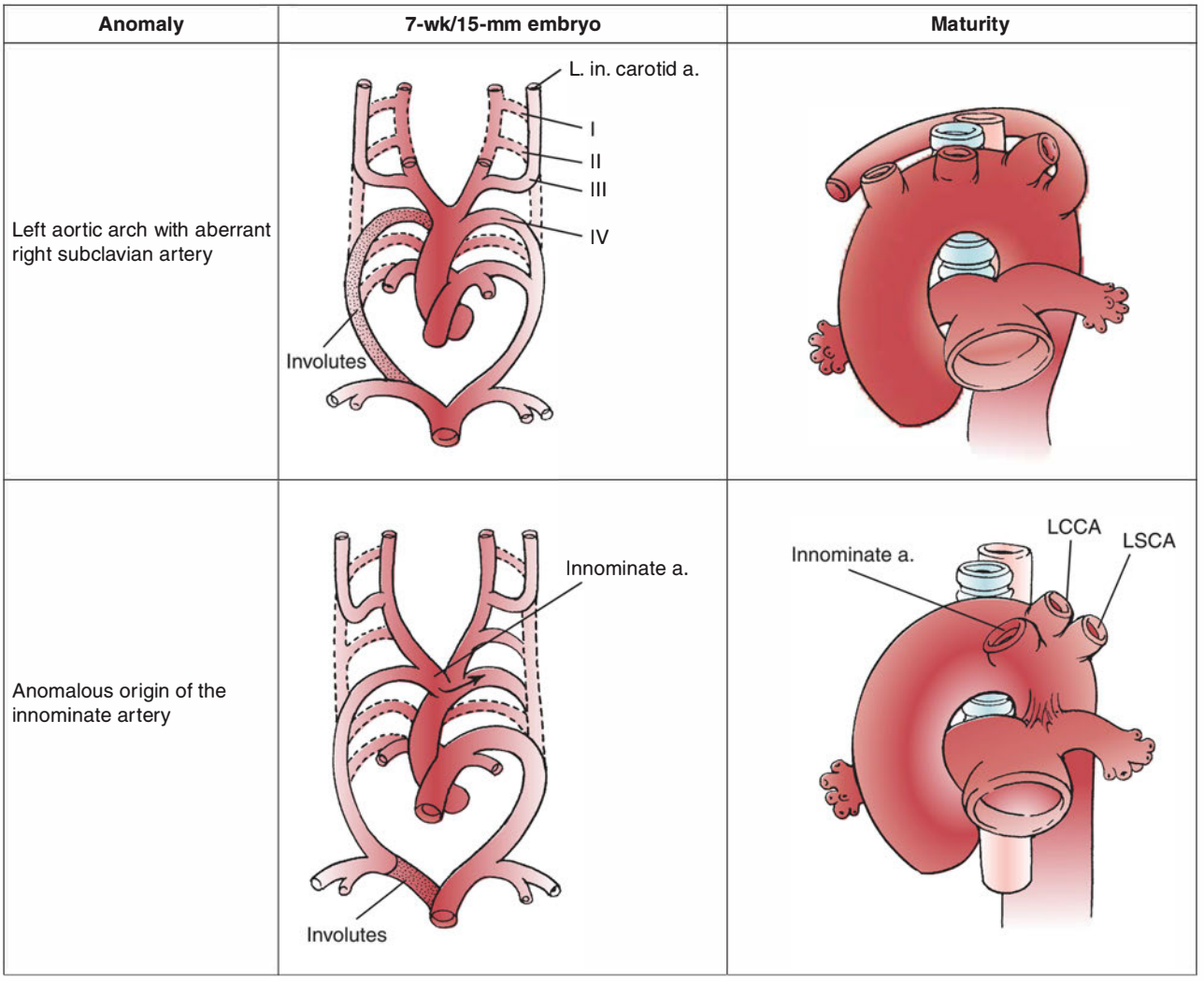


Fig. 78.2. (*continued*)

supraclavicular fossa. Infants with the type IB anomaly may also exhibit the compressive symptoms of dyspnea and stridor. Adults more commonly present with dysphagia. Some adults have presented with central nervous system symptoms and the subclavian steal syndrome as a result of stenoses of the left subclavian artery and origin of the vertebral artery.

Diagnosis

A vascular ring may be suggested on chest radiography by the presence of a pulmonary infiltrate, atelectasis, or unilateral or bilateral hyperinflation. The presence of a right aortic arch should raise suspicion.

Barium esophagogram remains a useful diagnostic measure. In cases of double aortic arch, the lateral view will reveal a posterior indentation in the esophagus. In the anterior projection, there are dual indentations, higher on the right and lower on the left. An aberrant right subclavian artery from a left aortic arch will also produce posterior esophageal indentation on the lateral view. On the anterior view, however, the aberrant right subclavian artery takes an oblique course descending left to right. The aberrant retroesophageal left subclavian artery from a right aortic arch will show a similar posterior indentation on the lateral view, but the oblique course on the anterior view runs in the opposite direction: from right to left. A circumflex aorta (right arch and left descending aorta) will have an impression on the right side of the esophagus on the anterior view and a deep, posterior indentation on the lateral view.

Historically, the clinical presentation and barium esophagogram have provided sufficient information to proceed to surgical treatment. The increasing availability and sophistication of magnetic resonance imaging (MRI) and computed tomography (CT), however, have elevated these noninvasive imaging techniques to the preferred diagnostic modality at most institutions. Both modalities provide detailed and complete depiction of the anatomy and compressive effects of a vascular ring. MRI, however, is expensive and requires sedation in the younger patients who are at the greatest risk from airway compression. Without sedation, motion artifacts frequently result in poor image quality. Traditional CT scanning with three-dimensional reconstruction can also provide excellent images

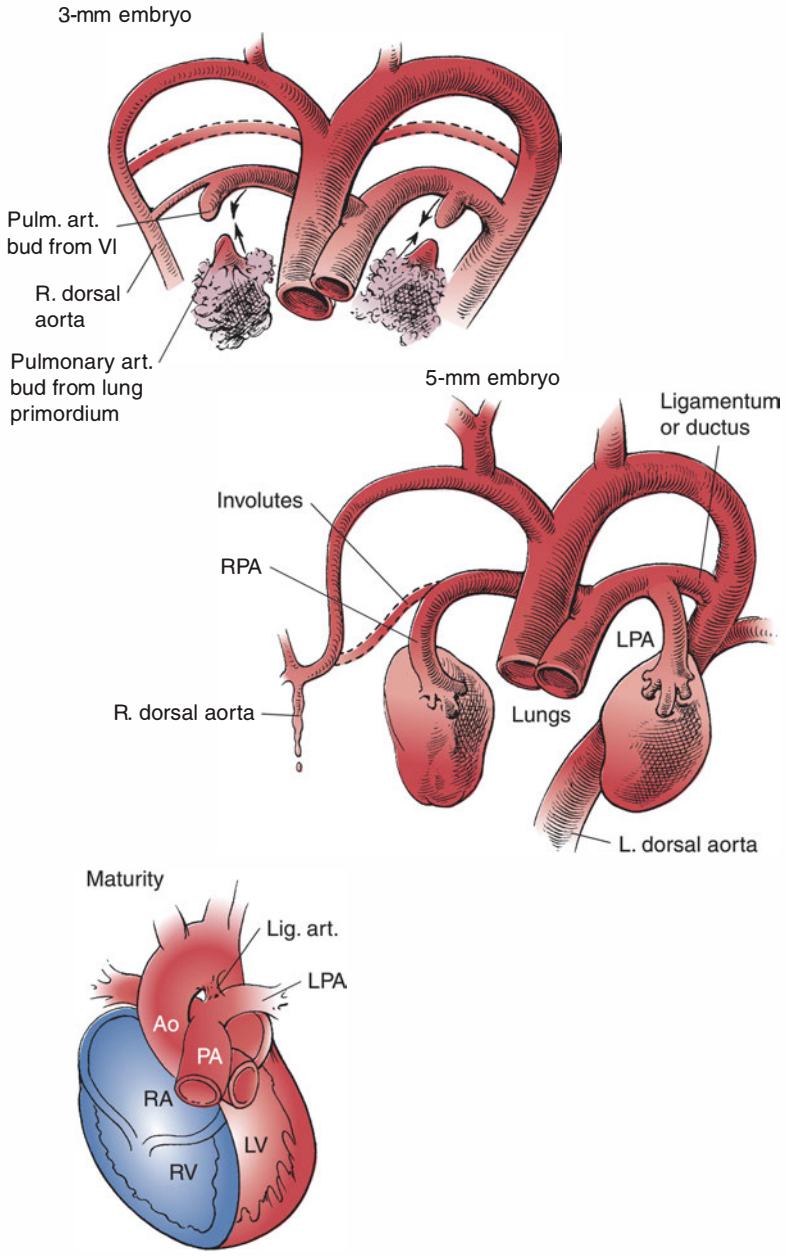


Fig. 78.3. Normal pulmonary artery development. The pulmonary arteries derive from two separate vascular buds, the sixth aortic arches (*left and right*) and the splanchnic plexus. These buds fuse to form the left and right pulmonary arteries. Ao, aorta; L., left; Lig. art., ligamentum arteriosum; LPA, left pulmonary artery; LV, left ventricle; PA, Pulm. art., pulmonary artery; R., right; RA, right atrium; RPA, right pulmonary artery; RV, right ventricle.

but is limited by the need for intravenous contrast and exposure to radiation and may also require sedation for adequate imaging. Rapid, multislice, helical CT scanning (if available) permits rapid scanning with significantly decreased radiation exposure. These scans can be done in <2 seconds in infants, obviating the need for sedation. Aortography is the most invasive technique and is rarely used. It can establish the completeness of a double aortic arch and identify areas of luminal irregularity. It cannot, however, distinguish between a double aortic arch with an atretic segment and a right aortic arch with a retroesophageal ligament. Transthoracic echocardiography is used to detect associated cardiac anomalies and is reasonably sensitive in evaluating vascular rings but appears deficient in determining atretic and nonluminal segments. Echocardiography is important in defining any other associated cardiac anomalies. As mentioned above, these are present in up to 20% of patients and are more common in association with right aortic arch (compared with double aortic arch).

Cervical arches may be suspected by the presence of a wide mediastinum, the absence of the aortic knob, and anterior tracheal deviation on chest radiography. Although angiography has been the standard for diagnosis, CT scanning and MRI now provide a noninvasive technique to confirm the diagnosis.

Indications for Surgery

Surgical correction of complete vascular rings is indicated when symptoms are present. There is no evidence that surgery has any benefit in the absence of symptoms, thus asymptomatic patients (children or adults) should not be exposed to the operative risk. Adults with minimal symptoms should be followed for significant progression prior to operative repair. Anomalies identified shortly after birth or within the first 6 months of life require surgical correction much more frequently than those discovered after the age of 6 months. Very mild symptoms presenting in the older infant or young child may resolve as the child grows. A watchful approach is appropriate in such cases.

Surgical Technique

Ninety percent of vascular ring cases can be corrected through a left posterolateral thoracotomy. Single-lumen general endotracheal anesthesia is applied.

Double aortic arch: In cases of double aortic arch, where flow is present in both arches, the dominant arch is preserved. Division is performed at the location on the ring that preserves the greatest brachial and cephalic flow. When an atretic segment of the vascular ring is present, division is performed at the atretic segment. Before vascular division, the carotid and radial pulses are monitored by the anesthesiologist to confirm flow after clamps have been placed.

After induction of general orotracheal anesthesia, the infant is placed in a full lateral position with the left side up (Fig. 78.5). Through a posterior lateral muscle-preserving incision, the left chest is entered through the third or fourth intercostal space. The lung is retracted anteriorly and inferiorly, exposing the posterior mediastinum. This position is retained with malleable retractors secured to the chest retractor with nonpenetrating towel clamps.

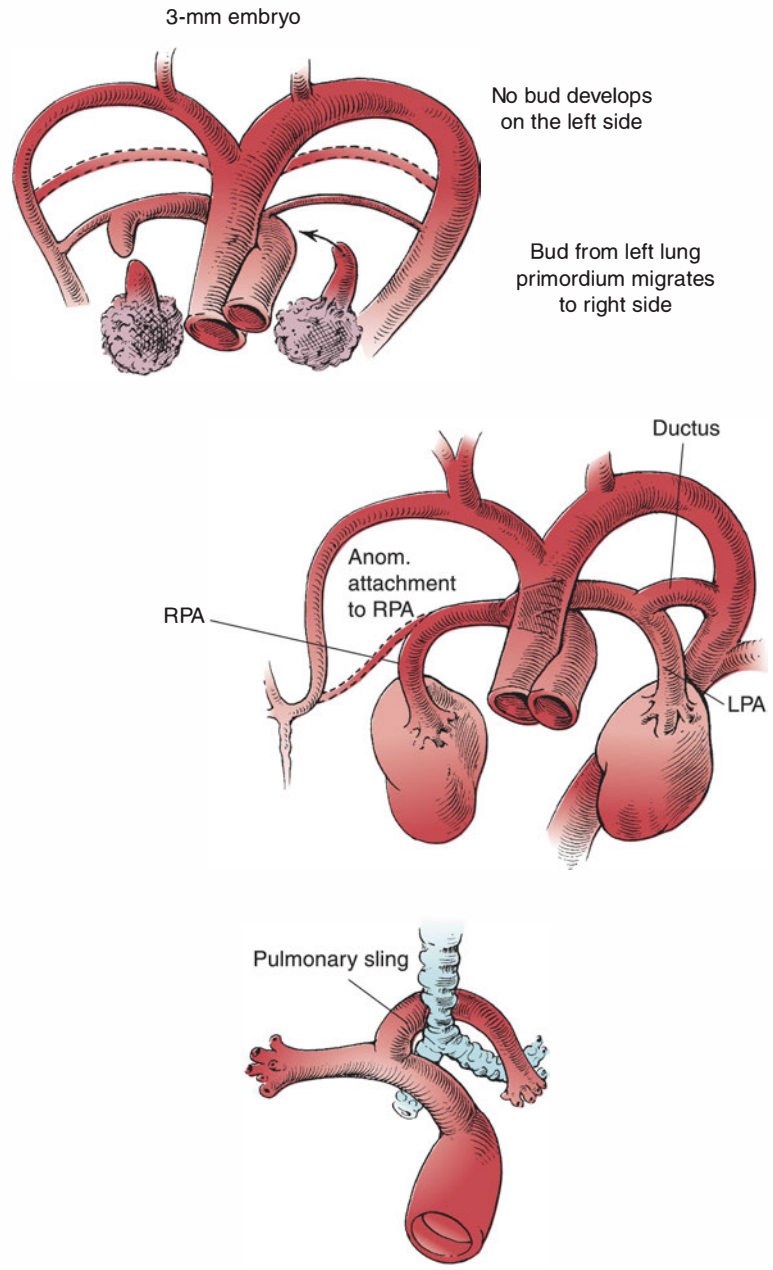


Fig. 78.4. Anomalous development of the left pulmonary artery and formation of the pulmonary artery sling. When the splanchnic bud of the left pulmonary artery fails to fuse with the left sixth aortic arch, it migrates posteriorly between the trachea and esophagus to fuse with the right pulmonary artery. This results in a compressive "sling" about the trachea. Anom., anomalous; LPA, left pulmonary artery; RPA, right pulmonary artery.

The descending aorta, anterior (left) aortic arch, left subclavian artery, and the vagus and phrenic nerves are identified before the pleura is opened (Fig. 78.5). The vagus nerve descends anterior to the left subclavian artery, crossing the left aortic arch, giving off the recurrent laryngeal nerve as it penetrates into the mediastinum medially around the ligamentum arteriosum. The posterior (right) aortic arch may or may not be visualized initially depending on the extent of mediastinal fat.

The pleura is incised from the descending aorta inferiorly to the left subclavian artery, and then elevated from the mediastinum anteriorly and posteriorly with stay sutures. The vagus nerve is elevated off the mediastinum with the anterior pleural flap exposing the branching of the recurrent laryngeal nerve. If necessary, the vagus and recurrent laryngeal nerves may be mobilized with the posterior pleural flap to achieve better exposure of the proximal anterior arch and prevent nerve injury. This is similar to the exposure of a patent ductus arteriosus. The ligamentum arteriosum (or ductus arteriosus) is identified between the distal anterior (left) aortic arch and the left pulmonary artery. The ligamentum is sharply dissected out in its entirety in preparation for ligation and division in standard manner. The posterior pleural flap is gently elevated off the mediastinum. With this, the posterior (right) aortic arch becomes visible as it penetrates the mediastinum posterior to the esophagus and joins the left (anterior) aortic arch to form the descending aorta. Opening the pleura extensively in this manner will provide wide exposure to the mediastinal structures. The anterior arch is dissected to the level of the left common carotid artery. The left subclavian artery is mobilized distally toward the thoracic outlet. The posterior arch is fully isolated from its mediastinal attachments, and all adhesive bands are sharply divided. At this point, the narrowest portion of the vascular ring is identified. In most cases, the narrowest segment will involve the anterior (left) arch either between the left carotid and left subclavian arteries (Figs. 78.6 and 78.7) or between the left subclavian artery and the posterior (right) arch. In 20% of cases, the posterior arch is smaller and can be divided via left thoracotomy at its junction with the anterior arch. If both arches are of equal size, division is planned for the most accessible point in the ring: usually between the left subclavian artery and the posterior arch.

The ligamentum arteriosum (ductus arteriosus) is divided in standard manner. Once the appropriate site for ring division is identified, vascular clamps are applied and all radial and carotid pulses confirmed. Intraoperative neuromonitoring, especially transcranial Doppler, provides immediate feedback about cerebral blood flow after clamping and prior to vascular division. The vessel is then divided and each end is closed with a running 4-0 polypropylene suture. Clamps are slowly removed to assure hemostasis. Dissection of the posterior arch in its course through the posterior aspect of the mediastinum is performed to allow for retraction of the posterior arch away from the posterior wall of the esophagus. Final inspection for hemostasis is performed and the pleura is closed. A single chest tube is then placed posteriorly along the mediastinum into the apex of the left chest.

When MRI or CT scan clearly shows that the right (posterior) arch is smaller than the left, a right thoracotomy is the

Table 78.2 Classification of Vascular Rings and Pulmonary Artery Sling

Classification	Relative prevalence in surgical series (%)
Group I. Complete vascular rings	**75**
IA. Double aortic arch	55
Right arch dominant	80
Left arch dominant	15
Arches equal	5
IB. Right aortic arch/retroesophageal ligament	45
Aberrant left subclavian artery	70
Mirror image branching	30
IC. Left aortic arch/right descending aorta/tight ligament	<1
Group II. Partial vascular rings	**20**
Left aortic arch	100
Aberrant right subclavian artery/left ligament	20
Innominate artery compression	80
Group III. Pulmonary artery sling	**5**

preferred approach. The smaller posterior arch is easily accessed through the right chest. The divided posterior segment of the arch does not retract behind the spine (as it does through a left thoracotomy) but remains readily controllable should significant bleeding occur.

Right aortic arch with retroesophageal ligament: In cases of right aortic arch with a retroesophageal ligament, the surgical approach is through a left thoracotomy. Division of the ring is achieved by division of the ligament. Through a posterolateral muscle-sparing thoracotomy, the vagus nerve is identified in the pleura overlying the mediastinum. The pleura is opened from the proximal descending aorta to the left subclavian artery posterior to the vagus nerve. The anterior flap of pleura is elevated to expose the left pulmonary artery, at which point the ligamentum arteriosum is identified. Several different potential origins of the ligament exist: (1) from a Kommerell's diverticulum off the posterior (right) arch, (2) directly from the right posterior arch, and (3) from the retroesophageal left subclavian artery. Dissection of the ligamentum is performed to the point of its origin while the recurrent laryngeal nerve is protected. The ligamentum arteriosum is divided in standard manner. All adhesive bands are released. In most cases, it is not necessary to excise the diverticulum as pexy to the prevertebral fascia usually prevents recurrent symptoms. Routine excision of Kommerell's diverticulum is advocated by some groups as it has been known to cause both independent tracheoesophageal compression and recurrent symptoms (Fig. 78.8A and 78.8B). With adequate mobilization, the structures will retract, releasing impingement on the trachea and esophagus. When Kommerell's diverticulum appears large or dilated, it should be resected to prevent subsequent independent compression of the trachea and esophagus. In these cases, transfer of the left subclavian artery to the left carotid artery or aorta is performed to relieve any sling-like effect of the posterior arch and left subclavian artery on the trachea (Fig. 78.8C).

Vascular rings requiring right thoracotomy: A right thoracotomy is indicated in correction of vascular rings in 10% to 20% of cases. As discussed, the most common of these indications is the double aortic arch with dominant left arch. Rarer cases include (1) left aortic arch with a right descending aorta and right ligamentum arteriosum between the right descending aorta to the right pulmonary artery and (2) left aortic arch with aberrant right subclavian artery and right ligamentum arteriosum. The surgeon should be alerted to these rare cases by a distinctive esophagogram demonstrating significant indentation high in the upper left posterior aspect of the esophagus. MRI and CT will also identify these cases. Similar principles apply to correct these anomalies. The recurrent laryngeal nerve recurs around the right-sided ligamentum arteriosum. Division of the ring is achieved by division of the ligamentum arteriosum. Isolation of the ligamentum arteriosum and caution with dissection of the recurrent laryngeal nerve are applied. Dissection of the arch of the aorta through the mediastinum posterior to the esophagus is performed to divide adhesive bands to the posterior wall of the esophagus.

Vascular rings requiring median sternotomy: Median sternotomy accounts for approximately 5% of surgical approaches to vascular rings. This approach is used when the vascular ring is associated with correction of another cardiac defect such as tetralogy of Fallot, in which 20% to 25% of cases have an associated right-sided arch. In most cases of tetralogy with right aortic arch, however, there is mirror image branching with the ligamentum arteriosum between the left innominate artery and left pulmonary artery and thus no vascular ring.

Surgical repair for cervical arches is rarely indicated but may be required for complications such as compressive symptoms, arch hypoplasia, and aneurysm of the arch. The surgical approach in these cases is dictated by the anatomy of the complicating feature and may range from division of the ligamentum arteriosum to aneurysm resection with graft interposition.

For a right arch with left descending aorta and associated arch hypoplasia and coarctation (the circumflex aorta), the approach is via median sternotomy and includes wide mobilization of the ascending aorta, retroesophageal arch (and all associated branches), and proximal descending aorta. The entire circumflex retroesophageal arch (and coarctation site if present) is excised and an anastomosis is performed between the distal ascending aorta and the descending aorta (Fig. 78.9A and 78.9B). The anastomosis is anterior to the trachea and esophagus. Although this technique can be performed off-pump, we recommend support with cardiopulmonary bypass. An alternative repair more suitable for an adult is an extra-anatomic bypass between the ascending and descending aorta via sternotomy.

Group II: Partial Vascular Rings

Partial vascular rings include (1) left aortic arch with retroesophageal right subclavian artery and left ligamentum arteriosum and (2) leftward origin of the innominate artery resulting in anterior tracheal compression.

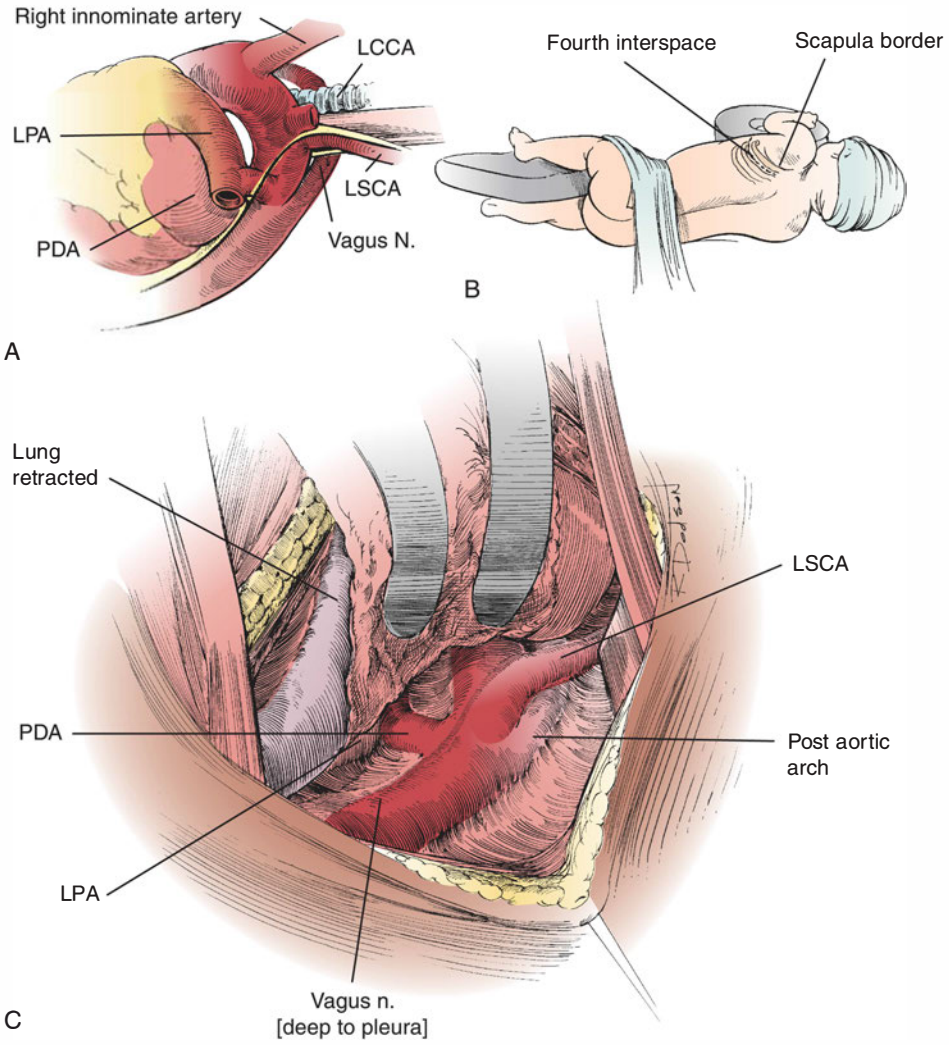


Fig. 78.5. Surgical approach, exposure, and anatomy of a complete vascular ring. (**A**) The patient is placed in the right lateral decubitus position with an axillary roll. The right leg is bent at the hip and knee. Legs are separated by pillows, and the left leg is straight. The incision is a gentle S-curve, at the tip of the scapula. With a muscle-sparing technique, the chest is entered through the fourth interspace. (**B**) The chest retractor is placed with the crossbar anteriorly. The lung is retracted anteriorly and inferiorly and is held into position with a moistened sponge secured with malleable retractors that are fixed to the retractor crossbar. (**C**) Before the pleura is incised, note the position of the vagus nerve, aorta, left pulmonary artery, and, if possible, the posterior (*right*) arch and ligamentum arteriosum. LCCA, left common carotid artery; LPA, left pulmonary artery; LSCA, left subclavian artery; n, nerve; PDA, patent ductus arteriosus.

Left Aortic Arch with Aberrant Right Subclavian Artery

The anomaly of left aortic arch with aberrant right subclavian artery is the most common anomaly of the aortic arch, found in 0.5% to 1.8% of the population. Most patients are asymptomatic. It forms as a result of involution of the entire right dorsal aorta. The right subclavian artery migrates posteriorly behind the esophagus as the left subclavian artery migrates cranially. Coarctation is often associated with this lesion. When symptoms do occur they are most commonly related to difficulty in swallowing. A barium esophagogram is usually sufficient for diagnosis. Surgery is rarely indicated for this anomaly. When symptoms persist, correction consists of left thoracotomy and mobilization and division of the retroesophageal right subclavian artery. An alternative approach favored by some surgeons is a right thoracotomy with division of the right subclavian artery and reimplantation into the right carotid artery or aorta.

Anomalous Innominate Artery with Tracheal Compression

Anatomy

This entity is described in patients with a normal left aortic arch and left-sided ligamentum arteriosum. In these patients, the innominate artery arises either partially or completely to the left of the trachea and in its course from left to right produces anterior compression on the distal trachea. The degree of tracheal compression varies considerably, with only a small proportion of patients developing symptoms of airway obstruction.

Clinical Presentation

Innominate artery compression occurs most commonly in children younger than 2 years of age. Primary symptoms are respiratory and include repeated bronchopulmonary infections, stridor, and apnea.

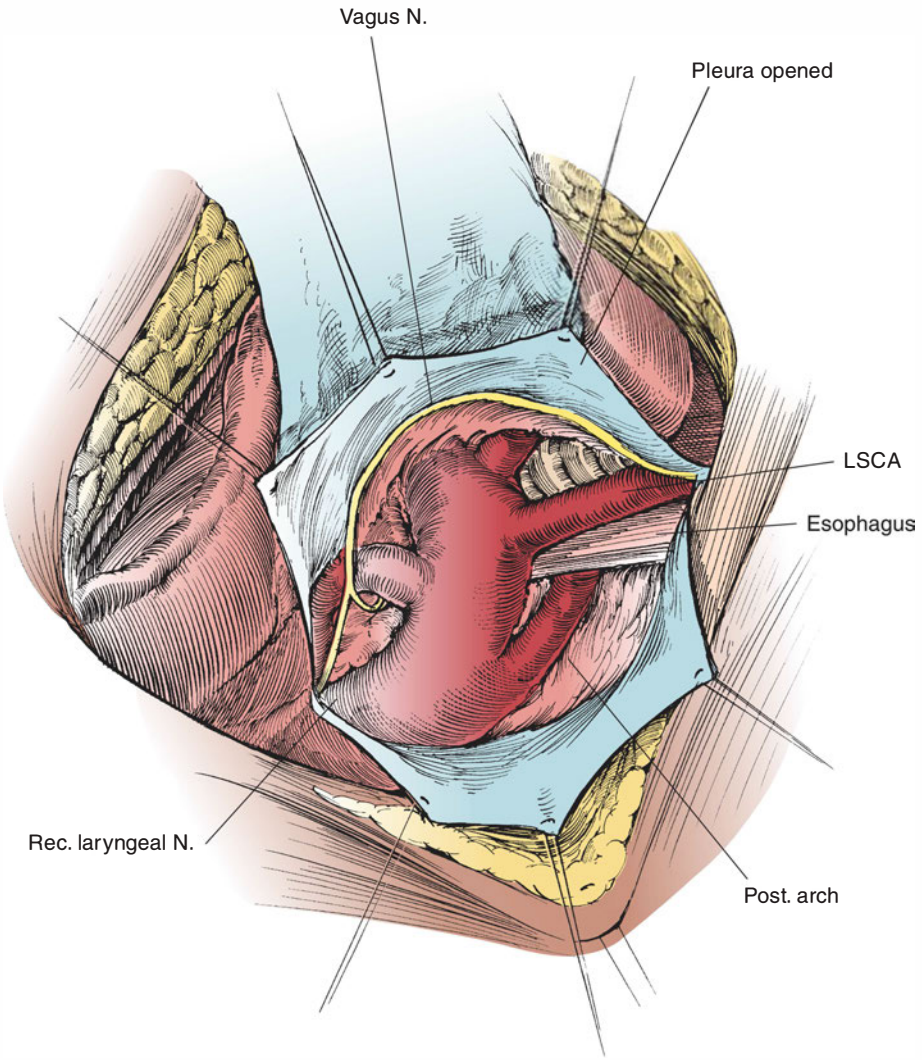


Fig. 78.6. Anatomy of a double aortic arch, right arch dominant. The vagus and recurrent laryngeal nerves are mobilized with the anterior pleural flap in this view. These structures may be mobilized with the posterior pleural flap if the division of the anterior (left) arch is expected proximal to the origin of the left common carotid artery. This will allow proximal exposure without undue tension on these nerves. Ant., anterior; Esoph., esophagus; L, left; LCCA, left common carotid artery; LSCA, left subclavian artery; n., nerve; PDA, patent ductus arteriosus; R., right; RCCA, right common carotid artery; Rec., recurrent; RPA, right pulmonary artery; RSCA, right subclavian artery.

Diagnosis

Diagnosis of innominate artery compression is best obtained with bronchoscopy, during which an anterior pulsatile indentation is noted compressing the trachea 1 to 2 cm above the carina. The trachea should be narrowed at least 50% to 75% for the symptoms to be attributed to this anomaly. The diagnosis can also be made with MRI or CT angiography.

Indications for Surgery

Surgical therapy is dictated by the severity of symptoms. Patients with mild symptoms are managed conservatively and allowed to improve with growth. Patients with a history of apnea, severe stridor, or respiratory distress or two or more episodes of bronchopneumonia or tracheobronchitis are candidates for surgery if significant tracheal compression is demonstrated.

Surgical technique: Several techniques for correction of innominate artery compression have been described. Mustard's technique requires division of the artery overlying the trachea. Langlois's technique involves division of the artery and reimplantation on the ascending aorta. We prefer Gross's technique of innominate artery suspension.

Although Gross used a left anterior thoracotomy, we prefer a right anterior thoracotomy through the second intercostal space (Fig. 78.10). The right lobe of the thymus is excised, and the innominate vein is mobilized to expose the innominate artery. The innominate artery should not be dissected from the trachea so that suspension of the artery exerts traction on the compressed segment of trachea restoring the lumen of the airway. Video-assisted rigid bronchoscopy is extremely valuable in assessing the effect of suture placement and tension on the tracheal lumen. Several 3-0 braided sutures are placed in the adventitia of the innominate artery at the level of the pericardium at a point directly beneath the sternum (Fig. 78.11). These sutures are then placed deeply into the periosteum of the posterior surface of the sternum. Alternatively, the ends of the sutures may be passed through holes bored in the sternum, tying the sutures over the anterior surface of the sternum. It is important to achieve a secure suspension to the sternum in either case. Two to four additional 3-0 sutures are placed more distally in the innominate

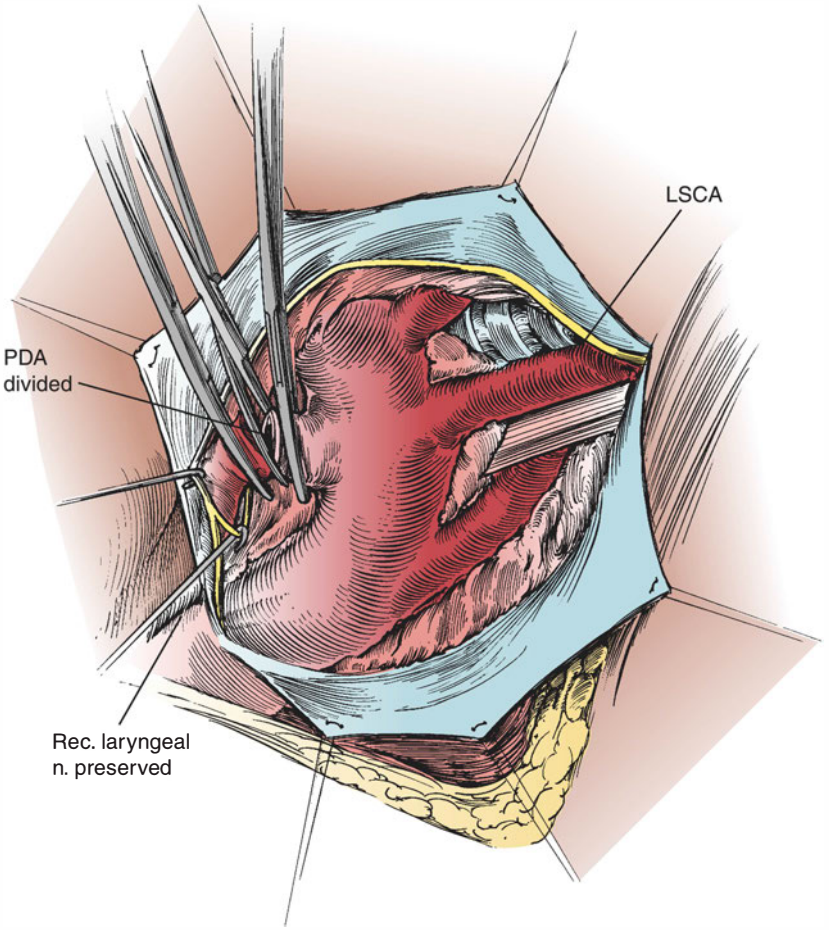


Fig. 78.7. Division of the anterior (*left*) aortic arch. The patent ductus arteriosus has been divided and oversewn. Vascular clamps are applied, with care being taken to make sure that the recurrent laryngeal nerve is not trapped in the clamp on the posterior surface of the left arch. The anterior (*left*) arch is divided and oversewn in two layers. The distal stump may be rotated and attached to the prevertebral fascia. Adhesive bands about the trachea and esophagus are divided sharply, relieving esophageal and tracheal compression. Ant., anterior; LCCA, left common carotid artery; LSCA, left subclavian artery; n, nerve; PDA, patent ductus arteriosus; Rec., recurrent.

artery and secured to the periosteum of the adjacent ribs such that the entire extent of the innominate artery producing compression on the anterior wall of the trachea is suspended (Fig. 78.12). Emphasis is placed on bronchoscopic guidance to assure relief of the tracheal obstruction. In the rare case in which innominate artery suspension fails to significantly improve the tracheal compression, reimplantation of the innominate artery via a median sternotomy should be considered. Reimplantation is advisable as the primary procedure when present with a chest wall deformity.

Group III: Pulmonary Artery Sling

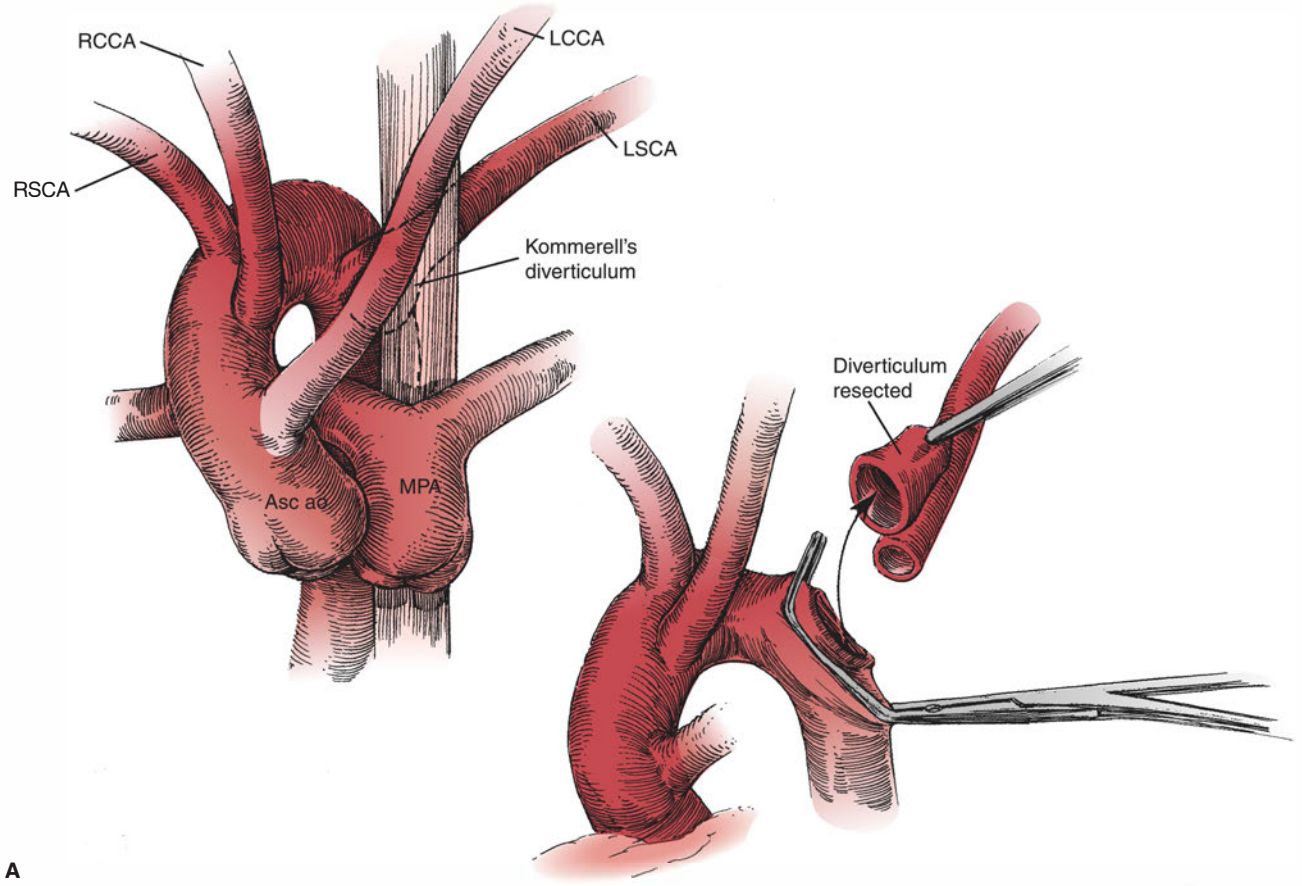
Anatomy

In cases of pulmonary arterial sling, the left pulmonary artery originates from the posterior aspect of the right pulmonary artery, proceeds over the right mainstem bronchus, between the trachea and the esophagus, and reaches the hilus of the left lung at a level lower than normal (Fig. 78.13). This lesion does not form a ring but forms a sling around the distal trachea and right mainstem bronchus, producing compression of these structures. The esophagus is not obstructed, but indentation on the anterior wall of the esophagus can be noted on esophagoscopy or barium esophagogram, a pathognomonic feature of this lesion.

Pulmonary artery sling anomalies are associated with tracheal stenosis in 33% to 64% of cases. In these cases, tracheal stenosis results from the absence of the normal posterior membranous portion of the trachea and proximal primary bronchi, replaced by complete tracheal rings. This ring deformity can occur in an isolated segment of the distal trachea or can be much more debilitating when it involves the entire length of the trachea. Predominately, this defect is limited to the area impinged on by the pulmonary arterial sling.

Clinical Presentation

Pulmonary artery slings produce symptoms of tracheal compression due to posterior impingement of the trachea. Infants generally present soon after birth with stridor that is episodic. They are often treated as asthmatics and may have a history of chronic respiratory infections. On examination, stridor, tachypnea, nasal flaring, inspiratory and expiratory wheezing, and intercostal retraction may be noted.



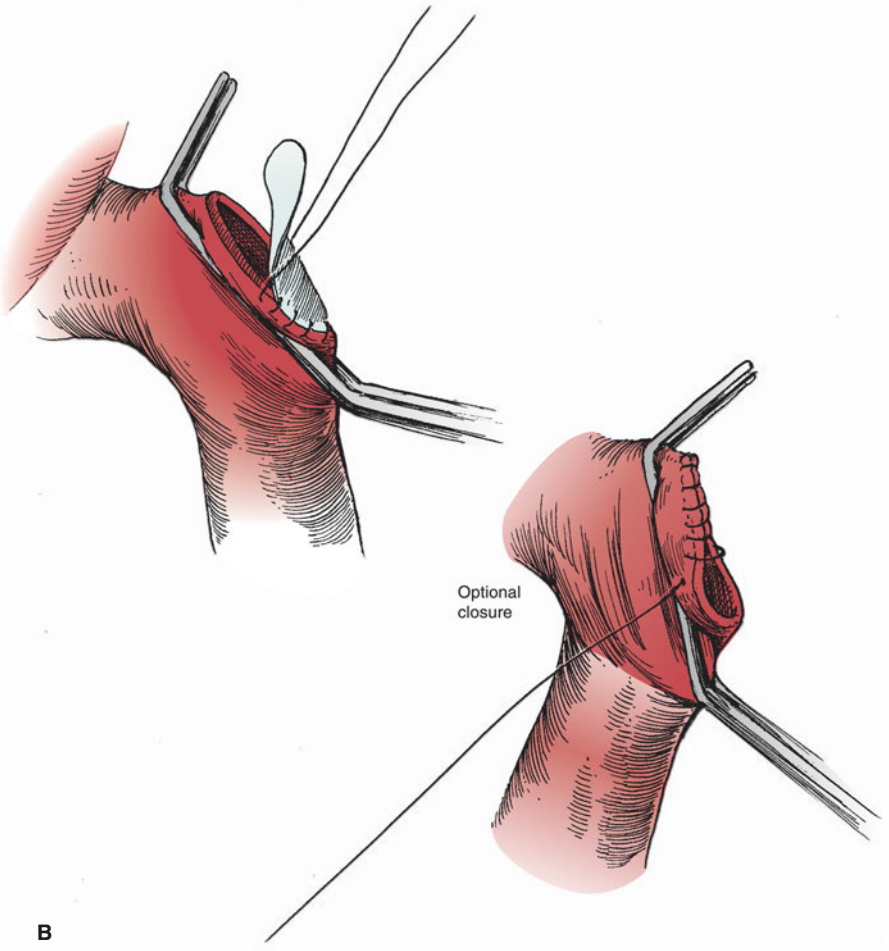


Fig. 78.8. (**A**) Anatomy of right aortic arch with retroesophageal left subclavian artery and associated Kommerell's diverticulum. (**B**) Patch closure or primary closure after resection of Kommerell's diverticulum. (**C**) Reimplantation of left subclavian artery into left carotid artery. Asc ao, ascending aorta; Esoph, esophagus; LCCA, left common carotid artery; LSCA, left subclavian artery; MPA, main pulmonary artery; RCCA, right common carotid artery; RSCA, right subclavian artery.

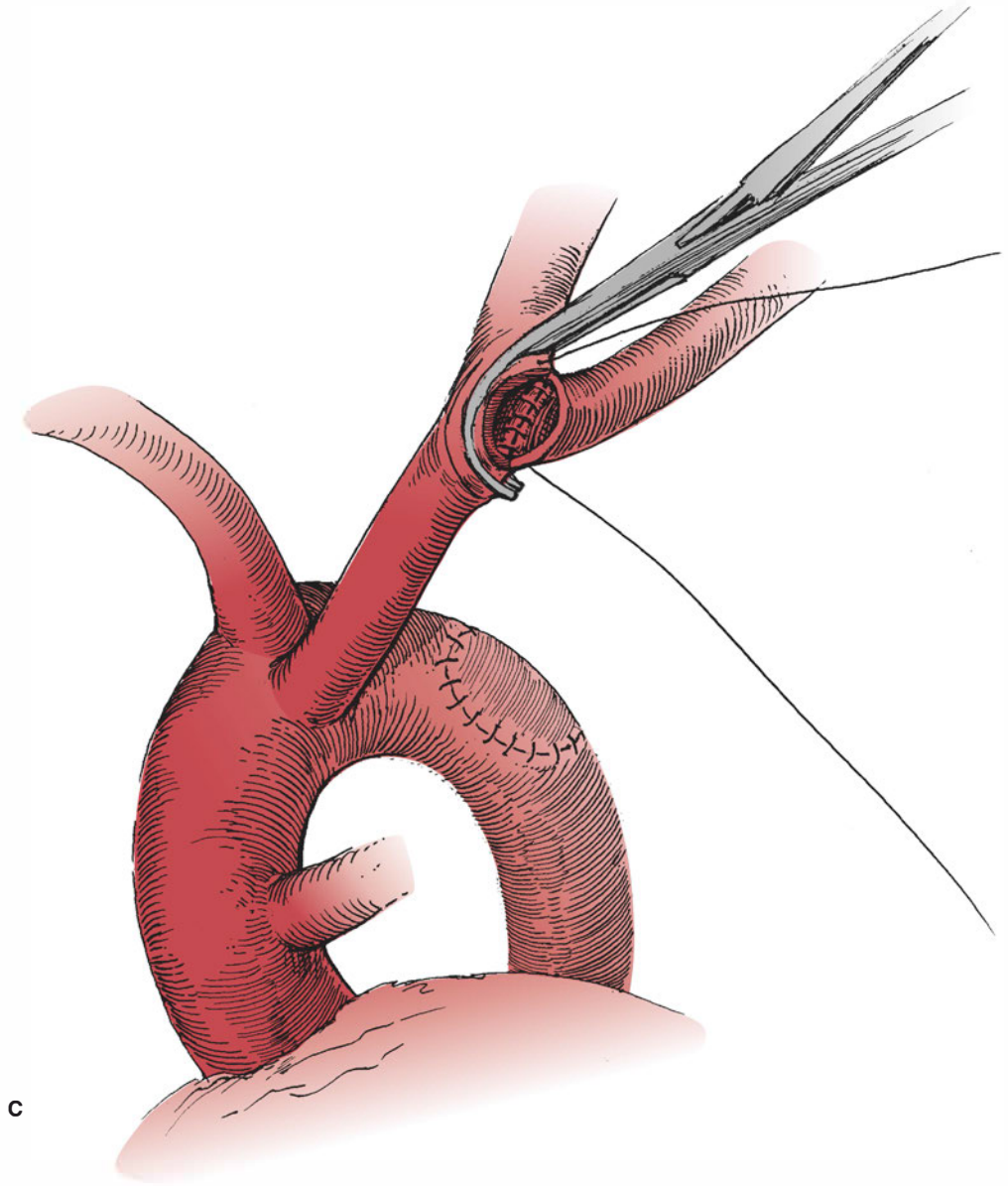


Fig. 78.8. (*continued*)

Diagnosis

In cases of pulmonary artery sling, chest radiography may demonstrate anterior bowing of the right mainstem bronchus, deviation of the lower trachea and carina to the left and unequal aeration of the lung. As with vascular rings, barium esophagography may be diagnostic. The position of the anomalous left pulmonary artery between the trachea and esophagus creates an anterior indentation in the esophagus when viewed in the lateral projection. MRI, however, provides more complete three-dimensional information and is the preferred diagnostic modality for this lesion.

Indications for Surgery

Patients with pulmonary artery sling and signs and symptoms of significant respiratory obstruction are candidates for operative repair. All patients should undergo preoperative bronchoscopic evaluation to assess the position, degree, and extent of any tracheal or bronchial constriction.

When workup demonstrates extrinsic tracheal compression without fixed stenosis, surgical repair entails relocation of the left pulmonary artery anterior to the trachea. This is achieved by reimplantation of the left pulmonary artery in its anatomically correct location on the main pulmonary artery (Fig. 78.13). Resection of redundant left pulmonary artery is usually necessary to prevent kinking after reimplantation. An alternative approach involves transection of the trachea with reanastomosis posterior to the undivided left pulmonary artery. When a fixed tracheal stenosis is noted, a sliding tracheoplasty is performed in addition to reimplanting the left pulmonary artery (Fig. 78.14A and 78.4B).

Surgical Technique

Patients are approached through a median sternotomy. Although cardiopulmonary bypass is not essential for reimplantation of the anomalous left pulmonary artery, its use does replace respiratory function and allows a wide anastomosis on empty and open vessels. The use of cardiopulmonary bypass also provides the option of dividing the trachea without dividing the left pulmonary artery. For this reason, we prefer a median sternotomy with cardiopulmonary bypass for all patients with pulmonary artery sling with or without fixed tracheal stenosis.

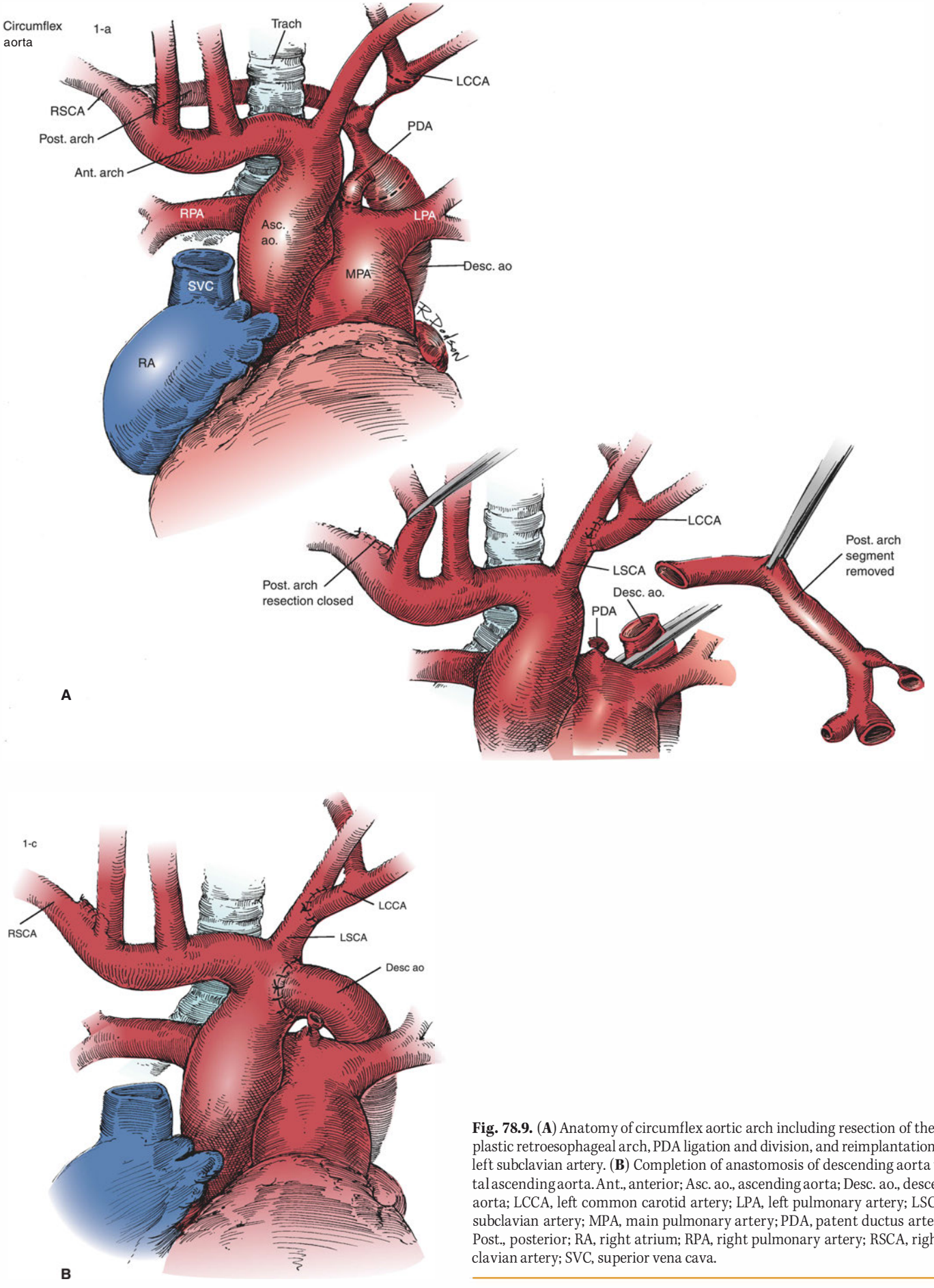


Fig. 78.9. (**A**) Anatomy of circumflex aortic arch including resection of the hypoplastic retroesophageal arch, PDA ligation and division, and reimplantation of the left subclavian artery. (**B**) Completion of anastomosis of descending aorta to distal ascending aorta. Ant., anterior; Asc. ao., ascending aorta; Desc. ao., descending aorta; LCCA, left common carotid artery; LPA, left pulmonary artery; LSCA, left subclavian artery; MPA, main pulmonary artery; PDA, patent ductus arteriosus; Post., posterior; RA, right atrium; RPA, right pulmonary artery; RSCA, right subclavian artery; SVC, superior vena cava.

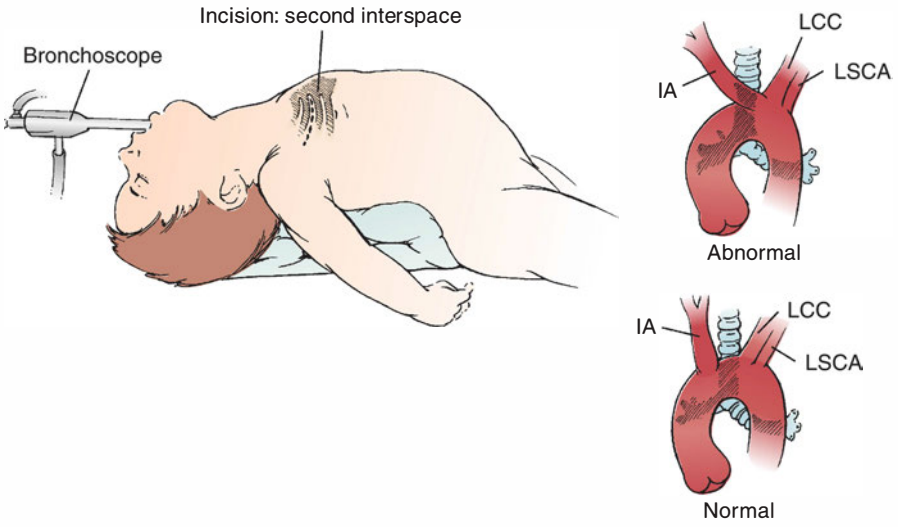


Fig. 78.10. Positioning and approach for relief of innominate artery compression. A right anterior thoracotomy through the second interspace is used. A ventilating fiber optic rigid bronchoscope with video attachment is placed before the incision is made. IA, innominate artery; LCC, left common carotid artery; LSCA, left subclavian artery.

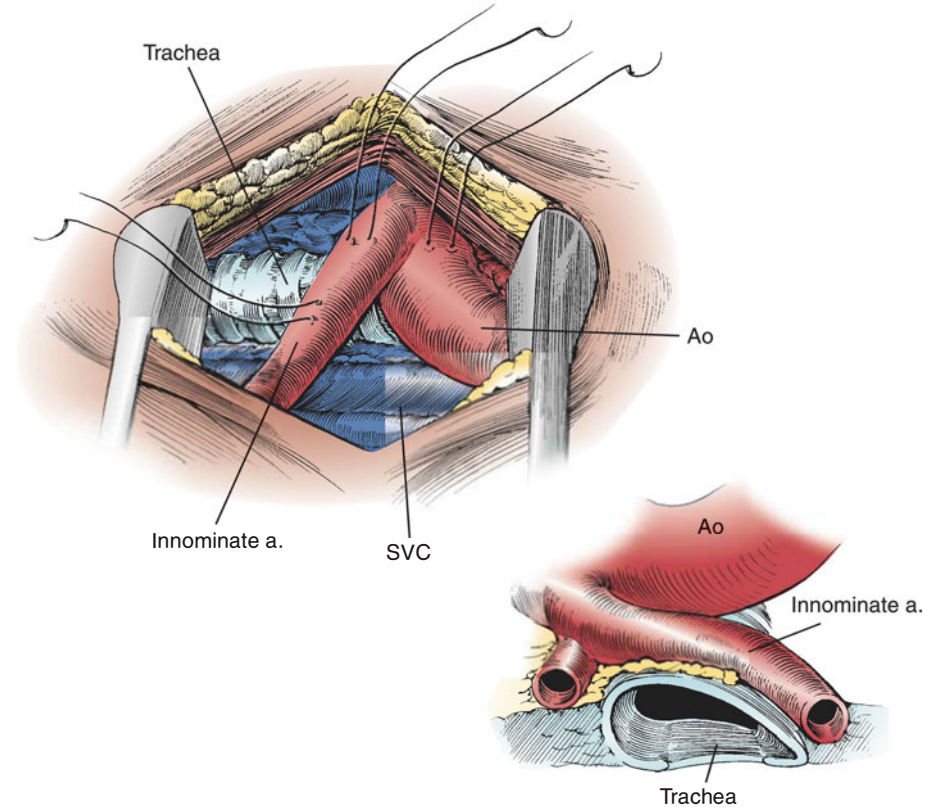


Fig. 78.11. Arteriopexy for innominate artery compression. The surgical exposure demonstrates the leftward origin of the innominate artery. Braided 3-0 sutures are placed in the adventitia of the vessel. The cross-sectional view reveals the mechanism of anterior tracheal compression. The adventitial tissue between the vessel and the trachea must not be dissected if satisfactory retraction of the anterior tracheal wall is to be achieved. a., artery; Ao, aorta; SVC, superior vena cava.

After standard median sternotomy, the pericardium and left pleura are opened. After appropriate heparinization, the patient is cannulated via the aorta and right atrium. Cardiopulmonary bypass is instituted, and the patient is cooled to 32°C. The proximal right and left pulmonary arteries are dissected free and the trachea is identified. The ligamentum arteriosum (ductus arteriosus) is divided and oversewn. At this point, options include division of the left pulmonary artery at its origin with reimplantation anterior to the trachea or simple transection of the trachea with anterior relocation of the undivided left pulmonary artery.

Whether or not tracheal compromise exists, we prefer to divide the left pulmonary artery and anastomose it to the left lateral aspect of the main pulmonary artery at a site approximating the take-off of a normal LPA. It is usually necessary to excise redundant LPA prior to completing the anastomosis (Fig. 78.13).

In patients with fixed tracheal stenosis (Fig. 78.14A), the trachea is transected at the midpoint of the stenosis. The superior portion of the stenotic trachea is then incised posteriorly and the inferior portion of the stenotic trachea is incised anteriorly. A sliding tracheoplasty is then performed using interrupted 5-0 polydioxanone suture (Fig. 78.14B). The tracheal repair is then tested under 40 cm H_2O to rule out air leaks.

Once the integrity of the tracheal anastomosis is ensured, ventilation is reinstituted and cardiopulmonary bypass is discontinued. After heparin reversal and satisfactory hemostasis, the sternotomy is closed in the standard manner.

POSTOPERATIVE CARE

Particular care must be given to respiratory management in all cases. In most vascular ring and pulmonary sling cases, extubation can occur immediately or within 24 hours of surgery. Occasionally, if postobstruction collapse or pneumonia was present preoperatively, prolonged intubation is necessary for tracheal suctioning, positive-pressure breathing, and oxygen administration. Humidified oxygen with bronchodilator therapy should be initiated. After extubation, continued administration of humidified air and bronchodilator therapy are required, as well as aggressive pulmonary toilet. Bronchoscopy is frequently necessary to assist with management of secretions and granulation tissue after slide tracheoplasty.

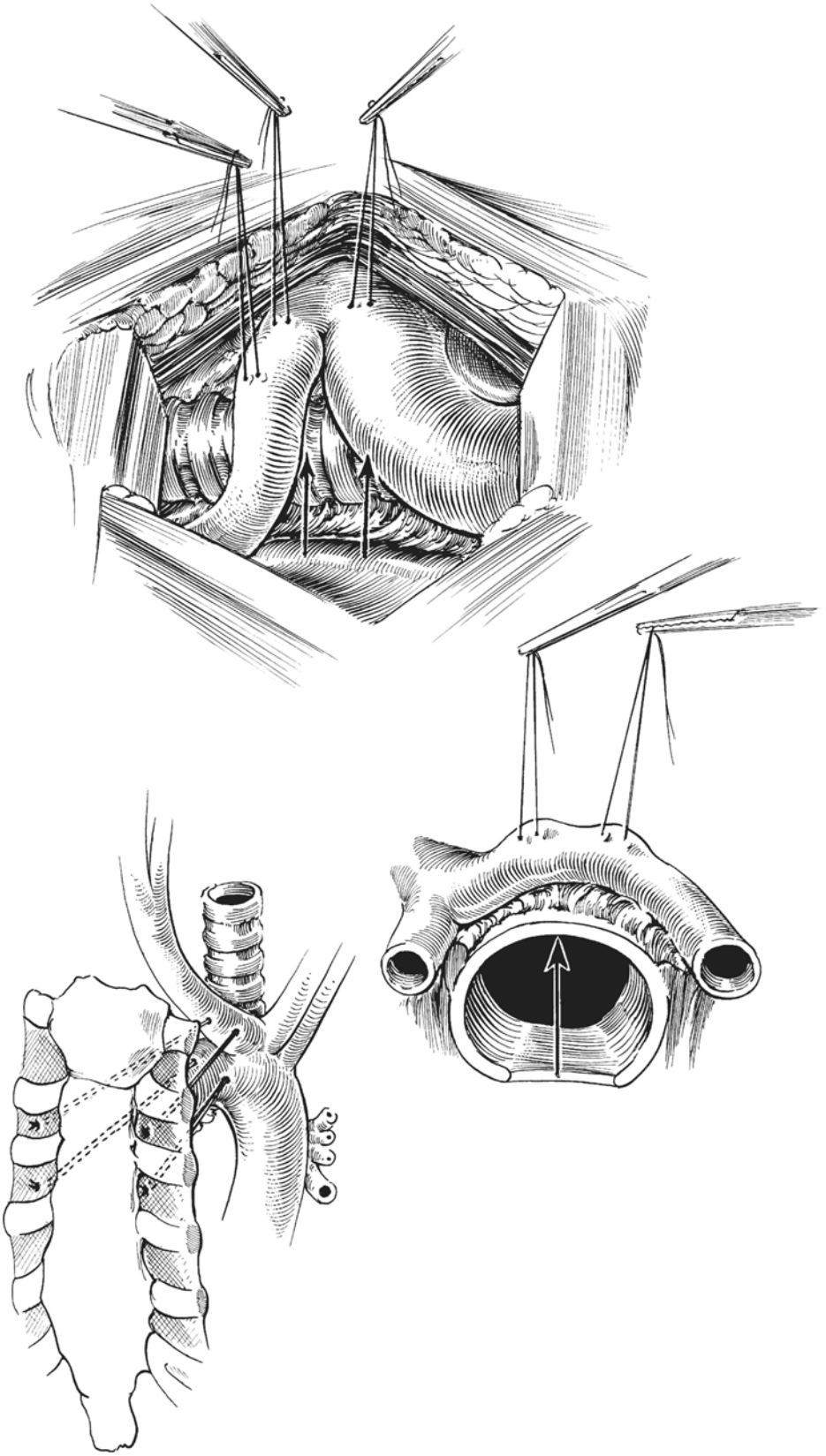


Fig. 78.12. Reduction of innominate artery compression. Retraction on the adventitial sutures elevates the innominate artery and the anterior wall of the trachea. The use of video-assisted bronchoscopy assures ideal suture placement for maximal tracheal decompression. Fixation to the sternum is demonstrated in the 30-degree view.

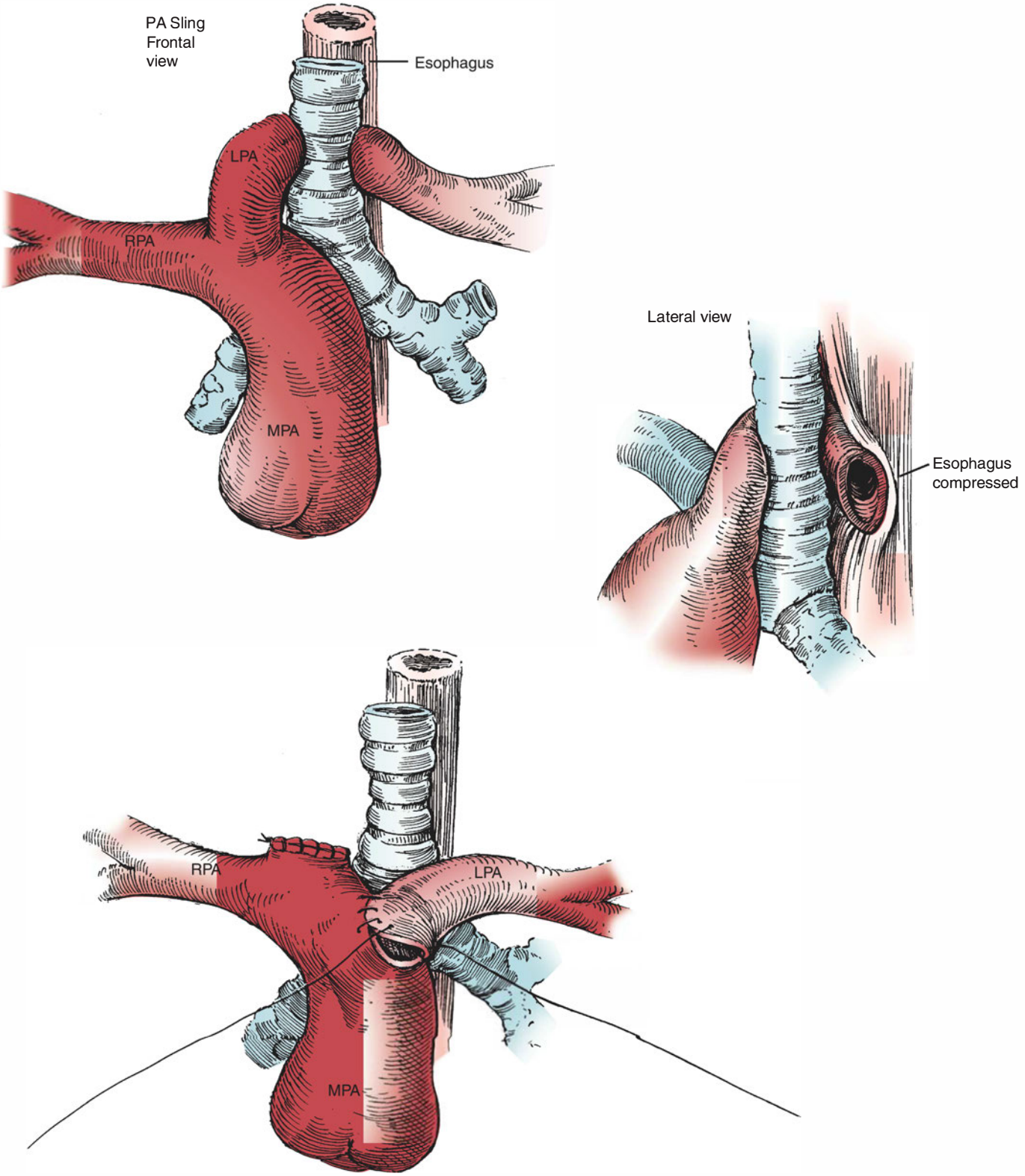


Fig. 78.13. Surgical anatomy of the pulmonary artery sling. Exposure via median sternotomy is shown. Localized tracheal stenosis is present in this case. Esoph, esophagus; LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery.

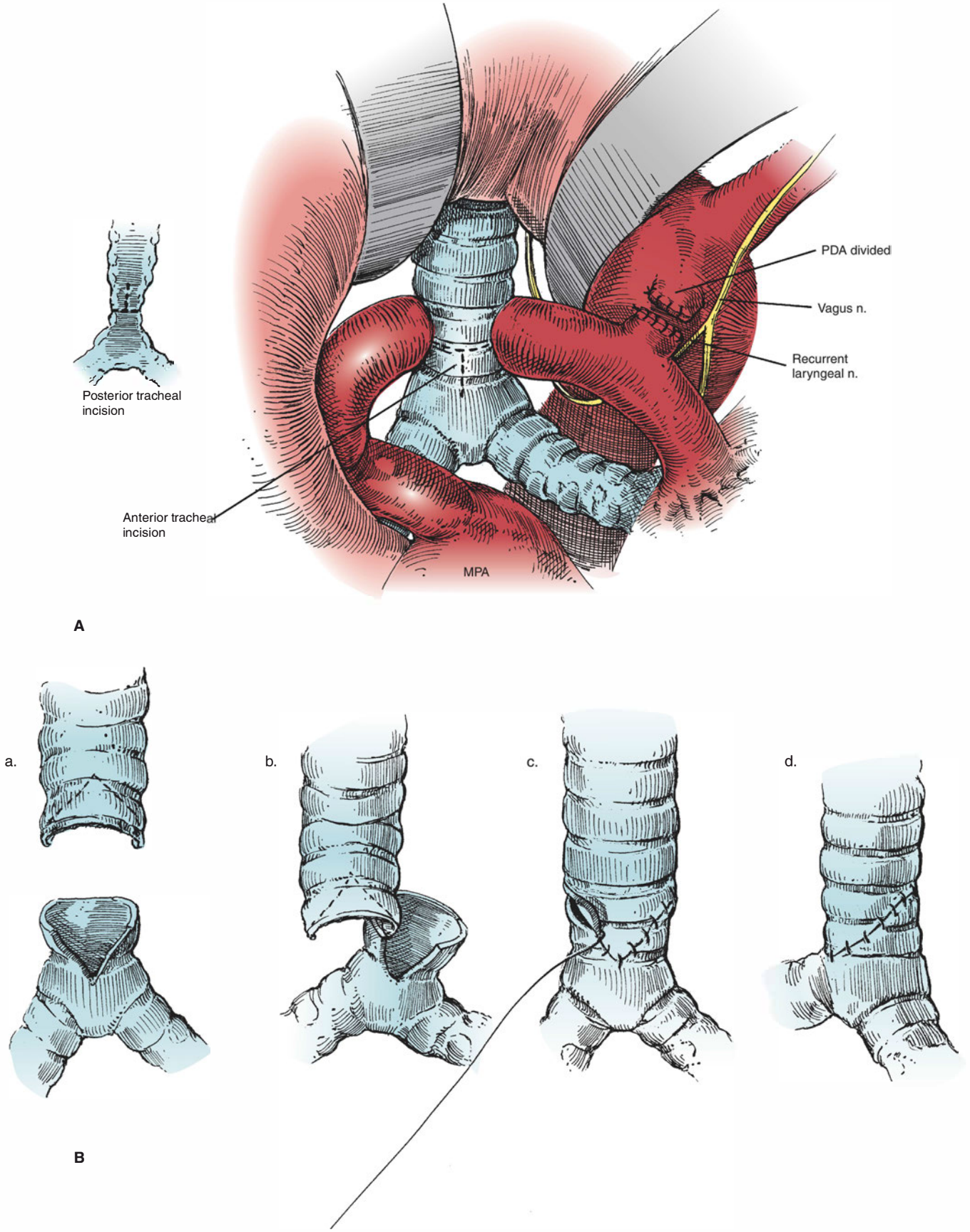


Fig. 78.14. (**A**) Location of tracheal incisions for sliding tracheoplasty. The trachea is divided at the midpoint of the stenosis and incisions are made anteriorly (inferior trachea) and posteriorly (superior trachea). (**B**) The superior and inferior portions are then slid over one another and anastomosed with interrupted absorbable suture. MPA, main pulmonary artery; n., nerve; PDA, patent ductus arteriosus.

Symptoms frequently are not resolved immediately, and it may take days to weeks for edema to resolve. Infrequently, recurring upper respiratory infections and chronic brassy cough may persist for as long as 1 to 2 years. Such prolonged postoperative symptoms are especially prominent in children who had onset of symptoms at age <6 months but were operated on late in their disease process.

VIDEO-ASSISTED THORACOSCOPIC SURGERY

Video-assisted thoracoscopic surgery (VATS) has been successfully applied to some patients with complete vascular rings. Safe use of this technique requires a significant VATS experience on the part of the operator as well as appropriate patient selection. The most suitable patients are those with a right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum (type IB) and the occasional patient with a double aortic arch with an atretic left arch. In these patients, a nonvascular segment of the ring can be approached endoscopically and safely divided. Endoscopic vascular clamps are not reliable enough to recommend this technique for division of a patent vascular segment. MRI can accurately detect segmental patency or atresia and thereby identify those vascular ring patients suitable for the VATS approach.

RESULTS

In cases of true vascular rings, mortality rate ranges from 0.5% to 6.0%. Most deaths are due to postoperative complications secondary to upper respiratory infections, pneumonia, and pulmonary failure in children undergoing delayed operation. Morbidity approximates 30% in children with complete vascular rings and 25% in cases of innominate artery compression. This morbidity most frequently includes chronic wheezing and upper respiratory infections. Infrequent complications include recurrent laryngeal nerve injuries and chylothorax.

Pulmonary artery sling repair performed with division and reimplantation of the artery demonstrates patency rates exceeding 75% and mortality rates under 5%.

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EDITOR'S COMMENTS

As noted in the chapter on imaging modalities, MRI scanning has become perhaps the most useful imaging technique for the evaluation of vascular rings and slings. Although barium swallow studies can give a fairly accurate assessment of arch anatomy, the relative size of the anterior and posterior arch in double aortic arch is not readily determined by this type of study.

Rapid multislice helical CT scanning has been very useful for the assessment of arch anomalies. The advantage of CT reconstruction is the ability to gain images in a very brief time, which may obviate the need for sedation or intubation with general anesthesia as for MRI scanning. Images are usually quite good and comparable to MRI images. However, the increased amount of radiation and the need for contrast are disadvantages to this technique.

In patients with aberrant left subclavian artery with right aortic arch and Kommerell's diverticulum, it is sometimes valuable to perform a pexy of the diverticulum or the base of the left subclavian artery to the prevertebral fascia to maintain complete opening of the ring and to mobilize the trachea and the esophagus in the middle portion to ensure that there is no adventitial tissue that may result in residual compression. As noted by the authors, resection of Kommerell's diverticulum is rarely necessary, although some centers have routinely done this, believing that greater relief of potential reobstruction or aneurysm formation is possible with this approach. If a resection of Kommerell's diverticulum is performed, we prefer to reimplant the left subclavian artery to the left carotid or the aorta to maintain flow to the left arm as suggested by the authors. Diverticulum resection in small infants can be associated with late stenosis of the reimplanted subclavian vessels; therefore, we prefer not to resect these diverticulae unless aneurysmal formation has occurred.

The situation with innominate artery compressive syndromes is a complex

(*continued*)

EDITOR'S COMMENTS (*continued*)

one. In most cases, the innominate artery compresses the trachea because of abnormal location more to the left on the aortic arch than normal. Often this is associated with abnormalities of chest wall symmetry with either pectus excavatum or a decreased anteroposterior diameter. We believe that these abnormalities often predispose patients to innominate artery compression, and simple innominate arteriopexy to the chest wall through a thoracotomy may not completely relieve the compression. In these cases, we believe that it is valuable to relocate the innominate artery on the ascending aorta more to the right, which completely relieves the obstruction even in an abnormal chest geometry. This procedure can easily be undertaken through a sternotomy incision, although a supraclavicular approach has also been described.

Although retroesophageal aberrant right subclavian artery is rarely associated with significant symptoms, we have seen patients who have had true dysphagia or chronic cough from this anatomy. Whereas ligation and division of the subclavian can relieve obstructive symptoms, we prefer to relocate the subclavian vessel from behind the esophagus over to the right side, either to the carotid or directly to the arch of the aorta. This maintains the patency of the subclavian vessel and prevents subclavian steal syndrome. This can generally be done through a right thoracotomy incision.

The anatomy of pulmonary artery sling is complex, and the results with repair of this defect are suboptimal. As noted by the authors, patients with pulmonary artery sling have had a very high mortality and a high occlusion rate of the left pulmonary artery when it has been divided and relocated anterior to the trachea. The causes for this are multifactorial; however, often these are small infants, and the pulmonary artery flow may not be adequate to maintain growth of the anastomosed vessel. If reimplantation of the left pulmonary artery to the main pulmonary artery is performed in pulmonary artery sling, it is often advisable to shorten the posterior aspect of the pulmonary artery slightly to prevent kinking and to patch augment the anterior aspect with pericardium or homograft material to decrease the risk of stenosis. In our experience, relocation of the pulmonary artery anterior to the trachea, because of the abnormal origin of the left pulmonary artery, can lead to kinking and stenosis of the origin of the left pulmonary vessel due to the elongated nature of the left pulmonary artery, which now sits anterior to the trachea. In some cases, even with simple translocation of the left pulmonary artery anterior to the trachea, it may be necessary to patch augment the origin of the vessel to prevent development of kinking or stenosis.

Much of the mortality in patients with pulmonary artery sling is related to airway complications, and, as noted in this chapter, direct attention to the airway is the primary component of surgical repair for this condition. We have seen several patients who have a tracheal origin of the right upper lobe bronchus with complete tracheal rings of the distal trachea to the bifurcation. Because of the early take-off of the upper lobe bronchus, the distal trachea is quite small and the bifurcation into the mainstem bronchi is also relatively small. In these patients, either extensive resection of the complete rings with slide tracheoplasty or rib cartilage patch tracheoplasty is necessary in addition to division of the trachea and relocation of the pulmonary artery anteriorly. These complex operations can be done with good results, and we prefer to use rib cartilage grafts rather than pericardial patches if sliding tracheoplasty is not feasible in most of these patients because of the rigidity of the tissue and the maintenance of a patent airway without anterior malacia from a pericardial patch. In more typical forms of pulmonary artery sling, simple resection of involved tracheal segments can completely relieve the obstruction. Mobilization of the trachea with hilar releases is often fully sufficient to prevent tension on the anastomosis, even with extensive slide tracheoplasty for long segment tracheal obstruction.

These patients must be diligently managed postoperatively to prevent secretions from pooling in the airway, and repeated bronchoscopic examinations are also necessary to examine the airway to clear secretions and to avoid the development of obstructive granulation tissue at the tracheal anastomosis. Associated cardiac defects such as ventricular septal defect or tetralogy of Fallot may also be present with a pulmonary artery sling complex and can be addressed at the time of the tracheal repair

TLS

79 Atrial Septal Defects

Richard D. Mainwaring, Peter Pastuszko, and John J. Lamberti

Substantial changes have occurred in the management of atrial septal defects (ASDs) since the approval of the Amplatzer ASD occluder device by the Food and Drug Administration in 2002. Today, the vast majority of isolated ASDs are closed in the cardiac catheterization laboratory. However, secundum-type ASDs are frequently a component of more complex congenital heart defects and need to be closed surgically as part of the procedure. Also, other forms of ASD would appear to remain in the surgical domain for at least the foreseeable future. Therefore, it is still imperative that surgeons performing pediatric heart operations understand the physiology of these defects, the indications for intervention, and the surgical techniques used for their repair.

Secundum-type ASDs account for approximately 80% to 85% of all ASDs. Sinus venosus defects (both the superior and the inferior type) and incomplete atrioventricular (AV) canal defects each constitute between 5% and 10% of all ASDs. Coronary sinus defects are a relatively uncommon form of ASD. The topic of incomplete AV canal—type ASDs is addressed in Chapter 82.

Secundum-type ASDs are three times more frequent in female than male patients. In addition, there is also a significant incidence of familial inheritance. These observations indicate that genetic factors play an important role in the formation of secundum ASDs; however, the exact gene locus responsible for this phenomenon has not been identified.

Most ASDs result in a left-to-right shunt (Q_p:Q_s) of about 2 or 3:1. The amount of flow across an ASD is determined by the relative ratio of diastolic compliance between the right and left ventricles because antegrade flow of the systemic and pulmonary venous return within the heart require the AV valves to be open. The amount of shunt is also influenced by the size of the interatrial communication, the presence or absence of pulmonary valve stenosis, and the pulmonary vascular resistance. Despite the increase in pulmonary blood flow seen with ASDs, most patients will have normal pulmonary artery (PA) pressures.

The chronic volume overload associated with an ASD has several adverse effects. The increase in flow through the left atrium (LA), right atrium (RA), and right ventricle (RV) results in enlargement of these structures. It is clear that cardiac enlargement, regardless of its etiology, will eventually translate into decreased quality of life as well as decreased life expectancy. With ASDs, the heart enlargement may take many decades before it leads to ventricular dysfunction or the onset of atrial arrhythmias. Similarly, the increase in pulmonary blood flow is initially well tolerated, but on a long-term basis may result in increased pulmonary vascular resistance.

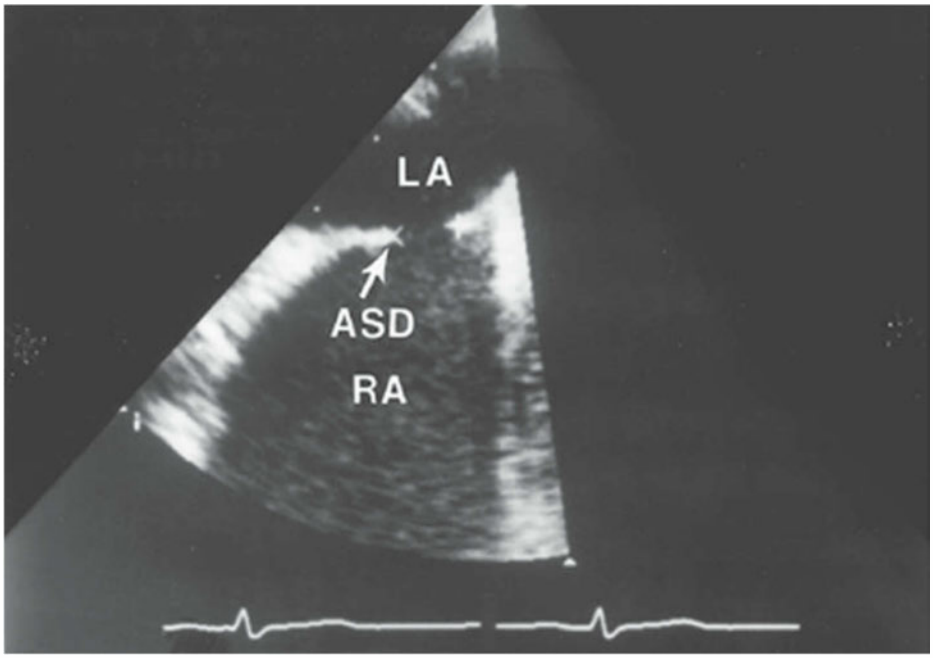
The natural history of an ASD has been well documented. Studies indicate that patients with unrepaired ASDs and a Q_p:Q_s of $>$1.5:1 will have a diminished life expectancy, averaging about 45 years. There is a great deal of variability in the natural history of these patients. Most infants and young children with ASDs are asymptomatic, demonstrating normal growth and absence of cyanosis. Some school-age children will tire a bit more easily than their peers or siblings and occasionally demonstrate perioral or periorbital cyanosis following exertion. As patients with untreated ASDs enter into their teens and 20s, they may have a gradual decline in their exercise capacity. Also during this time, women enter into their childbearing years. Women with an untreated ASD who become pregnant have a significant incidence of fetal (20% to 30%) and maternal (2%) mortality. As patients enter into their 30s and 40s, most experience a progressive decline in their exercise tolerance coincident with the deterioration in right ventricular function. Atrial arrhythmias are usually a late occurrence and may be the cause of palpitations. The development of arrhythmias may tip a previously well-compensated patient into congestive heart failure because of the loss of atrial contraction. Cyanosis is usually a very late and ominous sign of irreversible pulmonary vascular disease. Patients with a right-to-left shunt or a minimal left-to-right shunt are at risk for paradoxical embolus, although the actual risk is difficult to quantitate. The combination of right heart failure, arrhythmias, and cyanosis generally progresses and eventually leads to the eventual demise of the patient.

DIAGNOSIS

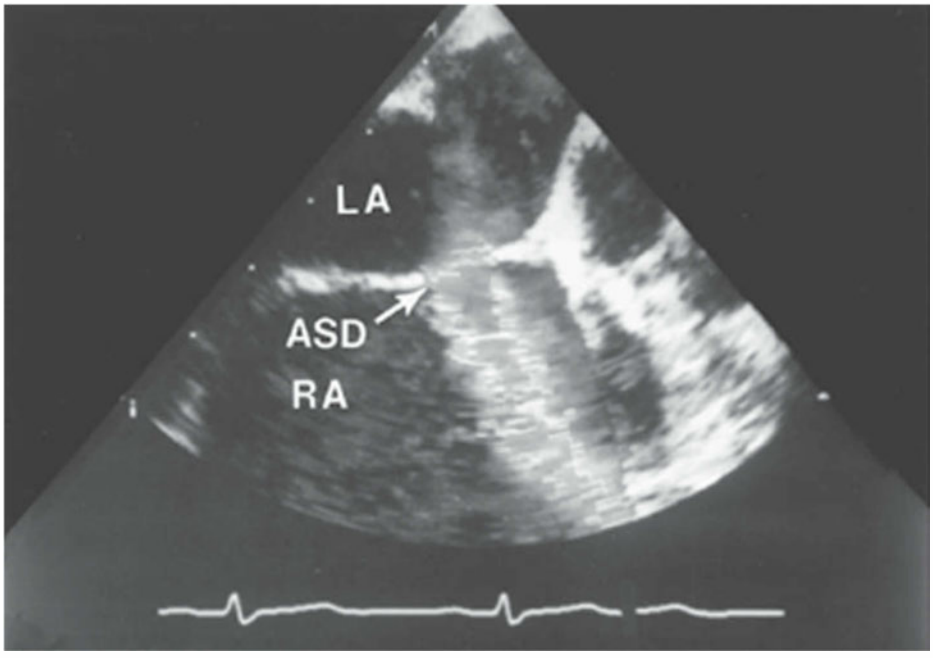
The diagnosis of ASD is usually suspected based on the physical examination. The history may not be particularly helpful in children but is an important part of the assessment in adults. Echocardiography is used to confirm the diagnosis and to delineate the pulmonary venous drainage (Fig. 79.1). Echocardiographic diagnosis is sufficient in most cases because the vast majority of patients identified with ASDs are infants and children. Diagnostic cardiac catheterization is indicated in two specific circumstances. In children, the need for catheterization arises when there remains doubt as to the complete diagnosis (Fig. 79.2). This can occur with abnormalities of systemic or pulmonary venous drainage or to exclude other associated congenital heart defects. Diagnostic catheterization is indicated in adult patients to assess pulmonary vascular resistance and to exclude the presence of acquired heart disease.

RECOMMENDATION FOR CLOSURE OF ATRIAL SEPTAL DEFECTS

The recommendation for closure of ASDs is based on the premise that the risk of the intervention is substantially lower than the risk incurred by following the natural history. ASD repair prevents the progression of cardiac and pulmonary changes and in younger patients restores a normal life expectancy. When the diagnosis of ASD is



A



B

Fig. 79.1. (**A**) Echocardiogram demonstrating a secundum-type atrial septal defect (ASD). The positions of the left atrium (LA) and right atrium (RA) are noted. (**B**) The ASD shown in panel A. The flow from the LA to the RA is shown by color flow (in this case in *black* and *white*).

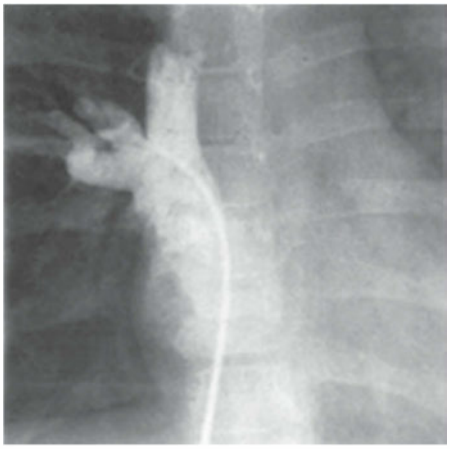


Fig. 79.2. Cardiac catheterization in a patient with a sinus venosus-type atrial septal defect. The entrance of the anomalous right upper lobe pulmonary veins into the superior vena cava is demonstrated.

made in infancy or early childhood, closure is generally recommended between 2 and 4 years of age. Regardless of the technique selected (surgery vs. interventional catheterization), these procedures are usually well tolerated and are typically associated with a rapid recovery. The psychosocial aspects of the hospitalization may also be better handled at this age.

Patients with an ASD detected in adulthood will usually benefit from closure of the defect and remain good candidates for whichever technique is selected. Assessment of the adult patient is based on a combination of clinical history, physical findings, echocardiographic information, and cardiac catheterization data. Patients who are fully saturated (i.e., have a left-to-right shunt by echo) and have RV enlargement consistent with a volume load from the shunt will almost certainly derive considerable benefit from closure. Conversely, most patients who have developed cyanosis should not be considered for closure because they rarely benefit from this intervention. Cardiac catheterization may better define those patients in the gray zone. Pulmonary vascular resistance approaching systemic or a shunt ratio barely exceeding 1 suggests that the patient is not a candidate for closure (approximately 10% of patients by the fourth decade will have reached this stage). The conventional teaching has been that surgical intervention is warranted in patients with a Q_p:Q_s of $>$1.5:1, but this criterion can probably be lowered to a Q_p:Q_s of 1.2 or 1.3:1 with the less invasive septal occluder devices. Closure should also be strongly considered in patients with a history of stroke even if the defect is very small, in order to preclude subsequent events.

Women who are discovered to have an ASD during pregnancy represent a unique clinical situation. During pregnancy, the circulating blood volume expands by about 50%, allowing even more blood to shunt through the ASD. Women who were previously well compensated may develop signs and symptoms of heart failure during the third trimester of their pregnancy. Every attempt should be made to avoid cardiac surgery in this circumstance because the risk to both mother and fetus is quite high. Most pregnant women can be successfully

managed using a combination of medical therapy and bed rest. However, on occasion a patient cannot be stabilized and must undergo urgent intervention. This mode of presentation was more common years ago than it is today, presumably because of improved methods of diagnosis.

SURGICAL TECHNIQUE

ASDs are usually closed through a sternotomy approach. In boys, this can be performed through a relatively short, low-lying midline incision. Our preference in girls has been to use a curvilinear, transverse (inframammary) skin incision as shown in Figure 79.3. This technique, once it has been learned, provides a superior cosmetic result without compromising in any way the exposure or versatility. Independent of the orientation of the skin incision, the sternum is divided vertically through the midline. The sternal retractor is then positioned for exposure. The retractor does not need to be opened widely for this operation because visualization of the ASD is relatively straightforward. Excessive retraction can result in sternal fracture.

The thymus is a relatively large structure in younger children and can present an obstacle to cannulation of the aorta. A portion of the thymus can be excised to improve exposure. This maneuver is not necessary in adult patients. The pericardium is then opened according to the intended operation. For the repair of a secundum-type ASD, we prefer to open the pericardium slightly to the left of midline with the anticipation that the patch will be cut to the appropriate size and shape once the defect has been visualized. For the repair of a sinus venosus-type defect, the anterior pericardium can be harvested and preserved. Some surgeons prefer to treat the pericardium with glutaraldehyde because this both strengthens the patch and makes handling it a bit more straightforward. The remaining pericardial edges are sutured to the drapes to form a pericardial well. Inspection of the anatomy is then conducted with several specific considerations:

1. The heart is inspected with regard to its size and configuration. Often the degree of cardiac enlargement will seem disproportionate to that seen on chest X-ray film.
2. The great vessels are inspected and palpated. The PA is usually enlarged consistent with increased flow through this vessel. A thrill at the level of the pulmonary annulus may signify the presence of pulmonary stenosis.
3. The systemic veins are inspected to ensure that there are no abnormalities. A small or absent innominate vein suggests the presence of a left superior vena cava (LSVC). In the case of a superior sinus venosus defect, the SVC is isolated, and the presence or absence of anomalous pulmonary veins is confirmed. The position of the azygos vein should also be identified.
4. The pulmonary veins on the right are visualized, with the intent to inspect the left pulmonary veins once on bypass. As indicated, in the case of a sinus venosus defect the anomalous pulmonary veins should be identified at this point.

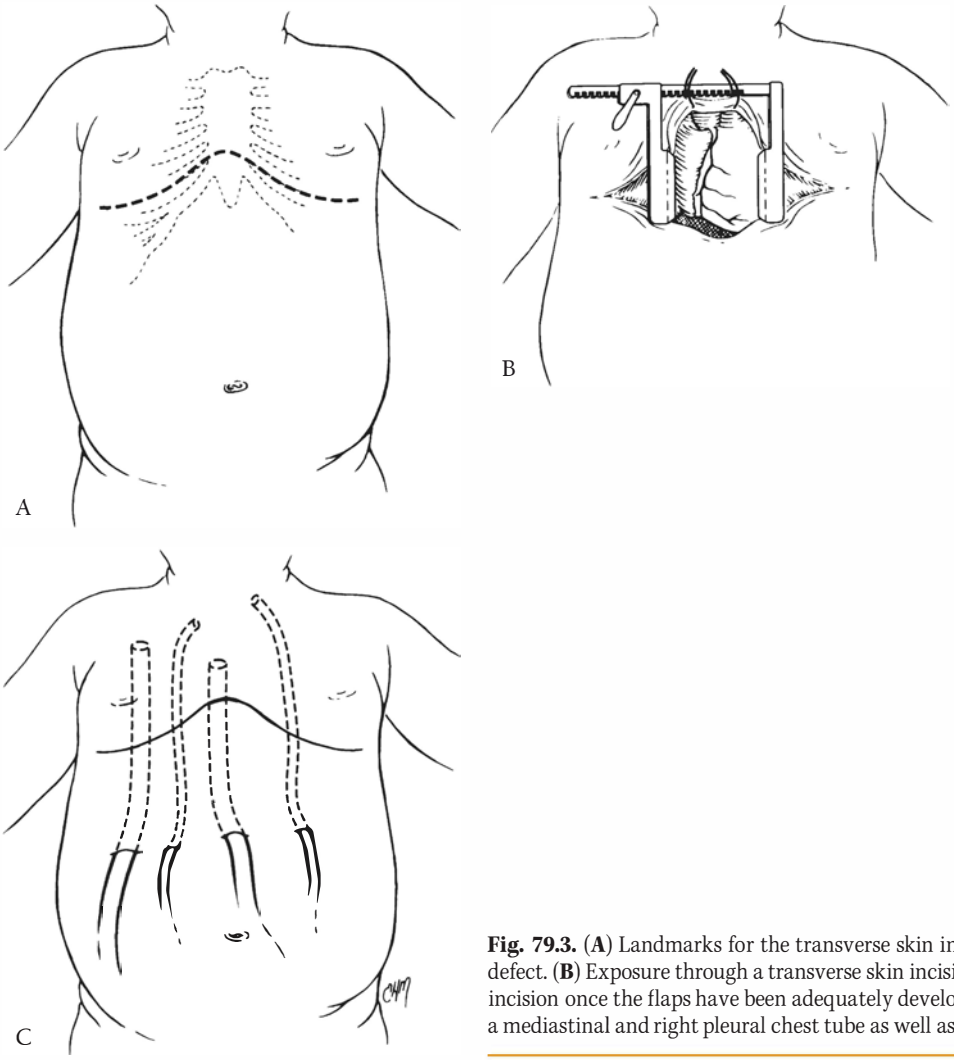


Fig. 79.3. (**A**) Landmarks for the transverse skin incision used in female patients with an atrial septal defect. (**B**) Exposure through a transverse skin incision is nearly identical to that of the standard vertical incision once the flaps have been adequately developed. (**C**) Closure of the transverse skin incision with a mediastinal and right pleural chest tube as well as two subcutaneous drains beneath the upper flap.

Cannulation is commenced by placing purse strings in the superior and inferior venae cavae as well as the aorta. If there are anomalous pulmonary veins in association with a sinus venosus defect, the superior vena caval cannulation site should be at the junction of the SVC and innominate vein in order to be well above the entrance of the anomalous veins. The patient is cannulated and cardiopulmonary bypass instituted. Normothermia is used in most cases of simple ASD. Mild levels of hypothermia (32 to 34°C) may be used when a longer cross-clamp time is anticipated (e.g., sinus venosus ASD with partial veins). Inspection of the heart is then completed. Specifically, the position of the left pulmonary veins and the presence of a LSVC can be ascertained by tilting the heart to the right. After inspection, caval tapes are placed around the superior and inferior venae cavae and a cardioplegia needle is inserted into the aortic root. The aortic cross-clamp is applied, and cardioplegia is delivered to achieve electromechanical silence. During this time, the caval tapes are tightened and the RA opened to collect the coronary venous effluent.

Surgical Repair of Secundum-Type Atrial Septal Defects

Secundum-type ASDs are approached through a standard right atriotomy (Fig. 79.4). Traction sutures can be placed at the edges of the atriotomy to facilitate exposure. The ASD is identified at its position just beneath the superior limbus. A few moments should be taken to identify the position of the coronary sinus and its relationship to the tricuspid valve. These two structures along with the tendon of Todaro form the triangle of Koch. The AV node should lie at the superior apex of this triangle and may not be too far from the medial border of the ASD. A right-angled clamp can be passed through the ASD into the pulmonary veins to confirm their position. Finally, the position of the inferior vena cava (IVC) cannula should be inspected along with its relationship to the inferior border of the ASD and the Eustachian valve. Misinterpretation of this anatomy can result in inadvertent baffling of the inferior vena caval flow across the ASD into the LA.

After this checklist of anatomic landmarks is completed, repair of the defect is undertaken. The majority of secundum ASDs in young children can be closed primarily, whereas in teenagers and adults the defects are usually large and the tissues less compliant, so that a patch repair is almost always indicated. Our preference has been to use autologous pericardium, although synthetic patches can also be used. Pericardial patch repair of an ASD is straightforward, and the technique described here is one of the several possible variations. Two separate sutures of 5-0 or 6-0 nonabsorbable vascular suture are placed at the superior and inferior ends of the defect. Each of these sutures is passed through the pericardium at a distance slightly exceeding the length of the ASD (Fig. 79.5). The pericardial patch is cut and lowered into position with the smooth side toward the LA. Each of the sutures is tied, and then each arm of the double-armed suture is run 90 degrees to meet its opposite suture. Just before completing this process, the left side of the heart is filled with blood by ventilating the patient to increase pulmonary venous return and thereby expressing any air remaining in the LA (Fig. 79.6). The patient is then positioned in a steeply head-down position and

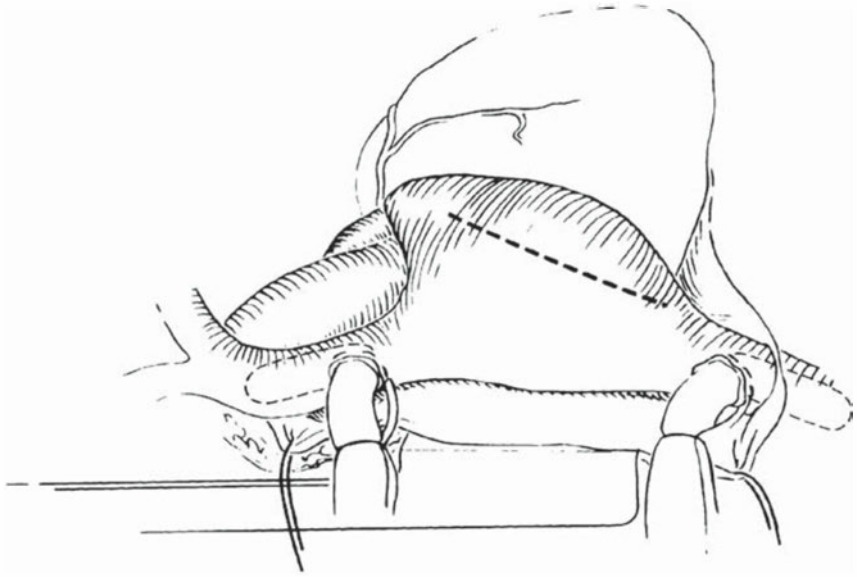


Fig. 79.4. View of the right atrium and superior and inferior venae cavae from the surgeon's perspective. Right-angled cannulas have been placed in each vena cava. The *dashed line* demonstrates the placement of the atrial incision for repair of a secundum atrial septal defect.

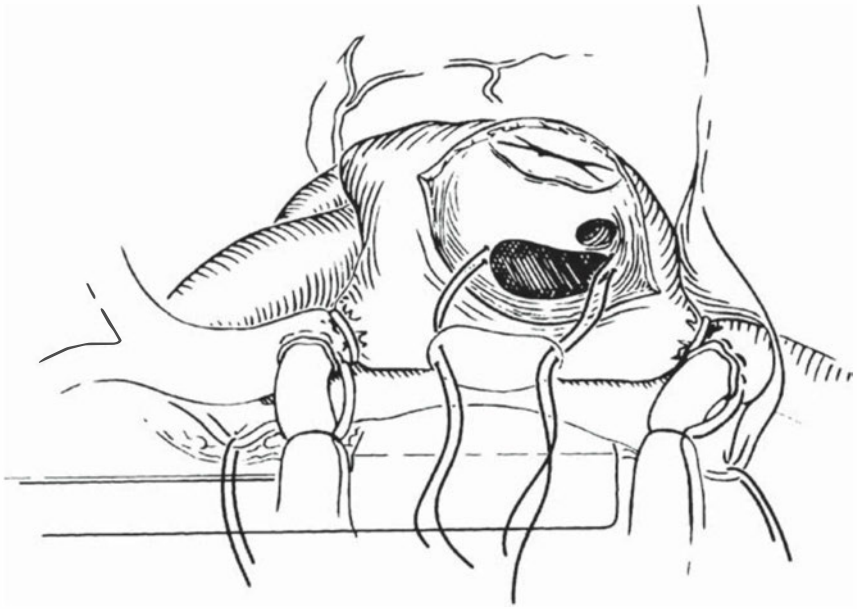


Fig. 79.5. Two separate sutures have been placed at either end of the atrial septal defect and then through the pericardium. The pericardial patch has been cut and is now lowered into position. Each of these sutures is then tied and run toward its opposite suture.



Fig. 79.6. Just before completion of the repair, the left heart is deaired.

the cross-clamp removed with the aortic root on suction. Rewarming is performed while the right atriotomy is closed with a double running 5-0 or 6-0 nonabsorbable vascular suture. The patient is weaned from cardiopulmonary bypass once rewarming is completed.

Amplatzer Repair of Secundum-Type Atrial Septal Defects

As we have previously stated, the majority of secundum-type ASDs are now closed in the catheterization laboratory. The original ASD devices were developed more than 30 years ago and were prone to late structural failures. The current design has proven to be both effective and durable (Fig. 79.7). The advantages of device closure are that it is less invasive than surgery and the recovery from the procedure is shorter. The limitations of the device include the fact that it requires a 7F or 8F sheath in the femoral vein, so that patients need to be at least 10 to 12 kg to accommodate the vascular access. Amplatzer device closure may not be feasible from a technical standpoint in patients with a deficient rim of tissue surrounding the ASD, particularly along the inferior aspect of the defect. For these reasons, there is still an occasional secundum defect that is best treated with surgical closure.

There are a number of early and late complications that have been reported following Amplatzer insertion. Systemic thromboembolism has been observed following implantation of an Amplatzer. Patients who experience this problem should undergo evaluation for hypercoaguable state (e.g., anti-thrombin 3 deficiency, protein C or S deficiency) and be anticoagulated. Repeat thromboembolism is a reasonably strong mandate for the removal of the device and conventional surgical closure of the ASD. Onset of new arrhythmia or heart block has also been described following deployment of the Amplatzer device. This finding may indicate that the Amplatzer is not seated properly. Dysrhythmias in this setting may potentially resolve over time but have also been associated with subsequent erosion of the device into adjacent structures. Thus, the presence of new arrhythmias should be investigated thoroughly with a high index of suspicion.

Complications following Amplatzer deployment may precipitate the need for urgent or emergent surgical intervention. The Amplatzer device may be malpositioned resulting in a residual ASD. In this setting, the patient can be taken to the operating room in the next 24 to 48 hours with the removal of the device and closure of the ASD. However, malposition of the Amplatzer with subsequent embolization of the device (typically to the aortic valve or ascending aorta) may result in hemodynamic compromise and require emergency surgery for extraction of the device and closure of the ASD. We have seen one patient who sustained a severe neurologic deficit following this scenario, possibly from thrombus forming on the malposed device.

Amplatzer devices can also erode into the LA, RA, base of the aorta, or into the free pericardial space. These complications may occur early (more frequent) but have also been reported years after device deployment (less frequent). The presentation of a patient with hemopericardium after an Amplatzer device should alert the surgeons to the possibility of erosion. This should be treated as a surgical emergency with cardiopulmonary bypass available prior to sternotomy. If erosion into the pericardial space is confirmed at surgery, the device should be removed and the ASD and atrial erosion site repaired. Erosions into the base of the aorta may manifest

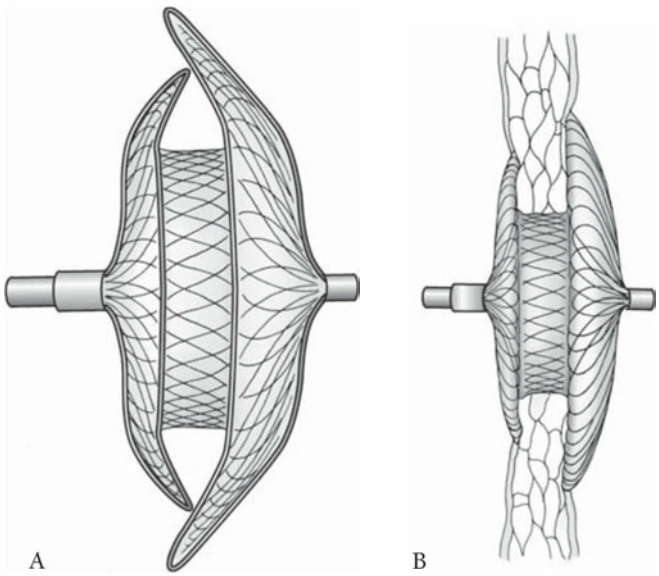


Fig. 79.7. Artist's schematic of the Amplatzer septal occluder.

itself in one of two ways; at times a patient will present with a new murmur and echocardiography detects the presence of an aorto-left atrial fistula. An aorto-left atrial fistula may also be unmasked when a malposed Amplatzer is removed. A fistula is then detected by the intraoperative TEE. In this circumstance, the device itself is acting as a "bottle stopper," and when it is removed, the fistula becomes apparent. To address the situation, the heart should be arrested (or rearrested), an aortotomy performed and the fistulous tract repaired with a small patch.

The widespread use of the Amplatzer device has created an interesting ethical dilemma. The use of this device represents a "self-referral" for cardiologists and thus has the potential for misapplication. In addition, the reporting of complications, while recommended, has not been mandated by the US Food and Drug Administration Manufacturer and User Facility Device Experience database. It is likely that complications are somewhat underreported. At various surgical meetings, informal surveys would suggest that catastrophes after Amplatzer implantation are far more frequent than the literature would indicate. Mandatory reporting of device complications would clarify the issue and facilitate safe use of these devices. So far, the call for mandatory reporting of Amplatzer complications has not been heeded. In summary, while Amplatzer device closure is an effective therapy for most secundum ASDs, cardiac surgeons should stay well versed in the field of urgent and emergent Amplatzer-device removal techniques.

Repair of the Superior-Type Sinus Venosus Atrial Septal Defect

The superior type of sinus venosus ASD is far more common than the inferior type. The hallmark of this diagnosis is the position of the ASD high in the RA above the superior limbus. The majority of the time the right upper and right middle pulmonary veins drain anomalously into the lateral aspect of the SVC. The distance between the entrance of the pulmonary veins into the cava and the entrance of the cava into the roof of the RA can usually be assessed from the outside by dissecting these structures in advance. The azygos vein is also identified in advance so that it is not confused with a pulmonary vein when these structures are inspected from the internal aspect of the cava. When the pulmonary vein entrance is near the cavoatrial junction, a simple patch repair can be performed to channel the pulmonary vein blood flow across the ASD into the LA. However, it is not infrequent that the pulmonary vein entrance is a considerable distance from the cavoatrial junction. In this circumstance, the "double-patch" technique is the preferred option. A lateral atriotomy is performed and extended cephalad across the cavoatrial junction onto the SVC (Fig. 79.8). Traction sutures may be helpful to facilitate exposure. A portion of previously harvested pericardium is used to baffle the anomalous veins to the LA. The patch is sutured in position with 5-0 or 6-0 nonabsorbable suture, beginning at the superior apex just above the entrance of the superiormost anomalous pulmonary vein (Fig. 79.9). Suturing is continued along the lateral border of the cava and then around the lateral and inferior border of the ASD. Deairing is performed before final completion of the suture line. The repair can be inspected for leaks by having the anesthesiologist ventilate the lungs (Valsalva maneuver). Because the pericardial baffle takes up a portion of the superior vena caval cross-sectional area, primary closure of the cava might result in compromise of the lumen. Therefore, a second pericardial patch is used to augment the cava along its anterior surface (Fig. 79.10).

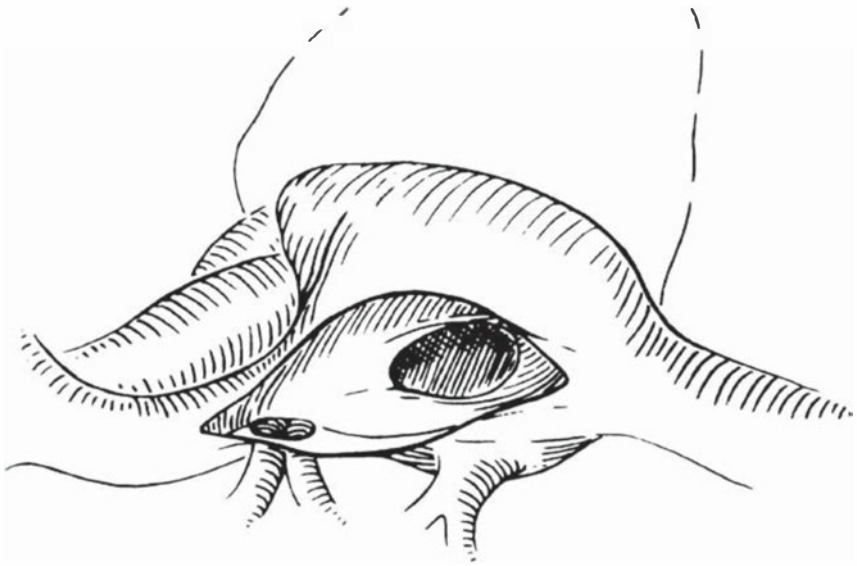


Fig. 79.8. A lateral atriotomy is performed extending onto the superior vena cava. The orifice of the anomalous right superior pulmonary vein is seen entering into the cava.

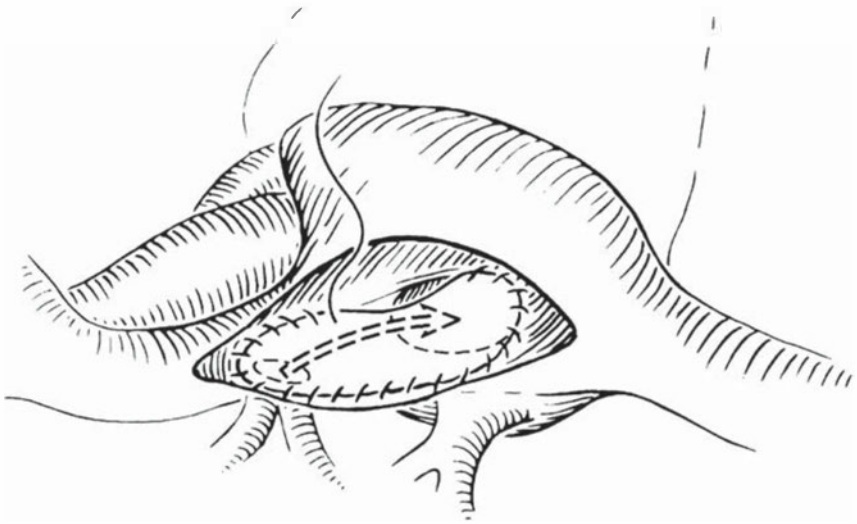


Fig. 79.9. A pericardial patch is sutured above the orifice of the anomalous vein and continued inferiorly to encompass the orifice of the sinus venosus atrial septal defect.

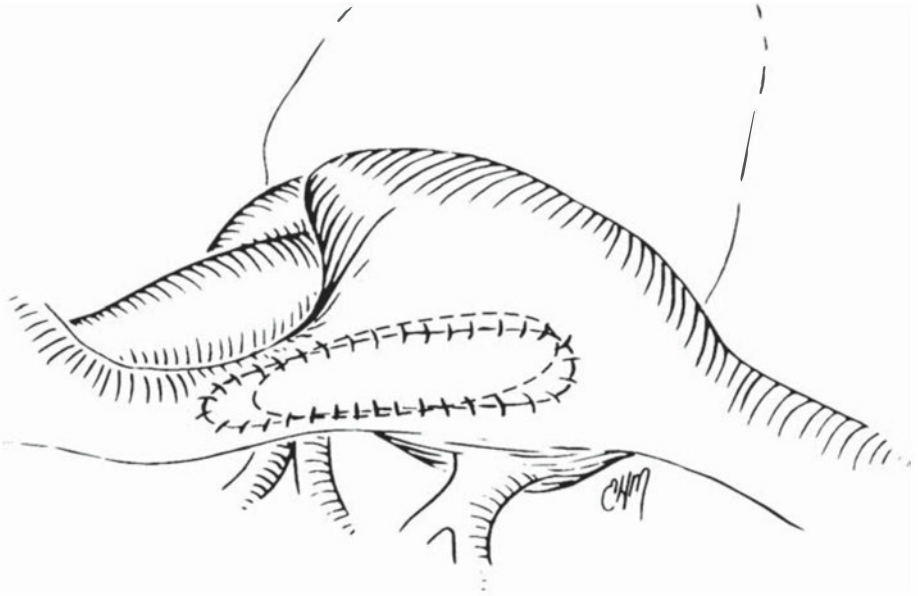


Fig. 79.10. The superior vena cava and right atrial junction are augmented with a second pericardial patch to prevent narrowing of the superior vena caval orifice at this point.

An alternative approach to the repair of a superior-type sinus venosus defect, particularly when the entrance of the anomalous pulmonary veins is high on the SVC, is the Warden procedure. Popularized by Warden and colleagues in the 1980s and 1990s, it was first described in an animal model by Gerbode in 1949. The first clinical experience was reported simultaneously by Lewis and Ehrenhaft in 1958. The technique involves division of the SVC just above the entrance of the uppermost pulmonary vein and reanastomosis of it to the atrial appendage. The proximal end of the divided SVC is closed and a patch is placed on the inside of the atrium effectively closing the SVC and baffling the pulmonary vein blood flow across the ASD into the left side of the heart.

The potential advantage of the Warden procedure is the avoidance of any manipulation of the SA node or disruption of its blood supply, thereby reducing the risk of postoperative arrhythmias. In those patients with sinus venosus ASD where the pulmonary veins enter high on the SVC, baffling the veins with a single patch is not a feasible option. Placement of such a patch would lead to an unacceptably high incidence of SVC obstruction. In those patients, the "double-patch" technique is the only suitable alternative. However, the course of the SA node artery is highly variable and an incision across the lateral aspect of the SVC–right atrial junction necessary for the placement of the second patch may result in disruption of the blood supply to the SA node. Warden et al., in their series of 40 patients, identified the location of the sinus node artery in 27 individuals, observing that in 10 of these patients (37%), an incision across the cavoatrial junction would result in injury to the SA node artery.

The decreased risk of injury to the blood supply of the SA node with the Warden procedure makes it our procedure of choice for the repair of the superior-type sinus venosus ASD, particularly if the pulmonary veins enter high on the SVC. Although not unanimous, evidence in the literature tends to support this approach.

Our standard exposure is via a median sternotomy. Thymectomy is routinely performed and the right pleural space is opened widely in order to fully visualize all of the anomalous pulmonary veins and assess their relationship to the azygous vein. The SVC is dissected completely and the azygous vein is ligated and divided. Standard cardiopulmonary bypass and mild hypothermia are utilized. In Warden's original description of the procedure, the SVC is cannulated with a straight venous cannula placed through the RA and the right atrial appendage. Alternative approach, and our preferred technique, is to cannulate the SVC directly, high at the SVC–innominate vein junction. Once the cardiopulmonary bypass is established, the SVC is divided just above the uppermost pulmonary vein. The proximal end of the SVC is then closed either primarily or with a patch (Fig. 79.11).

The RA is opened and the location as well as the size of the ASD is established. In some patients, the ASD may not be sufficient in size and may need to be enlarged, while in up to 18% of patients, the atrial septum is completely intact and an ASD needs to be created. A patch is then placed in the roof

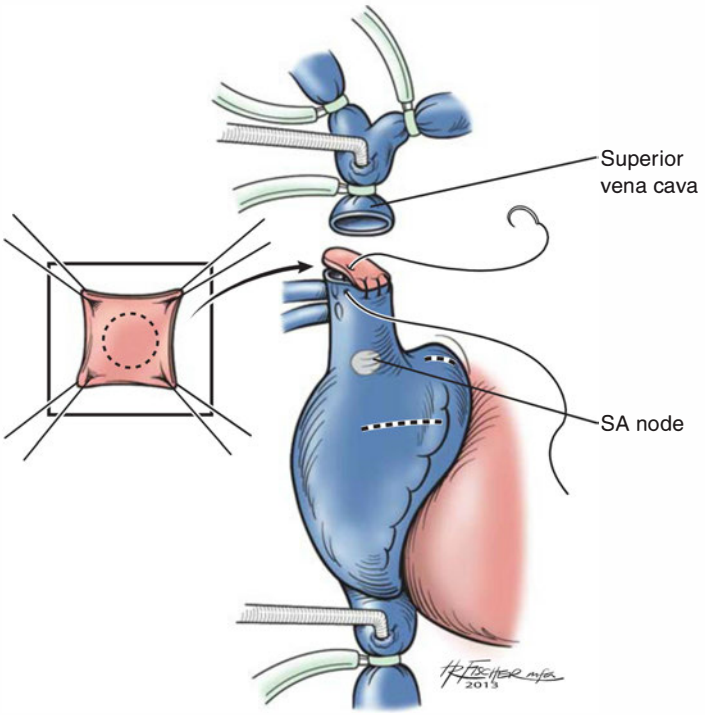


Fig. 79.11. The Warden procedure. The superior vena cava (SVC) cannula is placed high, at the SVC–innominate vein junction. The SVC is divided just above the uppermost pulmonary vein. The proximal end of the SVC is closed either primarily or with a patch (pictured).

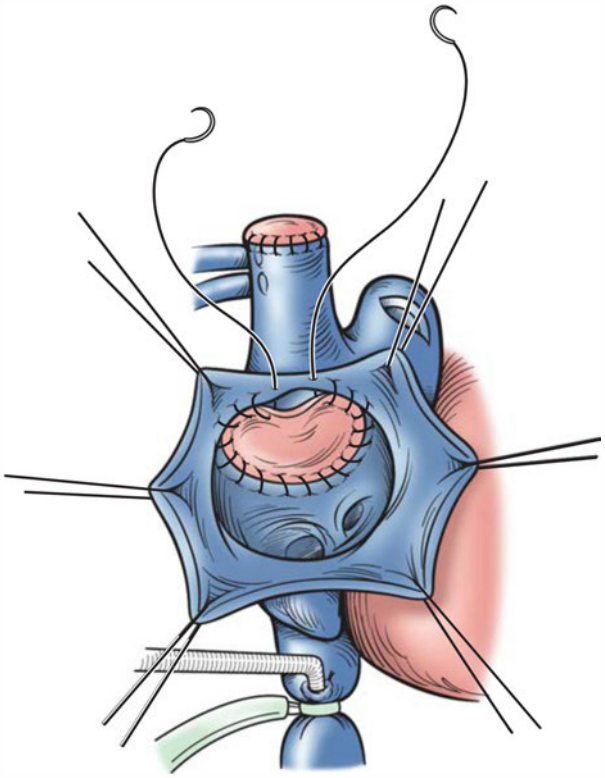


Fig. 79.12. After a standard atriotomy is performed, the superior vena cava (SVC) is closed from the inside with a patch incorporating the atrial septal defect (ASD), baffling the pulmonary vein blood flow across the defect to the left side of the heart. The atrial appendage is amputated and the muscle bundles in the area are carefully and thoroughly divided and resected.

of the RA to divert blood flow from the right upper and middle lobe veins across the ASD into the LA. The patch simultaneously closes the SVC from the remainder of the RA (Fig. 79.12). Our preferred patch material is glutaraldehyde-preserved autologous pericardium, although other materials, such as fresh autologous pericardium, bovine pericardium, or Gor-Tex, have been reportedly used with no discernable difference.

Once the patch is in place, the tip of the right atrial appendage is amputated. The inside of the atrial appendage at this point is inspected and any trabeculations are divided. The division and resection of these muscle bundles may be the most critical portion of this procedure, since leaving any of them intact clearly increases the risk of stenosis at SVC–RA anastomotic site. The length of the atrial appendage is usually adequate to permit creation of an anastomosis without tension. However, a number of technical modifications of this repair, aimed at reducing the risk of stenosis, have been described. Those have included anastomosis with interrupted sutures, patch augmentation of the anastomosis, or interposition tube graft placement in older patients. It is unclear whether any of these techniques are superior to the others (Fig. 79.13).

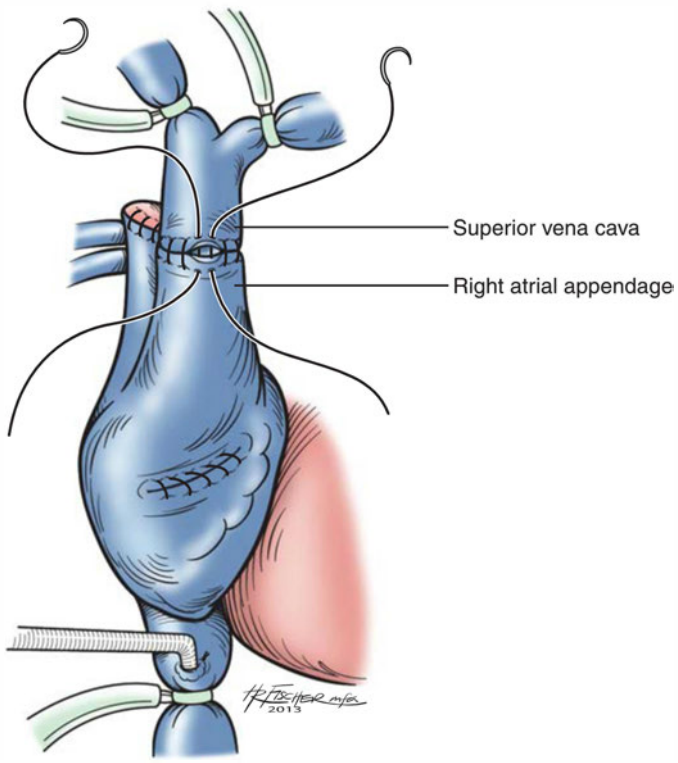


Fig. 79.13. The right atrium is closed and the SVC is anastomosed to the right atrial appendage with fine monofilament suture. If the SVC is small or there is a concern for tension on the anastomosis, the suture line may be interrupted or augmented with a patch.

Repair of the Inferior-Type Sinus Venosus Atrial Septal Defect

The inferior type of sinus venosus ASD is relatively rare and is often a surprising finding at operation. The hallmark of this defect is the absence of a rim of tissue from the IVC to the LA. This defect is positioned just above the inferior cavoatrial junction with the right lower pulmonary vein orifice quite visible through the ASD (Fig. 79.14). Thus, inferior sinus venosus defects are inferior and lateral in position as compared with secundum-type ASDs. It should also be noted that the Eustachian valve is often quite prominent in these patients, and care must be taken to create a suture line around the IVC that does not divert the IVC flow into the left by mistaking the Eustachian valve for the inferior rim of the defect.

Repair of the inferior-type sinus venosus ASD is accomplished by closing the atrial defect with a (pericardial) patch, leaving the inferior pulmonary vein on the left atrial side of the patch and the inferior cava on the systemic venous side of the patch. If it is known in advance (by echocardiography) that an ASD is an inferior sinus venosus type, the IVC cannulation can be performed a bit lower by incising the diaphragmatic pericardium

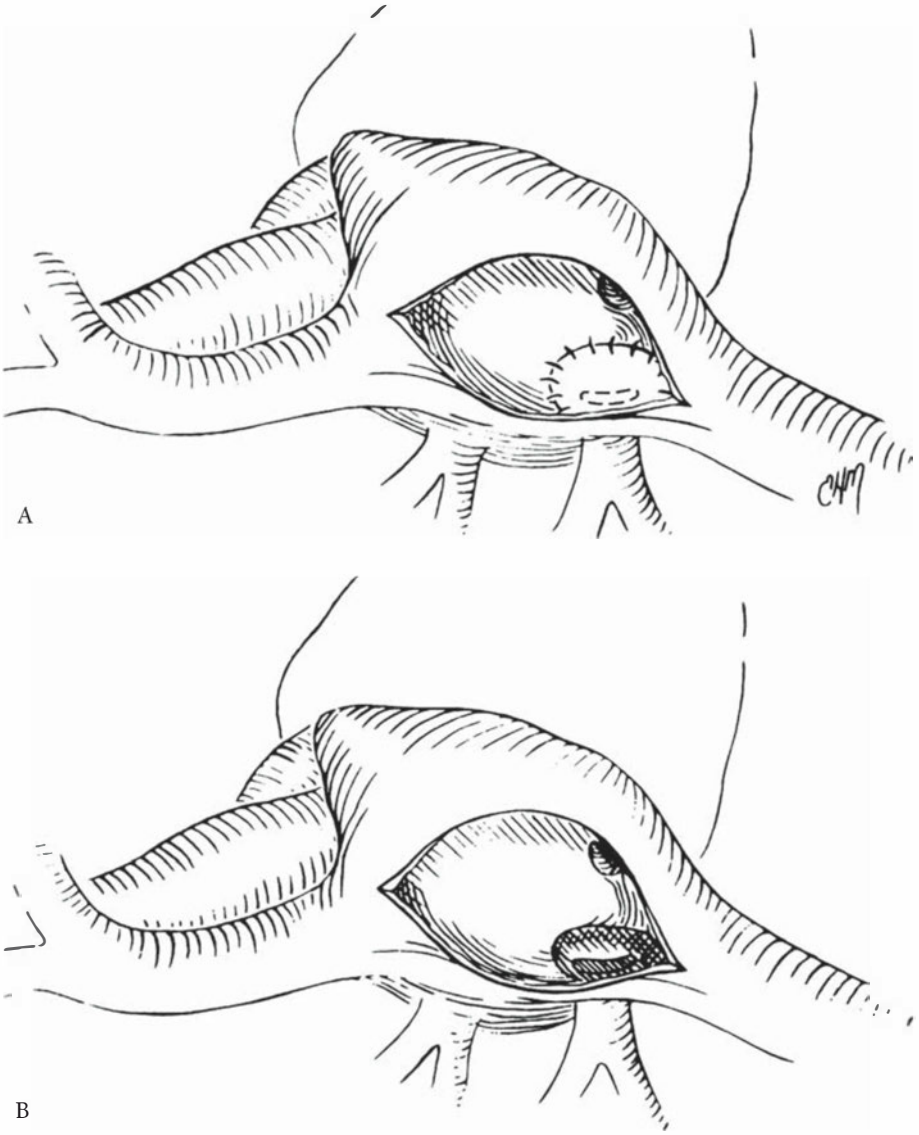


Fig. 79.14. (**A**) The position of an inferior type of sinus venosus atrial septal defect (ASD, being inferior and lateral to the fossa ovalis). Through the ASD, one can see the orifice of the anomalously draining right lower lobe pulmonary vein. (**B**) A pericardial patch is used to perform the repair.

overlying the cava. However, this differentiation between secundum and inferior sinus venosus defects is difficult to make by echocardiography, and statistically the secundum type is far more common. If the IVC is cannulated in a standard position, it may be discovered after the right atriotomy that the cannula overlies the defect. Repair can still be accomplished by clamping and removing the inferior cannula, placing a cardiotomy sucker through the cannulation site, and completing the inferior aspect of the suture line in this manner. The cannula can be replaced once the inferior corner has been completed. The pericardial patch is brought above the entrance of the right inferior pulmonary vein to maintain proper pulmonary venous drainage.

Repair of Coronary Sinus Atrial Septal Defects

Coronary sinus ASDs are relatively rare and are usually associated with complex congenital heart disease or anomalies of systemic venous return. Conceptually, these defects can be divided into those with and those without an LSVC. In the absence of an LSVC, a coronary sinus ASD exists when the coronary sinus is unroofed into the LA. The distal coronary sinus acts as a passageway for blood to shunt from the LA to the RA. When an LSVC is present, it can be the cause of abnormal shunting either when it enters directly or is unroofed into the LA. Coronary sinus ASDs may be associated with a secundum ASD or a common atrium but may also be seen with an intact atrial septum.

The goal in repairing coronary sinus ASDs is to separate systemic and pulmonary return and eliminate shunting at the atrial level. Because these defects are adjacent to the conduction system and the pulmonary veins, care must be taken in planning and performing these repairs in order to minimize morbidity.

Repair of a coronary sinus ASD without an LSVC is accomplished by what is referred to as a "roofing" procedure. The operation is performed using bicaval venous cannulation and a standard right atriotomy. If the atrial septum is intact, the fossa ovalis is incised for exposure. The unroofed coronary sinus is identified medial to the pulmonary veins. A pericardial patch is used to cover the defect (Fig. 79.15). The atrial septum is repaired either primarily or with a second pericardial patch.

Repair of a coronary sinus ASD with an LSVC may be accomplished through one of two principal methods. Selection of the

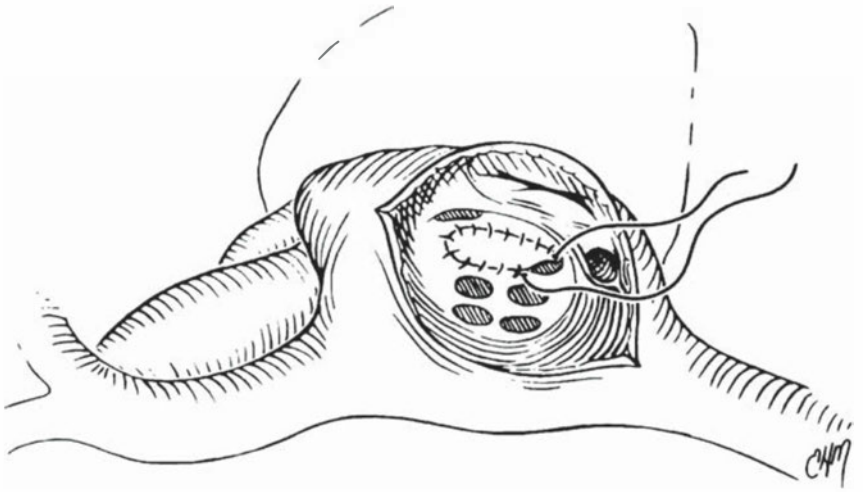


Fig. 79.15. The fossa ovalis has been incised to view the left atrial anatomy. The "unroofed" coronary sinus is shown in its position medial to the four pulmonary veins. A pericardial patch is used to repair the defect.

method depends on the anatomy of each particular case. When the LSVC is small, and particularly when there is a bridging vein, the simplest option is usually to ligate the LSVC and perform a roofing procedure as described previously. However, if the LSVC is large, an intra-atrial baffle technique can be considered. The fossa ovalis is incised to allow adequate inspection of the left and right atrial anatomy. A portion of the septum primum is excised under direct visualization to allow placement of the intra-atrial baffle. Autologous pericardium works quite well for this purpose. The suture line begins between the two AV valves along the rim of the ASD. Anteriorly, the suture line dips below the left atrial appendage and the orifice of the LSVC. Laterally, the suture line extends around the right pulmonary veins. Along the inferior border, the suture line follows the rim of the ASD; superficial bites are taken adjacent to the orifice of the coronary sinus (Fig. 79.16). If there is a common atrium, the orifice of the coronary sinus in the RA may be absent and thus the landmarks for the conduction system less certain. In this case, the baffle is sutured along the annulus of the tricuspid valve and then is carried out onto the right atrial wall to include the area of the conduction system on the left atrial side (Fig. 79.17).

Raghib syndrome refers to an LSVC to the LA with the absence of a coronary sinus and a low-lying ASD (near the position of an inferior sinus venosus ASD). This rare condition is physiologically analogous to a coronary sinus ASD with LSVC and a common atrium (Fig. 79.17). Anatomically, the two are quite distinct because in Raghib syndrome one cannot see the pulmonary veins or left atrial appendage from the RA. Simple closure of the ASD results in persistent desaturation because of the LSVC to the LA. Raghib syndrome should be suspected whenever there is a low-lying ASD with an LSVC. Correction can be accomplished through ASD repair and ligation of the LSVC (relatively simple) or excision of the septum primum and placement of an intra-atrial baffle (complex).

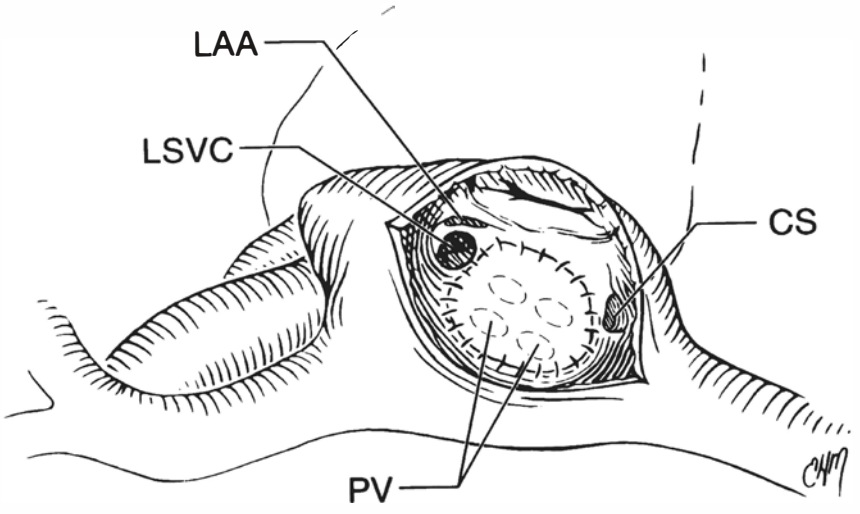


Fig. 79.16. The fossa ovalis has been excised to facilitate the placement of the intra-atrial baffle. The baffle directs pulmonary venous drainage to the mitral valve while the left superior vena cava (LSVC) and coronary sinus (CS) drain to the right atrium. LAA, left atrial appendage; PV, pulmonary veins.

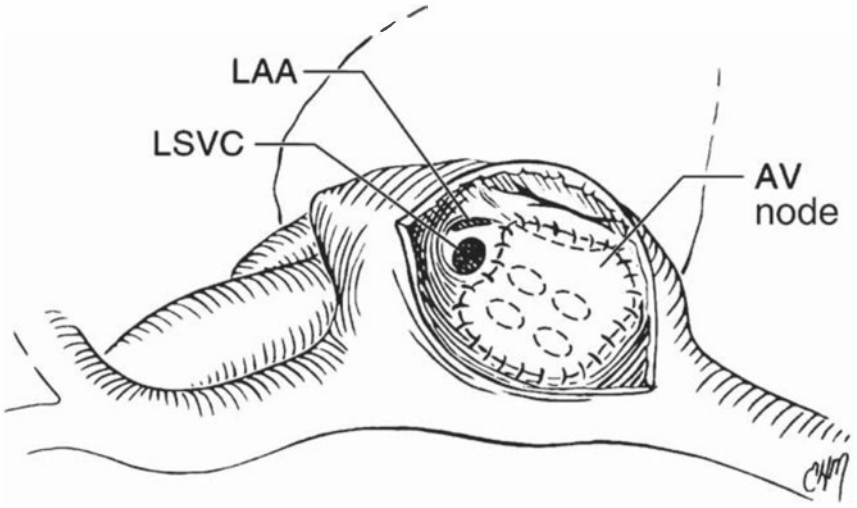


Fig. 79.17. Repair of coronary sinus atrial septal defect with a left superior vena cava (LSVC), common atrium, and absence of a coronary sinus in the right atrium. The presumed location of the atrioventricular (AV) node is shown. In this situation, the medial portion of the pericardial patch is sutured to the hinge point of the tricuspid valve to avoid the bundle of His and the AV node. LAA, left atrial appendage.

Repair of Scimitar Syndrome

Scimitar syndrome is a relatively rare group of findings that may be associated with an ASD. This condition is associated with anomalous venous drainage of the right lung to the IVC at the level of the diaphragm. This anomalous vein creates a curvilinear density adjacent to the right heart border and was likened to the shape of a Turkish sword (or scimitar). In its full-blown manifestation, scimitar syndrome also includes hypoplasia of the right lung, hypoplasia of the right mainstem bronchus, shift of the mediastinum to the right (dextroposition of the heart), pulmonary sequestration to the right lower lobe, and a secundum-type ASD. Not all of the patients have the complete spectrum of abnormalities, thus accounting for the considerable variability among patients with this syndrome.

Operative techniques for the repair of scimitar syndrome have traditionally been (1) baffling of the right pulmonary vein flow through the RA across a secundum ASD to the LA, (2) reimplantation of the anomalous pulmonary veins higher into the RA with baffling across the ASD, and (3) reimplantation of the anomalous pulmonary veins into the LA with closure of the ASD. These techniques have all been associated with failures due to kinking and obstruction of the pulmonary veins. A fourth alternative that may be used when it is apparent that the anatomy is not amenable to any of these techniques is to incise the walls of the RA and the right common pulmonary vein as they travel in parallel and then sew the edges together in a side-to-side anastomosis. This brings the right pulmonary vein confluence more superiorly in the RA and can then be baffled to the LA through the ASD without creating any point at which the baffle becomes narrowed or has an acute change in direction.

Atrial Septal Defect with Failure to Thrive

There is the occasional patient who will present in infancy with a secundum-type ASD and failure to thrive. These patients demonstrate poor eating and poor weight gain, and have the clinical appearance that one would normally associate with infants who have a large ventricular septal defect. It is also of note that these patients invariably have significant pulmonary hypertension, with PA pressures typically one-half to three-fourths systemic. The pulmonary vascular resistance tends to be quite elevated, so that the Q_p:Q_s is usually on the low side (1.5:2.0) for patients with ASDs. The RV is usually noted to be thick due to the increased PA pressures. Some of these patients will have Down syndrome, and others will have a dysmorphic appearance that may be part of an unnamed syndrome. It has been speculated that the primary abnormality in this setting is the pulmonary hypertension and that the ASD is almost an incidental finding.

Surgical closure of a secundum ASD in this setting is usually prompted by the poor weight gain in association with a congenital heart defect. The patients are quite frail due to their poor nutritional status and pulmonary hypertension. Thus, they should be approached with caution much like one would approach an infant with failure to thrive associated with a VSD. The operative technique is identical to the standard approach outlined previously. However, we recommend the additional placement of a PA catheter to measure PA pressures postoperatively. This measure at first may seem unwarranted but will serve as an early warning system for the first time the CO_2 levels increase and the PA pressures rise to systemic or suprasystemic levels. It has been our experience with this entity that many of these patients will continue to demonstrate poor growth after surgical repair of the ASD, but at least the congenital heart defect is removed from the potential list of causes.

CONCLUSION

ASDs are among the more common forms of congenital heart defect. Repair of an ASD is a curative procedure that restores a normal life expectancy. With modern techniques, these procedures can be performed with an extraordinarily low rate of morbidity and mortality.

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EDITOR'S COMMENTS

Although ASD (usually of the secundum type) is one of the most common congenital heart defects seen in any clinical practice, it is not a completely benign lesion. Even small ASDs can be associated with paradoxical embolus late in life, and the association of secundum ASDs and late development of pulmonary hypertension with elevated pulmonary resistance is well known. It appears that there are subsets of patients who in the face of a left-to-right shunt have a predisposition to development of pulmonary vascular disease. Thus, the development of pulmonary vascular disease does not necessarily correlate with the size of the defect or the age of the patient. For these reasons, we believe that even small ASDs should preferentially be closed early in life to prevent late complications. With the advent of occluder devices, it may be possible to close even small ASDs with very little morbidity and eliminate even minor risks of paradoxical emboli or volume overload of the RV with the consequent arrhythmias. As noted by the authors, when an ASD presents in adulthood with only a small shunt, the occurrence of significant pulmonary vascular disease must be suspected and closure might not be indicated if there is any bidirectional or right-to-left shunting.

ASD closure is now commonly being performed for secundum defects with occluder devices such as the Amplatzer device. As noted, there are still complications with these devices, which fortunately are rare, but include erosion into the aorta or through the atrial wall. In addition, as larger devices are being deployed for defects that were not previously amenable to device closure, the incidence of such complications may increase; therefore, continued longitudinal follow-up of patients with occluder devices is necessary. Certainly, a small proportion will eventually require removal and standard ASD closure.

The surgical techniques for the closure of ASDs are numerous. We prefer a vertical skin incision in both male and female patients, especially when the operation is performed in early childhood. These incisions are quite short and quite low to make them as cosmetic as possible. A partial sternotomy incision is generally used. We have avoided transverse incisions in young girls because it may be difficult to identify breast tissue, and if the incision cuts across any breast tissue, unsightly

(*continued*)

EDITOR'S COMMENTS (*continued*)

scars can occur. In addition, we have abandoned the thoracotomy incision for ASD closure in most cases because if the mammary pedicle is damaged, there may be asymmetric breast development. Thus, we believe that the most cosmetic incision may well be a very small low vertical incision in the midline over the xiphoid (2.5 cm in length), which avoids elevation of major skin flaps and usually heals with excellent cosmetic results.

The minimally invasive approach to secundum ASD repair through a small sub-xiphoid incision can be done with the use of electrical fibrillation and normothermic bypass, which avoids the need to cross-clamp the aorta. Even in larger secundum defects without a superior rim, where patch closure is necessary, the procedure can be performed through these small incisions without the need for cardioplegic arrest. Operations for atrial septal defects are generally done under normothermia at our institution because bypass and cross-clamp times are generally extremely short.

Many centers have used different approaches to close ASDs and other simple cardiac lesions with superior cosmetic results. A significant experience has been accumulated in making small axillary incisions for exposure of ASDs and VSDs and other centers have considered posterior thoracotomy approaches and minimally invasive and even robotic techniques through port access. All of these approaches have met with significant success and now, especially in females who require a surgical ASD closure, the selection of the incision for the optimal cosmetic result is generally the primary consideration.

Some comments are warranted on the multiple techniques for repair of sinus venosus-type ASDs of the superior and inferior types. The authors described a technique for the repair of the superior sinus venosus defect that also involves patching of the superior vena cava/right atrial junction. In the majority of cases, sinus venosus defects occur just at the superior vena cava/right atrial junction and the right pulmonary veins enter at that site. In these cases, approach through a right atrial incision as used for secundum defects with gentle retraction on the superior vena cava will expose the defects adequately so that direct patch closure can be performed without narrowing the vena cava as it enters the right atrium. Care must be taken, however, to tailor the patch appropriately so that it does not bulge into the superior vena cava or left atrium. The advantage of this approach is avoidance of an incision across the superior vena cava/right atrial junction, which might interrupt sinus node blood supply. In cases where the right pulmonary veins enter high in the superior vena cava well away from the right atrium we have preferred the use of the Warden-type repair as described in this chapter, in which the superior vena cava is divided and then reconnected to the right atrial appendage and the stump of the superior vena cava, to which the anomalous veins return, is baffled across the atrial septal defect to the left atrium. This approach avoids the need for an incision across the caval–atrial junction that may interfere with sinus node blood supply, and there have been many recent reports of excellent consistent results with this technique with a low incidence of late development of superior vena caval stenosis. If superior vena caval stenosis occurs at the suture line, and if the azygos vein is left open, decompression is often to the IVC, and the SVC can be dilated or stented in the catheterization lab if significant obstruction is present.

The authors described the techniques for repair of scimitar syndrome, which include reimplantation of the anomalous vein or baffling of the vein across an atrial septal defect to the left atrium. They presented a good summary of the various techniques that have been used. Recently, there has been a resurgence of interest in reimplantation of the right-sided pulmonary veins directly into the left atrium through a right thoracotomy incision without the use of cardiopulmonary bypass. An advantage of this technique is the fact that the geometry of the vein can be better assessed with the lung slightly inflated through a thoracotomy incision, which may prevent kinking of the vein as it enters the left atrium. This technique is most applicable when there is not a significant atrial septal defect present. Although it is possible to close the atrial septal defect through a right anterior thoracotomy incision and reimplant the anomalous vein, we believe that if bypass is necessary, a standard sternotomy incision is generally the simplest way to get to the areas of interest and avoids struggling with the division of the right pulmonary vein entrance at the level of the diaphragm. Although the few reports of direct reimplantation off bypass through a thoracotomy have suggested a very good patency rate, the development of stenosis or occlusion of the reimplanted vein over time needs to continue to be assessed before this technique can be adapted routinely.

The greatest complication after repair of scimitar syndrome is stenosis or kinking of the scimitar vein at the anastomosis to the right or left atrium. This can result in occlusion of the vein and late development of hemoptysis, which is extremely difficult to treat. In some cases, stenosis of the vein can be treated with stent implantation in older children. Because of the concerns of obstruction of the scimitar vein, I have most recently been interposing a short segment of PTFE graft between the scimitar vein and the anastomosis to the left atrium through a right thoracotomy approach to prevent stenosis of the suture lines. While follow-up remains short, at this point there appears to be some improvement in flow through the veins with the use of this technique. Essentially, the short segment of PTFE graft acts as a stent to prevent contraction of the suture lines.

It should also be noted that scimitar syndrome presenting in infancy with significant pulmonary hypertension has a very poor prognosis. Because of the high incidence of vein obstruction in scimitar syndrome in adulthood, some authors have recommended that these patients be followed without operative intervention. We still believe, however, that patients with a significant left-to-right shunt and no associated anomalies with no evidence of pulmonary vascular resistance should be considered for surgical therapy with attention to the technical details to prevent vein obstruction.

(*continued*)

EDITOR'S COMMENTS (*continued*)

In patients who present with scimitar syndrome in infancy with pulmonary hypertension, often associated with other left-to-right shunts, assessment of the aortopulmonary collateral circulation to lower portion of the right lung is imperative. It may be possible to palliate many of these children with occlusion of these aortopulmonary collateral vessels to decrease the left-to-right shunt and improve pulmonary hypertension in the newborn and infant. The subsequent need for the repair of the scimitar syndrome is then based on the relative size of the right lung and the relative magnitude of the left-to-right shunt that is flowing through the generally hypoplastic right lung. In some cases, if the flow distribution is markedly to the left lung and away from the hypoplastic right side, repair of the scimitar vein may not be indicated.

The authors commented on the presence of ASDs with failure to thrive in infancy. Although it is certainly not predictable that closure of a secundum ASD will alter the growth patterns of very young children (because generally these shunts are not large), I have seen several patients over the years who have had marked improvement after a moderate-sized secundum defect was closed in infancy. Care must be taken, however, to ensure that these patients do not have additional left-sided cardiac lesions that exacerbate the left-to-right shunt from a relatively small ASD and that can be unmasked after ASD closure. Nevertheless, in patients with no other cause for failure to thrive and a significant ASD, I believe that the low risk of closing the defect is warranted.

TLS

80 Ventricular Septal Defects

Stephanie Fuller

VENTRICULAR SEPTAL DEFECTS

Ventricular septal defects (VSDs) are among the most common congenital heart anomalies. Isolated VSDs represent approximately 20% to 30% of all congenital cardiac malformations and have a prevalence of 1 to 2 per 1,000 live births. There is a slight female predominance.

Roger first described the clinical signs and symptoms of a VSD in 1879 as a small, flow-limiting VSD with associated normal pulmonary artery. At the other end of the clinical spectrum is a large VSD with severe pulmonary hypertension and fixed pulmonary vascular resistance resulting in cyanosis with right-to-left shunting through the VSD, first described by Eisenmenger in 1897.

VSDs may be associated with a wide variety of other cardiac defects, including mitral valve disease, atrioventricular (AV) discordance, conotruncal abnormalities such as transposition or double-outlet ventricle, and hypoplasia of either ventricle. They are the most common heart defect in many chromosomal anomalies, including patients with trisomies 13, 18, 21 and with Holt–Oram syndrome. Isolated VSDs and those associated with patent ductus arteriosus and coarctation of the aorta are described in this chapter.

CLASSIFICATION OF VENTRICULAR SEPTAL DEFECTS

The ventricular septum forms between 4 and 7 weeks of gestation and involves fusion of several tissues, including endocardial cushion-derived mesenchyme, primary atrial septum, and muscular components of the atrial and ventricular septum. The muscular intraventricular septum forms by infolding of the ventricular muscle within the primitive cardiac tube. It then aligns with the conal septum that is positioned between the two outflow tracts. The membranous septum closes adjacent to the anteroseptal commissure of the tricuspid valve. VSDs may occur anywhere in the septum.

VSDs are classified based upon their location within the ventricular septum. Spontaneous closure rate and the presence of associated defects depend upon the location of the defect and the type of VSD. Of the many classifications of VSDs, the most widely accepted today are those of Soto and van Praagh (Fig. 80.1). The ventricular septum has three main components: (1) an inlet portion beneath the septal leaflet of the tricuspid valve and extending from the tricuspid annulus to the papillary attachments of the tricuspid valve chordae, (2) a trabecular or muscular portion that extends from the chordal attachments of the tricuspid valve to the apex of the ventricles and cephalad to the conal septum, and (3) a smooth-walled conal or outlet septum, which comprises the infundibular septum clasped between the anterior and posterior limbs of the septal band (trabecular septomarginalis) and extends up to the pulmonary and aortic annuli. The inlet and trabecular portions are often referred to collectively as the ventricular septum, in contrast to the conal septum, which is also called the "outlet" or "infundibular" septum.

Conoventricular (Perimembranous) Defects

Conoventricular defects are the most common isolated VSDs, comprising about 70% to 80% in most series. They are situated in the junctional area between the conal or outlet and inlet portions of the septum and may extend into the inlet, outlet, or both parts of the septum. They extend up to the tricuspid valve annulus in the region of the anteroseptal commissure of the tricuspid valve. The aortic valve is easily visible through the defect when viewed through the tricuspid valve at operation. Alternatively, these defects may be partially occluded or rendered restrictive by overlapping tricuspid valve tissue. Occasionally, the noncoronary aortic leaflet may prolapse through the defect resulting in progressive aortic incompetence.

The conduction system is intimately associated with this VSD. The bundle of His penetrates the right trigone of the central fibrous body at the anteroseptal commissure of the tricuspid valve. From there, it is closely related to the lower half (posteroinferior edge) of the defect, giving off the left fascicles along its course to the medial papillary muscle (also called the muscle of Lancisi or the papillary muscle of the conus). As it passes just inferior to the latter, only the right bundle remains, and this continues into the trabecular septomarginalis away from the edge of the defect. Conoventricular septal defects are usually repaired through the right atrium.

Conal Septal Hypoplasia (Outlet) or Conotruncal Defects

Conotruncal defects are also called supracristal, subarterial, subpulmonary, juxtaarterial, or infundibular. They account for 5% to 10% of isolated defects, except in the Asian population in whom they are more common accounting for up to 30% of VSDs. Typically, the defects are oval in shape and extend up to the pulmonary and aortic annuli. Because the normal subpulmonary conus is deficient, the pulmonary and aortic valves are separated only by a very thin rim of fibrous tissue that maintains fibrous continuity between the aortic and pulmonary valves. This results in aortic leaflet prolapse, which occurs in 40% to 50% of conal defects. The right aortic leaflet most commonly is sucked into the defect, resulting in aortic insufficiency. The conduction tissue is remote from the borders of this type of VSD, which is most easily repaired through a short transverse incision in the right ventricular outflow tract, or through the main pulmonary artery (Fig. 80.1). These defects will not close spontaneously

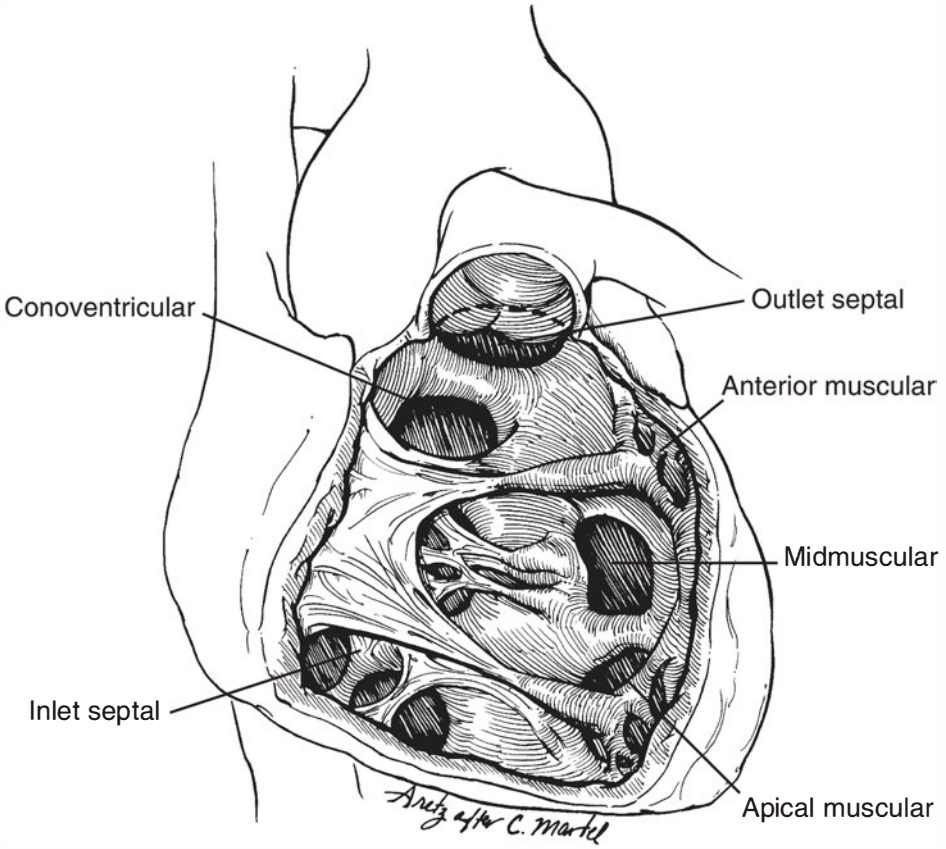


Fig. 80.1. The right ventricular free wall has been resected to show the ventricular septal defects: conoventricular (perimembranous); conal septal (subpulmonary or outlet); atrioventricular (inlet); muscular that may be midmuscular, anterior, or apical. The penetrating bundle is closely related to the inferior margin of the conoventricular defect and diverges away from this margin into the trabecular septomarginalis beneath the muscle of Lancisi.

but can be significantly restricted by a prolapsed aortic valve leaflet.

Inlet Septal Defects (Atrioventricular Canal-Type Defects)

Inlet septal defects account for about 5% of isolated defects. They are situated beneath the septal leaflet of the tricuspid valve, with the tricuspid valve annulus forming their posterior border. The tricuspid valve may straddle across the defect with attachments into the left ventricle. The conduction tissue is closely related to the posteroinferior border of the defect, up to the muscle of Lancisi, at which point the right bundle branch diverges down the trabecular septomarginalis to form the moderator band. The AV septum is intact, although occasionally the anterior leaflet of the mitral valve may be partially cleft. Inlet septal defects will not close spontaneously and are best repaired through the tricuspid valve.

Muscular Defects (Trabecular Defects)

Muscular VSDs account for 10% to 20% of isolated VSDs, may be single or multiple, and may occur in any part of the muscular septum. They may be associated with other types of VSDs. They are generally classified depending upon their location in the septum and may be divided into midmuscular defects (most common), apical or posterior muscular, and anterior muscular defects (Fig. 80.1). Muscular defects may have numerous openings of variable size on the right ventricular side but only a single opening on the left ventricular side of the septum. This is referred to as a "Swiss cheese" defect. The conduction tissue is generally remote from the edges of a muscular defect, with two notable exceptions: (1) when associated with a conoventricular defect, the penetrating bundle usually runs in the muscle bridge separating the two defects and may easily be injured if each are closed individually and (2) if a muscular defect is present in the inlet septum (i.e., the defect is separated from the tricuspid valve by a thin rim of muscle), the conduction tissue runs along the defect superior and anterior (leftward) margin—the AV node penetrates the interventricular septum at the anteroseptal commissure of the tricuspid valve and takes the most direct route to the medial papillary muscle along the superior border of the defect. Muscular defects often close spontaneously. Surgical approach is variable based upon the location of the defect.

PATIENT CHARACTERISTICS

Both the relative pulmonary vascular resistance and the size of the defect determine the hemodynamic effect of the VSD. In the early newborn period, when pulmonary and systemic vascular resistances are relatively equal, there is little or no shunting of blood across the defect. As the pulmonary vascular resistance declines in the first weeks of life, the degree of shunting across the defect increases. Shunting occurs largely during systole and is directed to the lungs and the left side of the heart, causing left heart overload. In infants with a hemodynamically significant VSD, increased pulmonary blood flow from moderate-to-large defects leads to symptoms and signs of heart failure and to left atrial and left ventricular dilation.

Infants and children may not develop symptoms early in life unless the VSD is large. Patients with symptomatic VSDs present similar to those of heart failure, including tachypnea, tachycardia, increased work of breathing and, in some cases, repeated pulmonary infections. Those patients with large VSDs are often too tachypneic to feed orally, experience recurrent chest infections and aspiration of gastric contents, and may have pulmonary hyperinflation syndrome or cardiac asthma. The latter results from systemic pressure in the segmental pulmonary arteries compressing the small bronchi and resulting in chronic air trapping. Many of these infants fail maximal medical management aimed at decreasing the symptoms of heart failure.

DIAGNOSIS

The size, number, and location of VSDs can be accurately defined by echocardiography. Estimates of pulmonary artery pressure can be made using Doppler imaging of the velocity of a tricuspid regurgitant jet, if present. Defects are considered "large" if they approximate the size of the aortic annulus or result in systemic pulmonary artery pressures. Chronic elevation in

pulmonary blood flow can alter the pulmonary vascular bed, leading to intimal proliferation and muscularization of the arterioles known as Eisenmenger syndrome, a condition where the increased pulmonary vascular resistance leads to pulmonary hypertension and right-to-left shunting across the VSD. Ultimately, this can result in cyanosis.

VSDs are also characterized by the degree of shunting by catheterization. A simple formula for calculating the Q_p:Q_s ratio using oxygen saturations measured by cardiac catheterization is as follows:

$$Q_p:Q_s = (\text{Ao\%}-\text{RA\%})/(\text{PV\%}-\text{PA\%})$$

"Medium"-sized defects result in a Q_p:Q_s ratio of 2:1 to 3:1 and a systolic pulmonary artery pressure that is 40 to 50 mmHg or about one-half that of the aorta. "Small" defects have essentially normal pulmonary artery pressures and a Q_p:Q_s ratio of <1.5:1.

INDICATIONS FOR OPERATION

More than 75% of small VSDs will close by fibrosis and muscular hypertrophy within the first 2 years of life. Closure of a VSD is recommended once a large VSD is detected or once a patient becomes symptomatic. Thus, infants with large VSDs presenting in the first few months of life with severe congestive heart failure should undergo prompt repair. Delaying surgery until the patient is "bigger" is not beneficial and often results in additional morbidity and mortality.

Historically, small infants with a large VSD were initially palliated by placing a constricting band around the main pulmonary artery, suturing this to the adventitia, and gradually narrowing the band circumference until the systolic pressure in the pulmonary artery distal to the band was reduced by 50%. This decreased the flow through the pulmonary artery, ameliorated congestive cardiac failure, and allowed the patient to grow to a large enough size for the band to be removed and the VSD to be safely repaired. However, as methods of myocardial protection advanced and the surgical skills and postoperative care of small infants improved, pulmonary artery banding was abandoned in favor of primary closure of the VSD except in rare cases.

When reviewing the indications for surgical closure of a VSD, there are four aspects to take into consideration: (1) characteristics of the defect, (2) the patient's age and symptoms, (3) pulmonary vascular resistance, and (4) associated cardiac and noncardiac defects.

Any VSD associated with failure to thrive should be closed. If a VSD is associated with prolapse of the aortic valve leaflet resulting in even mild regurgitation, this should be repaired irrespective of the symptoms or size of the defect. In addition, surgical repair is recommended for inlet or outlet types as they are not likely to close spontaneously. The same is true for residual VSDs typically >3 mm in size.

The exception to early primary closure in a symptomatic infant is the presence of multiple muscular defects, which is still associated with significant mortality when repaired in infancy. This is one of the few remaining indications for pulmonary artery banding. Debanding of the pulmonary artery and repair of the VSDs are done when the patient is about 6 to 12 months of age. Some muscular defects may be suitable for percutaneous catheter device closure by our cardiology colleagues. This can be done either before operation, intraoperatively with a hybrid approach or postoperatively. Apical and anterior muscular defects are often difficult to adequately close in the operating room and may be closed percutaneously.

Infants with symptoms controlled with medical therapy and large defects that have not appreciably decreased in size should be electively repaired between 6 months and 1 year of life. Most of these infants experience poor weight gain and failure to thrive. Many will develop increased pulmonary vascular resistance. These infants are considered to have "reactive pulmonary hypertension" and are at increased risk for developing pulmonary hypertensive crises in the postoperative period, particularly if their surgery is delayed.

Beyond 12 to 18 months of age, patients with large VSDs should undergo cardiac catheterization and be repaired if their pulmonary vascular resistance is <8 to 10 U/m^2 while inspiring 100% oxygen or on nitric oxide thus demonstrating reversibility or the Q_p:Q_s ratio exceeds 1.5:1 at rest. Pulmonary vascular resistance is seldom prohibitive during the first year of life in patients with an isolated VSD, except occasionally in patients with associated Down's syndrome. These patients tend to develop pulmonary vascular obstructive disease at an earlier age.

In a patient with high fixed resistance, the defect should not be closed. When small defects are discovered later in childhood, spontaneous closure is unlikely. These defects should be closed if any complications develop including endocarditis and psychological trauma including sports restriction.

ASSOCIATED CARDIAC AND NONCARDIAC PATHOLOGIC CONDITIONS AFFECTING CLOSURE

Infants with significant additional left-to right shunts, such as an atrial septal defect or patient ductus arteriosus, usually have intractable congestive heart failure in early infancy. Early primary closure of all defects is thus indicated. There is a slightly increased risk and morbidity associated with repair of these infants, which relates mainly to their poor preoperative condition. In contrast, neonates and young infants with associated coarctation of the aorta should first have the coarctation repaired through a left thoracotomy if feasible. If they cannot be weaned from the ventilator or continue to have symptoms after the coarctation repair, prompt repair of the VSD is advised during the same hospitalization. In the recent era, however, studies have also shown that the single-stage approach of simultaneous VSD and coarctation repair via a median sternotomy can be performed with comparably low morbidity and mortality.

There are a few caveats to this approach: (1) coarctation and associated multiple muscular defects in which pulmonary artery banding may be done at the time of coarctation repair via the same incision, (2) coarctation and associated large outlet septal (subpulmonary) defect in which simultaneous primary repair is indicated as this type of VSD does not usually undergo spontaneous closure, and (3) VSD and associated interruption, which are best simultaneously repaired though a midline approach.

About 25% of infants undergoing repair of isolated VSD have some noncardiac morphologic syndrome or pathologic condition (i.e., Down's syndrome, VACTERL syndrome, tracheoesophageal fistula, etc.) associated with their VSD. In very symptomatic patients, it is often difficult to quantify the contribution to symptoms of a medium-sized defect. Early closure of the cardiac defect is, therefore, reasonable.

TECHNIQUE OF REPAIRING VENTRICULAR SEPTAL DEFECTS

For premature infants or small patients younger than 2 months of age (or <3.5 kg in weight), the technique of profound hypothermic circulatory arrest may be used for repair. The right atrial appendage is cannulated with a single 16-Fr right-angled venous cannula and the ascending aorta is cannulated with an 8- or 10-Fr cannula.

The ductus arteriosus is ligated while the patient is being cooled. The ductus should always be ligated before the circulation is arrested to prevent air in the opened right heart from entering the aorta inadvertently through a small undiagnosed patent ductus arteriosus. Once the target temperature has been reached, the aorta is clamped, and a single dose of cold blood cardioplegia is given via a cardioplegia needle in the ascending aorta. The circulation is then stopped, and the venous cannula is removed once the patient has been drained.

For older infants, bicaval cannulation is used and the repair is done with either normothermia or mild hypothermia (32 to 26°C) and cold blood cardioplegia. A conventional method for cannulating the superior vena cava (SVC), in especially with very small infants, is to first cannulate the right atrial appendage with a small but straight venous cannula and commence cardiopulmonary bypass (CPB). After commencement of CPB, that cannula is passed into the SVC and a caval tape placed around the SVC. The ductus is ligated once CPB is commenced.

After the aorta has been clamped and the heart arrested with cardioplegia, the right atrium is opened with an oblique incision (Fig. 80.2). A pump sucker is placed in the left atrium via a patent foramen ovale or creation of an ASD via a stab incision in the atrial septum to keep the operative field dry. Alternatively and depending on the size of the patient, the pump sucker or LV vent can be placed through the left atrial appendage or the right superior pulmonary vein.

Occasionally, the defect is small enough to close using interrupted horizontal mattress suture of felt pledgeted 5-0 polypropylene sutures. More typically, some sort of patch material is needed for repair and polyester (Dacron) velour is the most commonly used patch material. Alternatively, fresh or glutaraldehyde treated autologous pericardium or polytetrafluoroethylene (Gore-Tex) surgical patch may be used. The advantage of using Dacron is the vigorous endocardial reaction that it stimulates, expediting the spontaneous closure of small residual defects that may remain after closure.

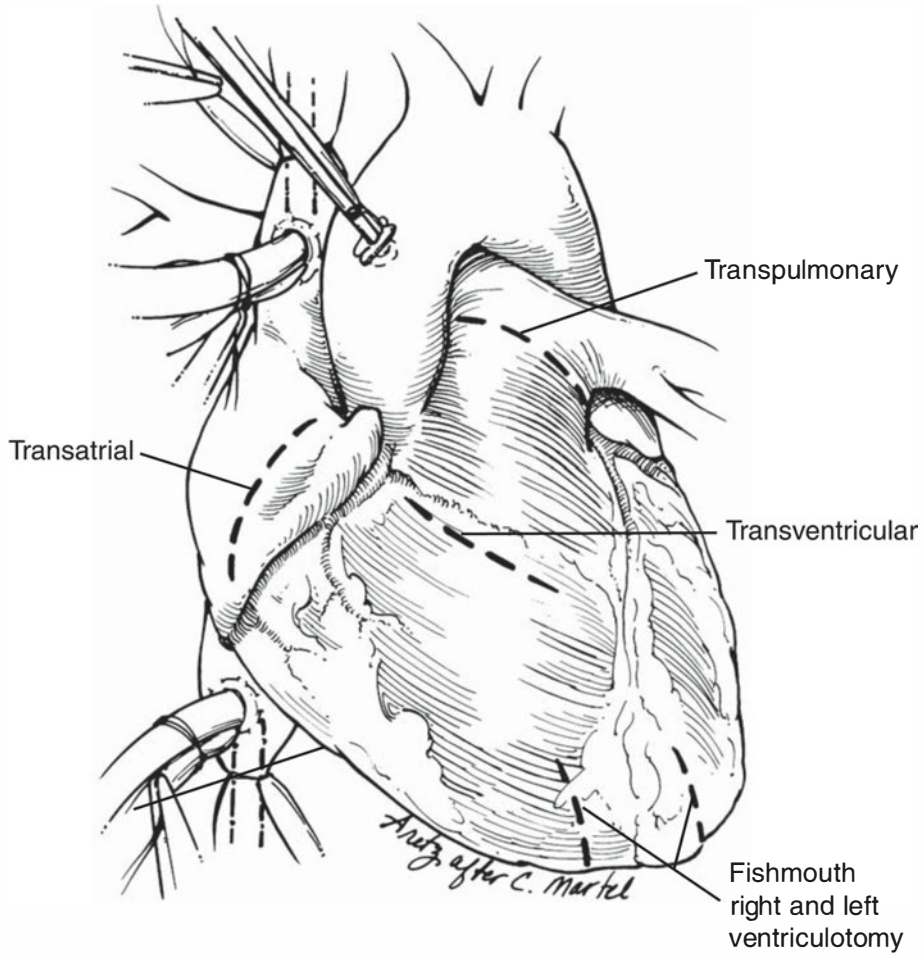


Fig. 80.2. The common incisions through which ventricular septal defects are repaired.

Repair of Conoventricular Defects

Conoventricular defects are usually repaired through the right atrium, which is opened with an oblique incision parallel to the right AV groove (Fig. 80.2). A small retractor is used to retract the tricuspid anterior leaflet to visualize the VSD. This retractor can then be rotated cephalad to expose the superior margin of the defect in the vicinity of the anteroseptal commissure. Gentle caudal traction on a small pump sucker placed through the VSD brings into view the superior and anterior margins of the defect. The anteriosuperior triangle where the aortic valve, tricuspid annulus, and parietoinfundibular fold meet is the most difficult area to expose and therefore the most common site of residual lesions. Attaching the VSD patch to trabeculated RV muscle in this area as opposed to true conus or even aortic valve annulus may result in residual "intramuscular" VSDs that are challenging to repair.

The VSD is repaired with a Dacron-felt patch, which is cut slightly larger than the defect but with a similar shape. Patch material can be sewn in place with continuous suture technique, although an interrupted technique with felt pledgeted sutures has been well described. As viewed by the surgeon through the tricuspid valve, a polypropylene suture is passed through the right side of the interventricular septum, close to the muscle of Lancisi, approximately 2 mm away from the edge of the septum. The suture is extended caudally until exiting through the septal leaflet of the tricuspid valve adjacent to the annulus. The other end of the suture is continued along the superior margin of the VSD with great care not to injure the tricuspid valve. Once this aspect is complete, this suture is continued as a horizontal mattress suture via the septal leaflet and medial margin of the patch that allows the patch to lie below the leaflet. The two ends of the suture are then tied on the right atrial side of the tricuspid valve with or without a pledget (Fig. 80.3). If the septum is very friable or sutures appear to have torn through the septal muscle, it is advisable to add interrupted pledgeted horizontal mattress sutures to secure the patch and avoid dehiscence.

Upon completion of the VSD patch, the sucker is withdrawn from the PFO and the left atrium instilled with saline solution as a deairing maneuver. Depending upon surgeon preference, the lungs are inflated as a Valsalva maneuver to further deair the left heart prior to PFO closure. The PFO is then closed with a single 5-0 polypropylene suture. The tricuspid valve is tested for

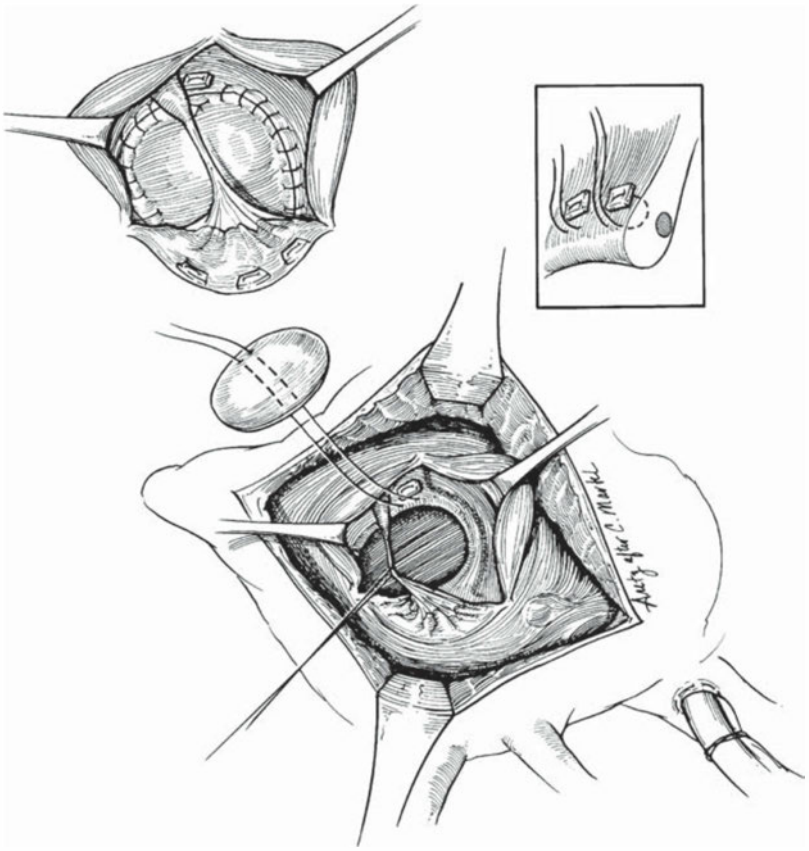


Fig. 80.3. Transatrial closure of a ventricular septal defect using a continuous suture technique. The first suture is placed just inferior to the muscle of Lancisi and about 3 mm from the edge of the defect. To the right of this suture, the conduction fibers are vulnerable up to the point where the suture is passed through the tricuspid septal leaflet. To the left of the first suture is the "safe" area. (*Inset*) The completed repair.

competence and repaired if necessary. The right atrium is then closed in a running manner, typically as an epicardial closure inverting any endocardial tissue.

In the case of abundant tricuspid valve chordae or attachments to the septum, the VSD is better visualized by incising the septal leaflet of the tricuspid valve approximately 2 mm from the annulus (Fig. 80.4). By reflecting the leaflet anteriorly, the VSD is exposed with clear, easily visualized margins. The leaflet is then reapproximated using a continuous suture, typically the same suture anchoring the Dacron patch to the septal leaflet. An alternative approach, although more challenging, is to divide the obstructing chordae to the septal leaflet and then repair by suturing the chordae to the septum or to the patch using Gore-Tex sutures.

An alternative approach to a VSD is through a right ventriculotomy. This approach is convenient in cases of RV muscle hypertrophy that require resection. It is also a useful approach in some cases in which the most anterosuperior aspect of the VSD is difficult to visualize through the tricuspid valve. In these cases, a vertical ventriculotomy is performed parallel to the left anterior

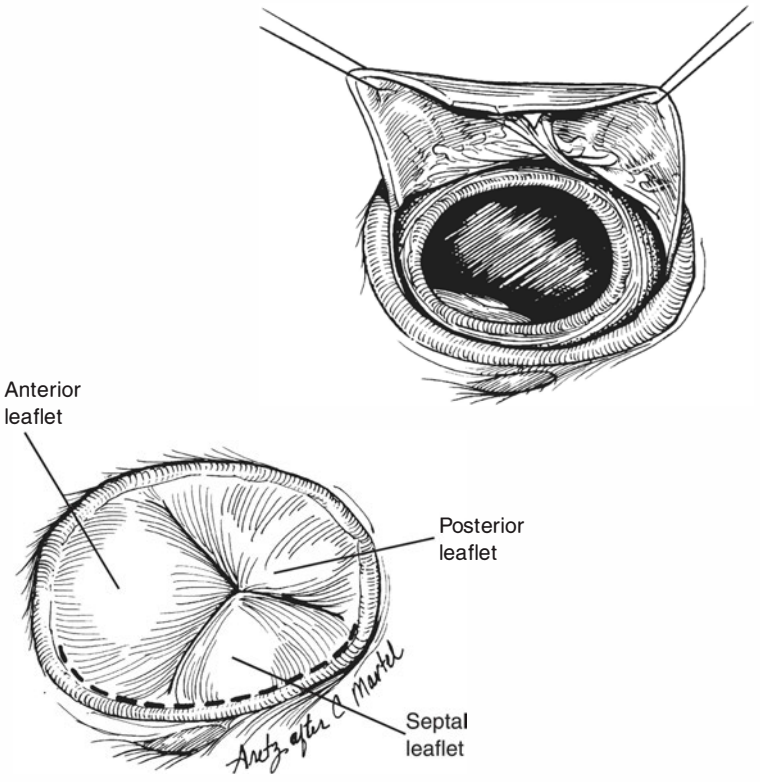


Fig. 80.4. The tricuspid leaflets may be detached 1 to 2 mm from the annulus to improve exposure in some conoventricular or inlet septal defects. Note that the atrioventricular node penetrates the right trigone of the central fibrous body at the anteroseptal commissure of the tricuspid valve.

descending artery. Great care must be used to inspect the surface of the ventricle for the coronary pattern. An oblique incision is recommended in situations in which the right coronary originates from the left anterior descending and crosses the RV outflow tract or when there is a "dual supply" left anterior descending with a large conal branch extending from the right coronary artery. The VSD is then closed in a continuous manner (Fig. 80.5).

Repair of Inlet Septal Defects

These defects are generally repaired through the right atrium similar to conoventricular defects. Detachment of the septal leaflet is rarely necessary because the inlet (AV canal type) defects are easily exposed through the tricuspid valve. In these cases, it is important to make the patch ovoid or even in a semi-lunar shape as the defect tends to be less circular. A circular patch can interfere with the tricuspid valve. It is important to differentiate an inlet septal defect from a muscular septal defect in the inlet septum because, in the latter, the bundle of His runs in the thin rim of muscle separating the VSD from the tricuspid valve annulus.

Repair of Conal Septal Defects

Defects in the infundibular septum are approached through the pulmonary valve via a transverse incision in the main pulmonary artery (Fig. 80.2) or through the aorta itself. Conal septal defects are juxtaarterial, the superior margin of the defect being the pulmonary and aortic valve annuli. These are frequently separated by a thin, fibrous rim that is inadequate itself to support sutures. In the repair, interrupted pledgeted sutures are placed through the annulus of the pulmonary valve so that the pledgets lie within the right ventricle below the valve (Fig. 80.6). The inferior margin of the defect is closed with a patch if necessary otherwise buttressed to the annulus with a pledgeted suture in those VSDs measuring <3 mm in size. In these cases, the conduction tissue is remote from the margins of the VSD. In the case of conal septal defects with aortic insufficiency due to a prolapsing leaflet, the aortic valve must be inspected. Typically, closure of the VSD alone resolves the Venturi or "windsock" effect of prolapse and associated insufficiency. The right coronary leaflet is most commonly involved and may be resuspended if necessary by performing a Trusler-type resuspension of the commissures. Yacoub describes concomitant resuspension of the valve leaflets with VSD closure.

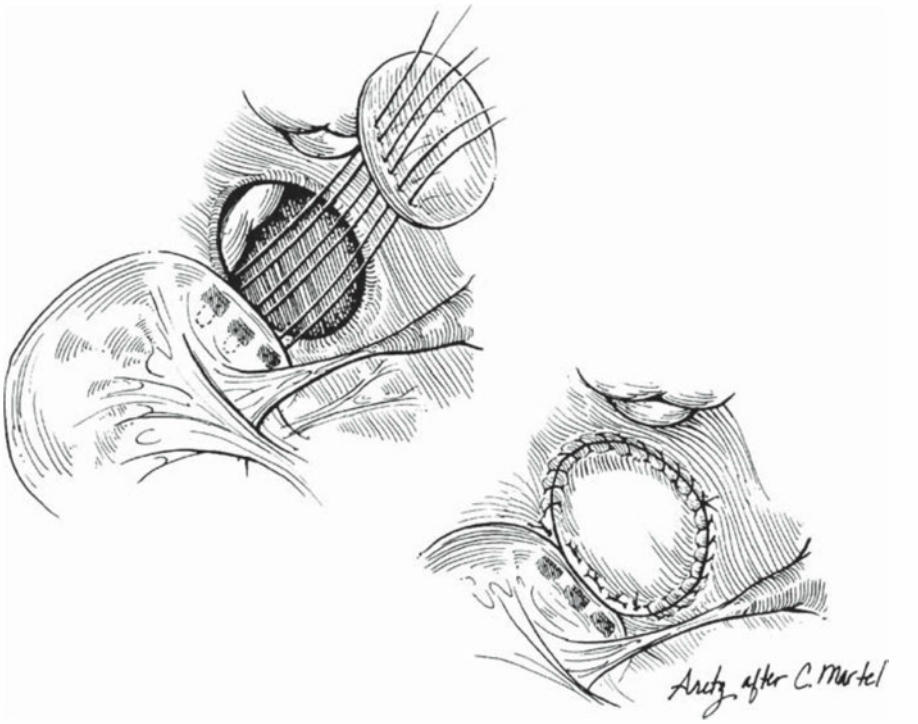


Fig. 80.5. Transventricular closure of a conoventricular defect. Initially, interrupted pledgeted horizontal mattress sutures are passed from the right atrial side through the tricuspid valve leaflet, and then through the Dacron patch. These are then tied down and the suture line completed with a continuous 5-0 polypropylene suture. The most vulnerable area is the transitional area between the septal leaflet and the muscle of Lancisi (medial papillary muscle). Sutures should be placed 3 to 4 mm from the edge of the defect in this area.

Repair of Muscular Ventricular Septal Defects

Midmuscular defects are closed in a similar manner to conoventricular defects, through the right atrium. When both a conoventricular and sizeable mid-muscular defect are present, one large patch may be used to close both defects in order to avoid injury to the conduction system. Anterior muscular defects are closed either through the tricuspid valve or via a ventriculotomy (Fig. 80.6). Multiple muscular defects maybe obscured by trabeculations in the right ventricle. Often these, trabeculations may be divided in order to adequately visualize the VSD. If the margins of the muscular defect cannot be identified through the tricuspid valve, it may be beneficial to pass a right angle clamp through the mitral valve and probe the ventricular septum carefully through the left side. Two pieces of Dacron or felt may then be used to "sandwich" the defect between the two layers of felt (Fig. 80.7).

Apical muscular defects and multiple "Swiss cheese" defects are the most difficult to repair. Many of these defects will close spontaneously. If they cause significant heart failure in infancy, PA banding is recommended. Upon repair, they are approached through the right atrium. However, if they cannot be repaired satisfactorily through the tricuspid valve, they may be approached via an apical left ventriculotomy. Upon doing so, the apical defects are well visualized and may be patch closed with running suture. An alternative therapy is the percutaneous approach with deployment of the Amplatzer device.

RESULTS OF OPERATION

Hospital mortality associated with the closure of isolated VSDs is <1%. Incremental risk factors for mortality are (1) multiple defects resulting in a Swiss cheese septum, (2) associated additional left-to-right shunts such as large atrial septal defects, and (3) severe associated noncardiac anomalies. Small patient size is no longer a risk factor for mortality, although prematurity and size <2 kg may be an indicator of increased morbidity.

Intraoperative transesophageal echocardiogram is performed routinely in the operating room upon separation from CPB to evaluate for the presence of a residual VSD, aortic or tricuspid valve competence, and ventricular function. If there is a question about the significance of a residual

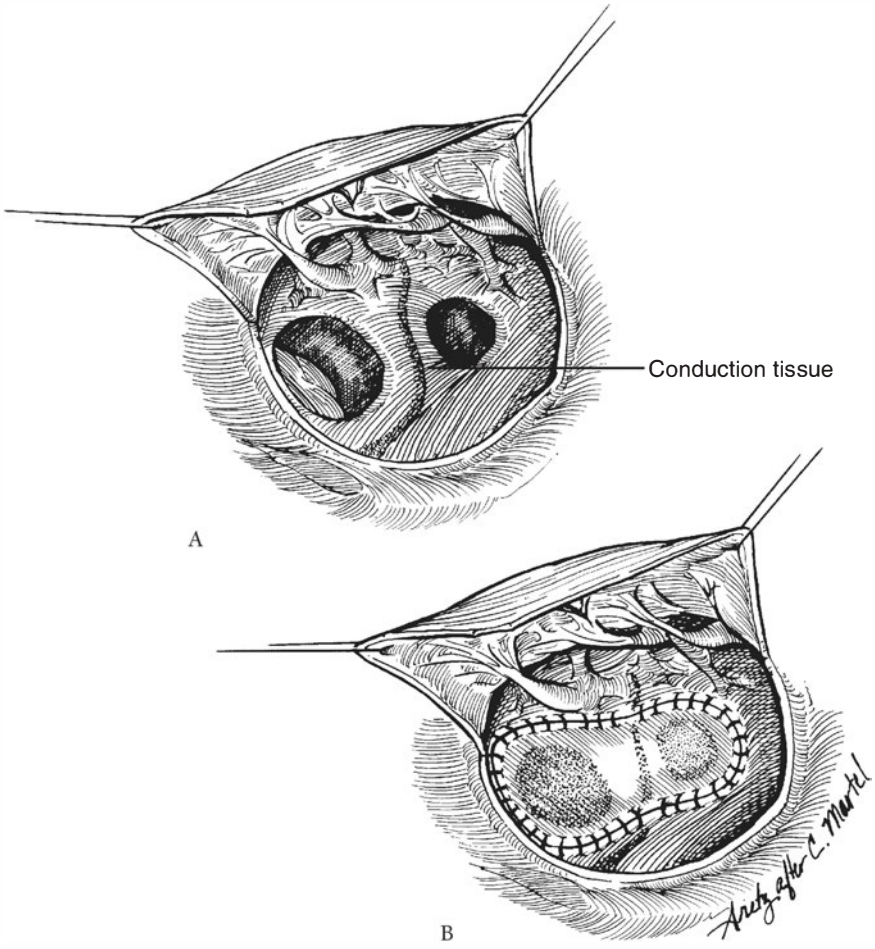


Fig. 80.6. Part of the anterior and septal leaflets of the tricuspid valve has been detached to improve exposure of a conoventricular and midmuscular septal defect. The penetrating bundle lies in the muscle bridge between the two ventricular septal defects. (**A**) The defects are closed using a single composite patch to avoid injury to the conduction tissue. (**B**) Once this has been completed, the tricuspid leaflets are sutured back on the annulus with a continuous 5-0 polypropylene suture.

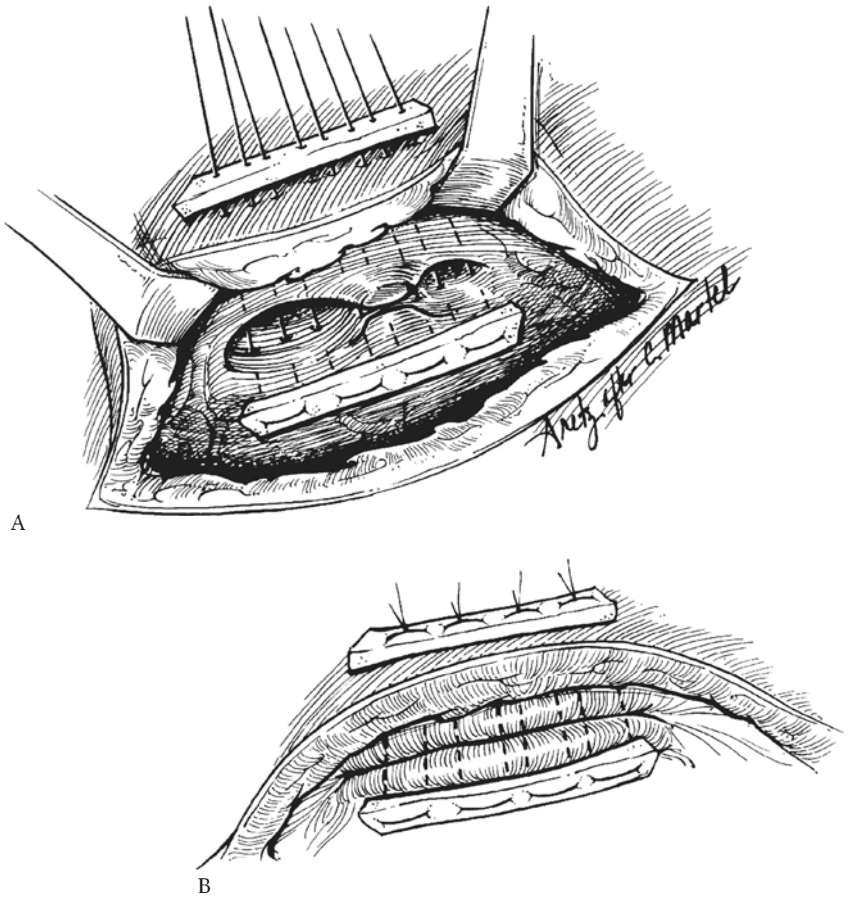


Fig. 80.7. Closure of multiple anterior muscular defects through a short vertical right ventriculotomy. (**A**) The defects are sandwiched between two strips of felt or pericardium, one placed inside and the other outside the right ventricle parallel to the left anterior descending coronary artery. (**B**) Interrupted horizontal mattress sutures are used.

VSD, direct measurement of pulmonary artery pressure and oxygen saturations can be made and the Q_p:Q_s estimated. Small residual defects (<3 mm) usually close spontaneously and only need reoperation in symptomatic patients with a Q_p:Q_s ratio of exceeding 1.5:1 or if pulmonary artery pressures remain significantly elevated. Further, cardiac intervention is required in fewer than 5% of patients and long-term survival is excellent after surgery. Complete heart block occurs in approximately 1% to 3% of patients. A right bundle branch block is usually present in patients undergoing right ventriculotomy for closure. The presence of a right bundle branch block postoperatively is not associated with any long-term morbidity.

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EDITOR'S COMMENTS

The knowledge of the anatomy and techniques of closure for the various types of VSDs is critical because VSDs, either isolated or in combination with other defects, are perhaps the most common malformation addressed surgically in congenital heart disease. The approaches to VSDs and techniques for avoiding interfering with the conducting tissue are well outlined in this chapter.

There are several types of VSD that demand specific attention. VSD of either the conoventricular or conoseptal hypoplasia variety associated with prolapse of the right or noncoronary leaflet of the aortic valve is a specific circumstance that can be approached by various operative techniques. It is generally believed that the loss of support for the aortic valve leaflet and commissure causes progressive prolapse of the valve leaflet into the defect and can cause progressive aortic insufficiency. Therefore, the presence of even mild aortic insufficiency in association with a VSD is considered an indication for operative intervention. In most cases, simple closure of the defect either primarily or with a patch can resuspend the aortic valve leaflets and cause either improvement or stabilization of the degree of aortic insufficiency. In unusual cases, more direct repair of the prolapsed aortic valve leaflet is required with techniques that include triangular resection of the central portion of the prolapsing in aortic valve leaflet or Trusler-type plication of the leaflet at the level of the commissural attachment. In some cases, the defects can be repaired primarily through the aorta with pledgeted sutures placed on the left ventricular side of the ventricular septum through the base of the aortic valve leaflet with plication of the sinus of Valsalva posterior to the aortic valve prolapse, recreating the support for the aortic valve leaflet and dealing with the prolapsed and enlarged sinus of Valsalva simultaneously (Yacoub MH, Khan H, Stavri G, et al. Anatomic correction of the syndrome of prolapsing right coronary aortic cusp, dilatation of the sinus of Valsalva, and septal defect. J Thorac Cardiovasc Surg 1997;113:253). Primary closure with pledgeted sutures can also occasionally be performed through the right atrium.

The approach to closure of conoseptal hypoplasia VSDs through the pulmonary artery is described in this chapter. As noted by Dr. Fuller, it is important when closing these defects to pay attention to suture placement in the base of the leaflets of the pulmonary valve in case later pulmonary autograft valve replacement is necessary. Although in this chapter the technique for suturing conal septal hypoplasia VSDs discusses suturing at the pulmonary annulus, it should be noted that there is no true pulmonary valve annulus since there is no fibrous trigone related to the pulmonary valve. The sutures are placed in the base of the sinuses of the pulmonary valve with care being taken not to interfere with the base of the valve leaflets. On several occasions, we have been able to use the pulmonary valve for autograft aortic valve replacement in patients who had previous closure of conoseptal hypoplasia defects, either by primary closure or with the use of a patch, if the pledgeted sutures in the base of the pulmonary valve leaflet do not distort the valve unduly.

A continuing controversy is the appropriate approach to a symptomatic infant with multiple VSDs. Although pulmonary artery banding and later repair remain a mainstay of therapy in these patients, the progressive right ventricular hypertrophy associated with pulmonary artery banding can often cause additional obscuring of the margins of the ventricular defect from the right ventricular side. In addition, after banding, recatheterization may not identify residual VSDs, which may have little flow in the presence of a tight pulmonary band. After debanding and closure of the identifiable VSDs, with regression of right ventricular hypertrophy, additional muscular defects may reappear. We have, therefore, advocated a primary approach to multiple VSDs in infancy to close the majority of the defects while ventricular hypertrophy is not severe. In most cases, the defects can be identified. For apical muscular defects, an apical right ventriculotomy may be suitable for closure. Apical left ventriculotomies have the disadvantage of cutting through significant myocardium of the systemic ventricle; however, apical left ventriculotomies have not usually been associated with significant left ventricular dysfunction. In addition, in some cases when multiple anterior muscular defects are associated with a conoventricular defect, it is possible to expose the muscular defects and identify them by passing a right-angled

(*continued*)

EDITOR'S COMMENTS (*continued*)

clamp through the conoventricular defect and exploring the ventricular septum. The entrance points in the right ventricle can then be identified and trabeculae divided to expose the margins of the defect for accurate closure. Most apical midmuscular and conoventricular and conoseptal hypoplasia defects can be closed with a high degree of certainty at primary operation. However, anterior muscular defects remain problematic, because they may be difficult to expose through the atrium. In some cases, these defects can be approached through the aortic valve or a right ventricular outflow tract incision. In situations of multiple muscular defects, the Toronto group has described suturing together the trabeculae with fine sutures working through the tricuspid valve. In this manner, the entrance points of defects in the right ventricle can be closed and even multiple muscular defects can satisfactorily dealt with.

In situations where the margins of the muscular defect cannot be readily identified, it may be possible to pass a right-angled clamp through the mitral valve from a transatrial septal approach across the defect into the right ventricle. A large polypropylene suture can then be brought back into the left ventricle and left atrium and a piece of Teflon felt cut larger than the anticipated left ventricular side of the defect. The patch can then be secured with the suture and brought through the mitral valve on to the left side of the septum. The suture is then brought through a similarly shaped Teflon-felt patch on the right ventricular side and tied. In this manner, the defect is sandwiched between the two layers of felt and the actual direct margins of the defect do not have to be identified. This technique can be particularly useful in "Swiss cheese"-type ventricular septal defects or occasional anterior muscular defects that are obscure.

The advent of transcatheter closure devices has aided surgical intervention for patients with multiple VSDs. In patients in whom residual defects are present or where there are multiple midmuscular defects that can be approached with a transcatheter device, either transcatheter closure before operative closure of remaining defects or after closure of the surgically accessible defects may be associated with a high degree of success.

There has been a great deal of interest in the use of Amplatzer and clamshell-type VSD closure devices. Although these devices can be deployed in the catheterization laboratory, the approach, across the tricuspid valve, may be difficult. There has been great interest in a hybrid approach in which a standard sternotomy is made, and using a perventricular approach with a needle in the right ventricle, a guide wire is passed across the defect under echocardiographic guidance and then an Amplatzer device deployed with the heart beating off cardiopulmonary bypass under echo guidance. These techniques have the advantage of being able to close even fairly remote apical muscular defects with high degrees of success and avoid the need for cardiopulmonary bypass. As additional experience is gained with these techniques, greater use of hybrid approaches is likely to occur, and even defects that are normally accessible through the right atrium may be closed with a perventricular approach to avoid the need for cardiopulmonary bypass.

Perimembranous VSD devices are being developed and may supplant surgical therapy for many typical conoventricular VSDs. However, the close association of the aortic valve with the superior rim of the VSD may result in some problems with these devices over time, and the incidence of heart block has not completely been identified with the current generation of transcatheter conoventricular closure devices. Nevertheless, the advent of the VSD occluder devices has made pulmonary artery banding for VSDs virtually unnecessary. An additional area of controversy is the approach to patients with coarctation of the aorta and large conoventricular defects. In infants in whom there is a degree of arch hypoplasia in association with a very large conoventricular defect, which is a common association, we have elected to primarily repair both defects through a midline sternotomy with patching of the aortic arch, which relieves the arch obstruction completely. Although in a small percentage of cases these large defects might close spontaneously, the avoidance of pulmonary artery banding and significant shunting early in life controls congestive heart failure immediately and has been associated with good outcomes in our patients.

The presence of residual VSD after attempted surgical closure remains a problem. Most commonly with conoventricular defects or malalignment defects, it can be difficult to expose the anterosuperior aspect of the defect, and residual defects can be present in this location. In addition, in patients with tetralogy of Fallot, conal muscle bundles can masquerade as the margin of the defect, and the patch may be attached more on the ventricular muscle than the superior rim of the defect, leaving an abnormal connection from the left ventricle to the right ventricle underneath the aortic valve and obscured by the trabecular muscle. Often these defects are best approached through the aortic valve, where the superior margin of the patch can be readily identified, and either direct primary closure with sutures to the base of the right coronary leaflet of the aortic valve or additional patch closure can be undertaken.

It has been shown that residual peripatch ventricular septal defects of <3 mm generally will close over time and rarely require surgical reintervention. However, there are situations where sutures placed in the base of the septal tricuspid valve leaflet in conoventricular defects can tear through the delicate tissue if not supported by buttressed sutures, and in these cases it is possible to develop a left ventricle to right atrial shunt. This particular circumstance seems to result in elevated right atrial pressures which can be associated with pleural effusions and chylothorax. The magnitude of shunt may be larger than a ventricular level shunt and many of these patients will require reoperation to resolve the pleural effusions even though the relative size of the defect is quite small.

TLS

81 Aortopulmonary Window

Moritz C. Wyler von Ballmoos, Michael E. Barnes, and James S. Tweddell

EPIDEMIOLOGY, EMBRYOLOGY, AND ANATOMY

Aortopulmonary window is a rare defect and accounts for about 0.1% to 0.2% of all structural congenital cardiac defects. Major academic centers could anticipate taking care of one or two infants with aortopulmonary window per year. Antenatal diagnosis of aortopulmonary window is challenging and delayed diagnosis may occur; the actual defect prevalence at birth is hence more difficult to estimate.

Aortopulmonary window is caused by failure of fusion of the two opposing conotruncal ridges that are responsible for separating the truncus arteriosus into the aorta and pulmonary artery. The aortopulmonary window, therefore, occurs between the two structures that normally result from septation of the truncus arteriosus, namely, the ascending aorta and the main pulmonary artery. Normal anatomy of the aortic and pulmonary valves separates this defect from the persistent truncus arteriosus. The similarity among conotruncal defects raises the question of a common underlying pathogenesis, but anatomic studies suggest that it develops by a different mechanism than truncus arteriosus. Furthermore, to date, no genetic association or environmental risk factors have been linked to aortopulmonary window.

A classification of aortopulmonary window based on the location has been proposed by Mori dividing aortopulmonary window into three types; proximal (type I), distal (type II), and total (type III). The classification introduced by the Society of Thoracic Surgeon adds a fourth, "intermediate" category accounting for the fact that this defect likely has a continuum of morphologies. For patients with aortopulmonary window without additional lesions, it is the size rather than the location of defect that impacts management and outcome (Fig. 81.1).

Origin of the right pulmonary artery from the ascending aorta and arch hypoplasia with interruption or coarctation are additional anomalies occurring with large aortopulmonary windows. With increasing size of the aortopulmonary window aberrations in flow result in abnormal incorporation of the right sixth arch, destined to become the right pulmonary artery, such that the right pulmonary artery arises from the rightward aspect of the ascending aorta. Similarly, with large aortopulmonary windows flow patterns can be disturbed such that there is preferential flow through the ductus arteriosus and diminished flow in the developing aortic arch resulting in distal arch hypoplasia including coarctation or interrupted aortic arch. It has been suggested that when associated with interrupted aortic arch, aortopulmonary windows are larger with greater distal extension. The Congenital Heart Surgeons' Society multi-institutional study of aortopulmonary window with interrupted aortic arch found that all types of aortopulmonary window were more or less equally represented among patients with interrupted aortic arch (Fig. 81.2). Aortopulmonary window is not associated with DiGeorge syndrome, suggesting that aortopulmonary window is a distinct malformation not related to abnormalities of the conal septum, such as interrupted aortic arch with ventricular septal defect (VSD), tetralogy of Fallot, and persistent truncus arteriosus.

In reports based on the cumulative experience at high-volume centers, aortopulmonary window was associated with other defects in 58% of cases, the most common being ventricular and atrial septal defect, interrupted aortic arch or coarctation of the aorta, tetralogy of Fallot, and transposition of the great arteries. Abnormal origin of the coronary arteries is also commonly associated with aortopulmonary window. The coronary arteries may arise from the edge of the defect, or the origin may occur just on the pulmonary artery side of the defect.

PRESENTATION, DIAGNOSTIC CONSIDERATIONS, AND INDICATIONS FOR SURGERY

Critical to timely diagnosis and early repair, antenatal diagnosis of aortopulmonary window was first reported in 2002. Simple aortopulmonary window may not be identified by fetal echocardiography because equal pressure in the ascending aorta and pulmonary root in the fetus results in minimal detectable flow through the defect. Posterior deviation of the outflow septum that is characteristic of patients with interrupted aortic arch with a ventricular septal defect and would prompt further interrogation of the arch by the fetal echocardiographer, is lacking in the fetus with aortopulmonary window. The antenatal diagnosis of aortopulmonary window with interrupted aortic arch has only recently been reported.

The presentation of patients with aortopulmonary window may be different depending on the size of the window, associated other defects, and age at presentation. Although small, restrictive aortopulmonary windows do occur; generally, the communication is large, and patients present in the first weeks of life when pulmonary vascular resistance drops and increased pulmonary blood flow results in congestive heart failure. The clinical presentation is then similar to that of other patients with left-to-right shunts, such as patent ductus arteriosus (PDA) or VSD. This presentation includes signs of congestive heart failure such as tachypnea, diaphoresis, poor feeding, and inadequate weight gain. In early infancy, cyanosis is usually not a prominent feature, but with large defects, bidirectional shunting can produce systemic desaturation. With delayed diagnosis and persistent pulmonary overcirculation, remodeling of the pulmonary vasculature, and ultimately pulmonary hypertension occur.

Physical examination demonstrates a tachypneic infant with accessory respiratory

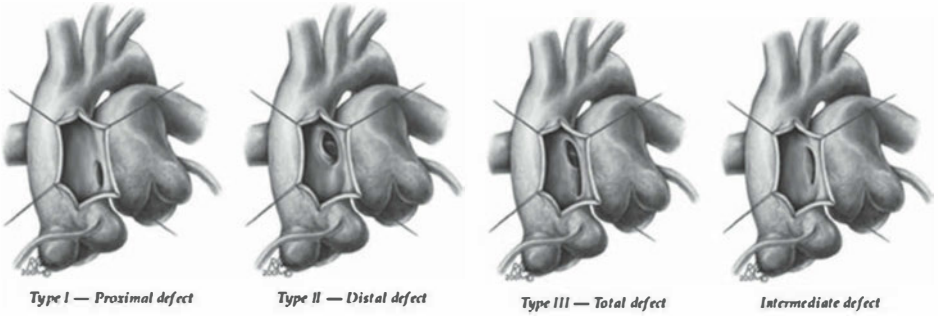


Fig. 81.1. The Society of Thoracic Surgeons' classification of aortopulmonary window. (Reprinted from Backer CL, Mavroudis C. Surgical management of aortopulmonary window: a 40-year experience. Eur J Cardiothorac Surg 2002;21:773-779, 202, with permission from Elsevier.)

muscle use. Cardiac examination reveals an enlarged heart, and similar to patients with PDA, the pulses are bounding. A systolic murmur and accentuated second heart sound that is not split can be heard along the left sternal border; however, unlike the situation with patients with a PDA, a diastolic component to the murmur is rare. Chest X-ray films reveal cardiomegaly and increased pulmonary vascular markings consistent with increased pulmonary blood flow. Patients with associated arch abnormalities have pronounced left-to-right shunting and PDA-dependent perfusion of the lower body. With impending ductal closure there will be worsening pulmonary edema and shock.

The diagnosis is routinely made with 2D Doppler echocardiography. The location and size of the communication as well as associated anomalies are carefully identified. Echocardiography of the conotruncal region will identify the defect and associated shunt and the diagnosis of aortopulmonary window is confirmed when two separate semilunar valves can be identified. For surgical planning, echocardiographic examination should identify the coronary anatomy, the size and distal extent of the aortopulmonary defect as well as associated lesions such as anomalous origin of the right pulmonary artery and interrupted aortic arch. Echocardiography as the sole imaging modality has been shown to be accurate and sufficient for preoperative evaluation of even complex congenital defects including aortopulmonary windows with or without associated other defects.

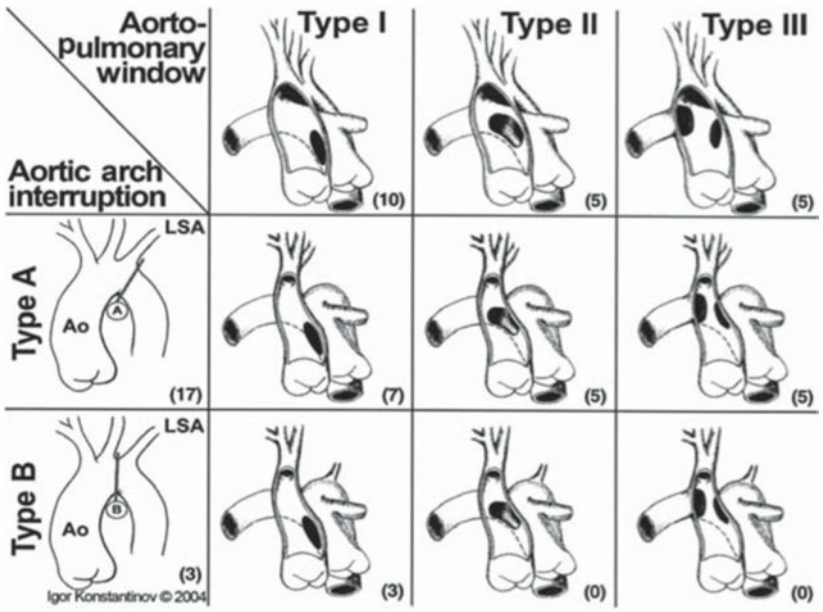


Fig. 81.2. The observed morphologies of aortopulmonary window with interrupted aortic arch. Type A interrupted arch predominated but the position of aortopulmonary window was more or less evenly divided between proximal and distal. (Reprinted from Konstantinov, IE, et al. Surgical management of aortopulmonary window associated with interrupted aortic arch: a Congenital Heart Surgeons Society study. J Thorac Cardiovasc Surg 2006;131:1136-1141, with permission from Elsevier.)

Although using cardiac catheterization to assess the origin of the coronary arteries is theoretically appealing, the large defect occurring just above the sinuses of Valsalva combined with the tremendous pulmonary flow makes assessment of coronary artery anatomy with catheterization impractical. Cardiac catheterization should thus be reserved for the patient who presents after early infancy and therefore is at risk for elevated pulmonary vascular resistance.

Those patients who are found to have an elevated pulmonary vascular resistance should undergo testing with pulmonary vasodilators to determine whether the pulmonary vascular resistance can be reduced prior to surgical correction of the defect. The use of magnetic resonance imaging for the diagnosis of aortopulmonary window has been reported in adults.

The presence of an aortopulmonary window is an indication for surgery. If left untreated, infants die of intractable heart failure or rapidly develop pulmonary vascular obstructive disease. Medical therapy is limited to the preoperative period and for patients with irreversible pulmonary hypertension, which poses the only contraindication to immediate repair of the defect.

PREOPERATIVE MANAGEMENT

Because of the challenges with early diagnosis of aortopulmonary window, it tends to present with symptoms of heart failure. Initial resuscitative efforts for patients presenting in shock are aimed at improving systemic output by limiting excessive pulmonary blood flow and are similar to those used in the patient with single-ventricle anatomy and unobstructed pulmonary blood flow or patients with truncus arteriosus. For the patient with a large aortopulmonary window, this often requires intubation and mechanical ventilation as well as sedation and sometimes neuromuscular blockade to achieve a balanced circulation. The use of hypercapnea and minimizing the fraction

of inspired oxygen (FiO_2) will increase the pulmonary vascular resistance, decrease left-to-right shunting, and improve systemic oxygen delivery. Inotropic support may be required. Prostaglandin infusion is necessary to maintain ductal patency in patients with aortopulmonary window and interrupted aortic arch or coarctation. These measures should be successful in restoring systemic output, and the patient should go to surgery without a metabolic acidosis.

SURGICAL TECHNIQUE

A median sternotomy incision is used for aortopulmonary window regardless of associated abnormalities. The anatomy should be carefully assessed (Fig. 81.3). The external extent of the aortopulmonary window and the coronary arteries should be identified. Coronary arteries involved in the defect can be seen arising from the area of the communication and coursing down the proximal aorta before reaching the myocardium. The position of the right pulmonary artery should be noted.

Simple Aortopulmonary Window

The right and left pulmonary arteries should be loosely encircled with snares so that once cardiopulmonary bypass is established, pulmonary flow can be controlled (Fig. 81.4). General anesthesia can produce a drop in pulmonary vascular resistance, resulting in excessive pulmonary blood flow at the expense of systemic perfusion. It is sometimes helpful to snare one of the branch pulmonary arteries to limit excessive pulmonary blood flow while continuing with preparation for cardiopulmonary bypass. The aorta should be dissected nearly circumferentially distal to the extent of the aortopulmonary window to allow for subsequent placement of the cross-clamp. After the administration of heparin, the aortic cannula is placed in the ascending aorta near the origin of the innominate artery (Fig. 81.4). If there is an associated atrial septal defect or VSD, bicaval cannulation should be undertaken; otherwise, a single venous cannula can be used. Cardiopulmonary bypass is begun and simultaneously the branch pulmonary arteries are snared. A left ventricular vent is placed through the junction of the right superior pulmonary vein and left atrium. A cardioplegia cannula is placed in the ascending aorta. For simple aortopulmonary window, moderate hypothermia to 32°C is adequate. The aorta is cross-clamped distal to the communication. Cardioplegic solution is infused while the pulmonary arteries are snared. The defect can be repaired via an incision in the window itself, through the aorta, or through the pulmonary artery (Fig. 81.5). An approach through the window is preferable because the origin of the coronary arteries can be easily assessed and the patch placed such that an abnormal coronary ostial origin is incorporated into the aorta. In addition, there is less potential for compromise of either of the great vessels or injury of the semilunar valves. The incision is initiated in the anterior–superior portion of the window, and, after the origin of the right coronary artery is identified, the incision is extended proximally, transecting the anterior half of the window. After the origins of the coronary arteries and the right pulmonary artery are identified, an appropriately sized patch of polytetrafluoroethylene (PTFE) or

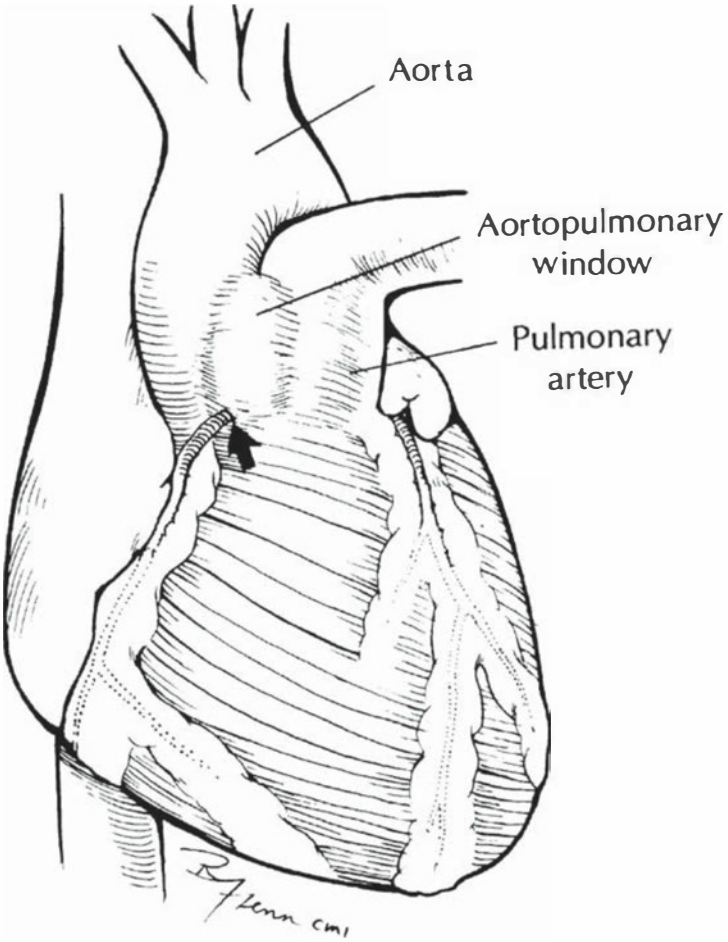


Fig. 81.3. External view of aortopulmonary window. The area of communication between the great vessels can be easily identified. The extent of the defect and the origins of the right pulmonary artery and right coronary artery should be identified. As in this figure, the right coronary artery (*arrow*) can be sometimes seen originating near the inferior margin of the defect and coursing proximally on the aorta before taking its normal position in the atrioventricular groove. This finding should prompt careful internal inspection for abnormal origin of the coronary artery from the inferior ridge of the defect or the pulmonary artery.

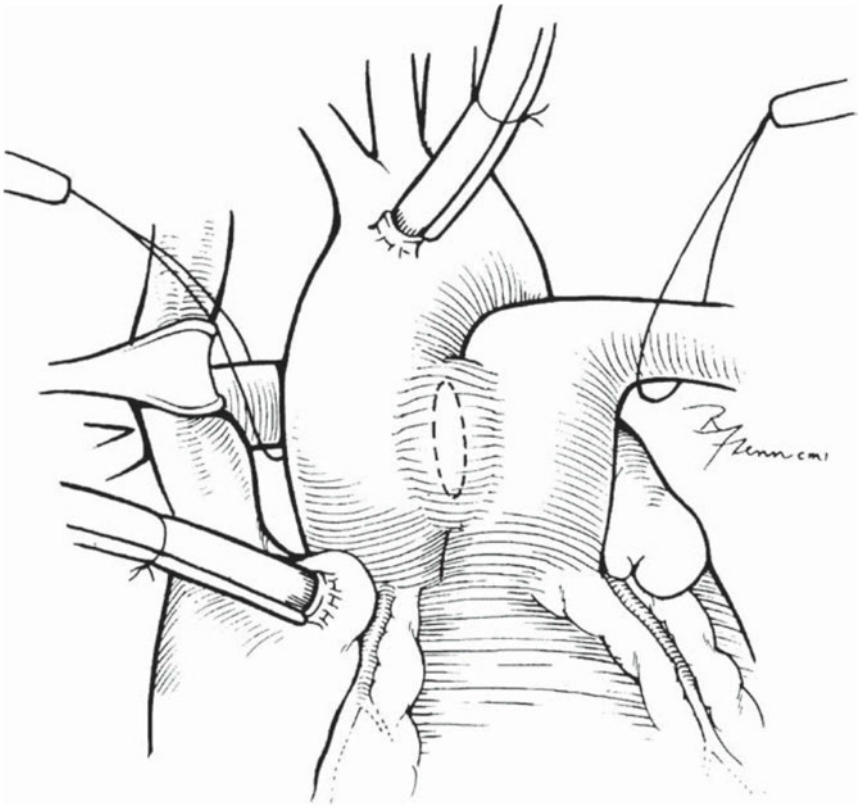


Fig. 81.4. Preliminary steps in the repair of a simple aortopulmonary window. The left and right branch pulmonary arteries are loosely encircled. The aorta is cannulated beyond the distal extent of the aortopulmonary window so that there is adequate room for a cross-clamp. Generally, a single venous cannula placed through the right atrial appendage is used for venous drainage. After cardiopulmonary bypass has begun, the branch pulmonary artery snares are tightened. A left ventricular vent (not shown) is placed through the right superior pulmonary vein.

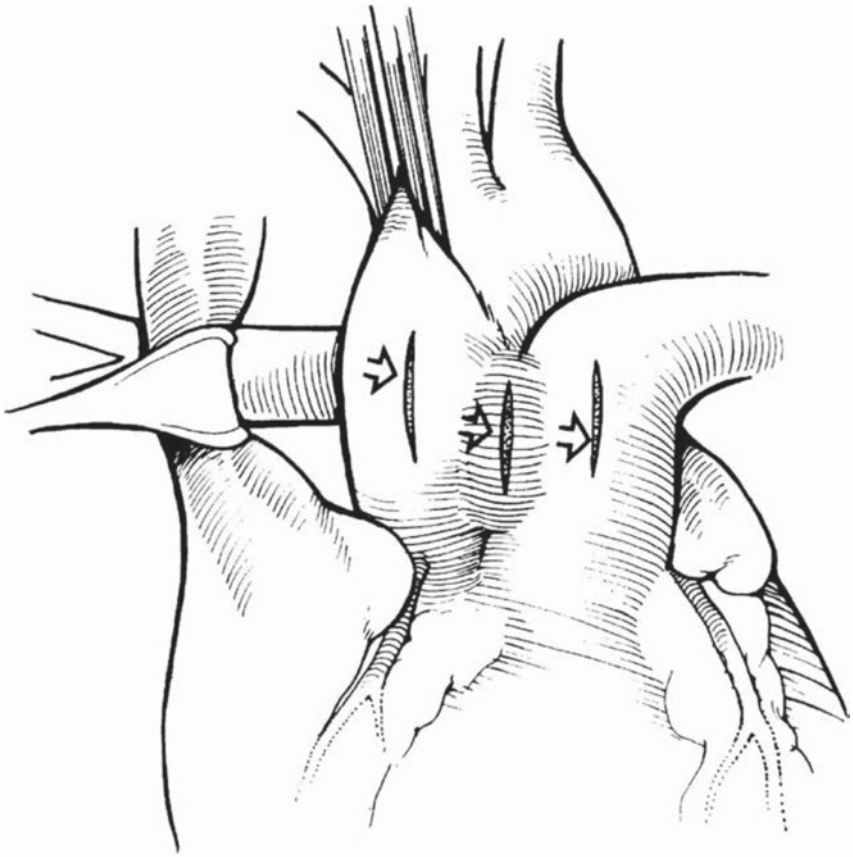


Fig. 81.5. Aortopulmonary window can be repaired via an incision in the window itself, through the aorta, or through the pulmonary artery.

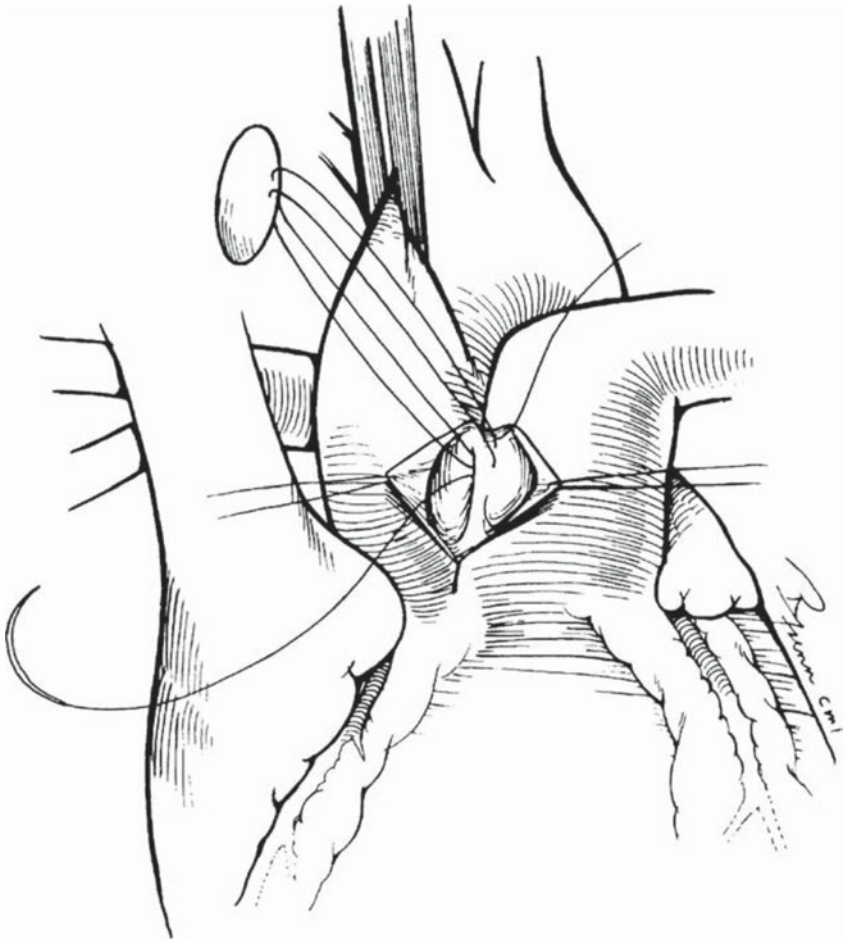


Fig. 81.6. Approach through the aortopulmonary window. An incision is initiated in the anterior–superior portion of the window, and after the origin of the right coronary artery is identified, the incision is extended proximally, transecting the anterior half of the window. After the origins of the coronary arteries and the right pulmonary artery are identified, an appropriately sized patch of polytetrafluoroethylene (PTFE) or pericardium is secured to the posterior rim of the defect.

pericardium is secured to the posterior wall of the defect using continuous suture (Fig. 81.6). The anterior incision in the window is then closed, incorporating the patch into the suture line (Fig. 81.7). Rewarming to normothermia is begun as the window is closed. The aortic root is deaired, and the cross-clamp is removed. Preparation for weaning from cardiopulmonary bypass includes placement of a pulmonary artery line through the right ventricular free wall as well as a left atrial line placed through the vent site. A milrinone infusion is initiated prior to weaning from cardiopulmonary bypass because this provides both inotropy and pulmonary vasodilation. Additional pulmonary vasodilators, such as inhaled nitric oxide, should be available especially in the older infant.

Repair of Aortopulmonary Window with Interrupted Aortic Arch

Aortopulmonary window with interrupted aortic arch is usually a large defect and is more frequently associated with abnormal origin of the right pulmonary artery (Fig. 81.8). A median sternotomy incision is used. Initial preparation is the same as for simple aortopulmonary window; again, the branch pulmonary arteries are loosely encircled with snares. Because of the large aortopulmonary communication, a single arterial cannula can be used. This is placed in the ascending aorta (Fig. 81.9). Flow to the distal half of the body will be through the aortopulmonary window and then via the ductus arteriosus. After cardiopulmonary bypass is established, the branch pulmonary arteries are snared and a left ventricular vent is placed. The patient is cooled over a period of at least 30 minutes to a bladder temperature of 18°C. During the cooling period, the aortic arch, head vessels, ductus arteriosus, and proximal descending thoracic aorta are mobilized. After reaching the target temperature, circulatory arrest is established, the head vessels are snared, a C-shaped vascular clamp is placed across the descending thoracic aorta at least 1 cm distal to the insertion of the ductus arteriosus, and cardioplegic solution is infused via the arterial cannula. With the branch pulmonary arteries, descending thoracic aorta, and head vessels occluded, cardioplegic solution will be directed into the coronary arteries. The entire procedure can be performed using deep hypothermic circulatory arrest, or alternatively continuous cerebral perfusion can be used by selectively perfusing the innominate artery. The ductus arteriosus is ligated near the pulmonary artery, and all ductal tissues is excised from the descending thoracic aorta. The undersurface of the proximal aortic arch is incised, and the incision is continued into the ascending aorta. The descending thoracic aorta is then brought up and anastomosed to the undersurface of the aortic arch and distal ascending aorta. Construction of the aortic anastomosis is facilitated by holding the descending thoracic aorta in a C-clamp (Fig. 81.10). This provides for close approximation while the anastomosis is performed. After reconstruction of the aortic arch, a cross-clamp is placed between the aortopulmonary window and the reconstructed arch and cardiopulmonary bypass is re-established. Alternatively, a single period of deep hypothermic circulatory arrest can be used for the entire repair. The window is approached as described. Again, the origins of the right pulmonary artery and coronary arteries are assessed (Fig. 81.11). A patch of PTFE or pericardium is placed to close

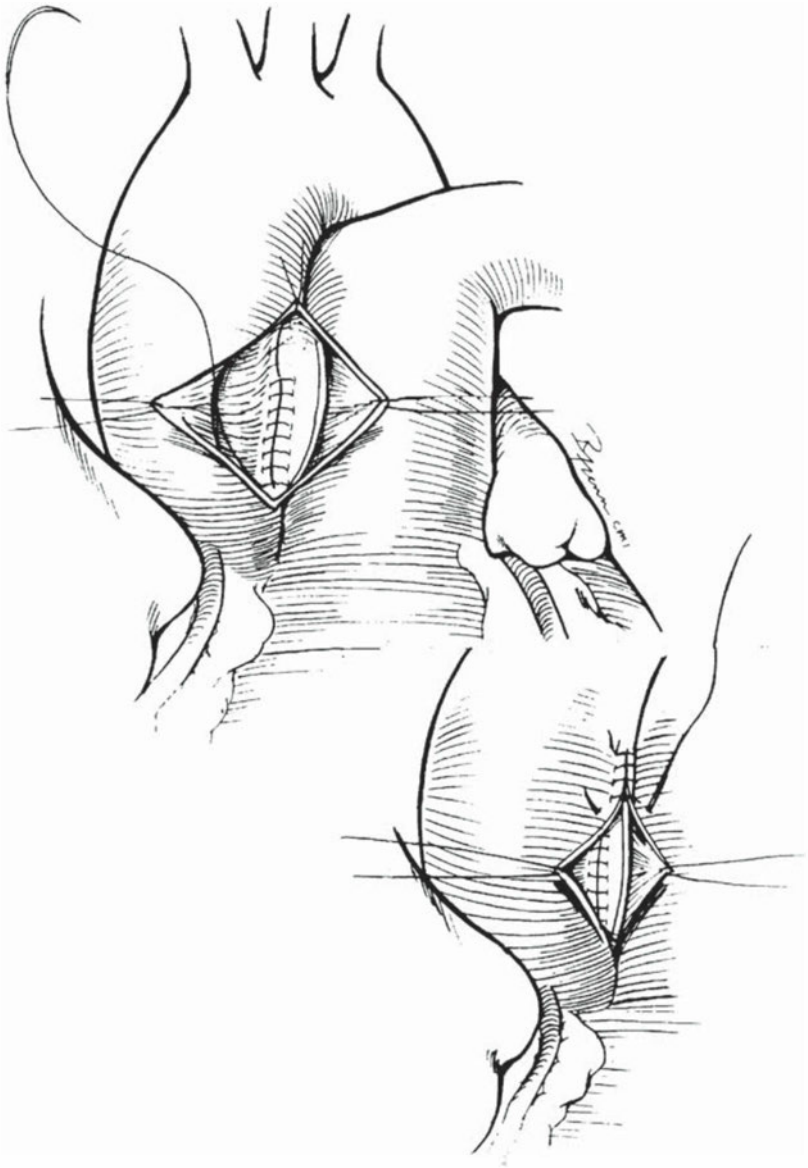


Fig. 81.7. Approach through the aortopulmonary window (continued). The patch is secured to the posterior margin of the aortopulmonary window using a continuous suture technique. Anteriorly, closure of the arteriotomy in the aortopulmonary window incorporates the patch.

the window such that the right pulmonary artery is incorporated into the main pulmonary artery trunk and the lumen of the right pulmonary artery is not compromised. The patient is rewarmed, monitoring lines are placed, and weaning from cardiopulmonary bypass is conducted as for simple aortopulmonary window.

An alternative approach to repair of aortopulmonary window with interrupted aortic arch may be necessary in cases in which there is little distal ascending aorta or proximal arch present. In this case, additional patch augmentation of the arch anastomosis and proximal great vessels may be necessary to complete reconstruction; this technique is described in Figs. 81.12–81.15.

Catheter-Based Approaches

The utility of transcatheter closure is limited by the large size of the defect, the small size of patients with correspondingly small femoral vessels, and the potential for complications related to anomalous origin of the coronary arteries that are not easily nor consistently identified prior to intervention. Nevertheless, device closure of aortopulmonary window may be suitable for small defects in which the risk of anomalous origin of the coronary arteries is low, specifically those with a more distal location.

SURGICAL COMPLICATIONS AND POSTOPERATIVE CARE

For simple aortopulmonary window and even aortopulmonary window with interrupted aortic arch, postoperative inotropic support should be minimal. As in other patients with large left-to-right shunts, there is potential for acute elevation of pulmonary vascular resistance with the development of critically low cardiac output after repair. If inotropic support becomes necessary, milrinone is a good first-line agent because of its vasodilatory effects on the pulmonary vasculature. Patients operated on in the first 2 weeks of life should be at low risk for pulmonary vascular resistance elevation and may be candidates for early extubation. Older patients may benefit from sedation and neuromuscular blockade for the first 12 to 24 hours. In higher risk patients, it is reasonable to monitor pulmonary artery pressure and trends in mixed central venous oxygen saturation. If pulmonary hypertension develops, then in addition to milrinone, inhaled nitric oxide should be started promptly. If ongoing dependence on inhaled nitric oxide complicates weaning from mechanical ventilation, then transitioning to sildenafil may be considered.

RESULTS OF SURGERY

As with other defects, the trend toward early repair along with the advances in diagnosis and perioperative management have substantially improved outcomes over the last decades. In the current era, early mortality following repair of uncomplicated aortopulmonary window approaches zero and long-term outcome should be excellent. Early morbidity includes pulmonary artery stenosis and residual aortopulmonary septal defects.

In a pooled analysis of 370 patients, from 22 reports over 6 decades the median age at surgery was 3 months (ranging from neonates to 27-year-old adults) and 58% of the patients had associated defects. There was one reported death during surgery in all patients with isolated aortopulmonary window (128 patients). No late death was reported for this group, but some of the patients needed reintervention for residual shunting or pulmonary artery stenosis in older series and in cases where simple ligation of the aortopulmonary window was performed. In the group with associated

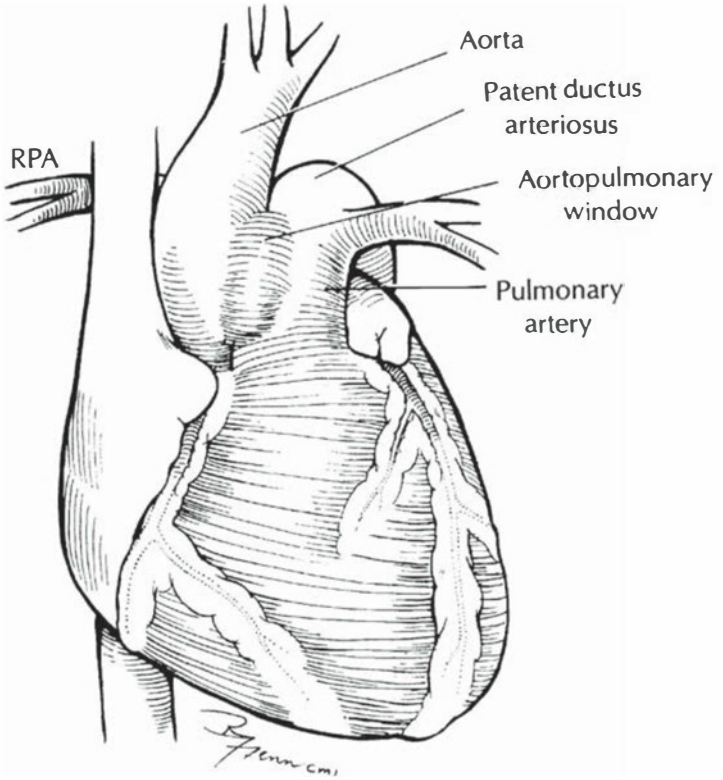


Fig. 81.8. External appearance of the aortopulmonary window and interrupted aortic arch. RPA, right pulmonary artery.

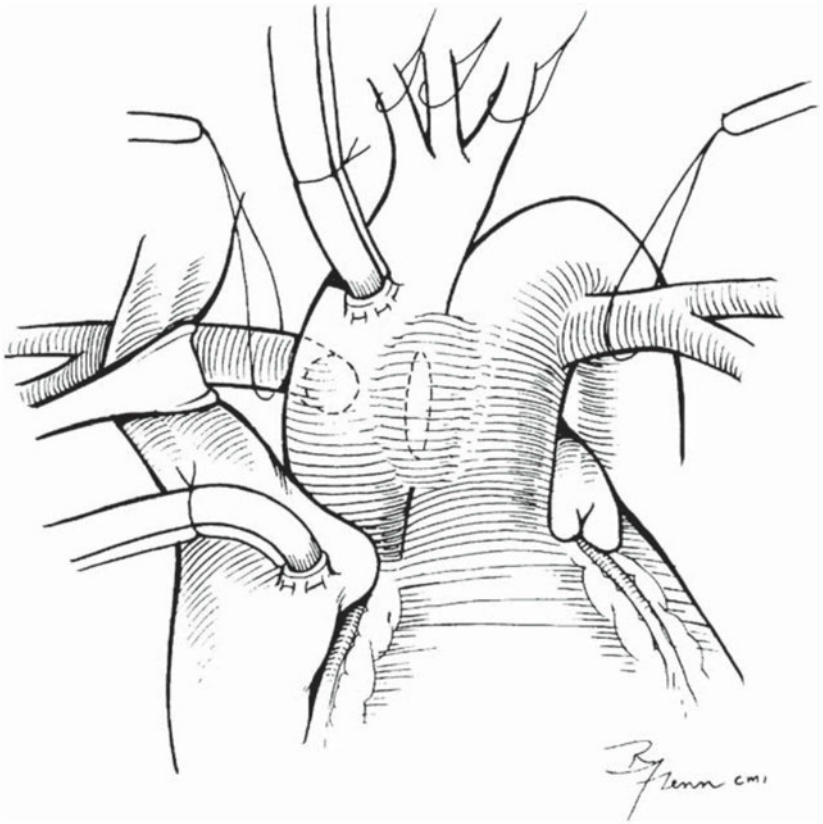


Fig. 81.9. Cannulation for repair of aortopulmonary window and interrupted aortic arch. The right and left branch pulmonary arteries are loosely encircled with snares. Unlike interrupted aortic arch with ventricular septal defect, a single aorta cannula is satisfactory because distal perfusion can be carried via the aortopulmonary window and a ductus arteriosus to the lower half of the body. Aortopulmonary window and interrupted aortic arch are rarely associated with ventricular septal defect, and a single venous cannula is usually all that is required. After establishment of cardiopulmonary bypass, the branch pulmonary artery snares are tightened. During the period of cooling, the brachiocephalic vessels are mobilized and loosely encircled with snares.

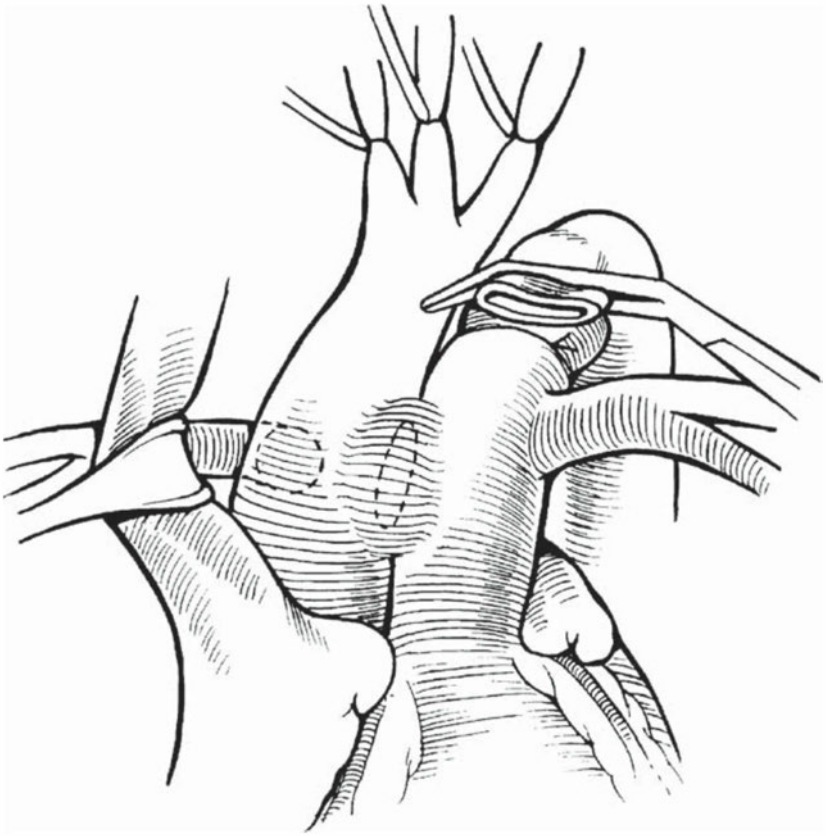


Fig. 81.10. Repair of aortopulmonary window and interrupted aortic arch. After the establishment of deep hypothermic circulatory arrest or continuous cerebral perfusion, a C-clamp is placed on the descending thoracic aorta approximately 1 cm distal to the insertion of the ductus arteriosus. The ductus arteriosus is ligated and divided. Residual ductal tissue is excised from the proximal descending thoracic aorta. By placing gentle traction on the C-clamp, the open end at the descending thoracic aorta is brought up to the undersurface of the proximal aortic arch and distal ascending aorta. An anastomosis is constructed between the descending thoracic aorta and distal ascending aorta/proximal arch with continuous suture.

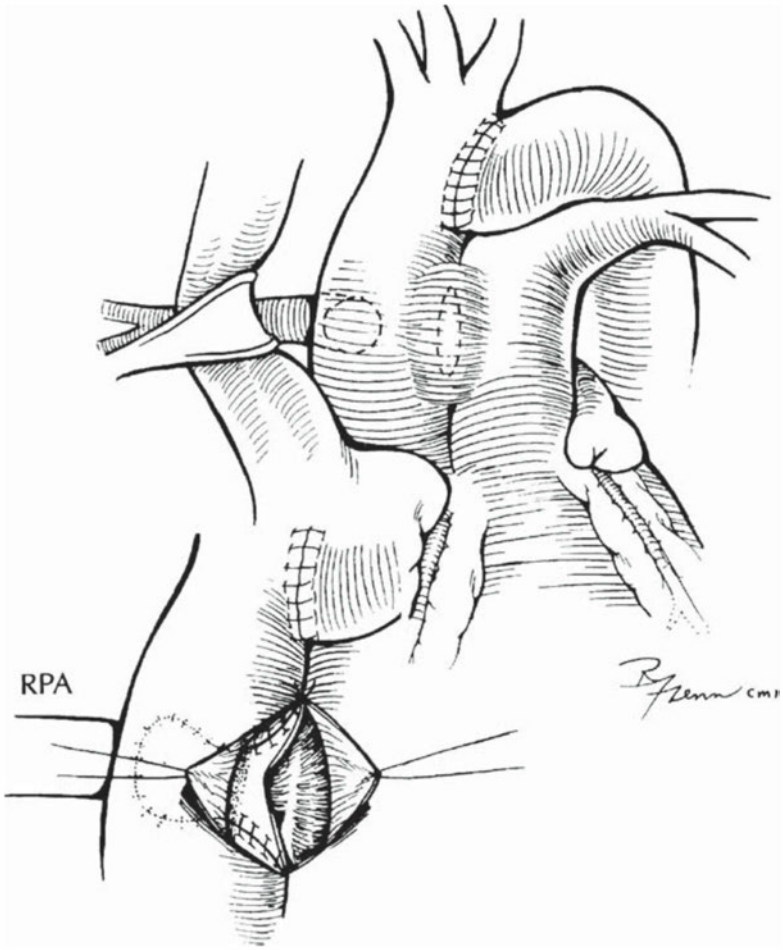


Fig. 81.11. Repair of aortopulmonary window and interrupted aortic arch (continued). After reconstruction of the aortic arch, full cardiopulmonary bypass can be reestablished, and an aortic cross-clamp is placed between the arch reconstruction and the aortopulmonary window. Alternatively, the entire procedure can be carried out using one period of deep hypothermic circulatory arrest. Again, the aortopulmonary window is approached via an incision in the window itself. A patch of polytetrafluoroethylene or pericardium is placed such that the origin of the right pulmonary artery is connected to the main pulmonary artery trunk and the lumen of the right pulmonary artery is not compromised. The anterior margin of the patch is incorporated into the arteriotomy closure.

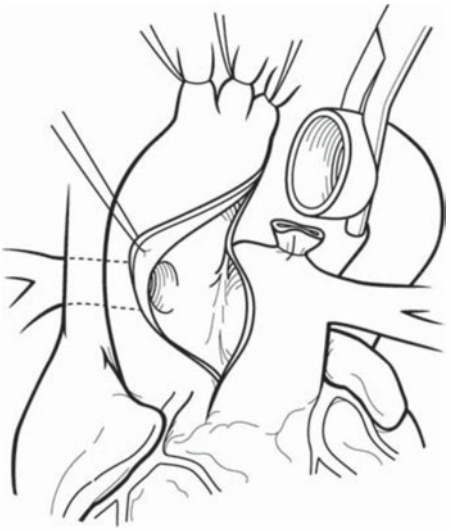


Fig. 81.12. An alternative approach to the repair of an aortopulmonary window with interrupted aortic arch may be necessary in cases in which there is little distal ascending aorta or proximal arch present. In this case, there may not be sufficient aorta distal to the aortopulmonary window to permit construction of an unobstructed anastomosis to the descending thoracic aorta. Cannulation is performed as shown in Figure 81.9, and perfusion techniques can include either continuous cerebral perfusion or deep hypothermic circulatory arrest. The ductus arteriosus is ligated, and all residual ductal tissues are excised from the mobilized descending thoracic aorta. An incision is made in the undersurface of the proximal arch initiated at the origin of the distal brachiocephalic vessel, in this case, the left subclavian artery, and extended proximally into the aortopulmonary window.

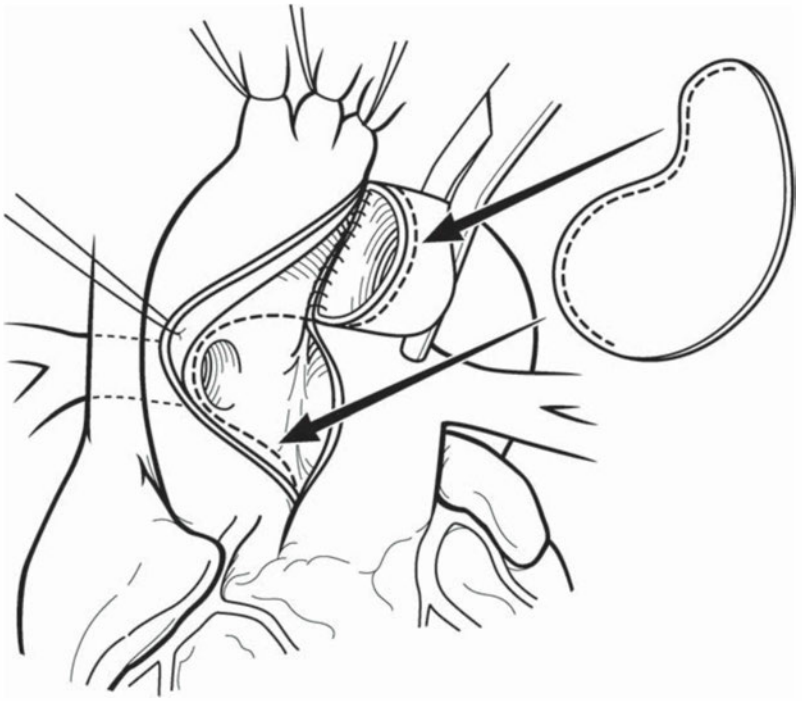


Fig. 81.13. The posterior one half of the circumference of the descending thoracic aorta is sutured to the posterior edge of the arch incision. A patch (either pulmonary homograft or pericardium) is fashioned as shown. The dashed line indicates the initial suture line. Reconstruction begins by suturing the patch to the anterior one half of the proximal descending thoracic aorta and then transitioning the suture line to the posterior wall of the ascending aorta such that the abnormally positioned right pulmonary artery is placed on the left side of the patch. The suture line then continues to the bottom of the aortopulmonary window.

defects, 35 patients (17%) died during or immediately after surgery. Seven additional patients (3.4%) died during follow-up that varied in length but was >2.5 years in all reports (median follow-up 6.5 years). Importantly, these estimates include the experience from six decades and in more recent series the mortality approaches zero even for cases of aortopulmonary window with associated defects. In three studies reporting long-term outcomes, actuarial survival was between 84% and 90% at 10 years and freedom from re-intervention was 43% to 70% at 5 to 10 years after the initial procedure including patients with associated defects. Among these studies, associations with higher morbidity and mortality were found for the combination of aortopulmonary window with another defect, for the simple ligation of the window, and for the transpulmonary repair technique.

Between 1983 and 2009, 25 patients with aortopulmonary window and associated lesions have undergone repair at the Children's Hospital of Wisconsin. Patients were divided into two categories based on the presence of important additional lesions. Simple aortopulmonary window ($n = 12$) included those patients with an isolated aortopulmonary window with or without an atrial level communication. Complex aortopulmonary window ($n = 13$) included

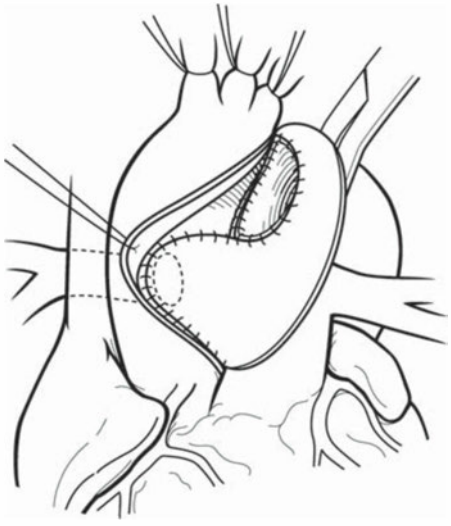


Fig. 81.14. The posterior suture line is in place. The anterior edge of the patch will now be secured to the edge of the ascending aorta.

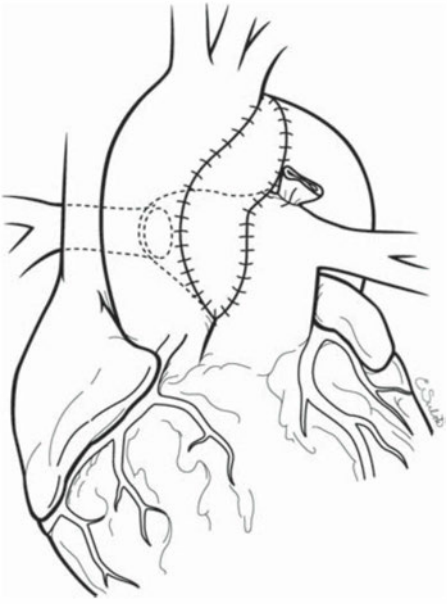


Fig. 81.15. Completion of the repair. The pulmonary artery edge of the aortopulmonary window is sutured to the centerline of the patch to complete closure of the main pulmonary artery. In this way, the patch augments both ascending aorta and main pulmonary artery and minimizes the potential for compromise of the caliber of either of these vessels or the right pulmonary artery. In addition, the patch allows augmentation of the arch reconstruction.

those patients with important additional lesions including interrupted aortic arch or coarctation of the aorta ($n = 8$), VSD ($n = 1$), pulmonary atresia with VSD and anomalous origin of the right coronary artery ($n = 1$), pulmonary atresia with intact ventricular septum and partial anomalous pulmonary venous return ($n = 1$), aortopulmonary window with d-malposed great vessels ($n = 1$), and congenital absence of the left pulmonary artery with pulmonary artery hypertension ($n = 1$). There were no deaths early or late in the simple aortopulmonary window group. In the complex aortopulmonary window group, there was one early death in the patient with aortopulmonary window, pulmonary atresia, and intact ventricular septum. There was one late death following lung transplantation in the patient with aortopulmonary window, absent left pulmonary artery, and pulmonary hypertension.

Long-term follow-up is indicated to look for the development of branch pulmonary artery stenosis. For patients with aortopulmonary window and interrupted aortic arch, long-term observation for recurrent coarctation is indicated.

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EDITOR'S COMMENTS

As noted by the author, aortopulmonary window is a rare congenital cardiac condition that presents in infancy with severe congestive failure and pulmonary overcirculation. Preoperative stabilization is imperative for low operative morbidity and mortality. Ventilatory maneuvers preoperatively can aid in stabilization of these patients. Because these maneuvers are often temporary, however, prompt resuscitation and early operation are indicated in virtually all infants with this condition.

Many operative techniques have been described for closure of aortopulmonary window. The one most commonly used, however, is the technique described here, in which the window is opened anteriorly, and after careful identification of the edges of the defect, an intra-aortic or intrapulmonary patch is placed to close the defect and separate the great vessels. In many cases, we have elected to divide the aortopulmonary window completely to carefully identify the origins of the coronary arteries and then patch the aortic and pulmonary sides separately. This can be especially useful if there is stenosis of the right pulmonary artery at its origin, which occasionally occurs. In these cases, the origin of the right pulmonary artery can be patched with the closure of the pulmonary artery.

In isolated cases of aortopulmonary window, in which the window is distally located and relatively small, it may even be possible to simply divide the defect or ligate the defect as one would a ductus arteriosus. These situations are rare, however, and can avoid the need for cardiopulmonary bypass. In this circumstance, direct identification of the origin of the coronary arteries is very important to avoid leaving a coronary coming from the pulmonary artery side of the defect.

In situations where aortopulmonary window is associated with interrupted aortic arch, we have elected to perform a more radical arch reconstruction. Because there is often very little distance distal to the aortopulmonary window and the take-off of the innominate artery and carotid vessels, we will generally ligate the ductus arteriosus, excise ductal tissue, and then make an incision into the left subclavian artery (if a type B interruption is present) or widely open the descending aorta (if a type A interruption is present), removing all ductal tissues. The aortopulmonary window is then opened and superiorly the incision is carried up into the origin of the left carotid artery for a short distance. The descending aorta is then anastomosed to the superior aspect of the incision in the ascending aorta and carotid, creating natural tissue approximation in this region. A generous patch of pulmonary homograft is then used to augment the undersurface of the aortic arch to take tension off the suture line and ensure that no kinking or narrowing of the anastomosis creates later arch obstruction and to avoid compression of the left main stem bronchus. This patch is then fashioned in such a way as to augment the ascending aorta and at its proximal portion to close the aortopulmonary window. With division of the aortopulmonary window, the pulmonary arterial side can be separately patched with a small patch of pulmonary homograft material to prevent any distortion or limitation of flow into the branch pulmonary vessels.

TLS

82 Coarctation of the Aorta

Karl F. Welke and Ross M. Ungerleider

Coarctation of the aorta can present as a severe and emergent problem in a neonate or as a subtle and essentially asymptomatic problem in an older child (or adult). This chapter focuses primarily on coarctation in neonates or infants because it is in that population that many of the important issues of coarctation are highlighted and best appreciated. In addition, this is the population that is most frequently treated with surgical repair of coarctation of the aorta. Between January 2007 and December 2010, 75% of the 3,667 patients who underwent surgical repair of coarctation of the aorta as recorded in the Society of Thoracic Surgeons Congenital Heart Surgery Database were under 1 year of age. Sixty-three percent of these patients were 30 days of age or younger.

Coarctation is a form of left ventricular outflow tract obstruction (LVOTO) and imposes an increase in afterload to the left ventricle. It is often but not necessarily found in association with a variety of other important cardiac defects, and these can have a crucial effect on the physiology of the defect and on the patient's presentation. In its "pure" form, coarctation is simply a constriction, or narrowing, of the aorta that usually occurs near the site of insertion of the ductus arteriosus. Because of this typical location, coarctation is often described as being "juxtaductal," and this term is used to distinguish the more common forms of coarctation from a less common form that can involve the aorta proximal to the ductus arteriosus and extend into the transverse aortic arch. This latter type is sometimes referred to as "preductal" coarctation, and because it will usually present early in infancy, it can also be referred to as "infantile coarctation." In practice, the use of the terms "juxtaductal" and "preductal" is not of critical importance as long as the extent of the coarctation is appreciated by the surgeon (Fig. 82.1).

Because of the obstruction to left ventricular (LV) outflow, as well as to distal aortic flow that is created by the coarctation, infants will generally suffer from severe LV failure manifested by poor distal perfusion and tachypnea. The LV outflow obstruction commonly results in pulmonary hypertension, and therefore "secondary" right ventricular (RV) hypertrophy is common in newborns with severe aortic coarctation. The LV failure may be reflected by a dilated, hypocontractile left ventricle with reduced output, and for this reason, the gradient across the coarctation site is not an indicator of the severity of the defect. The RV and LV features may be nicely demonstrated by transthoracic echocardiography. Furthermore, a well-performed two-dimensional echocardiogram will demonstrate the anatomy of the aortic arch and the great vessels and the discrete area of aortic narrowing near the ductus. Flow to the distal aorta can be severely restricted, and patency of the ductus arteriosus is often essential in the newborn to preserve perfusion to the lower body (Fig. 82.1). Therefore, neonates with coarctation should be started on an intravenous infusion of prostaglandin E_1 (PGE_1). Maintaining ductal patency with PGE_1 infusion will also allow decompression of the pulmonary circulation resulting from downstream obstruction of the LV. In neonatal coarctation, the RV may provide most of the perfusion in the descending aorta, and because the RV pressure may equal systemic pressure (especially if there is an associated ventricular septal defect [VSD]), there can be essentially no difference in the pressure above and below the coarctation. Therefore, the pressure gradient between the upper and the lower body will underestimate the severity of the coarctation. If no VSD is present, the descending aorta may be perfused with systemic venous blood from the RV and thereby results in the "differential cyanosis" that has been described for this lesion, with the lower body appearing more cyanotic than the upper body.

It is important to look for other commonly associated defects, which can occur anywhere along the "left heart/aorta complex." A bicuspid aortic valve is present in over 50% of patients with coarctation of the aorta. Other associated defects include mitral stenosis (often with mitral anomaly such as single papillary muscle), hypoplastic left ventricle (defined as an LV volume of <20 ml/m^2), endocardial fibroelastosis (a generalized scarring of the LV endothelium that appears as "brightness" on echocardiographic examination and that probably represents LV subendocardial ischemia), VSD (often with posterior malalignment of the infundibular septum that narrows the subaortic area), aortic stenosis (valvar or subvalvar), and atrial septal defect (ASD). All of these defects can be recognized by echocardiography, and it is not usually necessary to perform cardiac catheterization in these critically ill infants. The severity of the problem and the long-term prognosis are related to (1) age (young age, such as newborn, increases the risk), (2) the number and extent of associated defects, and (3) the actual anatomy of the defect (greater risk is incurred in patients whose defect extends proximal to the left subclavian artery).

Initial management of the newborn patient with aortic coarctation requires improving distal perfusion to the lower body by restoring the patency of the ductus arteriosus with an intravenous PGE_1 infusion. Intubation and mechanical ventilation may be necessary due to the 15% to 20% incidence of apnea that occurs with PGE_1 infusion. Once the patient has been stabilized, complete diagnostic assessment can be accomplished. This can be usually limited to an echocardiogram, which should demonstrate the anatomy of the aortic arch and isthmus, patency of the ductus arteriosus, the coarctation segment, and any important associated cardiac defects. It is not necessary to perform a cardiac catheterization for diagnostic purposes, but catheterization (or other form of imaging) should be done when there is any question about the arch anatomy, the nature of the coarctation, or the significance of a related defect that might require concomitant repair. Furthermore, cardiac catheterization has a

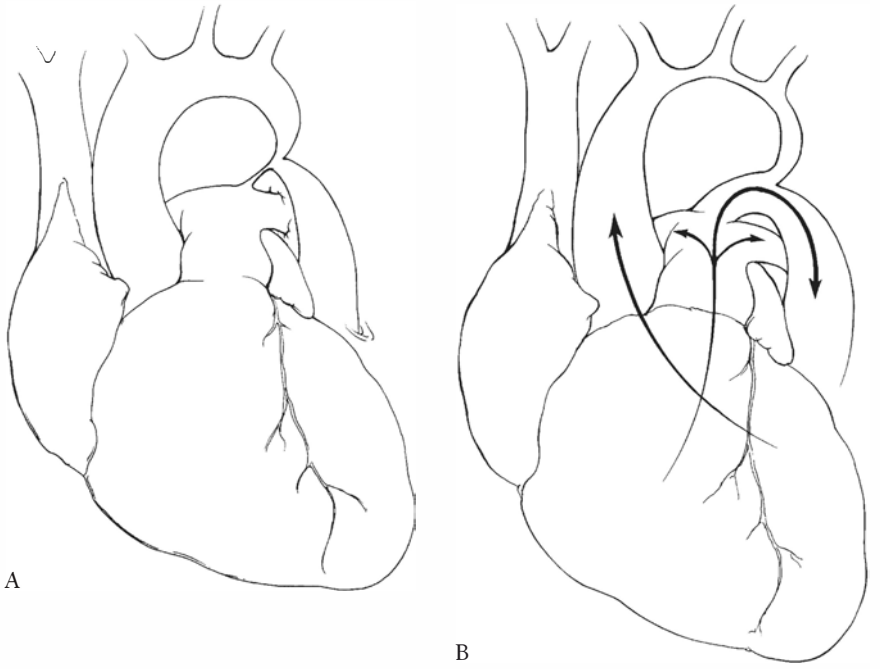


Fig. 82.1. (**A**) A juxtaductal coarctation with discrete narrowing at the level of the ligamentum arteriosum. (**B**) An "infantile" or "preductal" coarctation with distal aortic perfusion maintained across a patent ductus arteriosus. In these patients, the aortic arch is usually hypoplastic, as shown.

role when an intervention is desired before coarctation repair (such as a Rashkind atrial septostomy in patients with associated transposition of the great arteries who are not candidates for proceeding directly to the operating room). Operative repair should be considered once the diagnosis is confirmed. Coarctation of the aorta is an urgent problem in neonates, and prolonged medical management has a limited role reserved for unusual circumstances. There are currently several operative techniques for repair of aortic coarctation. Each has advantages and disadvantages, and the surgeon should be knowledgeable about each of these options. In most instances, repair is most easily and satisfactorily accomplished through a left thoracotomy, but occasionally median sternotomy is a useful approach and is discussed later.

SURGICAL APPROACH: GENERAL CONSIDERATIONS

The majority of coarctations can be exposed and repaired through a left lateral thoracotomy. Preferably, an arterial monitoring line should be placed in the right radial artery. Occasionally the right radial artery is the end vessel of an aberrant right subclavian artery, in which case, it will not provide accurate pressure data during clamping of the aorta. The left radial artery is less desirable as the flow to the left upper extremity will be compromised by clamping of the left subclavian artery during the repair. An additional arterial monitoring line or a blood pressure cuff can be placed on a lower extremity. A pulse oximetry monitor should be placed on a lower extremity as well. The patient is placed in the right lateral decubitus position and prepped for a left lateral, muscle-sparing thoracotomy. As visualization is mostly needed posteriorly, the fourth intercostal space can be exposed via the auscultatory triangle. If additional exposure is needed, the serratus anterior muscle can be divided. The lung is retracted anteriorly and the mediastinal pleura is opened over the area of coarctation and retracted with stay sutures. Alternatively, the aorta can be approached via an extrapleural dissection. The vagus and left recurrent laryngeal nerves are then identified. The extent of the dissection of the aorta and its branches depends on the type of repair to be performed. Specifics will be discussed in the sections below. The ductus arteriosus can be encircled and securely tied just before or after the vascular clamps are placed on the proximal and distal aorta. Most patients, even infants, can have several collateral vessels arising from the aorta near the coarctation site. A Satinsky-type clamp on the descending aorta is useful to control posterior collaterals. Additional collaterals can be controlled by a variety of techniques. Small hemoclips can be placed on individual collaterals and then removed after coarctation repair. Alternatively, gold (temporary) Yasargil cerebral aneurysm clips can be used to control collaterals. These atraumatic clips can be placed to lay out of the way of the repair and easily removed at the conclusion of the operation. Proximal control can be obtained with a C-clamp that is applied to include the distal aortic arch and left subclavian artery. Distal control can be obtained with a Satinsky-type clamp as noted above or if preferred a straight or angled Debakey clamp. We do not routinely give heparin as it is unnecessary in the absence of atherosclerotic disease; however, some surgeons administer a low or moderate dose of heparin prior to clamp placement. After the clamps have been placed and the ductus has been ligated, we have found it helpful to aspirate the isolated segment of the aorta using a 22-gauge needle on a 3 ml syringe. If this portion of the aorta remains decompressed following evacuation of blood, the operation proceeds. If the aorta re-distends, a search is made for additional collateral vessels.

At the conclusion of the repair, the distal clamp is removed first to allow for deairing and initial inspection of the suture line. After alerting the anesthesiologist due to the possibility of transient hypotension, the proximal clamp is removed. At this point, pulsatile lower extremity flow should be detected by pulse oximetry. There may be an initial gradient between the upper and lower extremity blood pressures due to lower extremity vasoconstriction or to a Coanda effect that favors flow up of the innominate artery as opposed to around the transverse aortic arch; however, this should resolve within 30 minutes. The residual blood pressure gradient should be 10 mmHg or less. If there is concern about a persistently high gradient, direct measurement of aortic pressure above and below the repair may be useful. Closing the mediastinal pleura after end-to-end or subclavian flap repair depends on personal preference. Some believe that closing the mediastinal pleura over the repair can cause compression of the repair site and thereby increase the incidence of recurrent coarctation. Others believe that closing the mediastinal pleura after coarctation repair may prevent the lung from adhering to the repair site, decrease the incidence of postoperative chylothorax, and create a circumferential wrap around the repair site, which will make balloon dilation safer if recurrent coarctation occurs. After patch aortoplasty repair, closing the mediastinal pleura over the repair may not be possible,

and it may cause compression of the newly enlarged aorta and produce a suboptimal outcome.

SUBCLAVIAN FLAP REPAIR

The technique of subclavian flap repair was once considered by many authorities to be the procedure of choice for neonates and infants. However, we do not favor it and recommend against its routine use. The technique requires division of the subclavian artery and turning it down as a flap to augment the area of coarctation (Fig. 82.2). Those who favor this procedure believe that it is simple and safe, and that the patch of subclavian artery will grow with the patient and therefore will lead to lower incidence of recurrent coarctation or late aneurysm formation. Unfortunately, recurrent coarctation does occur after this procedure with about the same incidence as after other commonly used procedures, and late aneurysm development has also been reported after this procedure. The subclavian flap procedure has the disadvantage that it requires permanent division of the left subclavian artery, and although this may be well tolerated in most patients, it can lead to long-term weakness of the left arm. Subclavian steal phenomenon has been described if the vertebral artery is left intact on the distal subclavian segment. Division of the left subclavian artery is problematic in the occasional patient with anomalous origin of the right subclavian artery below the coarctation site because in these patients, permanent loss of

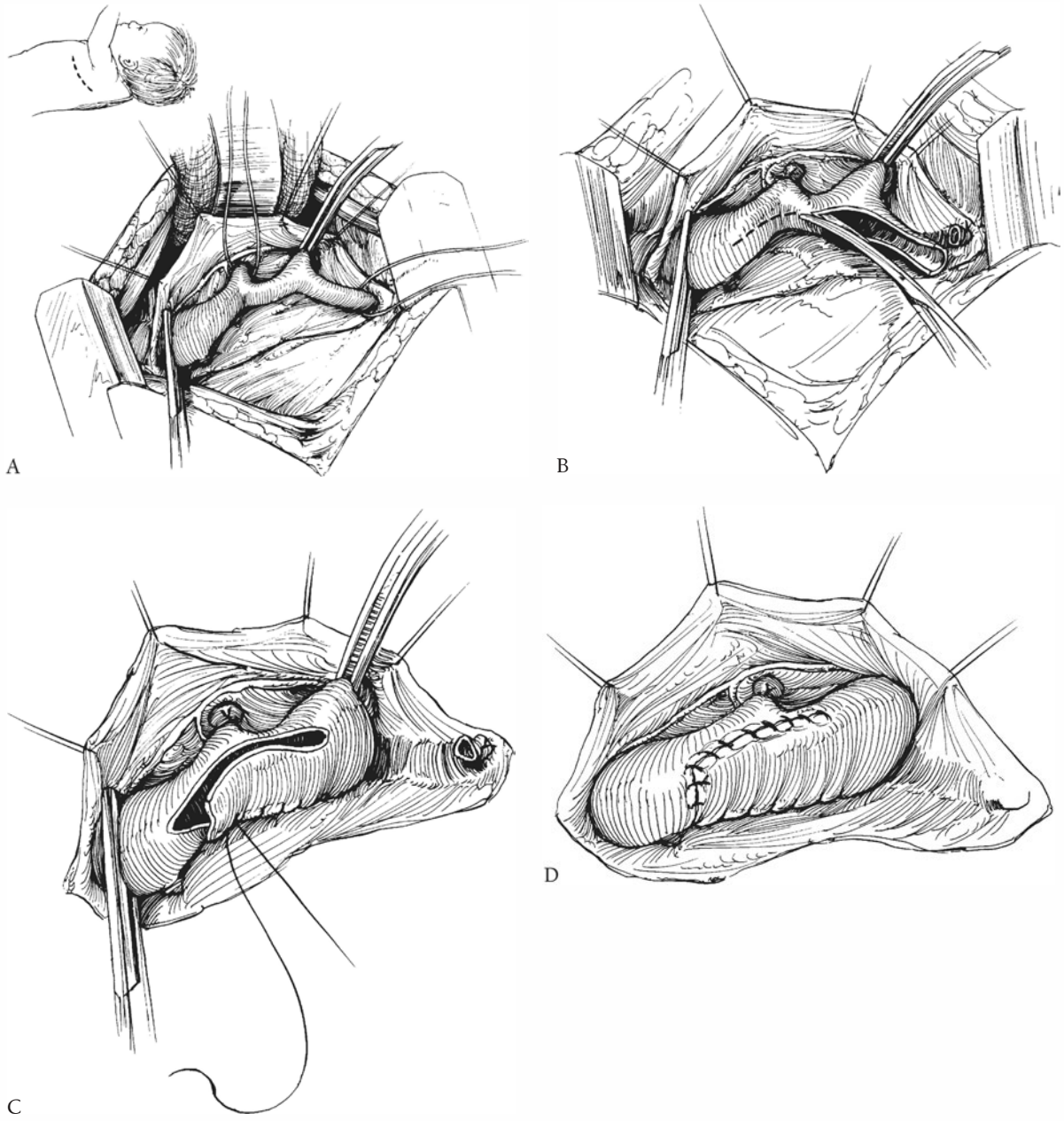


Fig. 82.2. (**A**) The coarctation is exposed through a left thoracotomy, and the aortic arch and descending aorta are controlled with clamps. The left subclavian artery and ductus arteriosus are encircled; collateral vessels can be temporarily controlled with hemoclips. (**B**) The ductus arteriosus is ligated, and the subclavian artery is ligated and divided as far distal as possible. It is also important to tie the vertebral artery (first branch off the subclavian artery) to reduce the possibility of late subclavian steal syndrome. An incision is then made through the subclavian artery and extended onto the descending aorta through the area of coarctation. (**C**) The subclavian artery is then sewn as a flap to cover the incision on the descending aorta. (**D**) The finished subclavian flap procedure provides augmentation of the juxtaductal area. The hemoclips previously placed on collateral vessels can be removed.

the left subclavian artery leaves no way to follow pressures above the coarctation site. Although some have suggested techniques for reimplanting the left subclavian as an augmentation patch without division and with relief of more proximal problems, these procedures are cumbersome, unnecessary, and less attractive than alternatives. Finally, and importantly, this procedure does not address hypoplasia of the aortic arch that is proximal to the left subclavian artery—a frequent finding in neonatal coarctation.

PATCH AORTOPLASTY

Several groups have advocated repair of discrete coarctation using a large patch of prosthetic material like Dacron, polytetrafluoroethylene (PTFE; Gore-Tex), or cryopreserved homograft (Fig. 82.3). Unlike the subclavian flap procedure, this technique does not require division of the subclavian artery, and the patch can be much larger than the subclavian flap patch. Furthermore, the patch can be extended proximally onto the aortic arch when necessary. Late aneurysm formation opposite the patch

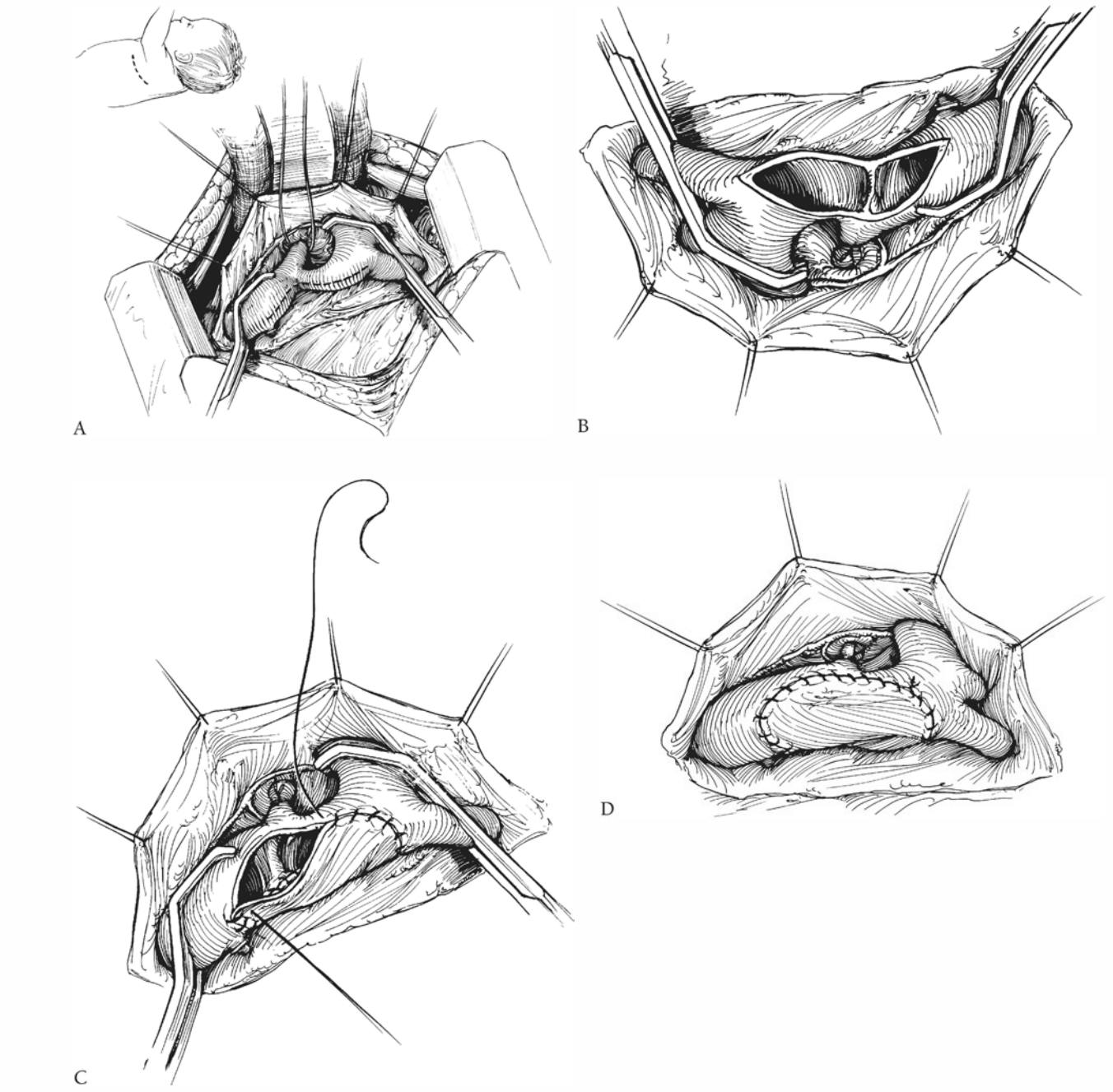


Fig. 82.3. (**A**) When the coarctation is discrete and juxtaductal, vascular occlusion clamps can be placed on the aortic arch and descending aorta after the anatomy is exposed through a left thoracotomy. The ductus arteriosus is encircled with a ligature. A Satinsky-type clamp on the descending aorta can be useful in controlling posterior collaterals. An incision is then made across the area of coarctation. (**B**) The coarctation ridge is left intact. This is usually the narrowest part of the aorta and is usually opposite the ligated ductus arteriosus. (**C**) A large patch of either prosthetic or homograft material can then be placed with running monofilament suture. This patch material should be large enough to restore a normal size to the aortic lumen. (**D**) The completed repair provides augmentation of the aorta in the juxtaductal region.

has been reported by some, and this has tempered enthusiasm for this technique. More recent studies have suggested that the occurrence of these aneurysms may be related to easily controlled technical factors such as the type of prosthetic material used and to how the coarctation ridge is managed. Most late aneurysms have been associated with the use of Dacron as compared with PTFE. Avoiding resection of the coarctation shelf has also contributed to reducing the incidence of this complication. Using a large patch of PTFE will compensate for the relatively minimal intrusion of the posterior ridge into the aortic lumen. Patch aortoplasty has also been described using a technique in which the coarctation tissue is resected and the back wall of the aorta is anastomosed, with a patch then placed over the anterior wall. However, this more complicated technique may not be justified, considering the good results with simpler patch aortoplasty without resection of the coarctation shelf. The recurrence rate after patch aortoplasty is very low, and this remains a very acceptable option for some patients. It is especially useful for recurrent coarctation, or in adult-sized patients, when mobilization of the aorta is limited.

END-TO-END AND EXTENDED END-TO-END ANASTOMOSES

The original coarctation repair was accomplished by resection of the coarctation region and end-to-end (ETE) anastomosis of the proximal and distal segments. This repair still plays a large role in the surgical treatment (Fig. 82.4). In infants, where the narrowing of the aorta can extend into the aortic arch (Fig. 82.1B), the anastomosis can be fashioned in a manner such that it uses the underside of the aortic arch to enlarge the anastomotic area (Fig. 82.5). This latter technique has been referred to as extended-ETE anastomosis. ETE anastomosis is the most common coarctation repair technique recorded in the STS Congenital Heart Surgery Database in a recent report of over 5000 patients, comprising over 75% of all reported cases. For ETE or extended-ETE repair, adequate mobilization of the proximal aortic arch to the level of the innominate artery, the proximal subclavian artery, and the descending thoracic aorta is important to achieve a tension-free repair. The aorta can be divided above and below the site of ductal insertion and the excess ductal tissue removed. It is usually only necessary to place a tie on the pulmonary end of the ductus; this will stay in place quite well after division of the ductus as long as enough tissue remains distal to the tie. The ETE anastomosis can be performed using a fine, running, nonabsorbable monofilament suture. The use of absorbable suture or interrupted rather than running anastomosis has not been consistently shown to decrease the incidence of recurrent coarctation. Care should be taken to not purse-string the anastomosis. The major advantage of the ETE techniques is that they do not require division of the subclavian artery nor do they use any prosthetic material. Furthermore, these techniques allow resection and removal of all remnant ductal tissue in the aorta, and this may play a role in limiting restenosis as well as the

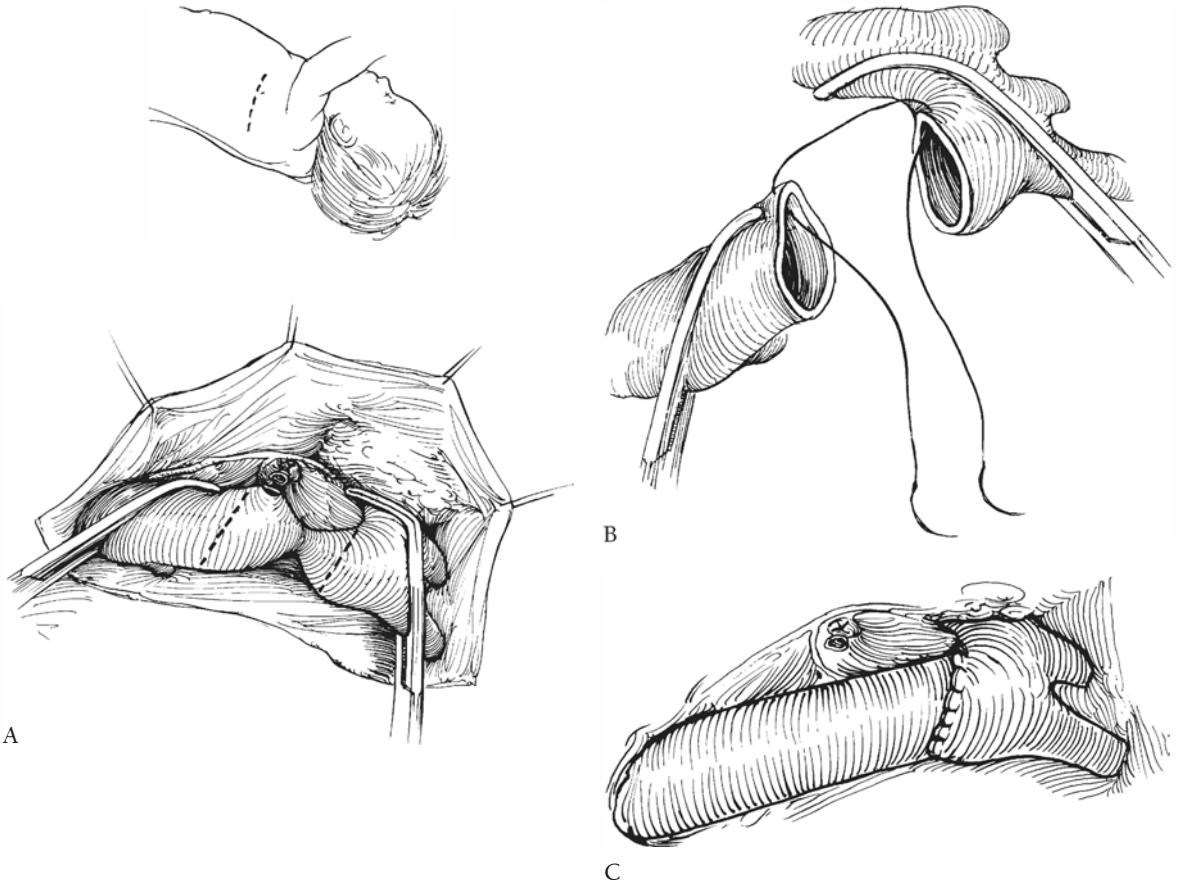


Fig. 82.4. End-to-end resection and anastomosis are easily accomplished through a left thoracotomy. (**A**) The proximal ductus arteriosus is securely tied. Vascular clamps control the aortic arch and the descending aorta. When present, collateral vessels can be controlled with hemoclips. The area of coarctation is excised and removed. (**B**) The two ends of the aorta are then anastomosed using a running monofilament suture. (**C**) This provides a normal appearance to the reconstructed aorta, and in most patients there is little difficulty in mobilizing the aorta to be reconstructed in this manner.

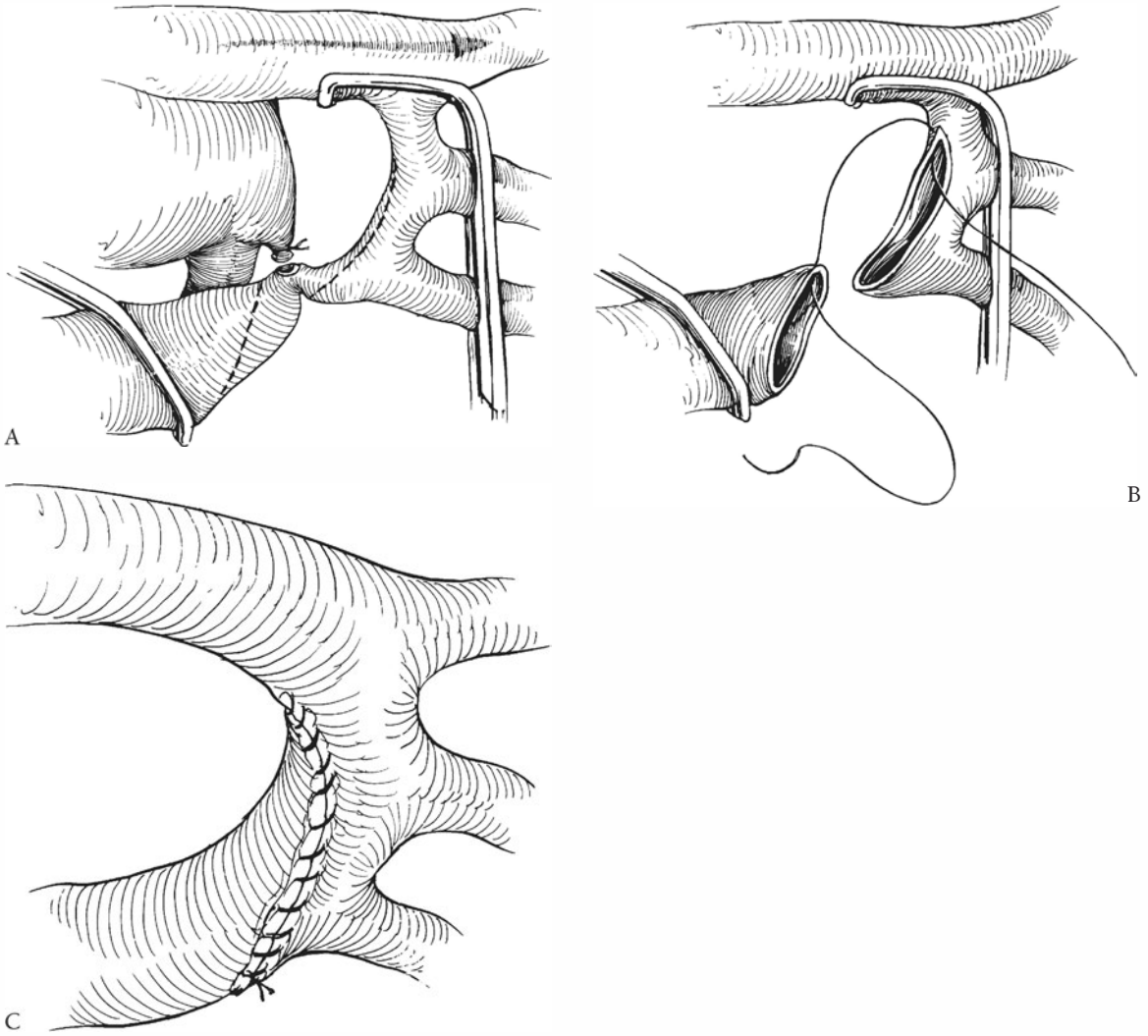


Fig. 82.5. An extended end-to-end resection can also be performed through a left thoracotomy. (**A**) It is necessary to place the proximal clamp across the left subclavian and left common carotid artery. This clamp should extend onto the ascending aorta and occlude part of the innominate artery so that the proximal incision on the underside of the aorta can extend proximally as far as the origin of the left common carotid (or farther, if necessary). The ductus arteriosus is ligated, and a distal clamp is placed on the descending aorta. It is important to mobilize the descending aorta as far distally as possible, and hemoclips can be used to control collateral vessels. These hemoclips can be removed at the completion of the procedure. (**B**) The incision on the underside of the aortic arch is carried as far proximal as necessary. The incision on the descending aorta is enlarged so that it will match the size of the proximal incision. (**C**) The two ends of the aorta are then anastomosed using running suture, and this provides augmentation of a hypoplastic aortic arch.

potential for degeneration of the aortic wall and aneurysm formation. The extended-ETE technique is also well suited to address transverse arch hypoplasia without the use of median sternotomy and cardiopulmonary bypass. It should be recognized, however, that there is a 10% to 15% recurrence rate of recoarctation after ETE repair, and aneurysms have been reported after balloon dilation of these recurrences. Nevertheless, the attractiveness of ETE techniques in removing the abnormal ductal tissue and in providing an essentially normal thoracic aorta, even in infants with hypoplasia of the transverse arch, has established these as our techniques of first choice in most cases of neonatal and infant coarctation.

INTERPOSITION GRAFT

Even though ETE repair is now our preferred method of repairing discrete aortic coarctation in neonates and young infants, this technique may not be suitable for repairing aortic coarctation in older children and adults or in recurrent coarctation. In these patients, it may be difficult to adequately mobilize the aorta proximal and distal to the coarctation segment to provide a tension-free ETE anastomosis. A prosthetic interposition graft consisting of Dacron or PTFE can be used for repair in these patients. This technique should be reserved for patients only when it is possible to place an adult-size graft (> 18 to 20 mm). Extra-anatomic bypass using prosthetic graft for the repair of aortic coarctation should be avoided except in unusual circumstances like complex recurrent coarctation. In many older patients, the use of endovascular stents deployed in the catheter lab is becoming an attractive and frequently applied option.

REPAIR VIA STERNOTOMY

Aortic coarctation can also be approached via a median sternotomy (Fig. 82.6). In these cases, the patient should be placed on cardiopulmonary bypass and cooled to a nasopharyngeal or rectal temperature of 16°C to 18°C. During the cooling period, the entire aortic arch and head vessels can

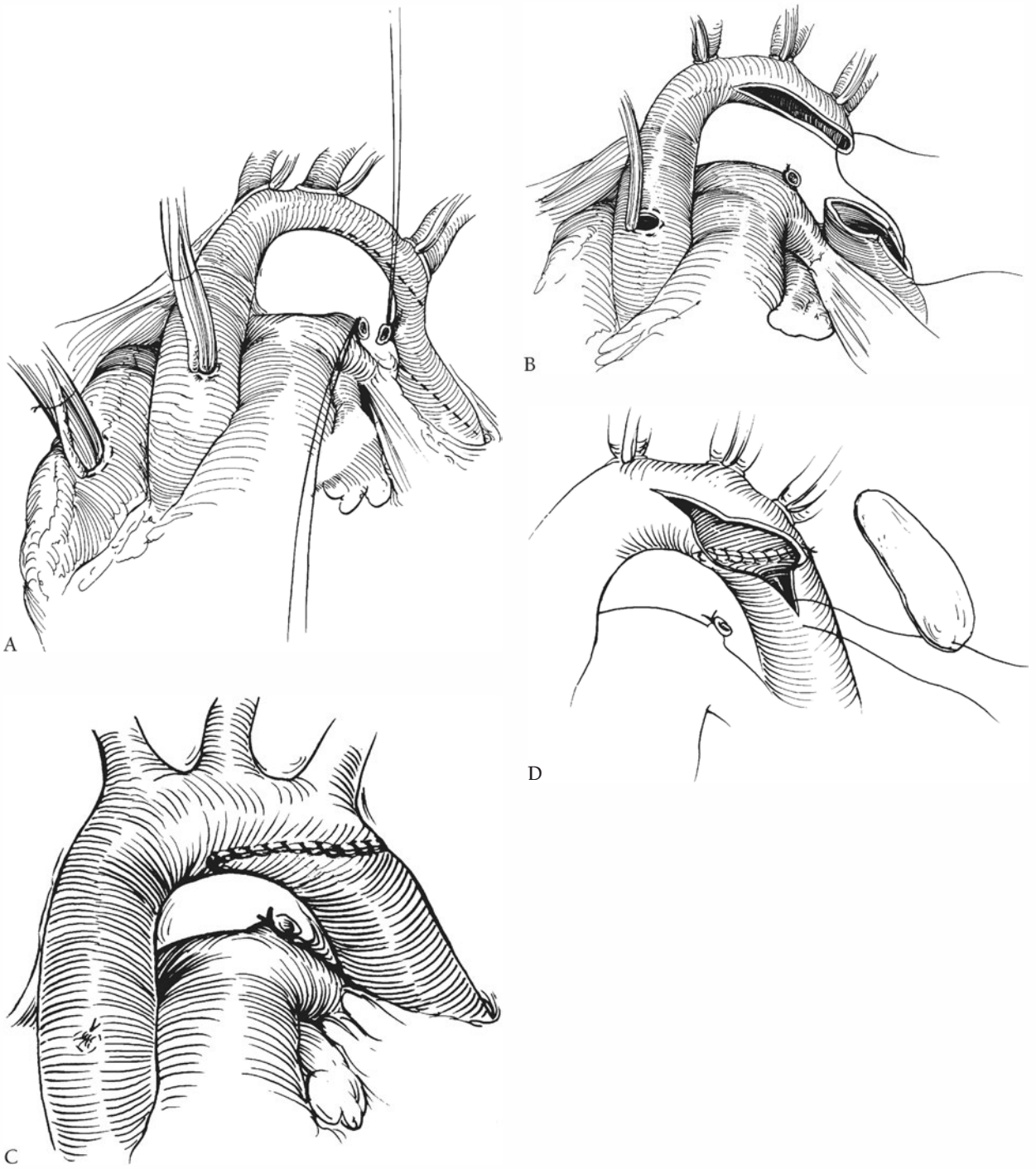


Fig. 82.6. Aortic coarctation, especially when it is associated with a hypoplastic aortic arch and other intracardiac defects, can be approached through a median sternotomy. (**A**) With the patient on cardiopulmonary bypass and cannulated through the ascending aorta and the right atrium, the aortic arch, the head vessels, and the descending aorta are aggressively mobilized. The ductus arteriosus can be divided between ligatures, and gentle traction on the ligature attached to the pulmonary arteries will retract the left pulmonary artery away from the area of coarctation. At the same time, gentle upward traction on the distal ligature will help to elevate the descending aorta so that it can be adequately dissected. (**B**) After hypothermic circulatory arrest is instituted, the aortic cannula is removed. Snares on the head vessels are secured, and an incision is then made in the underside of the aortic arch as well as on the descending aorta, as shown. (**C**) These two openings are then connected with fine, running monofilament suture, providing reconstruction of the aorta as well as the hypoplastic aortic arch. (**D**) If the two aortic segments are separated (such as may be present in interrupted aortic arch) such that significant tension is created with the anastomosis, the posterior wall of the aorta can be connected, and the anterior portion of the repair can be augmented with a patch (usually homograft), as shown. (**E**) The completed aortic repair. (**F**) Occasionally (such as in redo operations for coarctations), a sternotomy approach can be used to enlarge a recurrent coarctation. In this case, the patient is cooled on cardiopulmonary bypass and circulatory arrest is instituted. An incision is then made through the narrowest portion of the aorta. The incised area is then repaired with a patch and the patient is then replaced on bypass and rewarmed.

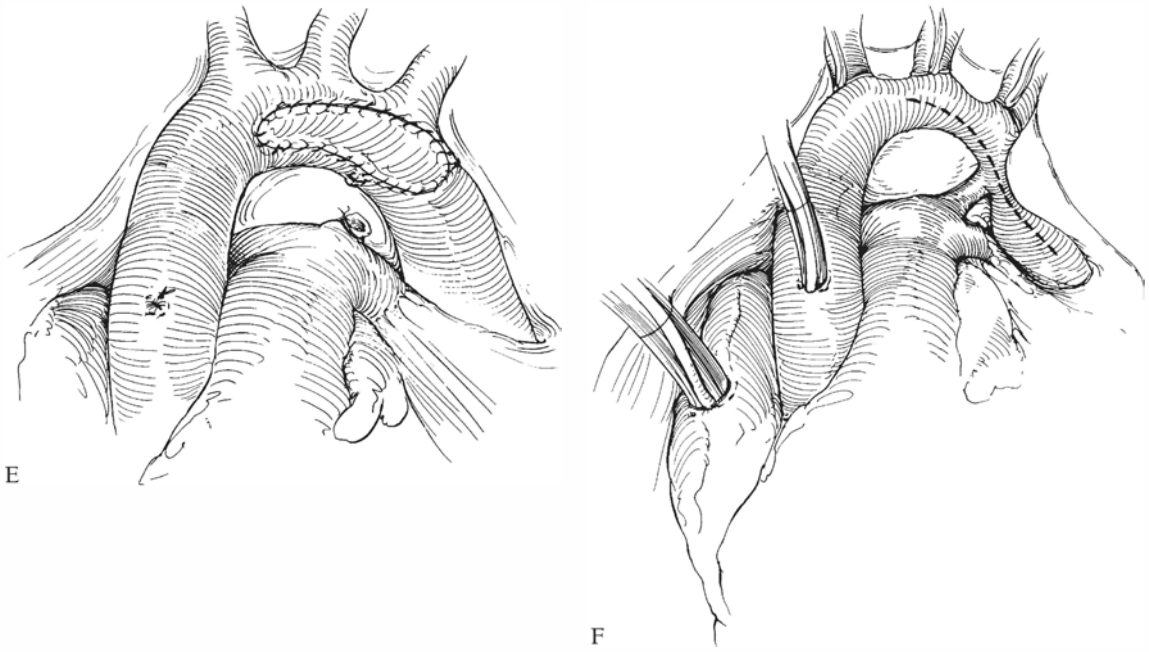


Fig. 82.6. Continued

be dissected from the surrounding tissue. Once the patient is on cardiopulmonary bypass, the ductus arteriosus can be ligated. In cases that approach aortic arch interruption (type A interruption), distal cooling may be limited from the aortic infusion cannula, and an additional arterial cannula can be placed in the main pulmonary artery and advanced across the ductus arteriosus into the distal aorta. A snare around the ductus, and this cannula (or alternatively, around the branch pulmonary arteries), can improve distal perfusion and cooling. After sufficient cooling, the patient's head is packed with ice, cardiopulmonary bypass is stopped, and the arterial cannula is removed. Snares placed around the previously dissected head vessels are occluded. (The surgeon also has the option of placing the arterial cannula into the innominate artery and snaring it so that low flow, continuous perfusion to the head can be provided. Some surgeons sew a shunt to the innominate artery to accomplish this, but we have found it quite convenient to simply advance the arterial cannula into the innominate if we choose this technique. In these cases, a clamp needs to be placed on the descending aorta to limit back bleeding into the surgical field from collateral flow. There are no compelling data to favor this technique over a short period of circulatory arrest.) The ductus arteriosus can be divided, and the distal aorta can be elevated by gentle traction on the distal ligature. This maneuver will facilitate dissection of the aorta well beyond the stenotic segment. In some instances, a vascular clamp placed as far distal as possible on the descending aorta will help to keep it elevated in the surgical field, improving the surgical exposure. All residual ductal tissues in the descending aorta near the ductal insertion should be excised. If the coarctation segment is discrete, a simple ETE repair can be performed (Fig. 82.6). If the coarctation is associated with arch hypoplasia, repair can be achieved by a combination of suturing the back wall together and using a patch to augment the arch and the repair site. Homograft is preferable in infants, especially if the entire aorta is being augmented, such as in stage 1 palliation for hypoplastic left heart syndrome (HLHS). Gore-Tex works well in older children, especially if the area is a recurrence and is surrounded by scar tissue. The sternotomy approach is convenient when the patient has significant associated cardiac defects that can be repaired at the same time or in patients with recurrent coarctation in whom control of the aorta to enable adequate access to the most proximal extent of the lesion would be challenging from the thoracotomy approach.

COMPLICATIONS

The most dreaded complication of coarctation repair is paraplegia. In an extensive study of aortic coarctation repairs in the STS Congenital Heart Surgery Database, there were no occurrences of paraplegia in close to 1000 reported cases, suggesting that the current incidence is less than previously reported and indicating that the risk of spinal cord injury is so low that it may be hard to study methods for limiting its occurrence. This has been reported to occur in 0.4% to 0.5% of patients undergoing coarctation repair. Risk factors are difficult to identify, but in some series, an aberrant retroesophageal right subclavian artery or a VSD and patent ductus arteriosus appears to put the patient at higher risk. Absence of adequate collateral vessels has also been implicated as a risk factor, but it is not always possible to delineate the adequacy of collaterals in infants, especially because they are often brought to surgery without preceding angiographic studies. Even with angiography, these tiny collaterals cannot always be visualized. Furthermore, the techniques for coarctation repair in infants are identical regardless of the extent of any collateral vessels. The use of shunts or monitoring of the distal aortic pressure has no role in infant coarctation but is commonly recommended for coarctation repair in older patients, where the use of left heart bypass for low distal pressures is appropriate and feasible. Although it would seem logical that the length of aortic cross-clamp time would relate to the incidence of paraplegia, this has not been supported by published information. Nevertheless, most surgeons attempt to limit the aortic cross-clamp time and generally try to perform the repair with clamp times of $<$20 to 30 minutes. Hypotension may also

be a contributing factor. Occlusion of the aorta with cross clamps will cause upper body hypertension; however, this should be allowed in order to maintain lower body perfusion which occurs through smaller collateral vessels. The presence of hyperthermia during aortic cross-clamping has also been linked to some cases of postcoarctectomy paraplegia. In neonates, rectal or nasopharyngeal temperatures of 34°C to 35°C can be achieved simply by turning the ambient room temperature down at the beginning of the procedure. Older patients may require active surface cooling with a cooling blanket. Although there has been some investigation into ways to prevent paraplegia (such as by identifying the arterial supply to the anterior spinal cord before surgery), this complication is not preventable by any reliable method, and families should always be informed of its potential. Other complications include hemorrhage, left recurrent laryngeal nerve palsy, chylothorax, postoperative hypertension (especially in older children), and abdominal pain (mesenteric arteritis). Hemorrhage is usually a result of tension on the suture line and can be mitigated by adequate dissection and use of a running suture to disperse the tension on individual loops. The left recurrent laryngeal nerve is visualized during mobilization of the ductus arteriosus or ligamentum arteriosum, and the potential for nerve injury can be reduced by careful dissection and judicious use of electrocautery. Chylothorax occurs in 2% to 5% of patients after repair of coarctation. This is usually caused by transection of small lymphatic vessels during the dissection for repair. In most instances, the leak will resolve with conservative management such as with dietary manipulation, but excessive drainage often requires reopening of the thoracotomy and suture ligation of the tissue that is leaking chyle. When we have seen patients with this degree of excessive drainage, it has not been necessary to give the patients cream stained with methylene blue, as suggested by some. Upon reopening the thoracotomy, the area that is "seeping" is obvious and easily controlled with figure-of-eight sutures. Abdominal pain may present in as many as one-third of patients who have hypertension after coarctation repair. Treatment for hypertension and abdominal pain includes β-blockers and "bowel rest" with intravenous fluids as necessary. It is less common for neonates and infants to require treatment of hypertension, and the increasing frequency of coarctation repair in the first year of life has largely reduced the incidence of hypertension as an important postoperative complication.

RESULTS

Despite the seriousness of this defect in neonates, surgical results are excellent. Hospital mortality in neonates after ETE or extended-ETE coarctation repair in the Society of Thoracic Surgeons Congenital Heart Surgery Database is 2.4%. For infants, the in-hospital morality rate is 1.0% and for children 1 to 18 years of age 0.3%. Surgical mortality is usually related to other factors such as associated defects, the size of the left ventricle, and the extent of the coarctation. Longer term survival appears to approach that of the general population. Depending on the age and weight of the patient at operation, the technique used, the quality of the initial repair, and the factors beyond control such as growth at the repair site, up to 20% of patients may develop a recurrent coarctation (defined as a gradient across the repair site of >20 mmHg). In most instances, this can be successfully dilated by balloon angioplasty, and surgical intervention is rarely preferred for recurrent coarctation. Late hypertension occurs mainly in older children and adults. Recurrent coarctation should be excluded as a cause of this finding. Late aneurysm formation is becoming less common and may be more often associated with balloon dilatation as opposed to primary coarctation repair techniques.

SPECIAL SITUATIONS

Coarctation and Ventricular Septal Defect

Aortic coarctation can be commonly associated with a VSD. In all cases, the coarctation should be repaired. These infants are usually in fairly significant congestive heart failure because of the LVOTO from the coarctation and the left-to-right shunt from the VSD. At the time of coarctation repair, the VSD can be dealt with by (1) pulmonary artery banding, (2) VSD closure (via a sternotomy with one-stage repair or with separate thoracotomy and sternotomy incisions during one operative setting), or (3) coarctation repair alone with nothing done for the VSD. In the latter instance, the patient can be observed postoperatively and referred for VSD closure as indicated for the treatment of that lesion as a separate entity. Previously reported Congenital Heart Surgeons Society (CHSS) data suggest that the safest approach is coarctation repair with pulmonary artery banding, but we prefer complete one-stage repair when it is apparent that the VSD will require surgical intervention. In particular, we recommend VSD closure concomitant with coarctation repair when the VSD is large (and especially when the LV outflow tract is small—but acceptable, including patients within the spectrum of interrupted aortic arch) or when the VSD is in a location that is not associated with spontaneous VSD closure (e.g., supracristal or posterior malalignment). We have done this through a sternotomy as well as through two separate incisions, and both provide excellent results. We do not favor pulmonary artery banding as palliation for a VSD, although this technique is applied successfully by some and should not be discounted in appropriate circumstances. In a review of the STS Congenital Heart Surgery Database, there was no advantage or disadvantage of one technique versus another for this combination of lesions and options.

Balloon Angioplasty for Native Coarctation

Balloon angioplasty has a limited role in the treatment of native coarctation in neonates. Results have been marginal with a high incidence of early recoarctation and development of aortic aneurysms at the coarctation site. Furthermore, there seems to be a disturbingly high incidence of paraplegia in patients who come to surgical coarctation repair after unsuccessful balloon angioplasty, and it is postulated that this may be related to the decreased stimulation for development of collateral circulation once the pressure gradient across the stenotic area has been partially relieved by angioplasty. The excellent results with surgical repair of native coarctation in neonates make balloon angioplasty a much less attractive treatment option in this population. Therefore, balloon angioplasty of native coarctation in neonates should be reserved for unusual circumstances where operative repair is not possible or desirable. In most cases, PGE_1 infusion can be continued until the complicating issue has been resolved and the child has been stabilized. In rare circumstances, such as a child presenting with cardiac arrest and resultant decreased ventricular function and a closed ductus arteriosus, balloon angioplasty may provide for a period of stabilization and recovery of myocardial

function after which the child can undergo surgical repair. When an infant requires nonsurgical intervention for an aortic coarctation, the use of an intravascular stent may be preferable to balloon angioplasty alone, since excessive dilation of the fragile aortic tissue can be avoided. Of course, when these patients recover from their complicating issues, they will ultimately need surgical enlargement of the stented portion of the aorta.

Balloon angioplasty and when indicated stent placement as an alternative to surgical repair have better long-term results in older patients. Although these procedures are not without risk of paraplegia and late aneurysm formation, the risk of recurrent coarctation is acceptable when compared with surgery. The minimum age for consideration for balloon angioplasty is 1 to 2 years of age. Patients over 20 to 25 kg are candidates for combined balloon angioplasty and stent placement. Smaller patients are generally not great candidates for stent placement due to the inability of the stents to be dilated to adult size.

Coarctation and the Hypoplastic Left Ventricle

Aortic coarctation commonly exists in the spectrum of lesions that constitute hypoplastic left heart syndrome. In the case of unequivocal hypoplasia of the left ventricle, the coarctation is repaired with patch augmentation as part of a standard Norwood procedure. Occasionally, the size of the left ventricle is borderline (approximately 20 ml/m^2) with regard to whether it is adequate to support the entire systemic circulation. Instead of staging toward a univentricular physiology, it is possible to repair the aortic coarctation by using a patch technique so that the ductus arteriosus can be left open (on PGE_1 infusion) after the procedure. This will serve to decompress the pulmonary hypertension that these patients may have postoperatively and to maintain systemic perfusion while the left ventricle recovers. If the left ventricle is inadequate to support the systemic circulation, right-to-left ductal blood flow will persist and the patient will need to be staged to a univentricular physiology. However, if the left ventricle is adequate to support the entire systemic circulation, blood flow in the ductus will change to left to right after the left ventricle recovers from the coarctation repair. The ductus arteriosus is then allowed to close by stopping the PGE_1 infusion. Occasionally, the ductus in these patients is large and will not close after cessation of the PGE_1. In these cases, it may be necessary to tie the ductus surgically.

Recurrent Coarctation

The incidence of recurrent coarctation ranges between 5% and 20%, depending on (1) the age of the patient at operation, (2) the extent of the original coarctation lesion, (3) the technique used for coarctation repair, and (4) the length of the follow-up period. Reintervention for these patients should be considered if the peak-to-peak gradient (by cardiac catheterization) across the coarctation site is >20 mmHg (a peak instantaneous gradient, by echocardiography, of >35 mmHg). Surgery can be challenging because of the adhesions and scar tissue around the aorta and lung from previous surgery. As a result, it can be very difficult and hazardous to gain proximal control of the aortic arch for the placement of a vascular clamp. Repeat ETE reconstruction is nearly impossible in these patients because the aorta cannot be mobilized adequately without placing excessive tension on the suture line. The treatment of choice for recurrent coarctation is balloon angioplasty. If balloon angioplasty is not successful and reoperation is necessary, patch aortoplasty should be considered. This can be done either through a thoracotomy or through a sternotomy on cardiopulmonary bypass with a limited period of hypothermic circulatory arrest. Alternatively, an extra-anatomic bypass graft from the ascending to the descending aorta can be used for complex recurrent coarctation, especially ones with a long segment of arch hypoplasia. We generally discourage this approach, believing that patch augmentation, even of a diffuse area of stenosis, is preferable to any extra-anatomic bypass. We have seen difficulty in patients treated with these types of extra-anatomic bypass if they present for future surgery. When this extra-anatomic solution is chosen, it can be performed either through a right thoracotomy or a median sternotomy without using cardiopulmonary bypass, depending on the area that is being bypassed.

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EDITOR'S COMMENTS

Although numerous techniques have been used for coarctation repair, all of the accepted techniques have been associated with excellent results with very low morbidity and mortality. The risk of death after coarctation repair now approaches zero for most children even with complex associated defects, except those associated with severe forms of LVOTO. Coarctation of the aorta in isolation, without associated congenital heart defects and of such severity as to not require immediate repair in infancy, is generally repaired in our institution when the patient is younger than 1 year of age. The exact timing of repair of coarctation in asymptomatic individuals remains controversial. Whereas later repair beyond 1 to 2 years of age is associated with a low incidence of late recurrence, the presence of a significant afterload on the ventricle with resulting hypertrophy may alter LV mass and compliance late in life. In addition, some patients, even after successful coarctation repair, continue to have proximal arterial hypertension, possibly related to an abnormal renin–angiotensin system or compliance differences in the aorta above and below the coarctation repair site. Many of these patients have refractory hypertension despite a completely unobstructed anastomosis.

The advent of successful balloon dilation for recurrent coarctation has permitted extension of early coarctation repair to younger than 1 year of age in the majority of patients and may help decrease the pressure load on the left ventricle. The subclavian flap technique for repair of coarctation of the aorta in infants, as popularized by John Waldhausen, has the advantage of being performed rapidly, and can be done with a single Satinsky-type partial occlusion clamp placed across the base of the ductus arteriosus incorporating the descending aorta and the arch of the aorta. The subclavian is ligated distally and divided, and the flap can be rapidly opened and sewn down across the coarctation site. I have not found it possible to resect much coarctation shelf in most of these children because there is often residual ductal tissue and the material is extremely friable. We have elected to use the subclavian flap only in situations in which a rapid anastomosis needs to be constructed or in children in whom ductal patency is being maintained after the coarctation repair is complete. Patch repair as advocated by the authors is another good choice. In patients in whom the ability of the ventricle to withstand coarctation repair is questionable because of LV volume overload, it is possible to perform a subclavian flap or patch repair, leave the ductus open, and then snare the ductus temporarily after repair while observing the effects on ventricular function and cardiac output or to leave the ductus patent and allow spontaneous closure after discontinuation of prostaglandin therapy. If patients with a relatively small LV volume continue to require ventilatory support after coarctation repair and ductal closure, conversion to a Norwood-type operation can be performed through the midline at a separate procedure.

Controversy continues regarding the use of patch material for coarctation repair in infants and older children. Although significant late problems have developed with the use of Dacron patches in the aorta, aneurysm formation has often been opposite the site of the patch, consistent with abnormalities of flow in the region of the coarctation repair. Thus, although PTFE patch has not been associated with late deterioration, the same concerns about late aneurysm formation must be considered. In addition, PTFE may bleed significantly when used proximally in the ascending aorta, and therefore, we have not utilized PTFE preferentially for coarctation repair except in exceptional circumstances. Like the authors, we have preferred to perform one-stage coarctation repair and VSD closure in children in whom the VSD is large and in whom there is associated arch hypoplasia and in those patients in whom the VSD is of a type not likely to spontaneously close. Care must be taken during coarctation repair in these children to tailor the pulmonary homograft patch adequately so that as it is brought down toward the ascending aorta, it is tapered so that a gentle take-off in size of the aorta is created to prevent kinking of the ascending aorta where the size changes abruptly to the patched arch.

In general, we have not elected to perform primary ETE anastomoses through a sternotomy incision because any tension on the anastomosis can result in bleeding that may cause stenosis of the repair site when repair stitches are used to control hemorrhage. In the same manner as for reconstruction for interrupted aortic arch, we have elected to augment the entire arch with a portion of pulmonary homograft material or to create an ETE anastomosis at the level of the subclavian and carotid vessel and then augment the undersurface with a patch of pulmonary homograft to take tension off the anastomosis. This has resulted in a lower incidence of recurrent coarctation in our patients.

Other centers have reported good results with a low risk of recoarctation with direct anastomosis of the descending aorta to the ascending aorta in patients with significant arch hypoplasia or with division of the isthmus beyond the left subclavian artery, transection of the aorta with an end-to-end anastomosis of the ascending to descending aorta, and then anastomosis of the proximal ascending aorta to the side of the newly reconstructed aortic arch. This approach is rarely necessary.

If subclavian flap repair or ETE anastomosis is performed, we have elected to routinely close the mediastinal pleura over the repair site in the hope of decreasing the possibility of late aneurysm formation and to allow better control of any disruption at the time of balloon dilation if recurrent stenosis occurs. Although pseudoaneurysm formation at coarctation repair has been extremely uncommon, we have seen this occur with primary disruption in at least one older individual. Control of the localized hemorrhage by mediastinal pleural closure can permit salvage of these patients even if aortic disruption occurs. If closure of the pleura will compromise the repair by compression, it is omitted.

The management of recurrent coarctation not amenable to balloon angioplasty remains problematic. In most individuals, it is not possible to create an ETE anastomosis after a previous coarctation repair by any of the usual techniques, and mobilization of the aorta can be difficult and create excessive tension on an ETE suture line. Therefore, recurrent coarctation may best be treated by patch augmentation of the coarctation site. We tend to use patch augmentation with either PTFE or pulmonary homograft in these individuals, usually with cardiopulmonary bypass and a brief period of circulatory arrest to decrease the risk of paraplegia in older children. Because these patients often have inadequate collateral supply, they appear to be at greater risk for the development of paraplegia postoperatively.

(*continued*)

EDITOR'S COMMENTS (*continued*)

The use of graft material for bypass of recurrent coarctation segments should be discouraged. Although in exceptional circumstances such grafts may be necessary, we have seen at least two individuals who presented late after graft bypass of recurrent coarctation segments with pseudoaneurysm formation and aortobronchial fistulae. The extensive scarring in the chest after these operations and the protrusion of the graft to the adjacent lung complicated repair of the pseudoaneurysm. Thus, if a graft is to be placed, we prefer to place it in the anatomic location with an ETE anastomosis and, if at all possible, to cover the graft with mediastinal pleura to prevent erosion into adjacent structures or to contain pseudoaneurysm formation.

Unusual circumstances in which recurrent coarctation is present in association with complex arch hypoplasia or previous extensive thoracotomies that have been performed occasionally require extra-anatomic bypass. In situations where the aortic arch and coarctation are not readily accessible due to previous surgery or there are other complicating factors, it is simple to create an ascending aorta-to-descending aorta bypass intrapericardially using an adult-size Dacron graft anastomosed to the side of the ascending aorta and then to the descending aorta posteriorly in the intrapericardial space above the level of the diaphragm. These extra-anatomic bypasses appear to have a decreased incidence of pseudoaneurysm formation compared with extra-anatomic bypass performed in the left chest and can readily relieve any residual gradients, usually without the need for cardiopulmonary bypass.

An area of controversy is the use of balloon dilation and stent implantation in native coarctation in adults. In some cases, this approach may be the most desirable because repair of coarctation in older adolescents or adults is associated with difficulty in mobilizing the arteries for direct anastomosis, and large collateral vessels in the chest wall may be associated with significant bleeding. Although the results with the use of stents for native coarctation appear to be good in the short term, long-term follow-up will be necessary to see whether the incidence of pseudoaneurysm formation or late gradients will limit the use of this technique. Nevertheless, in older patients the relative risk and benefit of catheter-based intervention versus surgical treatment would tend to favor catheter intervention, should there be a low incidence of late complications. Paradoxical hypertension, however, can occur with both catheter and surgical approaches.

The use of stents in younger patients has now been reported to be associated with recurrent stenosis requiring multiple stent redilations often requiring stent fracture as patients grow. The use of covered stents should be limited to patients who can have an adult-size stent implanted at the primary procedure.

It has now become common to use stents in recurrent coarctation in older individuals. The advantages of stent implantation are the decreased risk of recurrent obstruction and good relief of any residual gradients. Nevertheless, the long-term potential complications of a stent implanted into the wall of an abnormal aorta with abnormal compliance at the stent site have not been addressed and deserve further study. Nevertheless, the use of stents has significantly decreased the recurrence rate even after balloon dilation of recurrent obstruction at the coarctation repair site.

Postoperative complications after coarctation repair are infrequent. The most common complication, as noted, is paradoxical hypertension, which is often treated best by infusion of a short-acting β-blocking agent such as esmolol. The use of nitroprusside is less effective in controlling hypertension after coarctation repair, and very large doses of nitroprusside may be needed to control the blood pressure. In addition, nitroprusside may increase the force of ventricular ejection and predispose to possible aortic dissection in patients with abnormal aortic tissue. Therefore, we have elected to use esmolol as the primary treatment of significant hypertension postcoarctation repair. In most patients, the β-blocker can be discontinued within 24 hours after the operation without recurrence of hypertension.

Although enthusiasm has developed for the use of balloon dilation for native coarctation in infants in some centers, we discourage the use of this technique. The results of primary repair of coarctation have become excellent and result in a low incidence of recurrent coarctation or late aneurysm formation. The use of balloon dilation, which does not remove ductal tissue and may result in a primary disruption of the endothelium, seems an unattractive option in patients in whom an operative procedure may be associated with excellent early and late results. This is especially true because most infants who require coarctation repair have associated congenital anomalies that should be addressed promptly.

TLS

83 Interrupted Aortic Arch Complex

Richard G. Ohye, Takaaki Suzuki, Eric J. Devaney, Jennifer C. Hirsch-Romano, and Edward L. Bove

Interruption of the aortic arch (IAA) is a congenital anomaly characterized by complete discontinuity of blood flow between two portions of the aorta. This malformation may exist as a long-distance physical separation between adjacent segments or in the form of discontinuity between adjacent lumens of vessels that are otherwise connected externally. The latter anomaly is more commonly considered among discussions of coarctation of the aorta and is not discussed further here. IAA occurs uncommonly as an isolated lesion, being frequently associated with a number of complex intracardiac defects. Therefore, the diagnosis and management of IAA complex is best considered in combination with the entire cardiac anomaly. Recent advances in the management of IAA and the associated cardiac defects have resulted in a significant improvement in the overall outcome for patients with this complex anomaly. The approach of a single-stage repair of all coexisting defects simultaneously with the IAA has been shown to be a safe and effective management protocol.

ANATOMY

The aortic arch is that portion of the aorta between the innominate artery and the ductus arteriosus, and interruption of the arch may occur between any of the arch vessels. According to the classification originally described by Celoria and Patton (Fig. 83.1), type A occurs when the aorta is interrupted just distal to the origin of the left subclavian artery, between that vessel and the insertion of the ductus arteriosus itself. In this type, a fibrous cord is frequently found connecting the proximal and distal portions of the arch. The most common variety, type B, occurs between the origins of the left common carotid artery and the left subclavian artery. This type accounts for approximately two-thirds of all cases of IAA. In the rarest form, type C, the interruption occurs between the innominate and left common carotid arteries. This type is seen in only 5% of patients.

Associated cardiovascular anomalies are nearly always present in IAA. The most common condition is that of an isolated ventricular septal defect (VSD). In a multiinstitutional study by the Congenital Heart Surgeons Society (CHSS), which analyzed 250 neonates entered into the study over a 5-year period by 29 participating institutions, isolated VSD was present in 183 patients (73%). Other commonly associated lesions included truncus arteriosus, transposition of the great arteries (TGA) with VSD, and various forms of single ventricle. The frequency of associated anomalies, as found in the CHSS report, is shown in Table 83.1.

The VSD in patients with IAA is frequently of the malalignment type and is commonly associated with posterior deviation of the infundibular (outlet) septum. The displacement of the infundibular septum to the left of the posterior limb of the septal band results in narrowing of the left ventricular outflow tract and the potential for subaortic obstruction. Anomalous origin of the right subclavian artery from the descending aorta frequently occurs with IAA and is associated with a greater prevalence of subaortic obstruction secondary to reduced flow in utero through the left ventricular outflow tract and the aortic valve. Additional levels of left heart obstruction may also occur at the aortic valve leaflets, aortic annulus, mitral valve, and the ascending aorta.

PRESENTATION AND DIAGNOSIS

In the majority of patients, the diagnosis of IAA is first made on the discovery of signs and symptoms of congestive heart failure within the first few days of life. Lower extremity pulses may be poorly palpable or not palpable at all. In some cases, the diagnosis is not suspected until ductal closure occurs. When the ductus remains patent, flow to the lower body remains unobstructed, and the elevated pulmonary vascular resistance normally found in the newborn delays the expected increase in pulmonary blood flow through the VSD. This combination effectively delays the development of heart failure. Abrupt ductal closure, however, results in profound acidosis, cardiovascular collapse, and shock as lower body perfusion is severely reduced. Resuscitation with an infusion of prostaglandin E_1 to maintain ductal patency should be established when the diagnosis is made. If shock has occurred and there is associated renal and hepatic dysfunction, administration of dopamine is generally used as well. A period of a few days may be necessary to allow recovery of end-organ function before operative repair is performed. During this time, careful control of ventilation is needed to avoid hyperventilation, which serves to increase pulmonary blood flow and may worsen systemic perfusion further. In addition, treatment of associated conditions, including sepsis, necrotizing enterocolitis, and coagulation abnormalities, must be performed. Because 22q11 monoallelic microdeletion is a commonly associated condition in patients with IAA, occurring in 27% of patients in the CHSS report, careful control of calcium balance will often be necessary. All transfused blood should be radiated to avoid graft-versus-host disease until the diagnosis of 22q11 monoallelic microdeletion is definitively excluded.

The diagnosis of IAA can be accurately made from two-dimensional Doppler/echocardiographic studies. Cardiac catheterization with aortic angiography is rarely needed to define the anatomy. The exact site of the interruption in addition to the location of the branch vessels and the distance between interrupted segments should be determined. The presence of an anomalous origin of the right subclavian artery may be more difficult to diagnose by echocardiography but would not significantly alter the operative approach. In addition to the anatomy of the aortic arch, it is important to define the intracardiac anatomy in anticipation of a complete one-stage repair. The location

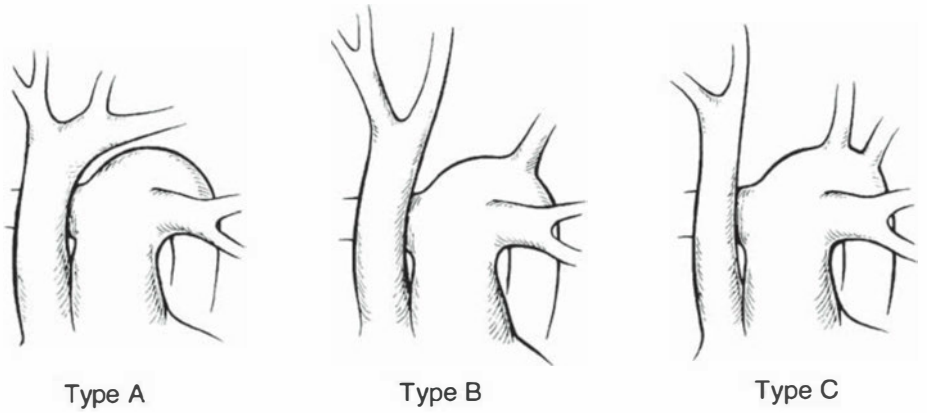


Fig. 83.1. The most commonly used classification of the three types of interruption of the aortic arch. In type A, the interruption occurs distal to the left subclavian artery. In type B, the arch is interrupted between the left carotid and the left subclavian arteries. In type C, the rarest form, the aorta is interrupted proximal to the left carotid artery.

and boundaries of the VSD, particularly in relation to the left ventricular outflow tract, aortic valve, and pulmonary valve, must be accurately seen. The majority of VSDs occur in the outlet portion of the septum, but other locations as well as additional defects must be sought. Although accurate measurement of a gradient across the left ventricular outflow tract is not possible in the presence of a nonrestrictive VSD and patent ductus arteriosus, some guidelines are helpful in predicting those patients who are likely to develop important left ventricular outflow tract obstruction after repair. When the measured ratio of the smallest diameter of the left ventricular outflow tract normalized to the diameter of the descending thoracic aorta at the level of the diaphragm is 1.0 as measured by echocardiography in diastole or is 0.6 when measured in systole, we have found that the risk of subaortic obstruction after closure of the VSD is high and that efforts to resect or incise that portion of the infundibular septum that is deviated posteriorly beneath the aortic annulus are beneficial in avoiding or reducing postoperative obstruction. Other groups have suggested other preoperative echocardiographic measurements, including cross-sectional area of the left ventricular outflow tract indexed to body surface area, subaortic diameter index, and subaortic diameter *Z* score. However, the optimal parameter, which consistently displays a high degree of sensitivity and specificity, remains elusive.

Table 83.1 Associated Cardiovascular Conditions in Neonates with Interrupted Aortic Arch

	Lesion number	Percentage
Isolated ventricular septal defect	183	73
Truncus arteriosus	25	10
Transposition of the great arteries/ventricular septal defect[a]	12	5
Aortopulmonary window	10	4
Single ventricle	9	4
Other	7	3
None	4	2

[a]Includes Taussig–Bing anomaly.
Source: Reprinted with permission from Jonas RA, Quaegebeur JM, Kirklin JW, et al. Outcomes in patients with interrupted aortic arch and ventricular septal defect: a multi-institutional study. J Thorac Cardiovasc Surg 1994;107:1099.

OPERATIVE MANAGEMENT OF INTERRUPTED AORTIC ARCH WITH ISOLATED VENTRICULAR SEPTAL DEFECT

The preferred operative management of a newborn with IAA and isolated VSD is a single-stage repair performed through a midline sternotomy approach. Nasopharyngeal and rectal temperature probes are placed. Arterial monitoring is achieved by the placement of a catheter in the umbilical or femoral artery, which has usually been accomplished in the intensive care unit before surgery. Additional noninvasive arterial monitoring should be done with a blood pressure cuff on the right arm to allow measurement of blood pressures above and below the repair site. If the right radial artery has already been cannulated, the cuff is placed on the leg. Venous access is accomplished through an umbilical or femoral venous catheter; internal jugular access is avoided in babies smaller than approximately 5 kg because of the increased risk of superior vena caval thrombosis. A midline sternotomy incision is made, and the presence or absence of thymic tissue is noted. If the thymus is present, it is totally or partially resected for exposure. Heparin is administered at this time, and the pericardium is opened. Purse-string sutures for cannulation are placed in the right atrial appendage, the ascending aorta at the base of the innominate artery, and the main pulmonary artery (Fig. 83.2). Alternatively, bicaval venous cannulation may be established at this time. A tourniquet is placed around the right pulmonary artery before bypass, and this vessel may be occluded at this time if significant hypotension exists. A Y-adapter is placed in the arterial line of the pump to allow cannulation of both the ascending aorta and pulmonary artery. Once bypass is initiated, a tourniquet is placed around the left pulmonary artery and is engaged (as well as the right if not already done) to direct all pump flow through the ductus and to the lower body. Alternatively, a tourniquet may be engaged around the ductus itself to exclude pump flow into the pulmonary circuit. This method of cannulation allows for even cooling of the upper and lower body in preparation for circulatory arrest. Bypass is initiated at a blood temperature of approximately 30°C, and progressive cooling to a nasopharyngeal temperature of 18°C or less is accomplished. Ice bags are placed around the head. Systemic vasodilatation with phentolamine (0.1 mg/kg) is given to facilitate even cooling between the nasopharyngeal

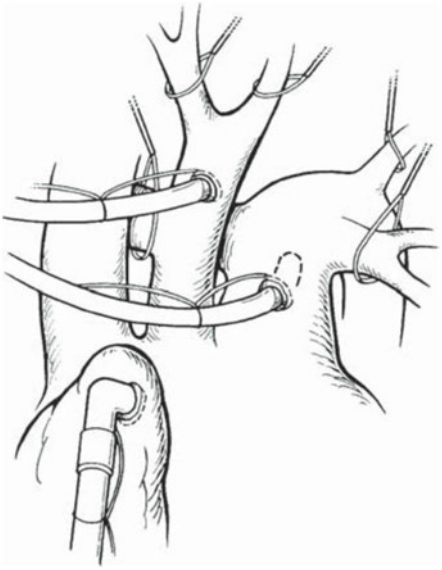


Fig. 83.2. Cardiopulmonary bypass and cannulation for repair of type B interruption of the aortic arch. Arterial cannulation is performed through the ascending aorta and proximal main pulmonary artery with the branch pulmonary arteries occluded with tourniquets. Venous return is through the right atrium. Additional tourniquets are placed around the head vessels for occlusion during the period of circulatory arrest.

and rectal temperatures. A minimal cooling period of 20 minutes is allowed, regardless of when the desired temperatures are reached.

During the interval required for cooling, the ascending and descending aorta and the brachiocephalic vessels are mobilized widely. Sufficient intercostal arteries are divided to permit a tension-free anastomosis. After appropriate cooling, the circulation is arrested, and the arch vessels are snared. Blood cardioplegia is delivered through a separate cannula after placement of a cross-clamp or through the arterial cannulation site to induce arrest. The ductus arteriosus is ligated on the pulmonary artery side and excised distally, with care being taken to remove all ductal tissues on the descending aorta. A vascular clamp on the descending aorta facilitates exposure and avoids the need to place excess tension with forceps on the vessel itself. When there is an anomalous right subclavian artery, ligation and division are usually required to permit an anastomosis without tension or compression of the trachea. In the case of a type B IAA, the descending aorta is sutured to an opening beginning at the base of the left common carotid artery, which is extended proximally along the underside of the ascending aorta (Fig. 83.3). Proper alignment is important, and care must be taken to avoid spiraling this incision. The anastomosis is performed with a continuous technique using 6-0 or 7-0 absorbable or nonabsorbable monofilament suture. In cases of type A IAA, the transverse aortic arch may be hypoplastic, as it is frequently seen with coarctation of the aorta. The anastomosis should be spatulated to enlarge the transverse arch using the distal aorta, bringing the suture line proximally to the level of the innominate artery to ensure that there is no residual narrowing.

The total elapsed circulatory arrest time at this point of the procedure is generally no more than 12 to 15 minutes, allowing ample time for VSD closure through an incision in the right atrium. Alternatively, bicaval cannulation can be utilized, rather than single atrial access. If this approach is employed, cardiopulmonary bypass may be resumed at this point for the VSD closure to minimize circulatory arrest time. Occasionally, when there is deficiency or absence of the infundibular septum and overriding of the VSD by the pulmonary valve, optimal exposure is best achieved through an incision in the main pulmonary artery. Transatrial exposure of the defect is usually accomplished easily with traction sutures placed on the anterior and septal leaflets of the tricuspid valve, and an appropriately trimmed patch of polytetrafluoroethylene material is placed with a continuous 6-0 polypropylene suture. When there is coexisting posterior deviation of the infundibular septum (Fig. 83.4), it has been our routine practice to perform a wedge resection of the septum through the VSD (Figs. 83.5–83.7). On occasion, an incision in the septum alone is performed, which is usually sufficient to enlarge the left ventricular outflow tract. The atriotomy is closed, and the cannulae are reinserted for cardiopulmonary bypass if the VSD was closed under circulatory arrest, with air being evacuated through a generous needle hole in the ascending aorta. Rewarming is accomplished, during which time epicardial pacing wires are placed on the right atrium and ventricle. After bypass is discontinued, one or two additional catheters are placed in the right atrium through the cannulation purse string. Left atrial pressure monitoring lines may also be placed. Routine sternal closure over a mediastinal drain is performed; occasionally, sternal closure is delayed if there is hemodynamic compromise or significant edema.

An alternative to deep hypothermic circulatory arrest for the repair of IAA is regional cerebral perfusion. For this technique, a limited amount of flow is delivered to the brain through the innominate artery. The ascending aorta cannula is placed near the base of the innominate artery. The patient is cooled and otherwise prepared in the same manner as for circulatory arrest. Cerebral oxygen saturation by near infrared spectroscopy may be employed to monitor cerebral perfusion, and a right radial arterial line can be inserted to monitor perfusion pressure. At the time that circulatory arrest would be initiated, the aortic cannula is advanced up by the innominate artery and snared in place. Flow is started at 5 cm^3/kg/min and gradually advanced to 30 to 50 cm^3/kg/min, while

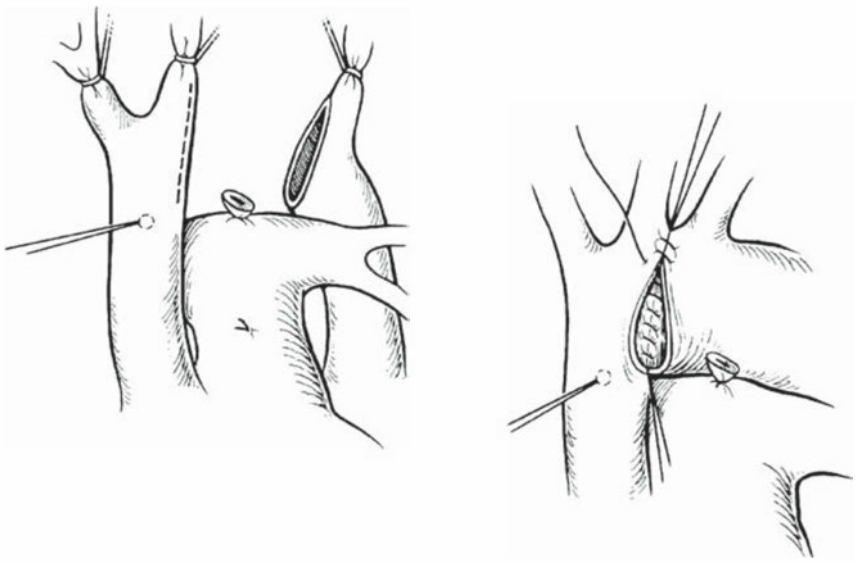


Fig. 83.3. Direct anastomosis between the descending thoracic aorta and the ascending aorta is performed with a continuous suture technique after all ductal tissue is resected. The anastomosis is begun at the base of the left carotid artery.

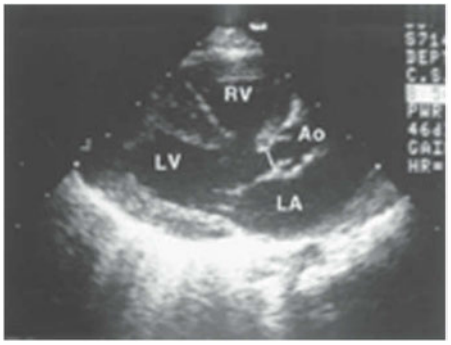


Fig. 83.4. An echocardiogram in the long axis view demonstrating a malalignment ventricular septal defect with posterior deviation of the infundibular septum typically seen with interruption of the aortic arch. (Reprinted with permission from Bove EL, Minich LL, Pridjian AK, et al. The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 1993;105:289.)

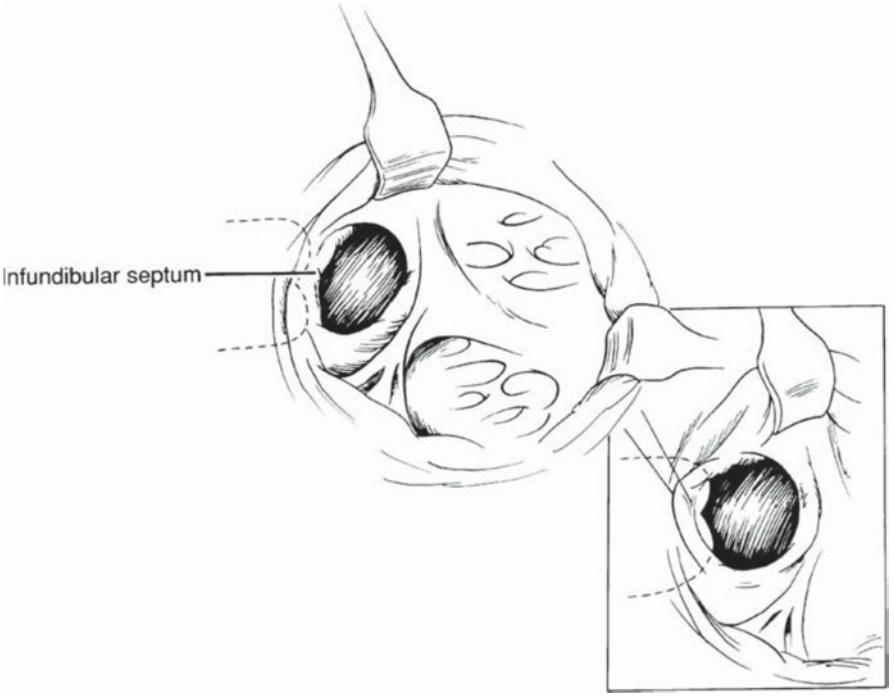


Fig. 83.5. View of the ventricular septal defect through the tricuspid valve. A traction suture in the infundibular septum facilitates exposure of the aortic valve. (Reprinted with permission from Bove EL, Minich LL, Pridjian AK, et al. The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 1993;105:289.)

monitoring cerebral oxygen saturation for a return to baseline (and pressure if a right radial arterial line is present). The arterial cannula in the pulmonary artery is removed, the venous return is collected by the venous cannula(e), and the remainder of the procedure is identical to that performed during circulatory arrest. While many centers have adopted this approach in an effort to provide better neurodevelopmental outcome, currently there are no data to suggest a benefit, particularly when compared with circulatory arrest times less than 20 to 30 minutes.

SPECIAL CIRCUMSTANCES

Interrupted Aortic Arch with Aortic Valve Hypoplasia or Atresia

When left ventricular outflow tract obstruction secondary to hypoplasia or atresia of the annulus of the aortic valve is present in conjunction with a VSD, one of two modifications of the Damus–Kaye–Stansel procedure may be used to bypass the aortic valve, depending on the specific anatomy. The approach is tailored to achieve an optimal surgical result with respect to a widely patent, tension-free aortic reconstruction, while avoiding compression of adjacent structures, such as the left main stem bronchus and pulmonary artery. One option is to divide the distal ascending aorta and repair the IAA by direct anastomosis. This reconstructed distal aortic segment, which may be augmented with a patch of allograft material, is then sutured end-to-end to the divided proximal main pulmonary artery. The proximal ascending aorta is then sewn end-to-side into the main pulmonary artery. This method of repair can be facilitated by performing a Lecompte maneuver. Alternatively, the ascending aorta may be transected at the level of the divided main pulmonary artery. The IAA is repaired forming the posterior wall for a modified Norwood-type augmentation patch of the aorta, as is described below for IAA with single ventricle. This reconstructed aorta is then sewn end-to-end to the proximal pulmonary artery, incorporating the proximal ascending aorta, as in a modified Norwood

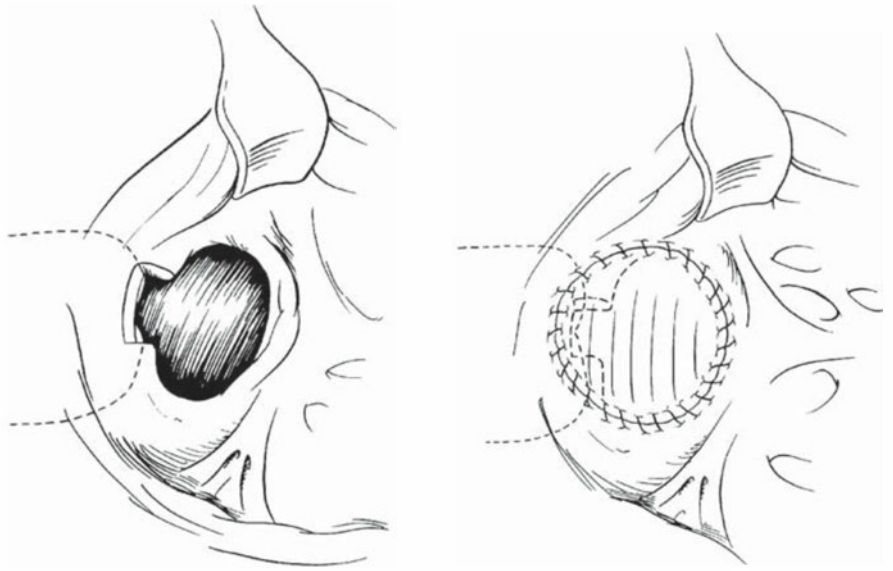


Fig. 83.6. Resection of the infundibular septum is performed until the aortic valve is reached. The ventricular septal defect is then closed with a patch. (Reprinted with permission from Bove EL, Minich LL, Pridjian AK, et al. The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 1993;105:289.)



Fig. 83.7. The appearance of the ventricular septal defect and the displaced infundibular septum as seen from the lateral view. Note the position of the traction suture in the septum (*upper left*) and the position of the ventricular septal defect patch (*lower right*). (Reprinted with permission from Bove EL, Minich LL, Pridjian AK, et al. The management of severe subaortic stenosis, ventricular septal defect, and aortic arch obstruction in the neonate. J Thorac Cardiovasc Surg 1993;105:289.)

procedure. Following either technique of aortic reconstruction, the VSD is closed to channel left ventricular blood to the pulmonary valve and a conduit is interposed between the right ventricle and the pulmonary artery bifurcation. While we prefer a single-stage approach, the pulmonary artery bifurcation may alternatively be closed with a patch, and pulmonary blood flow is provided through a modified Blalock–Taussig shunt, deferring complete repair until a later date.

The third alternative for patients with IAA and aortic valve hypoplasia, with or without VSD, is a Ross/Konno procedure. The ascending aorta is divided and the proximal ascending aorta and aortic valve are resected. The IAA is repaired, generally by forming a posterior wall at the site of the interruption, with an anterior Norwood-type patch augmentation. The pulmonary autograft is harvested in the usual manner and a standard Ross procedure is performed. For the Konno portion of the operation, it is often not necessary to fully divide the ventricular septum and place a patch to enlarge the left ventricular outflow tract. It is generally sufficient to divide the aortic annulus and simply incise the septum up to, but not through the endocardium of the right ventricular outflow tract (Fig. 83.8). A wedge of muscle can also be resected from the septum to further enhance unobstructed left ventricular output.

Interrupted Aortic Arch with Truncus Arteriosus

When interrupted aortic arch is associated with truncus arteriosus, significant modifications in the technique of repair are required. Nearly all interruptions are type B, although a single patient with type A has been seen in our center (Fig. 83.9). The ascending aorta distal to the origin of the ductus arteriosus and the pulmonary arteries are usually quite small in this condition. The arterial cannula is placed into the distal ascending aorta as it will be subsequently augmented, and the branch right and left pulmonary arteries are occluded at the onset of cardiopulmonary bypass. During cooling in preparation for circulatory arrest or regional cerebral perfusion, the innominate, left carotid, and left subclavian arteries are mobilized and encircled with snares for subsequent occlusion, as described in the preceding section. The remaining ascending and proximal descending aorta are also mobilized. Once circulatory arrest or regional perfusion is established, the ductus arteriosus is excised, and the orifices of the right and left pulmonary arteries are excised from the aorta with a button of adjacent arterial tissue that is as large as is feasible. The large opening in the proximal ascending aorta resulting from the excision of both the ductus arteriosus and the pulmonary artery bifurcation is extended superiorly along the medial side of the ascending aorta and into the base of the left carotid artery (Fig. 83.10). Primary end-to-end anastomosis with the descending aorta is then begun, using a running suture technique and placing the descending aorta and left subclavian artery distal to the ascending aorta beginning at the base of the left carotid artery. The large defect remaining below this portion of the anastomosis is then reconstructed and further augmented with a patch of allograft tissue beginning from the sinus of Valsalva and carrying it upward into the lower aspect of the primary arch anastomosis (Fig. 83.11). In this manner, two potential problems are avoided. First, placing the descending aorta too far proximally onto the ascending aorta may result in obstruction to the left pulmonary artery or compression of the left mainstem bronchus and residual aortic obstruction between the truncal valve and the anastomosis of the aortic arch. Second, this technique also maintains the potential for growth because neither the suture lines nor the prosthetic material are placed circumferentially. After completion of this aspect of the procedure, the VSD is then repaired in the usual manner generally on full cardiopulmonary bypass. The right ventricle-to-distal pulmonary artery continuity is established with a cryopreserved allograft or valved bovine jugular conduit.

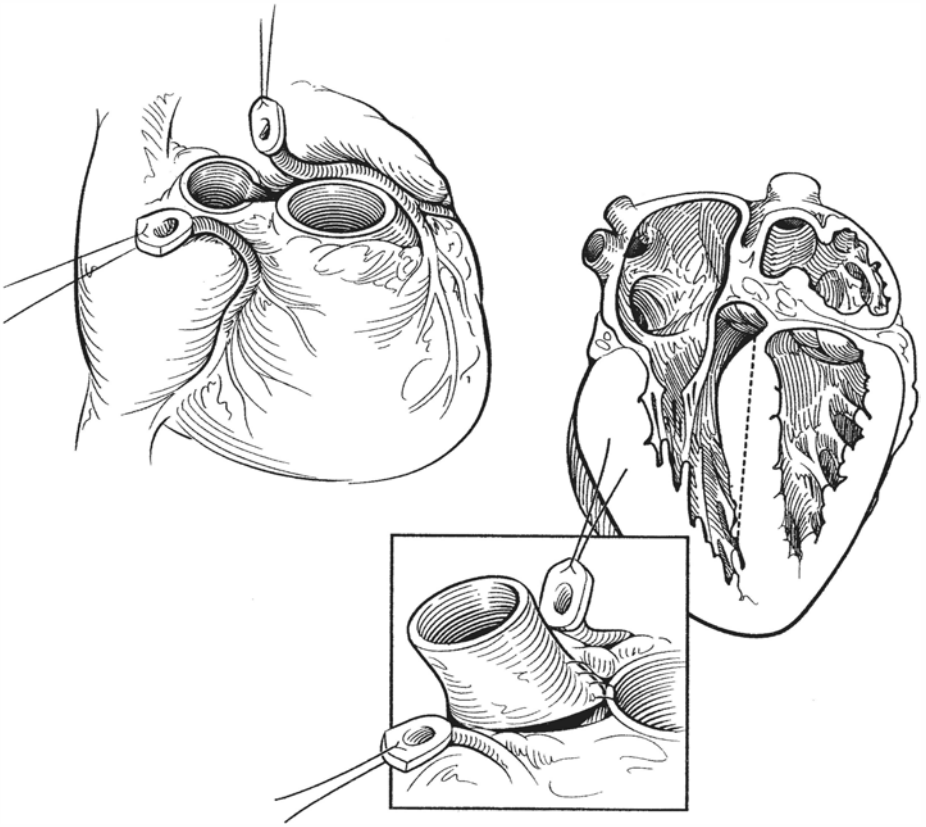


Fig. 83.8. After removal of the aortic valve and harvesting of the autograft, a partial thickness septal incision is performed to enlarge the aortic annulus (*left*). If necessary, a septal myomectomy may be performed for subaortic stenosis (*right*). In the region of the incision, the autograft is anastomosed directly to the endocardium of the right ventricular outflow tract (inset).

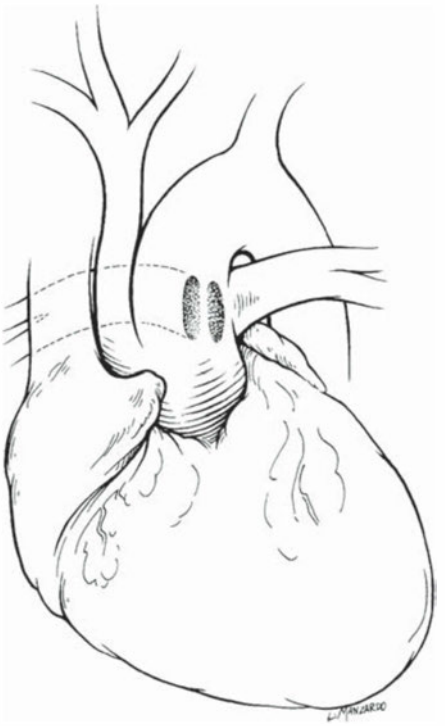


Fig. 83.9. A truncus arteriosus associated with type B interruption of the aortic arch. Note the small ascending aorta distal to the origin of the pulmonary arteries and the ductus arteriosus. (Reprinted with permission from Baue AE, Geha AS, Hammond GL, et al. eds. Glenn's Thoracic and Cardiovascular Surgery. 6th ed. Norwalk, CT: Appleton & Lange; 1994:1216.)

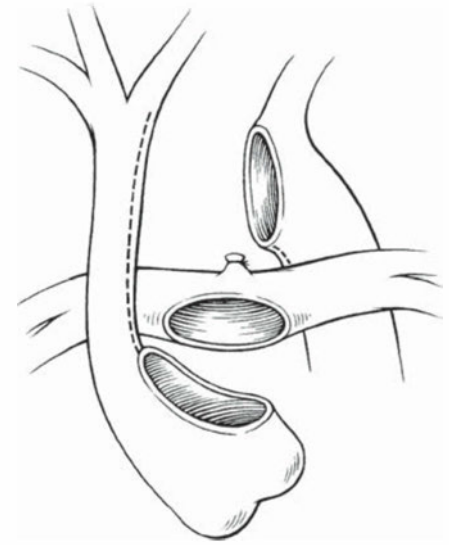


Fig. 83.10. Appearance of the great vessels after removal of the pulmonary arteries and ligation of the ductus arteriosus. The distal ductal tissue is excised from the descending aorta, and the ascending aorta is opened from the pulmonary artery excision site to the base of the left carotid artery. (Reprinted with permission from Baue AE, Geha AS, Hammond GL, et al. eds. Glenn's Thoracic and Cardiovascular Surgery. 6th ed. Norwalk, CT: Appleton & Lange; 1994;1216.)

Interrupted Aortic Arch with Transposition of the Great Arteries

Single-stage arterial switch and IAA repair are performed for TGA with IAA with few modifications. The arch anastomosis is performed first in a manner identical to that described in the section on IAA with VSD. Because the ascending aorta will be transected for the arterial switch and relocated posterior to the pulmonary artery bifurcation, the division is performed a few millimeters distal to the sinotubular ridge to foreshorten the ascending aorta and avoid kinking.

Interrupted Aortic Arch with Single Ventricle

When IAA coexists with single ventricle, the technique of repair must be individually tailored to the specific anatomy. In the usual situation, IAA is found with TGA and tricuspid atresia or with double-inlet left ventricle (Fig. 83.12). In these conditions, there will be unrestricted pulmonary blood flow and a restrictive or potentially restrictive outlet (bulboventricular) foramen that will result in subaortic obstruction. Therefore, the initial operative procedure must be designed to relieve all levels of systemic outflow tract obstruction and to control pulmonary blood flow. This is best accomplished with a modified Norwood procedure, which augments the entire ascending aorta and aortic arch with a patch of cryopreserved pulmonary allograft material (Fig. 83.13). The ascending and descending portions of the arch are sutured directly only along their posterior walls and the undersurface is opened distally for at least 1 to 2 cm in length. The ascending aorta is divided above the valve and opened posteriorly as well. The allograft patch is then used to augment the entire aorta, which is then sutured end-to-end to the proximal main pulmonary artery, incorporating the proximal end of the ascending aorta to ensure unobstructed coronary blood flow (Fig. 83.14). A 3.5-mm modified Blalock–Taussig shunt is routinely used, with a 4-mm shunt reserved for infants >3.8 to 4.0 kg. This technique of aortic reconstruction bypasses the restrictive outlet foramen with the pulmonary valve, allows appropriate size matching of the main pulmonary artery and augmented ascending aorta, and

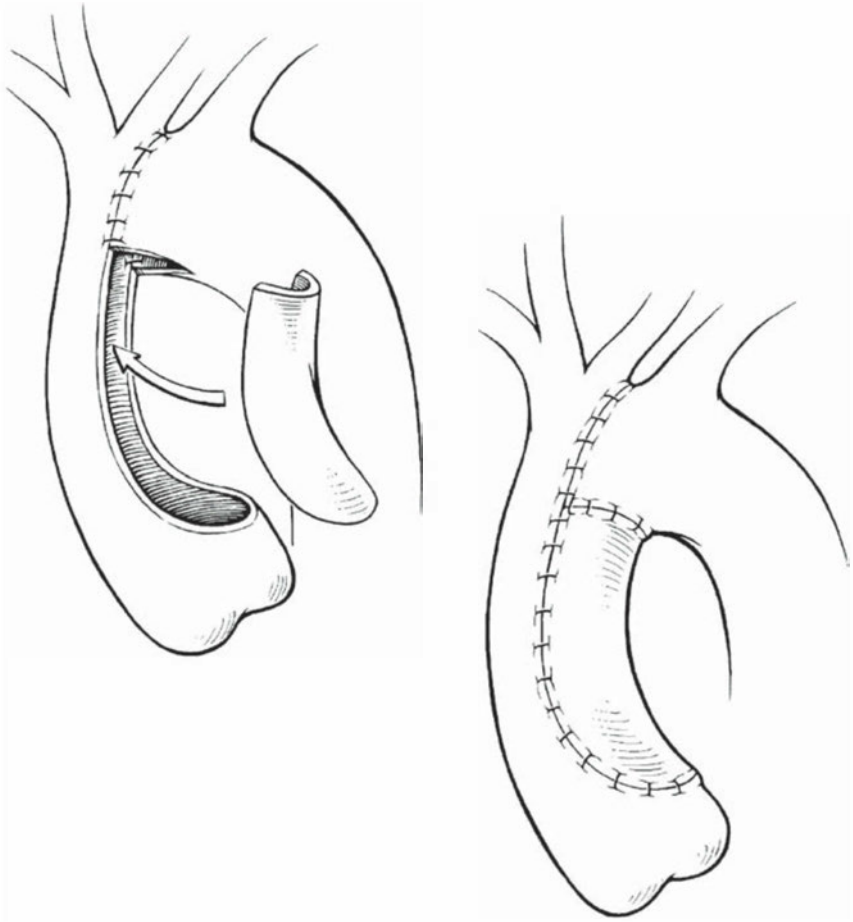


Fig. 83.11. A direct anastomosis is made between the descending aorta and the distal ascending aorta–proximal left carotid artery. The proximal ascending aorta is augmented with allograft tissue or prosthetic material. (Reprinted with permission from Baue AE, Geha AS, Hammond GL, et al. eds. Glenn's Thoracic and Cardiovascular Surgery. 6th ed. Norwalk, CT: Appleton & Lange; 1994;1217.)

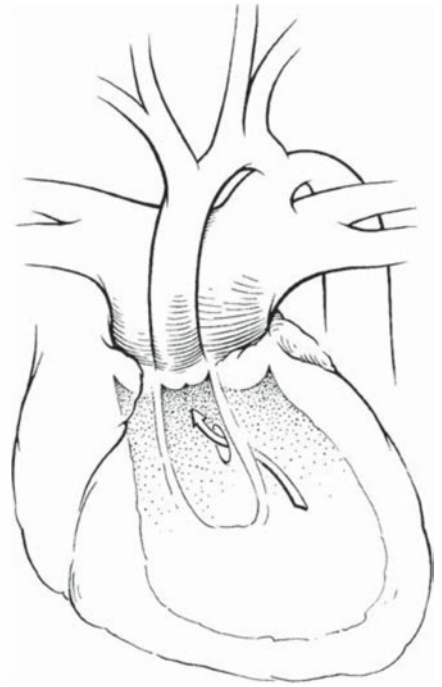


Fig. 83.12. The typical appearance of tricuspid atresia with transposition of the great vessels and coarctation of the aorta. The aortic blood flow is dependent on the bulboventricular foramen. Although the illustration shows a coarctation, the approach is similar for patients with arch interruption. (Reprinted with permission from Mosca RS, Hennein HA, Kulik TJ, et al. Modified Norwood operation for single left ventricle and ventriculoarterial discordance: an improved surgical technique. Ann Thorac Surg 1997;64:1127.)

avoids circumferential suture lines or prosthetic grafts in the arch repair.

POSTOPERATIVE MANAGEMENT

When single-stage repair of IAA and associated intracardiac defects is performed, the early postoperative management is similar to that for any neonate or young infant undergoing a complex cardiac repair. Mechanical ventilation is generally required for 2 to 4 days, and extubation is carried out after excess edema is mobilized. If weaning from the ventilator is unsuccessful, diaphragm paralysis from phrenic nerve injury, upper airway obstruction secondary to left recurrent laryngeal nerve injury and resultant vocal cord paralysis, and tracheal compression from the reconstructed aortic arch must all be investigated. Inotropic support with low- or intermediate-dose dopamine (5 to 10 μg/kg per minute) is routinely used and should be all that is necessary to maintain hemodynamic stability. When more significant levels of support are required, a detailed search for residual hemodynamic lesions must be carried out. Doppler/echocardiography is useful in diagnosing ventricular dysfunction, residual VSD, left ventricular outflow tract obstruction, tamponade, and atrioventricular or semilunar valve regurgitation. Residual aortic arch obstruction may be diagnosed or highly suspected by simple four-limb blood pressure recordings, although if cardiac output is low, gradients may be underestimated. Cardiac catheterization should be used if any doubt remains as to the cause of low cardiac output.

In patients with a single ventricle, who have undergone aortic arch repair and a systemic-to-pulmonary artery shunt, systemic output is dependent on the delicate balance between the systemic and pulmonary vascular resistances. Residual arch obstruction is extremely poorly tolerated and usually results in rapid deterioration. Excessive pulmonary blood flow is generally not a problem provided the correct size shunt is placed. Reduction of systemic vascular resistance with the appropriate medications is of benefit in improving oxygen delivery to the tissues.

OUTCOMES

Survival

Early and late survival for patients with IAA and VSD treated by single-stage repair has steadily improved over the past decade. The results for 60 consecutive neonates undergoing simultaneous arch and intracardiac repair (excluding single ventricle patients) between 1986 and 1994 at the University of Michigan are shown in Table 83.2. Although this series includes patients with coarctation in addition to IAA, the management and outcomes were sufficiently similar, especially considering all were younger than 1 month of age at the time of repair. Early mortality was 11.6% for the entire group; overall mortality (early plus late) was 15%. Not surprisingly, those patients with isolated VSD had a better

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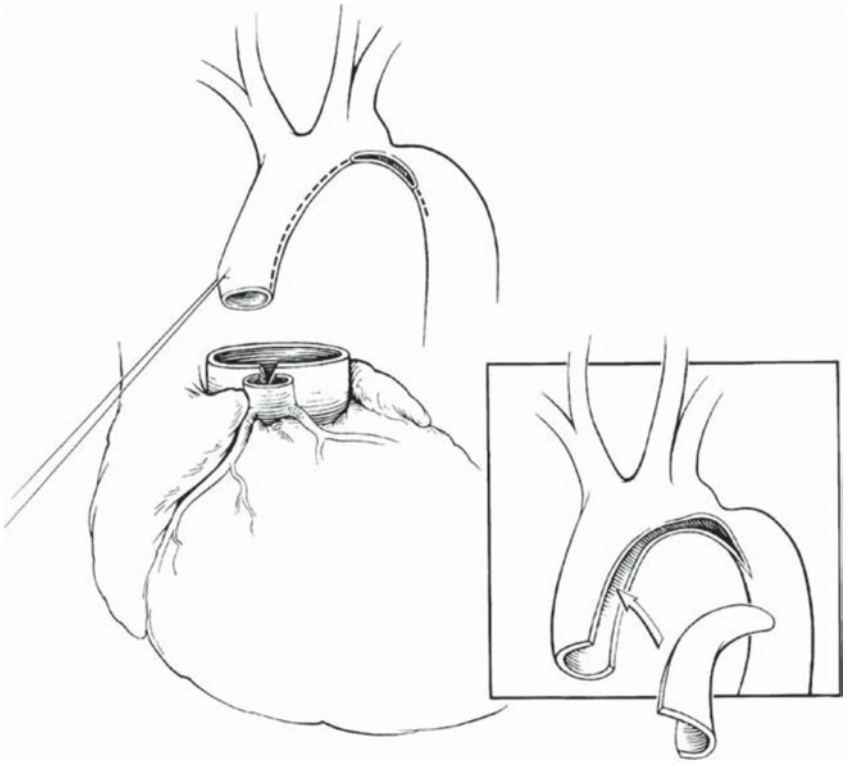


Fig. 83.13. Modified Norwood operation for single ventricle with ventriculoarterial discordance. Both great vessels are transected, and the ductal tissue is excised. The entire ascending aorta is opened and augmented with a patch of allograft tissue. (Reprinted with permission from Mosca RS, Hennein HA, Kulik TJ, et al. Modified Norwood operation for single left ventricle and ventriculoarterial discordance: an improved surgical technique. Ann Thorac Surg 1997;64:1127.)

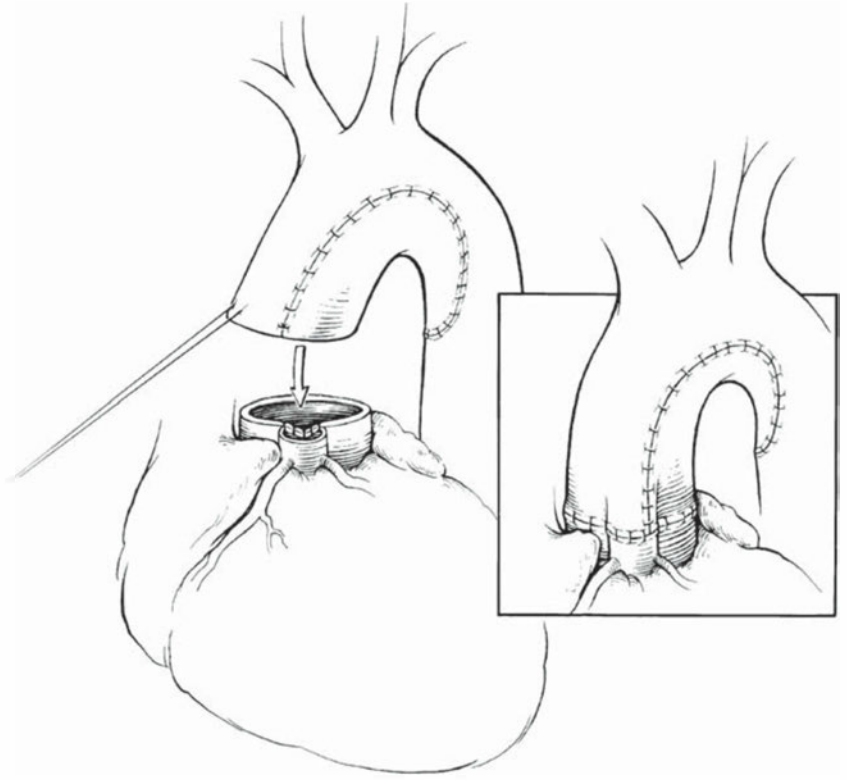


Fig. 83.14. The aorta is anastomosed in an end-to-end manner to the pulmonary artery, incorporating the proximal ascending aorta into the suture line. Pulmonary blood flow is provided through a systemic-to-pulmonary artery shunt or a cavopulmonary connection. (Reprinted with permission from Mosca RS, Hennein HA, Kulik TJ, et al. Modified Norwood operation for single left ventricle and ventriculoarterial discordance: initial management with an improved surgical technique. Ann Thorac Surg 1997;64:1128.)

outcome than those with more complex intracardiac defects. Among 37 neonates with coarctation or IAA and isolated VSD, there were three early deaths (8%) and only one late (noncardiac) death. For the 23 neonates with coarctation or IAA and more complex anomalies, there were four early deaths (17%) and one late death.

The results reported from the multi-institutional study by the CHSS were less optimistic. Among 174 neonates with IAA and VSD undergoing repair, survival was 73%, 65%, and 63% at 1 month, 1 year, and 4 years, respectively. In contrast to the series from Michigan, myotomy or myectomy in the presence of narrowing of the subaortic area was found to be a risk factor for death in the CHSS report. Paradoxically, the CHSS data showed that repair without concomitant procedures to address subaortic narrowing (myotomy/myectomy, Damus–Kaye–Stansel) was a risk factor, as was subaortic narrowing itself, leaving no clear solution to that difficult problem. In a more recent IAA follow-up study by the CHSS, data from 447 IAA patients at 33 institutions from 1987 to 1997 revealed a 21-year freedom from death of only 60% overall.

Recurrent Arch Obstruction

Late complications are specific to the type of intracardiac defect repaired or to the expected difficulties that arise after palliation for those patients with single ventricle lesions. The patient should always be monitored for residual or recurrent arch obstruction, and treatment is usually indicated for resting gradients in excess of 30 mmHg. Residual gradients of 20 mmHg occurred in only 2 of 53 late survivors (3.7%) of simultaneous neonatal arch repair for either IAA or coarctation and intracardiac repair of associated defects performed in our institution between 1986 and 1994. No patient has required reoperation for recurrent arch obstruction (mean follow-up, 23 months; range, 1 to 78 months) and two patients found to have gradients in excess of 20 mmHg satisfactorily treated with balloon dilatation. Both of these patients had coarctation of the aorta, were repaired early in our series, and neither had an extended arch anastomosis, which is now routinely used.

These results compare quite favorably with other published rates of recurrent arch obstruction.

In the CHSS study of late outcomes for 447 patients undergoing IAA repair, there were 119 patients who required a total of 158 interventions for recurrent arch obstruction. Competing risks analysis revealed that at 15 years, 32% had died

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Table 83.2 Early and Late Mortality for Single-Stage Repair of Interrupted Aortic Arch and Associated Intracardiac Defects

Group	Number	Early mortality	Late mortality	Weight in kg (mean ± SEM)	Age in days (median)
Coarctation/ventricular septal defect	19	1 (5%)	0	1.4–4.7 (3.1 ± 0.2)	1–29 (11)
Interrupted aortic arch/ventricular septal defect	18	2 (11%)	1	1.3–4.0 (3.0 ± 0.02)	2–15 (6)
Coarctation/ventricular septal defect and complex	23	4 (17%)	1	2.1–4.0 (3.1 ± 0.1)	2–24 (8)

SEM, standard error of the mean.
Source: Reprinted with permission from Sandhu SK, Beekman RH, Mosca RS, et al. Single-stage repair of aortic arch obstruction and associated intracardiac defects in the neonate. Am J Cardiol 1995;75:370.

without a first arch reintervention, 29% had undergone a first arch reintervention, and 39% remained alive without an arch reintervention. Of those patients who had undergone a first arch reintervention, at 15 years 22% had died without a second reintervention, 31% had undergone a second arch procedure, and 47% remained alive without a second arch procedure.

Recurrent Left Ventricular Outflow Tract Obstruction

Left ventricular outflow tract obstruction remains a significant source of late postoperative morbidity and mortality. From 1991 to 2001, 27 neonates underwent primary repair for IAA and an isolated malalignment-type VSD at our institution. Fifteen of these patients with the smallest subaortic areas were felt to be at risk for early or late postrepair subaortic obstruction. Consequently, these patients underwent transatrial myectomy or myotomy of the infundibular septum concomitant with the VSD closure and IAA repair, as described above. Those patients requiring myectomy/myotomy (Group I) had significantly smaller subaortic diameters (3.7 ± 0.9 mm), when compared with those who had only IAA with VSD repair (Group II, 4.5 ± 0.7 mm; $P = 0.0231$). This remained significant when indexed to BSA (0.83 ± 0.16 vs. 0.99 ± 0.13 cm $(BSA)^{0.5}$; $P = 0.012$). There was no difference in the mean aortic *z*-value between groups. There were two hospital deaths in Group I and one in Group II. No late deaths have occurred. No patient in Group II has required re-operation. Six patients required nine reoperations for left ventricular outflow tract obstruction, all of whom were in Group I and underwent myectomy/myotomy at the initial operation. Five of those patients underwent resection of a new subaortic membrane. Only one patient required myectomy for recurrent muscular subaortic obstruction. The mean interval between initial operation and first reoperation was 3.7 ± 4.1 years (range 0.5 to 9.5 years). Three patients required a second reoperation, primarily related to aortic valvar stenosis. These data reflect the continuing improvement in hospital survival (89%, 24/27) for single-stage IAA with VSD repair in the neonatal period. They also support an approach tailored to the degree of subaortic narrowing, with resection or incision of the infundibular septum at the time of primary repair for those patients felt to be at risk for residual or recurrent subaortic stenosis.

While these patients continue to be at risk for other levels of left ventricular outflow tract obstruction, this approach was very effective in preventing or prolonging the interval to recurrent muscular subaortic stenosis when compared with other published series. The late follow-up of the CHSS IAA cohort revealed that 69 patients required 100 reinterventions for recurrent left ventricular outflow tract obstruction. Competing risks analysis demonstrated that 15 years after repair, 33% had died without a first reintervention for left ventricular outflow tract obstruction, 18% had undergone a first reintervention and remained at risk, 1% had undergone a first reintervention and were no longer at risk, and 48% remained alive without a first reintervention for left ventricular outflow tract obstruction. Of those patients having a first reintervention for left ventricular outflow tract obstruction, competing risks analysis demonstrated that at 15 years following the initial reintervention, 13% had died without a second reintervention, 44% had undergone a second reintervention, and 43% remained alive without a second reintervention for left ventricular outflow tract obstruction.

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EDITOR'S COMMENTS

As noted by the authors, the results with repair of IAA in isolation or in association with other complex congenital heart disease have increasingly improved such that now the majority of infants with these conditions have satisfactory single-stage complete repair. The earlier poor results with neonatal single-stage repair led several surgeons to advocate pulmonary artery banding and arch reconstruction with artificial grafts or with the carotid artery to reestablish arch continuity. However, the late problems with the development of subaortic obstruction and the necessity of replacing these conduits during growth have led to almost universal adoption of single-stage primary repair as the treatment of choice for this condition. In rare cases, patients with interrupted aortic arch may be hemodynamically severely unstable or patients may be of very low birth weight. In such rare cases, it is possible to consider using a hybrid strategy for initial palliation with placement of bilateral pulmonary arterial bands and stenting of the ductus arteriosus. This approach may allow for growth of the ascending aorta and improvement in other organ system dysfunction prior to a complete operative repair. Dr. John Brown has reported good results with arch reconstruction with carotid artery turndown and PA bandings in a recent series. Minor variations in operative technique are used in many centers. We have elected in virtually all cases to avoid a direct circumferential primary anastomosis of the descending aorta to the ascending aorta and have used direct anastomosis over one half of the circumference of the arch in its most superior aspect with incision into the head and neck vessels and then augmentation of the undersurface of the arch with a generous patch of pulmonary homograft material. In this manner, native aorta is primarily anastomosed to allow for growth, and the arch is additionally enlarged to prevent any tension on the anastomosis. When bleeding is encountered in IAA repair with primary anastomosis, additional sutures to control bleeding may result in narrowing at the area of reconstruction; use of pulmonary homograft to augment the arch prevents this problem. With this technique, we have not found it necessary to divide an aberrant right subclavian artery in type B interruption and have left the right subclavian in place, incising the left subclavian artery at its origin as well as the left common carotid artery and sewing the carotid and subclavian together in their superior extent as part of the arch reconstruction.

The most difficult issue in repair of IAA with associated VSD is the issue of the subaortic infundibular septum, which may cause significant subaortic obstruction. As noted by the authors, it is not readily possible to identify those patients preoperatively who have significant subaortic obstruction by either echocardiography or catheterization because most flow goes across the VSD to the pulmonary arteries preoperatively. The difficulty in identifying patients with significant outflow tract obstruction may account for the fact that in the CHSS data, the narrowness of the subaortic region as determined by echocardiography did not correlate with outcome, leading the group to advocate primary repair in all circumstances. In spite of this recommendation, we have elected not to perform primary repair in some infants with very small subaortic regions, especially when the aortic valve annulus itself is severely hypoplastic.

A small number of infants have very significant hypoplasia of the aortic valve annulus and associated bicuspid aortic valve with very severe subaortic outflow tract obstruction. In these patients we have elected to perform a Yasui procedure, as described in this chapter, baffling the VSD to the pulmonary artery and reconstructing the aortic arch as in the Norwood operation with connection of the right ventricle to the pulmonary bifurcation with a homograft conduit.

The incidence of development of subaortic obstruction after complete repair of IAA with associated VSD is significant. Even with resection of some subaortic muscle, recurrent obstruction can occur. The muscle is difficult to excise in very small infants, and the resection, if carried superiorly enough, can damage the aortic valve. In addition, once the resection is performed, the superior most extent of the VSD patch is difficult to anchor because there is little tissue remaining in this location. Alternate methods of dealing with the subaortic muscle have been described. When possible, we prefer to place the VSD patch on the left ventricular side of the muscle superiorly and then bring the suture line to the right ventricular side on the inferior margin to avoid the conducting tissue. In this manner, the pressure of the left ventricle pushes the subaortic infundibular muscle away from the outflow tract during systole and seems to cause less stimulus for hypertrophy of this muscle, which can then develop into later outflow tract obstruction. If exposure is good, certainly resection is a reasonable approach. Starnes has recommended closure of the VSD via the pulmonary artery with pulling of the infundibular muscle to the right ventricle to address the subaortic outflow tract obstruction. The follow-up with all of these techniques has been relatively short, and the possibility of late development of residual outflow tract obstruction remains a concern.

The authors of this chapter have described an excellent result with myomectomy in their series of interrupted aortic arch repairs with reoperation necessary primarily for the development of subaortic membranes. We have seen a significant incidence of recurrent subaortic obstruction by membrane, but in addition, a significant number of these patients have aortic arch hypoplasia and aortic valve abnormalities and may come to a Ross–Konno operation relatively early.

Regardless of the technique for dealing with the subaortic infundibular muscle and interrupted aortic arch, a significant incidence of recurrent left ventricular outflow tract obstruction and need for reoperation has been reported in most series. Although patients with even small aortic annulus and subaortic area can successfully undergo single-stage complete repair as a neonate, it is not infrequent for these patients to have a bicuspid aortic valve and recurrent outflow tract obstruction, which requires a Ross–Konno operation as a second procedure often within the first 6 months of life. The results with these repairs, however, have continued to improve such that the overall survival with repair of interrupted aortic arch now approaches 95% in many centers.

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84 Left Ventricular Outflow Tract Obstruction and Valvar Aortic Stenosis

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VALVAR AORTIC STENOSIS

Pathology

Valvar aortic stenosis in children is most commonly congenital in etiology. The leaflets are thickened and dysplastic, with variable degrees of commissural fusion. The valve is typically bicuspid in morphology but may be tricuspid or even unicuspid. Stenosis of the right and left cusps is associated most frequently with fusion. Obstruction results from decreased leaflet mobility and a reduction in effective orifice size. Small annular size may also be present, further impeding left ventricular ejection.

Clinical Presentation and Diagnosis

Neonates with critical aortic stenosis develop symptoms of congestive heart failure and impaired systemic perfusion consequent to closure of the ductus arteriosus. Prompt diagnosis and institution of therapy are, therefore, necessary to prevent rapid deterioration and death.

Older children present less acutely, often with the finding of an asymptomatic heart murmur on routine physical examination. Feeding difficulty in infants and decreased exercise tolerance in older children may be observed as the severity of obstruction increases. Later symptoms include exertional angina, congestive heart failure, and syncope. Physical findings generally are limited to the cardiovascular examination: a harsh systolic ejection murmur at the right upper sternal border radiating to the neck, S_4 gallop, and poor upstroke of the carotid pulse. An ejection click may indicate the presence of a bicuspid valve.

Chest X-ray is usually nondiagnostic. The electrocardiogram may show left ventricular hypertrophy. Echocardiography is the principal diagnostic method for the evaluation of left ventricular outflow tract obstruction (LVOTO). Two-dimensional echocardiogram allows assessment of the LVOT and aortic valve morphology including leaflet number, degree of thickening, and mobility. In addition, left ventricular cavity size, the presence and severity of left ventricular hypertrophy, and ventricular function can be assessed. Color Doppler imaging accurately identifies the level of obstruction, allowing a distinction between valvar, subvalvar, and supravalvar stenosis. Doppler measurement of blood flow velocity across the valve provides an estimate of the peak pressure gradient. Progression of disease and timing of intervention, therefore, can be determined in a noninvasive manner in most cases.

With accurate echocardiographic assessment, cardiac catheterization is rarely required in the diagnostic evaluation of aortic stenosis. Direct simultaneous measurement of left ventricular pressure and aortic pressure is the most accurate method for assessing the outflow tract gradient. However, identification of the precise level of obstruction may not be possible. Elevation of left ventricular end-diastolic pressure indicates impaired diastolic function. Left ventriculography allows assessment of ventricular systolic function and may outline the valve leaflets, providing some assessment of morphology.

Treatment

Neonatal Critical Aortic Stenosis

Critical aortic stenosis diagnosed in the newborn period constitutes a medical emergency. A neonatal presentation indicates severe outflow obstruction that requires urgent intervention. Initial stabilization includes endotracheal intubation and inotropic support. Prostaglandin infusion will establish or maintain patency of the ductus arteriosus and improve systemic perfusion. Emergency aortic valvotomy previously was the treatment of choice, performed soon after resuscitation. In the current era, percutaneous transcatheter balloon aortic valvotomy has supplanted surgical valvotomy in most centers. Transcatheter valvotomy is performed in the catheterization laboratory, with the advantages of rapid relief of obstruction and avoidance of cardiopulmonary bypass and aortic cross-clamping. Regardless of the technique, neonatal aortic valvotomy is considered a palliative procedure for most patients, with 41% undergoing reintervention within 10 years.

In a small subset of neonates with critical aortic stenosis and small left ventricular size, it may be difficult to determine whether a biventricular approach with aortic valvotomy/replacement or a single-ventricle approach with the Norwood procedure is more appropriate. Several studies have attempted to identify preoperative predictors for the suitability of single ventricle versus biventricular repair in this challenging population. However, none of these studies have established a universally adopted set of criteria, and patient triage to a single versus biventricular pathway is the subject of a current multi-institutional study sponsored by the Congenital Heart Surgeons Society.

Neonates with Multilevel Left Ventricular Outflow Tract Obstruction

Neonates with multiple levels of LVOTO are a complex group. Surgical options must be tailored to the particular anatomic considerations (Fig. 84.1). Neonates undergoing single-ventricle palliation can be managed with a staged approach, consisting of a Norwood or modified Norwood operation. Hybrid palliation, with implantation of a ductal stent and concomitant bilateral pulmonary arterial banding, is another recently developed option. This strategy, however, is currently reserved for marginal candidates with contraindications to standard Norwood-type palliation, or as a bridge to heart transplantation, with few centers using Hybrid palliation as the preferred approach. Heart transplantation is an attractive strategy for infants with severe atrioventricular valve regurgitation,

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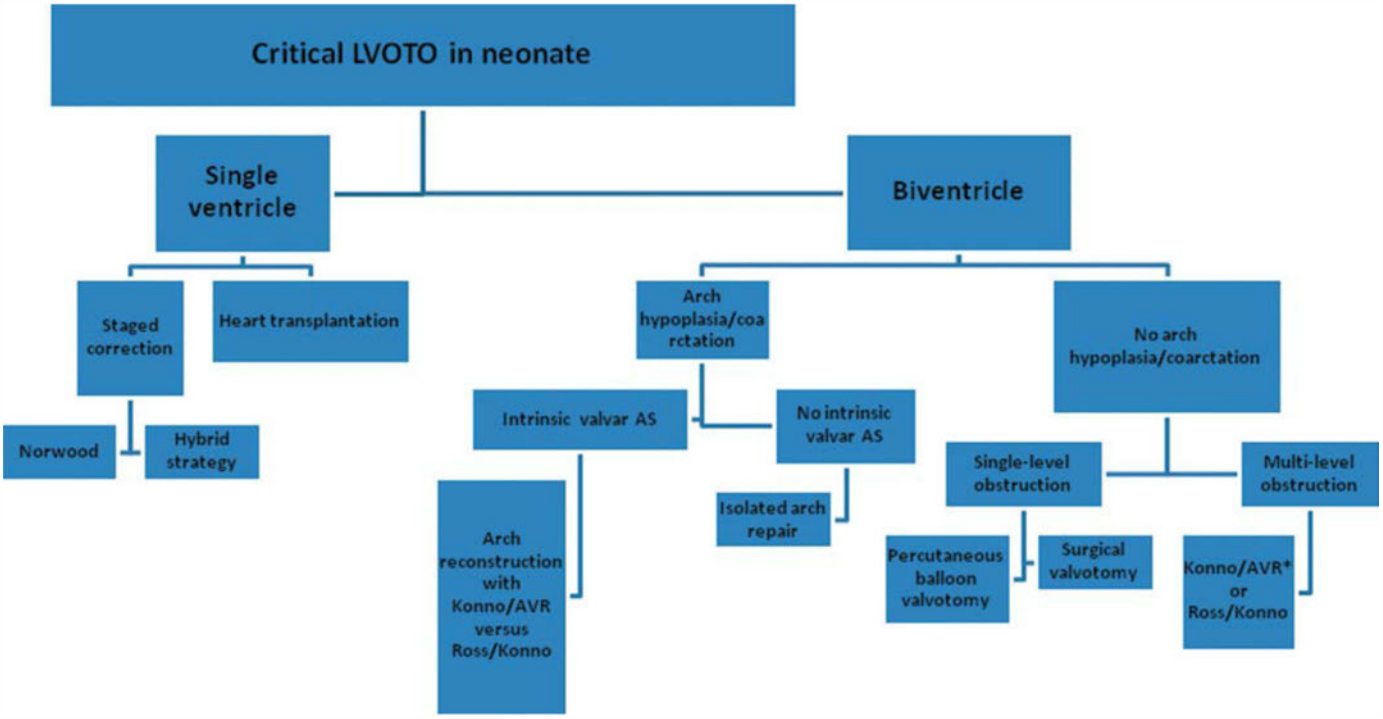


Fig. 84.1. Treatment algorithm for neonates and infants with critical left ventricular outflow tract obstruction.

visceral heterotaxy, or poor systemic ventricular function.

Neonates undergoing biventricular repair have several potential options depending upon the associated intracardiac and arch pathology. Infants with combined valvar aortic stenosis and aortic arch obstruction can be managed with standard patch aortoplasty concomitant with either a neonatal Ross operation or a mechanical aortic valve replacement with an annular enlargement procedure (Konno aortoventriculoplasty). Our institutional preference has been to manage these patients with aortic valve replacement and Konno, as results with the neonatal Ross operation have been poor, especially in neonates with associated mitral valve abnormalities. The presence of a ventricular septal defect adds an additional level of complexity, with options including those discussed above, arch repair with closure of the ventricular septal defect, or Yasui reconstruction.

Valvar Aortic Stenosis in Older Children

The objectives of surgical treatment of aortic stenosis are relief of symptoms and reduction of the risk of sudden death. Sudden death is related directly to the severity of obstruction and correlates with the peak systolic gradient, although it is reported to be <1%. Stenosis is considered severe when the mean echocardiographic gradient is ≥40 mmHg, moderate when it is between 25 and 40 mmHg, and mild when it is <25 mmHg. Surgery is indicated in any symptomatic patient regardless of severity and in asymptomatic patients with severe stenosis. Although surgery is not recommended for asymptomatic patients with mild stenosis, there is controversy about the management of asymptomatic patients with moderate stenosis. Surgical management should be considered on an individual basis. The authors generally recommend surgery in patients who demonstrate reduced left ventricular function, a strain pattern on electrocardiography, or abnormal exercise testing.

The two options for surgical management of aortic stenosis are valvotomy and aortic valve replacement. Valve replacement in children poses unique challenges that are not seen in the adult population. The smallest mechanical and bioprosthetic valves available (15 to 19 mm) may be too large for the annular size of smaller children. Even when replacement is feasible, unless an "adult-sized" valve is implanted, somatic growth eventually results in recurrent outflow obstruction due to patient–prosthesis mismatch. In addition, re-replacement with an appropriately sized valve may be limited by restricted annular growth from the original prosthesis. These limitations may necessitate concomitant performance of an annulus-enlarging procedure such as a Konno aortoventriculoplasty. Posterior enlargement procedures such as the Nicks or Manougian procedures are often insufficient in children. These options are discussed in detail in later sections.

The choices of prosthetic valves for children are more limited than are those for adults. Bioprostheses do not require anticoagulation, but calcific degeneration occurs at an accelerated rate compared with adults, limiting their use in children. Mechanical prostheses maintain their structural integrity and generally have excellent freedom from reintervention or re-replacement, but require anticoagulation with warfarin. Anticoagulation can be achieved safely in children, but issues of compliance, inconsistent diet, and potential for injury increase the risk of bleeding and thromboembolic complications in this patient population. A recent paper by Alsoufi and colleagues demonstrated that the use of either homograft or bioprosthetic valves in children was associated with an increased risk of valve-related reoperation (82% at 15 years) compared with both mechanical valve implantation and the Ross operation. However, long-term survival was excellent (86% at 15 years) in patients having homograft or bioprosthetic valve replacement, underscoring that these valve types can be useful in female patients or those in whom anticoagulation cannot be administered.

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Allograft replacement of the aortic root became popular in the early 1990s. Small valves with excellent early hemodynamic performance were readily available, and it was hoped that degeneration would not occur. Subsequent reports, however, demonstrated severe accelerated degeneration that required early reoperation in younger children. Severe calcification and inflammatory reaction of surrounding tissues complicate allograft removal and place adjacent structures, including the mitral valve and the bundle of His, at risk during reoperation.

Pulmonary autografting and reconstruction of the right ventricular outflow tract with an allograft (the Ross procedure) are often the best choice for aortic valve replacement in small children. The autograft has growth potential, does not develop calcific degeneration, and does not require anticoagulation. There are, however, a number of important limitations. The aortic and pulmonary annulus sizes must be similar; stretching of the autograft to fit a dilated aortic root will result in neoaortic insufficiency. The presence of a bicuspid aortic valve may increase the propensity for autograft failure given the associated aortopathies in this population. The operation is technically challenging, requiring longer periods of cardiopulmonary bypass and aortic cross-clamping compared with other techniques for valve replacement. Allograft reconstruction of the right ventricular outflow tract necessitates eventual reintervention (either surgical or transcatheter) to relieve conduit stenosis from somatic growth and calcific degeneration. Finally, it has been recognized that the pulmonary autograft develops important dilation over time out of proportion to somatic growth, and that this dilation eventuates in progressive neoaortic insufficiency.

In summary, there is no ideal substitute for aortic valve replacement in children with valvar aortic stenosis. Aortic valvotomy is, whenever feasible, the procedure of choice in this population. Although often not definitive, adequate palliation is usually achieved. Small residual gradients and mild aortic insufficiency are acceptable. Valvotomy can defer the need for valve replacement significantly, allowing insertion of a larger prosthesis at a later time and reducing the total number of re-replacements required over a patient's lifetime.

Surgical Procedures

Aortic Valvotomy

Surgical aortic valvotomy is usually performed via a median sternotomy. A single right atrial cannula provides venous drainage, and the aortic cannula is placed as distally as possible. Some surgeons advocate closed valvotomy that is performed by passing dilators of increasing size (usually up to 1 mm larger than the preoperative echocardiographic aortic annulus diameter) through the aortic valve; the dilators are introduced through a purse string in the left ventricular apex. This technique can be performed through a left thoracotomy without the need for cardiopulmonary bypass. The authors believe that open valvotomy with direct visualization of the aortic valve allows more precise leaflet separation, resulting in better relief of gradient and a decreased risk of significant aortic insufficiency.

The aorta is clamped, and cardioplegia is delivered via the aortic root. A transverse aortotomy is made just above the sinotubular junction, with care taken to avoid injury to the right coronary artery and the aortic valve leaflets. The valve is inspected carefully. Fused commissures are opened precisely with a scalpel (Fig. 84.2). The incisions are not extended to the aortic wall, since this may result in loss of support and aortic insufficiency (Fig. 84.3). Excessive fibrous tissue, if present, is excised from the leaflets. Fibrous attachments between the base of the leaflets and the aortic wall are divided to maximize leaflet mobility. Despite these steps, the effective orifice may be inadequate, particularly in smaller bicuspid valves. Ilbawi and colleagues described a technique of extended valvotomy for these patients. Circumferential incisions are made above the true commissures and raphes. They reported a low incidence of aortic insufficiency and significantly lower gradients compared with standard valvotomy. The aortotomy is closed precisely to avoid supravalvar narrowing, and the cross-clamp is removed. The patient is weaned from cardiopulmonary bypass with mild inotropic support (dopamine 5 μg/kg/min and milrinone 0.5 μg/kg/min). The adequacy of repair is assessed by transesophageal echocardiography and direct measurement of pressure in the left ventricle and ascending aorta. Modified ultrafiltration is performed before decannulation.

Early mortality in older infants and children undergoing aortic valvotomy is reported currently to be <2%, and late mortality is rare. Reintervention for progressive regurgitation or restenosis may be required, but this generally occurs later than in neonates. The reoperation rate in children over 1 year of age at the time of the initial valvotomy is 2% at 10 years but then increases 3.3% per year.

Aortic Valve Replacement

Aortic valve replacement is required when valvotomy is not sufficient to reduce the transvalvar gradient adequately, or in patients with greater than mild aortic insufficiency. In larger children with adequate annular size, simple replacement of the valve is performed as it is in adults. Prosthesis selection should be individualized to the patient's lifestyle and activity level. The authors generally avoid porcine and bovine bioprostheses as well as

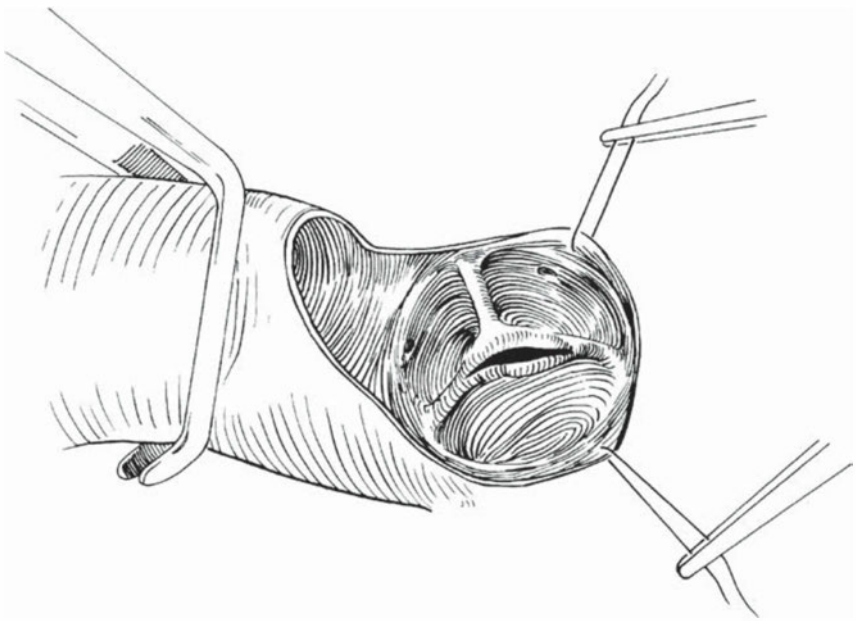


Fig. 84.2. Inspection of a stenotic aortic valve approached via an aortotomy.

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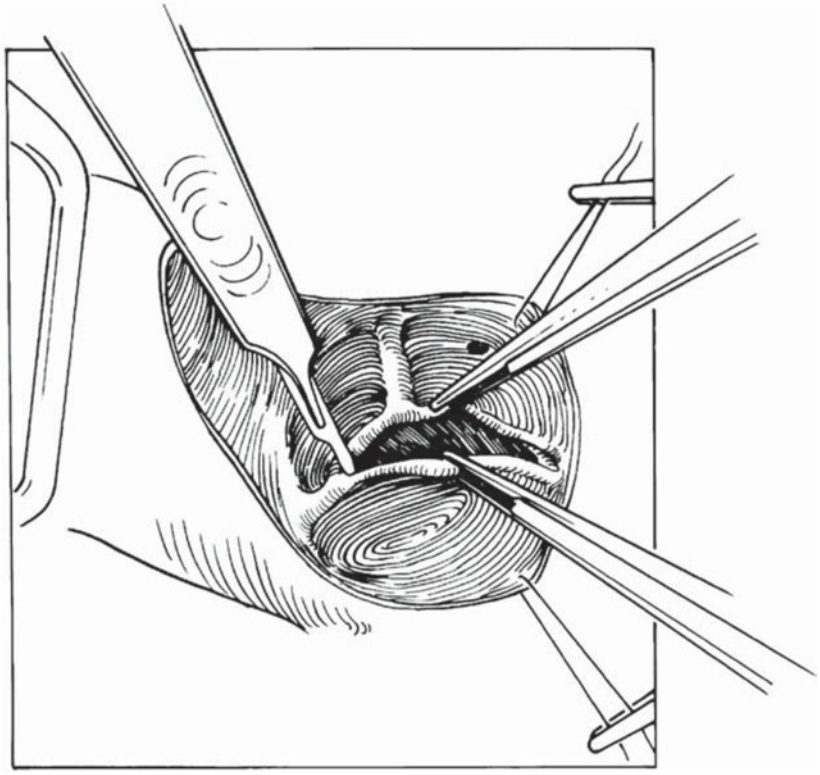


Fig. 84.3. A valvotomy is performed by incising the fused commissures. Care is taken to avoid incisions in false raphes, which may result in valve insufficiency.

allograft implantation because of the rapid degeneration and early failure observed in children. The remaining options, therefore, are limited to mechanical prosthesis and pulmonary autograft.

Aortic valve replacement with a mechanical prosthesis is performed via a median sternotomy. A single right atrial or two-stage venous cannula provides venous drainage, and the ascending aorta is cannulated distally. The aorta is clamped, and in the absence of significant aortic insufficiency, cardioplegia is infused in the aortic root. After aortotomy, additional cardioplegia is delivered every 30 minutes through a retrograde coronary sinus catheter or by direct coronary ostial perfusion. A left ventricular vent is placed through the left atrial appendage or the right superior pulmonary vein.

A transverse aortotomy is made and is extended into the noncoronary sinus of Valsalva. The aortic valve leaflets are excised. Interrupted pledgeted mattress sutures are placed circumferentially around the annulus. In smaller patients, intra-annular placement may be preferable to avoid coronary ostial obstruction by the sewing ring. The sutures are passed through the sewing ring, and the valve is parachuted into place. After the sutures are tied, the prosthetic leaflets are assessed carefully to ensure unhindered mobility. The aortotomy is closed, and the aortic clamp is removed. The patient is weaned from cardiopulmonary bypass with mild inotropic support. Transesophageal echocardiography is used to assess prosthetic function. Modified ultrafiltration is performed before decannulation.

Hospital mortality after mechanical aortic valve replacement in children is 0% to 5%. Early complications include permanent heart block in 3% and acute endocarditis in 2% of these patients. Late complications relate primarily to anticoagulation. In reports of long-term follow-up, valve thrombosis occurs in 0% to 2% of patients. This sometimes can be managed pharmacologically with thrombolytic agents but frequently requires urgent surgical thrombectomy or valve replacement. Embolic events are reported in 2%, and significant bleeding episodes occur at a rate of 0.15% per patient year. Freedom from reintervention or re-replacement is approximately 86% at 20 years but is increased in younger patients having smaller prostheses.

Ross Procedure

Ross reported aortic valve replacement with a pulmonary autograft and allograft reconstruction of the right ventricular outflow tract in 1967. In the absence of significant size discrepancy or connective tissue disease, the Ross procedure is the preferred technique for aortic valve replacement in small children. In larger children, it is frequently preferred over mechanical prosthesis to avoid the need for anticoagulation.

The approach is via a median sternotomy. The venae cavae are cannulated individually, and the ascending aorta is cannulated distally. The aorta is clamped, and in the absence of significant aortic insufficiency, cardioplegia is infused in the aortic root. Cardioplegia may also be delivered retrograde through a coronary sinus catheter and by direct coronary ostial perfusion after an aortotomy. Additional cardioplegia is delivered every 30 minutes during the cross-clamp period. A left ventricular vent is placed through the left atrial appendage or the right superior pulmonary vein.

The aorta and the pulmonary trunk are separated, and the pulmonary trunk is opened transversely just proximal to the bifurcation. The pulmonary valve is inspected to identify any pathology that would preclude its use as an aortic valve replacement. Transection of the pulmonary trunk is then completed. A right-angle clamp is passed carefully through the leaflets into the right ventricular outflow tract, and a site for proximal transection is identified in the infundibular free wall approximately 5 mm below the level of the valve. A transverse infundibular incision is made and carried to the infundibular septum at each end. The infundibular septum is scored with a scalpel blade. A plane can be developed between the subconal muscle and the underlying interventricular septum. Dissection in this plane completes the harvest, and the autograft is stored in normal saline solution before implantation. The resulting posterior raw surface is cauterized. In their practice, the authors also apply a thin layer of biological sealant to ensure hemostasis. In dissecting the autograft, care must be taken at the leftward extent of the septal dissection to avoid injury to the first septal perforating branch of the left anterior descending coronary artery. The pulmonary artery is sized, and an appropriate allograft is thawed and prepared for reconstruction of the right ventricular outflow tract.

The aorta is transected just above the sinotubular junction. The aortic valve leaflets are excised, and the left and right coronary arteries are excised with a generous button of aortic wall. The autograft is oriented by alignment of the commissures.

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A proximal anastomosis is then constructed. Interrupted simple braided sutures are used for smaller patients, although a continuous technique with polypropylene sutures can be used in larger children and teenagers. The coronary buttons are anastomosed to incisions in the respective autograft sinuses of Valsalva. The distal aortic anastomosis is constructed with a running polypropylene suture. Air is evacuated from the left side of the heart, and the aortic clamp is removed. Continuity between the right ventricle and the pulmonary artery is restored by interposing the previously thawed allograft in the reperfused, beating heart. The patient is weaned from cardiopulmonary bypass with mild inotropic support. Transesophageal echocardiography is used to assess autograft and allograft function as well as left ventricular wall motion. Modified ultrafiltration is performed before decannulation in children with an operative weight below 20 kg.

The results of the Ross procedure are reasonable in carefully selected pediatric patients. Early mortality is 0% to 6%, occurring primarily in infants under 5 months of age. However, patients with concomitant mitral valve disease or aortic arch hypoplasia, even when judged to have adequate biventricular physiology, fare poorly with the Ross operation. A recent paper by Shinkawa and colleagues from the University of Michigan reported a 36% early mortality in this subgroup. Similarly, Hickey and colleagues reported a 31% early mortality rate in infants younger than 3 months, and an actuarial 1-year survival <50% for neonates. Complications occur infrequently and include bleeding, arrhythmia, heart block, and stroke. Actuarial survival for all patients is 84% at 1 year and 77% at 5 years. Aortic root dilatation is common late after the Ross procedure in children and warrants careful echocardiographic follow-up.

Konno Aortoventriculoplasty

Annular enlargement is required in children with small aortic annular size requiring aortic valve replacement. Nicks and colleagues and Manougian and Seybold-Epting described techniques for posterior annular enlargement that have been used successfully in adults. However, the resulting increase in annular size is generally inadequate to allow insertion of even a small prosthetic valve in small children. Konno and coworkers described a technique of anterior enlargement that more effectively increases annular size and relieves coexistent subvalvar stenosis.

The approach is via a median sternotomy. The venae cavae are cannulated individually, and the ascending aorta is cannulated distally. The aorta is clamped, and in the absence of significant aortic insufficiency, cardioplegia is infused in the aortic root. Additional cardioplegia is delivered every 30 minutes during the cross-clamp period. A left ventricular vent is placed through the left atrial appendage or the right superior pulmonary vein.

A vertical aortotomy is made and is carried onto the right ventricular outflow tract well to the left of the origin of the right coronary artery. Care is taken to avoid injury to the pulmonary valve. The aortic valve leaflets are excised, allowing visualization of the left and right ventricular aspects of the infundibular septum. An incision is made across the aortic annulus into the infundibular septum. Injury to the conduction tissue is avoided by placing this incision to the left of Lancisi muscle. A diamond-shaped patch of Dacron is fashioned, and the inferior portion is sutured to the edges of the septal incision with interrupted pledgeted mattress sutures. An appropriately sized prosthetic valve is then inserted as was described for aortic valve replacement. Anteriorly, the valve sutures are passed through the prosthetic patch. The superior portion of the patch is used to close the ascending aorta. The right ventricular free wall is enlarged with a patch of bovine pericardium. Air is evacuated from the left side of the heart, and the aortic clamp is removed. The patient is weaned from cardiopulmonary bypass with mild inotropic support. Transesophageal echocardiography is used to assess prosthetic function, patch leaks, and left ventricular wall motion.

A modification of this technique can be used for annular enlargement in conjunction with the Ross procedure. A Ross–Konno is an ideal procedure in infants or children under 2 years of age, whose size precludes implantation of at least a 21-mm prosthesis or larger. The pulmonary autograft is harvested with a triangular portion of right ventricular free wall that is used as the septal patch.

In light of the complex form of LVOTO seen in pediatric patients who require the Konno or the Ross–Konno procedure, early and late results are quite good. On average, the annulus is enlarged to twice the original size, often allowing insertion of an adult-sized prosthesis (23 or 25 mm) in most patients over 3 years of age. Operative mortality for the Konno procedure with prosthetic valve replacement is 5% to 15%. Ten-year actuarial survival is 92%. Ten-year freedom from reoperation with a mechanical prosthesis is 80% to 89%. The linearized rate of reoperation is approximately 3.9% per year and is increased among patients undergoing a Konno to correct aortic valve pathology in conjunction with annular hypoplasia. An important and often underappreciated complication of the Konno procedure (with or without concomitant Ross operation) is pulmonary regurgitation, which occurs at a cumulative incidence of 10% at 16 years postoperatively. Operative mortality for the Ross–Konno procedure is 0% to 7% even in children under 1 year of age. Postoperative complications include bleeding, arrhythmia, heart block, and left ventricular dysfunction. No permanent effects on ventricular function are present at long-term follow-up.

SUBVALVAR AORTIC STENOSIS

Subaortic stenosis in children results from either a discrete fibrous membrane or, less commonly, diffuse, fibromuscular tunnel-like stenosis (Fig. 84.4). The discrete subaortic membrane is probably an acquired lesion that is rarely seen in infants, albeit with "anatomic precursors." Often, a ring of fibrous tissue is present that is adherent to the septum anteriorly, extending posteriorly to the right and left fibrous trigones and to the anterior mitral leaflet. Associated subaortic anomalies are present in 31% of these patients, including anomalous

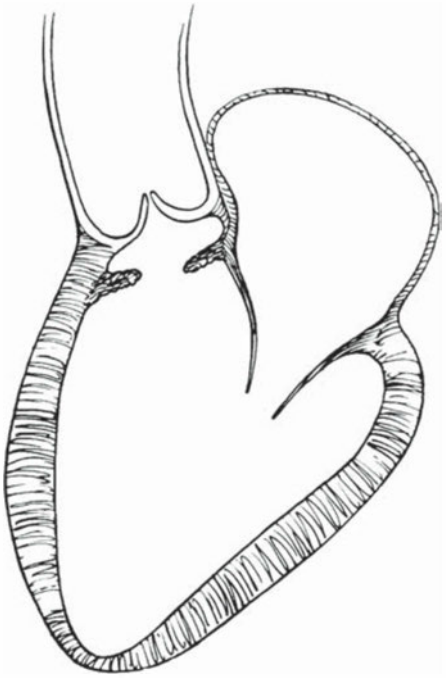


Fig. 84.4. Sagittal section of the heart with a discrete subaortic membranous obstruction.

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septal insertion of the mitral valve, accessory mitral valve tissue, anomalous papillary muscles, and anomalous muscular bands. In addition to outflow obstruction, aortic insufficiency develops in more than 50% of these patients from turbulence-induced leaflet deformity or direct attachment of the membrane to the aortic leaflets.

Tunnel-like subaortic stenosis is a congenital lesion in which the subaortic region is diffusely hypoplastic. Thickened septal myocardium and endocardial fibrosis contribute to outflow tract obstruction.

Clinical Presentation and Diagnosis

Patients with discrete subaortic stenosis usually present in childhood with an asymptomatic murmur detected on physical examination. Chest pain, syncope, and congestive heart failure may develop with worsening obstruction. A systolic ejection murmur without an ejection click is heard on auscultation. A diastolic decrescendo murmur may be heard at the left lower sternal border when a discrete subaortic membrane causes aortic insufficiency. Tunnel-like fibromuscular stenosis often presents in infancy but is otherwise indistinguishable from a discrete membrane on the basis of signs and symptoms. The evaluation is similar to that elucidated for valvar aortic stenosis, with echocardiography being the main diagnostic test. Chest X-ray is usually nondiagnostic, and electrocardiography shows left ventricular hypertrophy.

Treatment

Discrete Fibrous Subaortic Stenosis

Surgical excision of a discrete fibrous subaortic membrane is indicated for relief of symptoms, for severe obstruction (mean gradient >30 mmHg), and when moderate-to-severe aortic insufficiency is present. Asymptomatic patients with mean gradients <30 mmHg and without significant aortic insufficiency do not require surgery but should be followed with serial echocardiograms to detect progression. There is controversy about asymptomatic patients with moderate obstruction and mild aortic insufficiency. Some centers advocate prophylactic excision in all cases, citing the low risk of surgery and the benefit of preventing aortic insufficiency. However, clinically important progression is not universal, and factors determining at-risk substrates are debated. Furthermore, the efficacy of early surgery is controversial because of the variable outcomes in patients undergoing early surgical intervention coupled with a high postoperative prevalence of both recurrent stenosis and aortic regurgitation even after successful relief of subaortic obstruction. Patients with a mean echocardiographic gradient >30 mmHg, close proximity of the membrane to the aortic valve, and membrane extension to the mitral valve are at higher risk for progressive obstruction and should undergo surgical repair. Older age and a gradient above 30 mmHg are independent risk factors for progressive aortic insufficiency and are indications for repair.

The surgical approach is via a median sternotomy. A single right atrial or a two-stage venous cannula provides venous drainage, and the ascending aorta is cannulated distally. The aorta is clamped, and in the absence of significant aortic insufficiency, cardioplegia is infused in the aortic root. A transverse aortotomy is made and is extended into the noncoronary sinus of Valsalva. Edwin and colleagues recently reported a novel hexagonal six-point traction technique to provide improved exposure of the subaortic region. This method involves the placement of three commissural traction sutures along with three additional traction sutures placed at the nadir of each aortic valve leaflet. The nadir sutures not only elevate the subaortic region and improve visualization but also keep the aortic valve leaflets retracted against the aortic wall during the resection. The aortic valve leaflets are retracted gently, allowing visualization of the membrane in the left ventricular outflow tract.

The membrane is grasped anteriorly with a skin hook or a traction suture. A radial incision is made into the membrane, initiated just to the left side of an imaginary line descending from the right coronary ostium, extending just into the underlying septal myocardium. The membrane is then excised circumferentially in a counterclockwise direction by blunt dissection (Fig. 84.5). Care is taken to avoid injury to the anterior leaflet of the mitral valve and the aortic valve leaflets (Figs. 84.6 and 84.7).

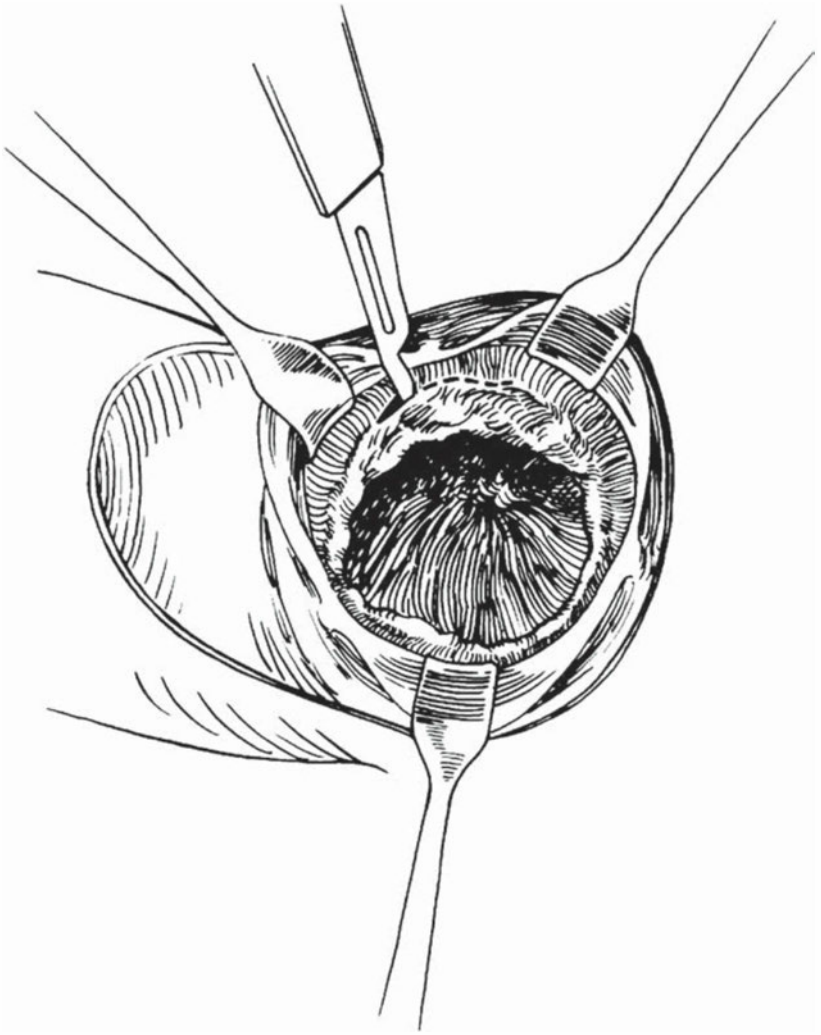


Fig. 84.5. The endocardium of the membrane is incised.

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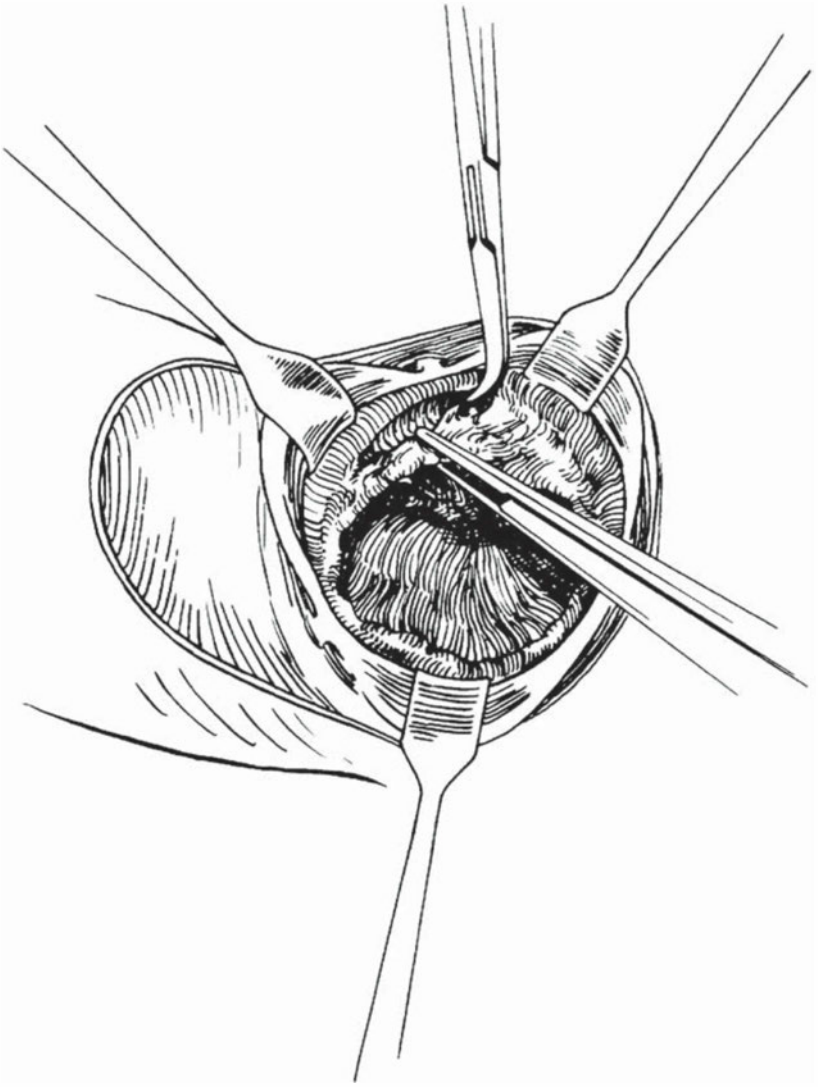


Fig. 84.6. Blunt dissection is initiated. The membrane is enucleated, not excised.

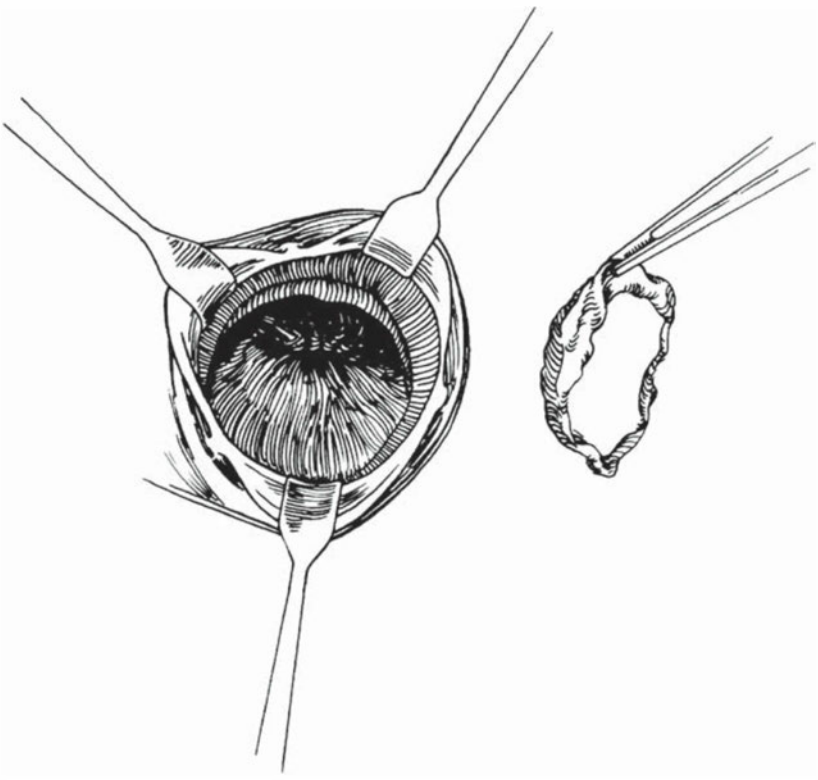


Fig. 84.7. The enucleated membrane is sometimes removed as a complete, circumferential ring.

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Resection of additional septal muscle is performed to ensure complete excision of the membrane. Yacoub and colleagues advocate the additional resection of fibrous tissue in the left and right fibrous trigones.

The aortotomy is closed, and the aortic clamp is removed. The patient is weaned from cardiopulmonary bypass. Transesophageal echocardiography is used to assess relief of obstruction and aortic insufficiency. After resection of discrete subaortic membrane, mortality approaches zero. Permanent heart block and iatrogenic ventricular septal defect occur in fewer than 5% of these patients. Reoperation for recurrent obstruction is required in 15% to 20% of patients at 5 years. Multiple studies have suggested that the risk of recurrent obstruction is increased in patients with higher LVOT gradients following initial resection, thus advocating for aggressive pursuit of any residual LVOT gradient documented by intraoperative echocardiographic or direct pressure measurements. Aortic insufficiency progresses in 25% of patients at 10-year follow-up and is more likely in patients with a higher mean echocardiographic gradient and in patients whose preoperative mean echocardiographic gradient exceeded 30 mmHg.

The indications for excision of recurrent discrete subaortic stenosis are the same as those for the initial operation. Re-resection with more aggressive septal myectomy is the authors' procedure of choice. In the case of a second recurrence, a modified Konno procedure is performed as described below for the treatment of tunnel-like stenosis.

Diffuse Fibromuscular Subaortic Stenosis

Indications for surgery in patients with tunnel-like subaortic stenosis are the same as those for patients with a discrete subaortic membrane, but relief of the obstruction is technically more challenging. Furthermore, the results following surgical treatment are less favorable in this subgroup of patients compared with those obtained following resection of discrete subvalvar stenosis. Resection of fibromuscular tissue from the septum may be possible, but this is often inadequate because of the long circumferential narrowing of the outflow tract. Conal enlargement with a modified Konno procedure is the preferred approach in these challenging patients.

The modified Konno procedure is performed via a median sternotomy incision. The venae cavae are cannulated individually, and the ascending aorta is cannulated distally. The aorta is clamped, and cardioplegia is infused in the aortic root. A transverse aortotomy is made just above the sinotubular junction. The subaortic area is inspected through the retracted aortic valve leaflets. A transverse incision is made in the right ventricular infundibulum, allowing visualization of the infundibular septum. A right-angle clamp is passed through the aortic annulus, and a safe site for septal incision is identified. A full-thickness longitudinal septal incision is made through the right ventriculotomy, and subaortic fibromuscular tissue is resected. The resulting ventricular septal defect is closed with a Dacron or Gore-Tex patch. The right ventriculotomy is closed with a bovine pericardial patch, and the patient is weaned from cardiopulmonary bypass in the usual manner. Early mortality was 0% in several large series of patients who underwent a modified Konno procedure. Excellent relief of obstruction is obtained, with no permanent heart block and no hemodynamically significant residual ventricular septal defects. More long-term follow-up is required, but freedom from reoperation at 3 years has been found to be 95% to 100%.

Placement of a left ventricular apicoaortic valved conduit has been described for patients with severe outflow tract obstruction considered not amenable to conventional repair. The authors believe that nearly all patients can be treated more definitively with resection and a modified Konno procedure.

SUPRAVALVAR AORTIC STENOSIS

Supravalvar aortic stenosis is the least common form of LVOTO. The underlying biochemical mechanism is abnormal elastin synthesis that is related to mutations in the chromosome region 7q11.23. This may occur in association with Williams syndrome, as an autosomal-dominant familial trait, or as a sporadic finding.

The pathologic lesion in supravalvar stenosis is a thickening of the aortic wall that results from collagen deposition and smooth muscle hypertrophy in the media as well as hyperplasia and fibrosis of the intima. The narrowing is localized to the sinotubular junction in 85% of cases, with either an hourglass or a discrete membranous morphology. Diffuse hypoplasia is present in 15% of cases and involves the entire ascending aorta, occasionally extending to the transverse arch and the descending aorta.

Although the site of stenosis may be localized, it is important to recognize that the pathologic process involves the entire aortic root, may coexist with important valvar aortic stenosis, and can also include peripheral pulmonary artery stenoses. Coronary ostial stenosis, most commonly involving the left coronary artery, occurs in 25% of these patients.

Clinical Presentation and Diagnosis

Patients with supravalvar aortic stenosis typically present in the first year of life with feeding or exercise intolerance, angina, or syncope, although cardiovascular findings are present in less than one-third of these newborns. The presence of characteristics ascribable to Williams' syndrome should increase the index of suspicion. About 10% of patients are asymptomatic.

As with other causes of LVOTO, chest X-ray findings are nonspecific. The electrocardiogram shows left ventricular hypertrophy. ST-segment and T-wave abnormalities consistent with ischemia may be seen in patients with coronary artery involvement. Two-dimensional echocardiography is the usual method of diagnosis in patients with supravalvar aortic stenosis. Decreased aortic caliber, increased wall thickness, and abnormal leaflet mobility are identified. Localized stenosis can be distinguished from the more diffuse pattern, and the extent of distal involvement can be assessed. Coronary ostial involvement as well as concomitant subaortic stenosis can also be detected. Cardiac catheterization is performed routinely to assess patients with supravalvar stenosis when surgical repair is being considered. It is the only method for accurate measurement of the peak systolic gradient. Aortography distinguishes the localized type from diffuse hypoplasia and accurately defines the distal extent of narrowing. Coronary ostial stenosis can also be detected and characterized.

Treatment

Surgical repair of supravalvar aortic stenosis is indicated in symptomatic patients as well as in asymptomatic patients with a peak catheter-measured systolic gradient >50 mmHg. The goals of repair are to relieve outflow obstruction and restore normal geometry to the aortic valve and sinuses of Valsalva. Expectant management of asymptomatic patients with lower LVOT gradients is acceptable since lesion regression has been documented in an important proportion of these patients coupled with a similar overall survival in patients treated with and without operative intervention.

The approach is via a median sternotomy. A single right atrial cannula provides venous drainage, and the aortic cannula is placed as distally as possible. The aorta is clamped, and cardioplegia is delivered via the aortic root. Surgical repair

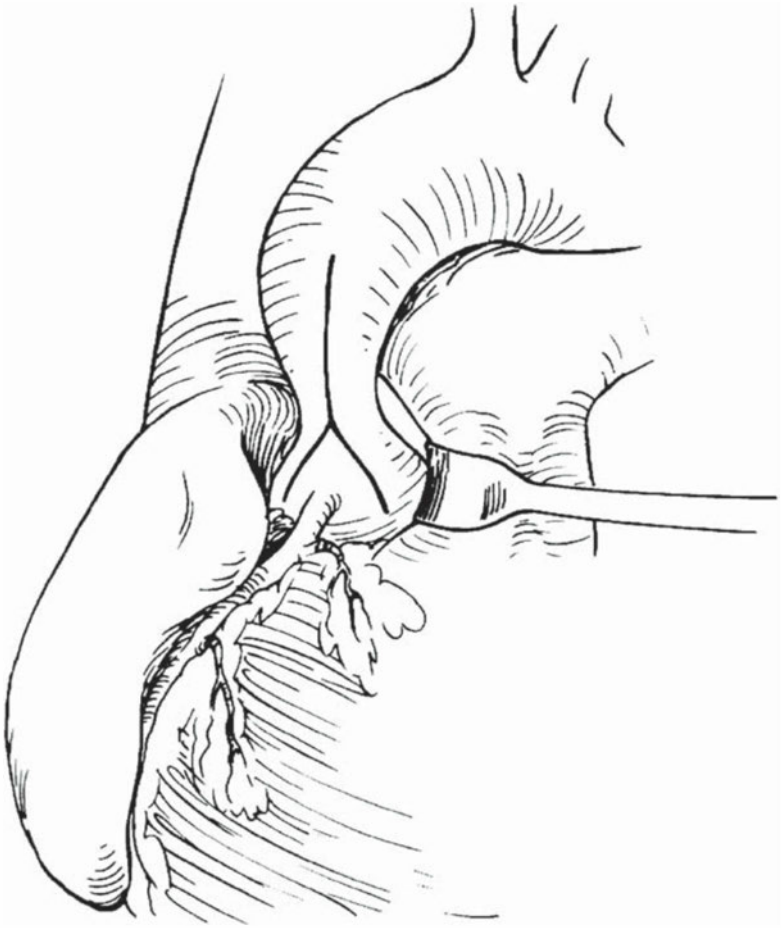


Fig. 84.8. Supravalvar aortic stenosis repair as described by Doty begins with an inverted Y-shaped incision.

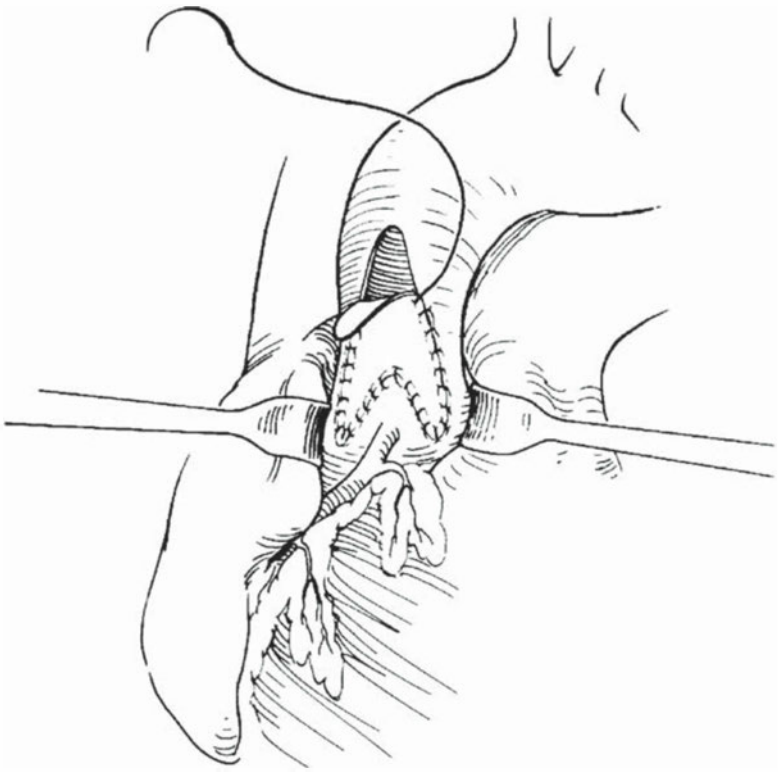


Fig. 84.9. The aorta is enlarged with a bifurcated patch.

techniques have evolved considerably since the first description by McGoonin 1961, mainly in an effort to obviate distortion of the aortic root and sinuses and restore a more normal geometric configuration. McGoon and colleagues first performed a longitudinal aortotomy, beginning above the area of narrowing and extending into the noncoronary sinus of Valsalva. The stenotic ridge is excised, and a diamond-shaped patch of bovine pericardium, Dacron, or Gore-Tex is used to augment the ascending aorta. With the recognition that distortion of the aortic valve and sinuses of Valsalva may contribute to outflow obstruction, techniques were developed to maintain greater symmetry of the sinuses. Doty and associates described an inverted Y-incision into the noncoronary and right coronary sinuses of Valsalva (Fig. 84.8). The incision in the right sinus is placed well to the left of the origin of the right coronary artery. A pantaloon-shaped patch enlarges both the ascending aorta and the sinuses (Fig. 84.9). Brom advocated augmentation of all three sinuses. The aorta is transected at the area of narrowing. Incisions are made into each sinus of Valsalva. The sinuses are augmented with triangular patches, and the augmented root is reanastomosed to the ascending aorta. Myers and Waldhausen and their associates described an innovative technique to achieve three-sinus augmentation using autologous tissue, termed the slide aortoplasty (Figs. 84.10–84.12). The aorta is transected at the area of narrowing, and incisions are made in the sinuses as with Brom technique. Three incisions are made in the ascending aorta opposite the aortic valve commissures, and the aorta is reapproximated. Variations of this technique include excision of the stenotic segment and the use of an autologous ring of pulmonary artery to enlarge the sinuses.

Regardless of the technique, the coronary ostia should be inspected carefully to identify any obstruction that may be present. When ostial wall thickening is present, a Brom repair is used, with the extension of the left coronary sinus incision into the left main coronary artery. Patch repair of the sinus is continued onto the coronary artery, with resultant coronary osteoplasty. Coronary obstruction from leaflet adherence to the stenotic ridge is relieved by careful separation of the fused leaflet and resection of residual tissue around the ostium. Diffuse narrowing of the left main coronary artery requires coronary artery bypass. A saphenous vein graft may be preferable since progressive arteriopathy may involve the subclavian artery and limit internal mammary artery flow.

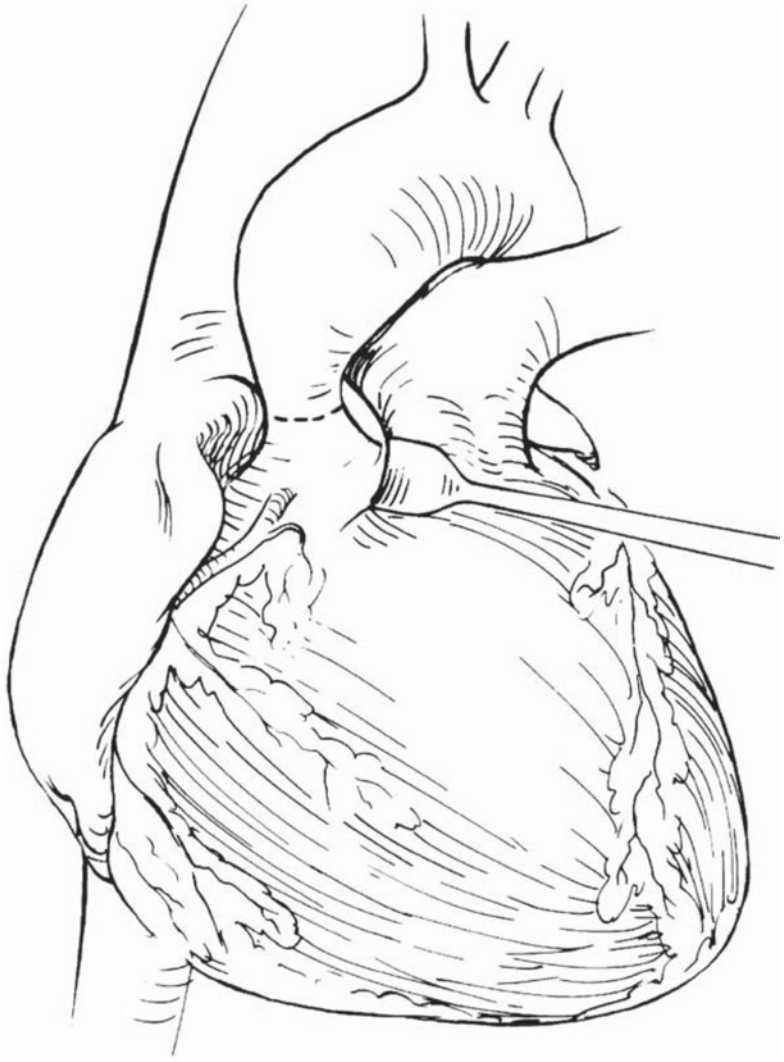


Fig. 84.10. Supravalvar aortic stenosis repair as described by Myers starts with transection of the aorta at its narrowest point.

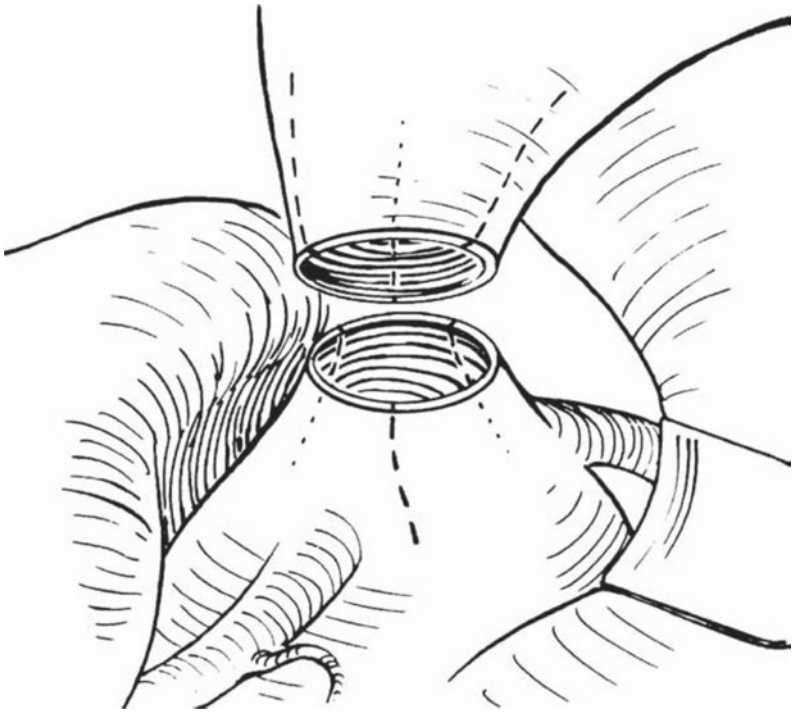


Fig. 84.11. Three vertical incisions are made in the proximal and distal aorta.

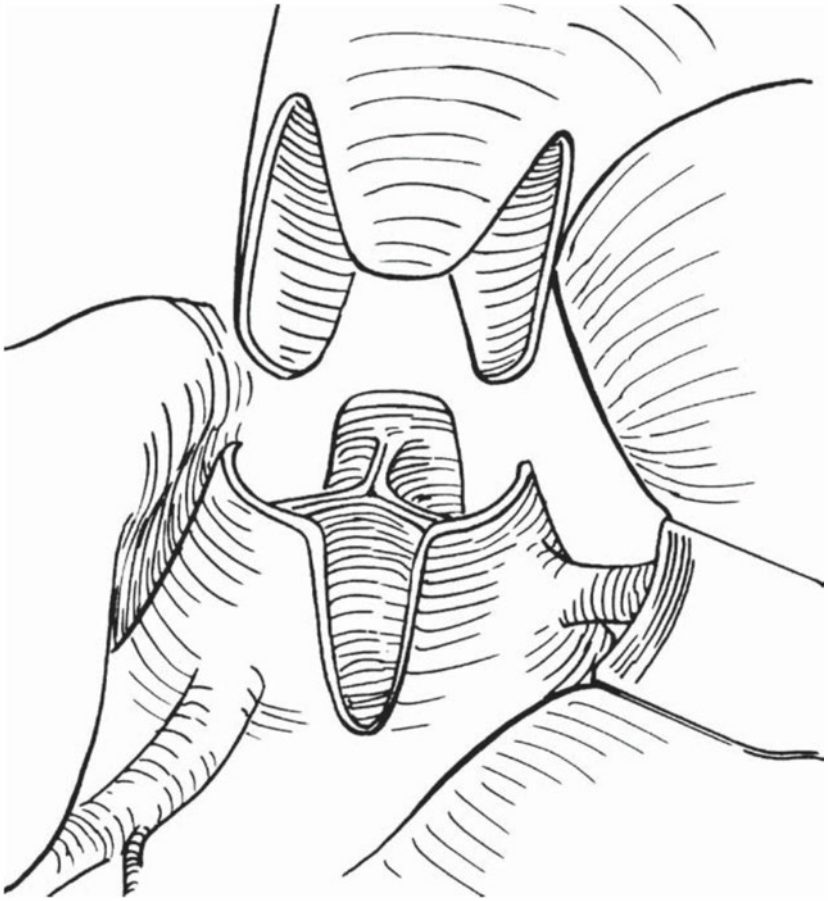


Fig. 84.12. The incisions are out of phase, allowing interdigitation of the proximal and distal flaps.

Patients with diffuse hypoplasia pose a greater surgical challenge. A combination of the techniques discussed above must be tailored to the specific findings that are encountered. Involvement of the transverse arch and descending aorta requires extensive augmentation using deep hypothermic circulatory arrest or low-flow bypass with continuous cerebral perfusion.

It is difficult to assess the results of surgery for supravalvar aortic stenosis because of the variability of pathologic findings, the diversity of the techniques employed, and the relatively small number of patients in each reported series. No consistent differences in short- or long-term outcome were noted between techniques augmenting one, two, or all three sinuses of Valsalva, though Kaushal and colleagues and Metton and colleagues reported superior outcomes (less aortic insufficiency or stenosis, and reduced reoperation rate) with the Brom technique. Freedom from reoperation at 10 years is 70% to 85%, and 10-year actuarial survival is 90% to 95%. Reoperation and late death most commonly are related to progressive valve dysfunction, not recurrence of supravalvar stenosis. Surgery for diffuse supravalvar stenosis is less successful. Early mortality is >10%, and rates of reoperation and late death also are increased. Poor aortic growth persists in both the localized and the diffuse forms despite successful relief of obstruction, and so long-term serial monitoring is required.

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EDITOR'S COMMENTS

Balloon dilation techniques have essentially supplanted surgery as the primary treatment for critical aortic stenosis of newborns and infants. These catheter interventions have a mortality rate that is virtually identical to surgical mortality rates for the same condition. Maintenance of ductal patency during the procedure is useful in aiding distal perfusion and for resuscitation after the procedure. Thus, surgical approaches to valvar aortic stenosis are relegated to cases in which initial balloon attempts have failed or there are other associated conditions. In a newborn with critical aortic stenosis, even direct exposure of the aortic valve is difficult, and usually valvotomy is performed in a relatively blind manner either antegrade with a dilator or retrograde from above with a clamp or dilator. Thus, there is little benefit to operative dilation in these patients over balloon dilation except for the benefit of supporting the patient on cardiopulmonary bypass. Surprisingly, even though the aortic valve in these infants is a structure that often appears very dysmorphic, late aortic valve development appears quite good in many cases, and the aortic valve can have a long-lasting improvement in orifice area and essentially normal anatomic and physiologic function. In most patients, however, eventual additional valve procedures will be necessary because of either restenosis or progressive calcification of a bicuspid valve.

Recently, there have been reports from Dr. John Brown in a surgical series of aortic valvotomy in critical aortic stenosis in newborns and in young infants with excellent results. In Dr. Brown's center, the results of surgical balloon valvotomy appear to be superior to the results with balloon aortic valvuloplasty in terms of delaying reoperation for valve repair or replacement and limiting the amount of aortic regurgitation. In spite of these excellent results, now a very few centers have the option of a primary surgical approach to aortic stenosis, with catheter intervention essentially having subplanted surgical therapy for this condition.

Patients who have only moderate improvement in valve function with balloon dilation can undergo repeat dilation and the most common complication is production of aortic regurgitation. Patients can then be stabilized, with aortic regurgitation, often for several months or years before valve replacement with the Ross procedure is necessary. Thus, the need for Ross aortic valve replacement in neonates is extremely unusual. In older infants and children in whom balloon dilation has been unsuccessful in relieving obstruction, direct aortic valvotomy may be valuable. As noted by the authors, incision into a true raphe should be avoided if possible. However, raphes can be debrided to aid mobility of the leaflet and to improve the effective orifice area of the valve. This often can be accomplished without significant development of regurgitation. In patients in whom there is significant fibrotic thickening of the aortic valve leaflet edges, debridement and thinning of the valve leaflets can be undertaken, and the incision in the commissures can be extended back to the annulus and then along the annulus for a short distance to improve effective orifice area and to prevent tethering of the valve leaflets at the commissural attachments.

Discrete subaortic stenosis is a relatively common finding either in isolation or in association with previous cardiac repairs. It is not uncommon for patients with interrupted aortic arch and ventricular septal defect closure to develop acquired subaortic stenosis from the development of a subaortic membrane. In addition, certain patients with atrioventricular canal defects can develop these membranes after repair, and the defect has also been reported after isolated closure of ventricular septal defect. Thus, these subaortic membranes appear to be always an acquired phenomenon related to abnormalities of geometry of the left ventricular outflow tract and turbulent blood flow in this region. An important feature is the fact that these membranes are actually attached to the endocardium of the left ventricular outflow tract, and enucleation can be performed simply by

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EDITOR'S COMMENTS (*continued*)

incising the junction of the membrane with the endocardium and identifying the endocardial layer. The membrane often can then be completely removed with blunt dissection with an endarterectomy spatula. On occasion, the membrane can extend up onto the aortic valve leaflets, causing some thickening and valve immobility and can be associated with aortic insufficiency. In these cases, it is valuable to excise the membrane and to debride the membrane from the valve leaflets. Because the membrane is an acquired structure, it is distinct from the endocardial surface of the valve leaflet and often can be peeled off completely without any disruption of the valve's intrinsic structure. The major problem with removal of subaortic membranes is the fact that recurrence is common. The high incidence of recurrence is not surprising considering that these are acquired defects related to the geometry of the outflow tract, and simple removal of the membrane does not alter this anatomic feature. I, therefore, believe it is important to perform a myotomy and myectomy of the left ventricular outflow tract wherever possible in association with resection of subaortic membrane to alter the geometry of contraction of the outflow tract, eliminate additional sources of obstruction, and potentially decrease the turbulent flow in the outflow tract, which can cause recurrence.

In spite of our preference to perform an extensive myotomy and myectomy in patients with subaortic obstruction, there have been reports primarily from the Hospital for Sick Children in Toronto that resection of subaortic muscle does not change the late recurrence rate after resection of subaortic membrane. However, we believe that the risk from myomectomy is low and interference with the left bundle conduction in the left ventricle may change the geometry of left ventricular contraction in a favorable way that may limit the development of recurrent obstruction and therefore continue to use this technique.

Tunnel subaortic stenosis is a more diffuse process and often presents a difficult surgical problem. In most cases, there is some hypoplasia of the aortic valve annulus or abnormality of the aortic valve in association with diffuse hypoplasia or tunnel obstruction of the outflow tract. On rare occasions, however, the aortic valve disease may be minor, and a direct approach with septoplasty can provide adequate relief of obstruction. In our experience, however, such patients are unusual. One difficulty with septoplasty is that in true diffuse subaortic obstruction, the obstruction extends all the way to the base of the aortic valve leaflet, and therefore, the superior extent of the septoplasty incision has to stop before the aortic annulus level is reached. Thus, there may always be a bridge of muscle tissue beneath the aortic valve leaflet that will become a potential source of obstruction. If there is an area of relatively normal outflow tract below the aortic valve, septoplasty can be very effective. In patients in whom septoplasty alone fails to relieve the obstruction because of the superior extent of the narrowing or associated aortic valve disease, we believe that a Ross–Konno autograft valve replacement supplies the best chance for long-term durability and complete relief of obstruction. Apicoaortic conduits are avoided because of the problems with late valve degeneration and abnormal coronary artery flow patterns. The use of the Ross–Konno operation has virtually eliminated the need for apicoaortic conduits in our experience.

Supravalvar aortic stenosis commonly associated with Williams syndrome represents an unusual condition but one in which good surgical results have been obtained with multiple techniques. We often have found it impossible to perform a simple patch of the ascending aorta with end arterectomy of the thickened supravalvar ridge in these patients because the ridge is often adherent to the commissural attachments of the aortic valve and excision of the ridge may result in damage to the valve commissures. The Doty technique using a Y-shaped patch has been useful, but it does not generally address the narrowing of the orifice into the left coronary ostium that is formed by the commissural attachment to the supravalvar ring posteriorly and is often associated with thickening of the endothelium and intima around the orifice of the coronary artery. Thus, we have used either a trifurcated patch to enlarge all three coronary sinuses or the technique described by Myers in which the aorta is primarily anastomosed into the base of the aortic sinuses. Although all of the techniques described can result in excellent relief of gradients across the left ventricular outflow tract, we believe that the primary anastomosis with attention to all three of the aortic sinuses permits the most optimal flow in the coronary arteries.

In many patients with Williams syndrome, there is significant hypoplasia of the ascending aorta and the aortic arch. In these cases, interdigitation of the distal aorta into the aortic sinuses for repair of supravalvar aortic stenosis is not possible. I have found it quite advantageous to patch the entire arch of the aorta with a triangular patch of pulmonary homograft material and then insert triangular patches into each of the coronary sinuses individually. The opening up of the sinus and sinotubular junction is quite effective with this technique, and then the size match is appropriate for anastomosis to the ascending aortic and arch reconstruction. In addition, if there is ostial stenosis of the left or right coronary or both, the triangular patches of homograft material can be carried down into the origin of the coronary artery itself, creating essentially an arterioplasty of the proximal coronary artery in addition to enlarging the aortic sinus and relieving the supravalvar obstruction. It is important when patching the sinuses in patients with supravalvar aortic stenosis not to oversize the patches, which can splay out the sinotubular ridge and create the substrate for potential aortic insufficiency. Often these patients have significant pulmonary stenosis and branch pulmonary artery hypoplasia, and transection of the aorta to perform this operation gives good exposure to place a T-shaped patch on the main pulmonary artery and onto the branch pulmonary arteries to the hilum of the lung.

The approach to hypertrophic subaortic stenosis in infants and children is primarily surgical. The early results with surgical resection of the subaortic septal myocardium have been excellent and the long-term results good. Thus, in children with significant gradients remaining despite medical management with calcium-channel blockers or beta blockers, surgical intervention should be considered the primary treatment. We have not found

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EDITOR'S COMMENTS (*continued*)

pacing to be particularly beneficial in children, and in fact there is controversy as to the beneficial effects of pacing for this condition. Extensive resection of septal muscle has resulted in virtually complete relief of gradients and good long-term outflow tract obstruction relief in children, and the incidence of significant conduction disturbance has been quite low.

The Mayo Clinic group has shown that extensive myomectomy for hypertrophic subaortic stenosis results in excellent relief of gradients long term with improvement in exercise performance and a decreased need for medical management.

When we perform left ventricular myomectomy for subaortic stenosis or for hypertrophic cardiomyopathy, we extensively debride all of the muscle to the left of the vertical incision in the ventricular septum all the way to the mitral valve to remove as much muscle as possible from the outflow tract in the area away from the conduction tissue. When associated mitral valve disease is present, valve replacement can be considered; however, the problems of prosthetic valves in children warrant a very conservative approach to valve replacement. We have elected to perform extensive muscle resection of the septum in association with primary mitral valve repair when feasible.

A particularly difficult group of patients are those with critical neonatal aortic stenosis and mitral valve disease. In some cases, the degree of aortic stenosis can mask the abnormalities of the mitral valve and after balloon aortic valvuloplasty patients may still have significant aortic stenosis and require a Ross–Konno type operation. If the mitral valve abnormality is not identified preoperatively, it can be extremely difficult to deal with the mitral valve after a Ross operation and mitral stenosis and insufficiency can cause pulmonary hypertension which decreases the durability of the right ventricular outflow tract conduit reconstruction. In patients with severe mitral abnormalities including arcade mitral valve in association with critical aortic stenosis that does not respond to balloon valvuloplasties, it is occasionally necessary to consider supra annular mitral valve replacement and Ross–Konno operation as a combined procedure. We have performed this in a few patients with good results, although the patients have often a complicated postoperative course due to relative left atrial hypertension and pulmonary hypertension. In cases in which the aortic annulus is significantly hypoplastic and the mitral valve is severely abnormal, some patients may be best served by a Norwood-type single-ventricle strategy.

TLS

85 Anomalies of the Sinuses of Valsalva and Aortico-Left Ventricular Tunnel

Luca A. Vricella and Duke E. Cameron

INTRODUCTION

Anomalies of the aortic root typically present as either enlargement or anomalous communication between supravalvular aorta and left ventricular (LV) cavity. In the former (sinus of Valsalva [SoV] aneurysm), the process develops progressively and is often associated with connective tissue disorders that manifest diminished tensile strength of the aortic wall. The latter (aortoventricular tunnel) is usually a perturbation in cardiac development that is nearly always clinically evident in the newborn period or in early infancy.

Aortic root dilation is also a late finding in children who have undergone repair of various forms of congenital heart defects, such as common arterial trunk, transposition of the great arteries, or bicommissural aortic valve. With regard to SoV aneurysm, we will focus on isolated dilation of a single sinus since diffuse aortic root enlargement (seen more commonly in association with other forms of congenital, atherosclerotic, or connective tissue disease) is detailed elsewhere in this book.

ANEURYSM OF THE SINUS OF VALSALVA

Morphologic Considerations

Each coronary sinus is limited inferiorly by the curvilinear hinge point of the corresponding aortic valve cusp. The intercommissural triangles are inferior to the festoon-like line of ventriculoaortic continuity (commonly referred to as "annulus"), whereas the sinotubular junction delineates the circular superior margin of the aortic root.

The sinuses of Valsalva are normally thinner than the tubular portion of the aorta, and this macroscopic finding is usually associated with a lesser degree of histologic representation of the tunica media. In patients with SoV aneurysm, this normal characteristic is accentuated. Thinning of the aortic wall with disconnection of the media from the aortoventricular junction is seen histologically and increases over time.

Figure 85.1 schematically demonstrates the anatomic relations of the aortic root with the adjacent cardiac structures as seen from the operating surgeon's viewpoint. These relations account in turn for the different modes of clinical presentation that are seen with pathologic enlargement of the SoVs. Dilation of the right SoV (most common) will, therefore, typically progress in the direction of the right ventricular outflow tract, whereas the noncoronary sinus will involve either left or right atrial chambers. Isolated enlargement of the left SoV (with possible rupture into the left atrium) is the rarest form of this pathology. Although typically SoV aneurysms involve one of the three sinuses as a well-defined diverticular outpouching, two or three sinuses can be affected simultaneously by the pathologic process. A ventricular septal defect (VSD) is observed in 25% to 50% of cases.

Aneurysms of the SoVs are rare. They are observed in 0.1% of autopsy series and in 0.14% to 0.96% of large operative cohorts; they are also five times more common in patients of Asian descent.

Clinical Presentation

Pathologic SoV enlargement is often diagnosed incidentally in asymptomatic patients. Clinical presentation can occur as an aortic catastrophe (free intrapericardial rupture), acutely with endocarditis or intracardiac rupture (within right or left atrium, right ventricle), or with a more indolent clinical course during childhood. In this last mode of presentation, progressive dilation, distortion, and subsequent loss of cusp coaptation can lead to clinically significant aortic valvar insufficiency.

Rupture or fistulization almost always involves the right coronary sinus, while a minority of patients have involvement of the noncoronary (up to 30%) or left coronary (<2%) sinuses. Perhaps, in the largest reported surgical series (149 patients with SoV aneurysm), intracardiac rupture was seen at presentation in just <50% of cases. Aortic root rupture into a neighboring cardiac chamber is furthermore a rare event in the first two decades of life. If we extrapolate from data available in young patients with connective tissue disorders, this is not surprising. In a meta-analysis of 286 patients age <20 years with Marfan syndrome, Knirsh and coworkers reported aortic dissection in only five patients (1.7%) and rupture in three patients (1.0%). All but one patient (14 years old) were 19 years of age at the time of the acute event involving the aortic root. When intracardiac rupture of a SoV aneurysm occurs, a mean survival period of 3.9 years has been reported in untreated patients.

When symptomatic, patients may present with chest discomfort, palpitations, dyspnea on exertion, or florid congestive heart failure, with diastolic hypotension and pulmonary overcirculation from acute left-to-right shunting between aorta and right-sided chambers. Of patients with fistulization, >60% will be symptomatic at presentation. A precordial continuous murmur is usually heard. The clinical picture can be that of sepsis in the setting of endocarditis (up to 20% of patients). Dyspnea or even cyanosis can result from progressive right ventricular outflow tract obstruction by an enlarging SoV aneurysm that protrudes into the right ventricle. Atrioventricular conduction abnormalities are present in 10% of patients.

The diagnosis is easily confirmed by transthoracic echocardiography, with cardiac catheterization reserved for patients with significant risk factors for coronary artery disease.

Surgical Indications and Technique

Indications for surgery in patients with uniform dilation of the aortic root are

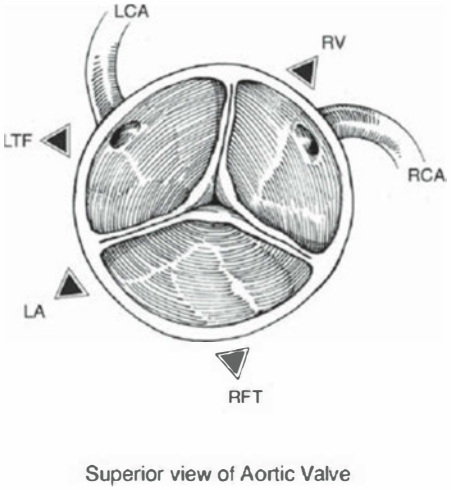


Fig. 85.1. Superior schematic view of the aortic root with adjacent structures LA, left atrium; LCA, left coronary artery; LFT, left fibrous trigone; RA, right atrium; RCA, right coronary artery; RFT, right fibrous trigone; RV, right ventricle.

fairly well established. Enlargement beyond 5.0 cm, or enlargement >1.0 cm per year, is a well-accepted indication for surgical intervention in asymptomatic patients. Aortic regurgitation from distraction of the commissures or asymmetric sinus enlargement with ventricular dilation is also an accepted indication for surgery, as is aortic dissection. Threshold for intervention can be lowered for young adults with lesser degrees of enlargement when there is a family history of aortic dissection or rupture. In our practice, we have applied the same adult criteria for intervention to child, as the risk of aortic rupture in the first decade is quite low. An exception is the Loeys–Dietz syndrome, where rupture and dissection can occur in much smaller aortas and at a much earlier age.

In the case of isolated enlargement of a single sinus, indications are not as well defined. Strong consideration to surgical intervention should be given in asymptomatic patients because of the high likelihood of progressive increase in size and the possibility of rupture or endocarditis. The latter is obviously an indication for urgent surgical correction.

A transesophageal echocardiogram is routinely performed after induction of anesthesia to confirm preoperative diagnosis, assess valvular competence, and rule out the possibility of a concomitant septal defect.

The operative approach is via median sternotomy, with bicaval venous cannulation and cardiopulmonary bypass and moderate hypothermia (24 to 28°C). A right superior pulmonary vein vent is inserted, and the heart is arrested with either antegrade or retrograde administration of cold blood cardioplegia solution; direct intracoronary delivery is used in case of ruptured aneurysm or significant aortic regurgitation, or with retrograde coronary perfusion. If the tip of the "windsock" is palpable through the right atrium, it can be compressed manually to prevent runoff of antegrade administered cardioplegia. We use continuous topical cooling as well as carbon dioxide flooding of the surgical field to minimize retention of air within the left-sided chambers.

A transverse aortotomy is carried out and the root anatomy is assessed. An oblique right atriotomy is performed next, allowing for the identification of both ends of the aneurysm or of the fistulous tract in case of rupture (Fig. 85.2). In case of protrusion or rupture into the right ventricle, exposure can be obtained through a right atriotomy or a limited ventriculotomy (Fig. 85.3). When the fistulous tract or diverticulum is in the right ventricular infundibulum, the lesion can also be exposed through a transverse pulmonary arteriotomy. The defect must be repaired through the aortic root, using a patch of autologous or bovine pericardium to exclude the aortic inlet into the aneurysm. Primary closure predisposes to a higher risk of recurrence (as high as 20%) or aortic valve regurgitation from deformation of the root. In case of fistulization, the opening on the atrial or ventricular side should be addressed as well. The ventricular or atrial aspect of the fistula can be closed primarily, but a patch should be used to incorporate closure of a coexisting VSD (Fig. 85.3). Great care should be taken in avoiding the atrioventricular conduction system at the time of VSD closure.



Fig. 85.2. Intraoperative photograph of unruptured aneurysm of the right sinus of Valsalva with "windsock" morphology, seen through a right atriotomy. A stay suture is retracting the aneurysm caudally. (Courtesy of Dr. David Yuh, Yale University School of Medicine, New Haven, CT.)

In tri-sinus enlargement of the aortic root and a functionally normal aortic valve, we attempt aortic root replacement with valve preservation. The techniques involved in aortic valve-sparing root replacement are detailed elsewhere in this book and have been successfully applied to pediatric patients as well. Need for aortic valve replacement has been reported in 30% to 50% of patients with significant preoperative aortic valve regurgitation, but with reproducible aortic valve repair techniques such as leaflet free edge suspension and mid-leaflet plication to address prolapse, the need for valve replacement has decreased in more recent series.

Alternatives to valve preservation in case of diffuse enlargement and unrepairable aortic regurgitation are valve replacement with mechanical, xenograft, or homograft prostheses. The Ross procedure (pulmonary valve autotransplantation) can be considered as a potential option in patients without stigmata of connective tissue disorders.

Results

In a large contemporary series, Au and coworkers reported long-term results on 53 patients operated for SoV aneurysms; there was no operative mortality, and overall survival was 83% at 15 years. Over a 32-year period, 22 patients underwent repair of single SoV aneurysm at the Johns Hopkins Hospital. Nineteen patients presented with intracardiac rupture; congestive heart failure was the most frequently presenting symptom. Operative survival was 95%, and 5- and 10-year survival was 84.9% and 59.4%, respectively. The lower long-term survival in our series reflected older age at presentation, more concomitant cardiac conditions, and more acquired rather than congenital etiologies, as are seen more frequently in the Western patient population. Factors such as bacterial endocarditis, aortic valve replacement, and concomitant VSD do not appear to adversely influence operative and long-term survival. However, the preoperative presence of a VSD or significant aortic insufficiency did affect the rate of late reintervention for aortic regurgitation. In particular, prolapse of the involved cusp with leaflet fibrosis and insufficiency appeared to jeopardize long-term viability of aortic valve repair. Need for late aortic valve replacement has been reported in 25% of patients at 10 years and was typically in patients with VSD on presentation and residual aortic regurgitation at the time of discharge. Need for late reintervention for

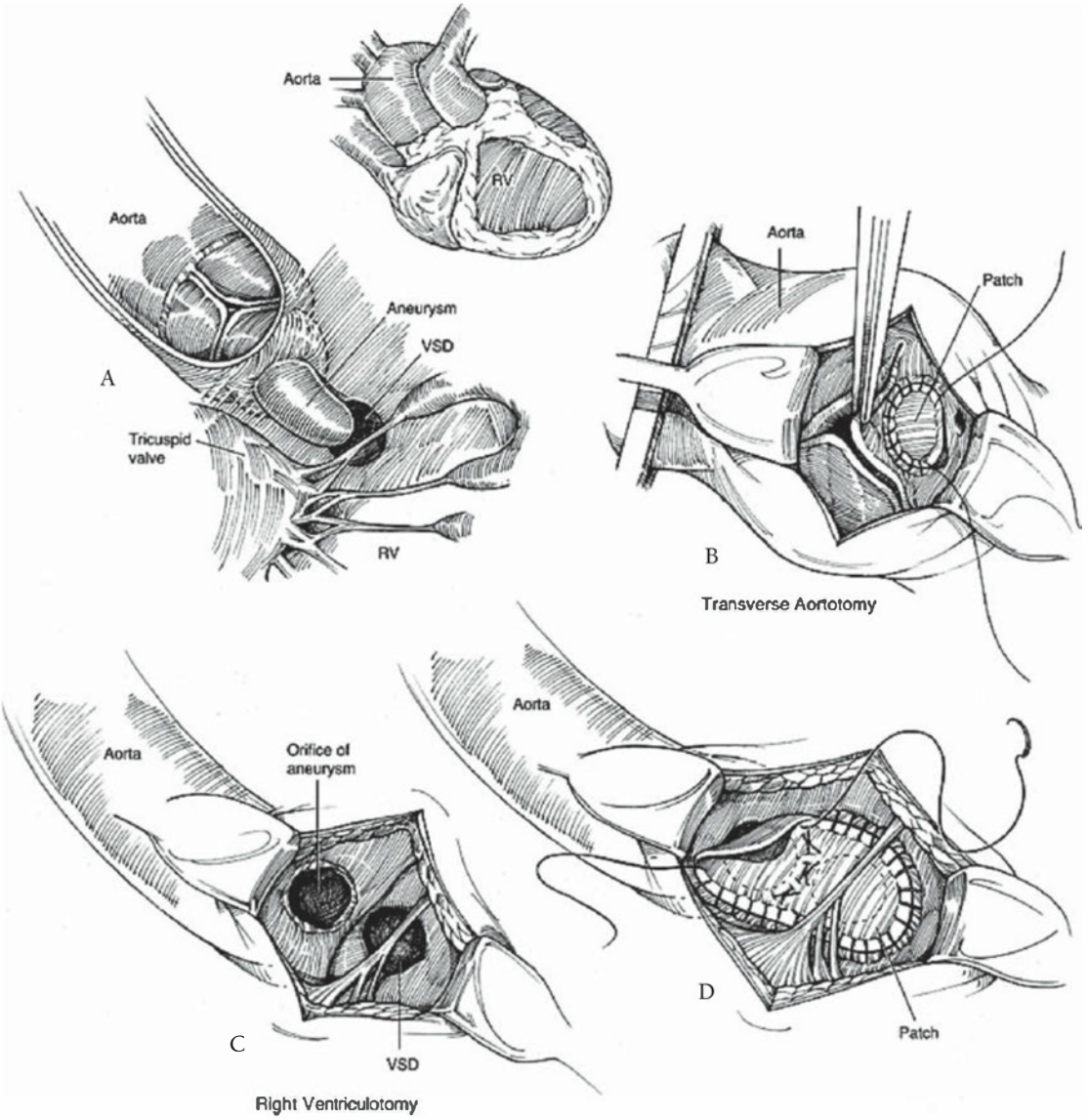


Fig. 85.3. (**A**) Unruptured sinus of Valsalva aneurysm originating from the right coronary sinus and associated with a ventricular septal defect (VSD). (**B**) Patch repair of the aneurysm with autologous pericardium through a transverse aortotomy. (**C** and **D**) Transventricular aneurysm and VSD closure with synthetic patch. RV, right ventricle.

recurrent regurgitation, prosthetic valve dehiscence, endocarditis, and thrombosis all impacted survival negatively.

AORTICO-LEFT VENTRICULAR TUNNEL

Morphology

Aortico-LV tunnel (ALVT) is an exceedingly rare condition. It represents 0.001% of all congenital cardiac anomalies; less than 150 cases were reported in the world literature up to 2012.

In this developmental abnormality, an endothelialized paravalvular communication exists between aortic root (typically the right coronary sinus) and the LV, or very rarely leading to the right ventricle by means of a ventricular septal aneurysm. The lesion can often be appreciated as a bulging and pulsatile mass, visible externally between the aorta and the pulmonary artery. Although the trajectory of the tunnel has a variable course, the supravalvular inlet is usually located in the right coronary sinus above, below, or at the level of the right coronary ostium. The ventricular opening is readily visible on retraction of the aortic valve cusps and typically located below the right coronary leaflet. The paravalvular tract is directed inferiorly and leftward into the LV. The proximal opening is usually larger and can be slit-like or oval in appearance. The tunnel can be either aneurysmal or serpiginous and narrow. There are, therefore, two distinct portions of the tunnel: (1) aortic, located between aortic opening and interventricular septum and (2) intracardiac, located within the interventricular septum and extending to the LV opening. The intracardiac portion is within the septum that forms the posterior wall of the right ventricular outflow tract. The variable morphologic features of ALVT have been summarized as types I to IV of the Hovaguimian classification (Fig. 85.4).

The aortic valve is usually competent or mildly regurgitant. Coronary anomalies can be seen in 30% of patients; relatively simple cardiac defects (VSD, bicommissural aortic valve, aortic stenosis among others) are observed in up to 45% of cases.

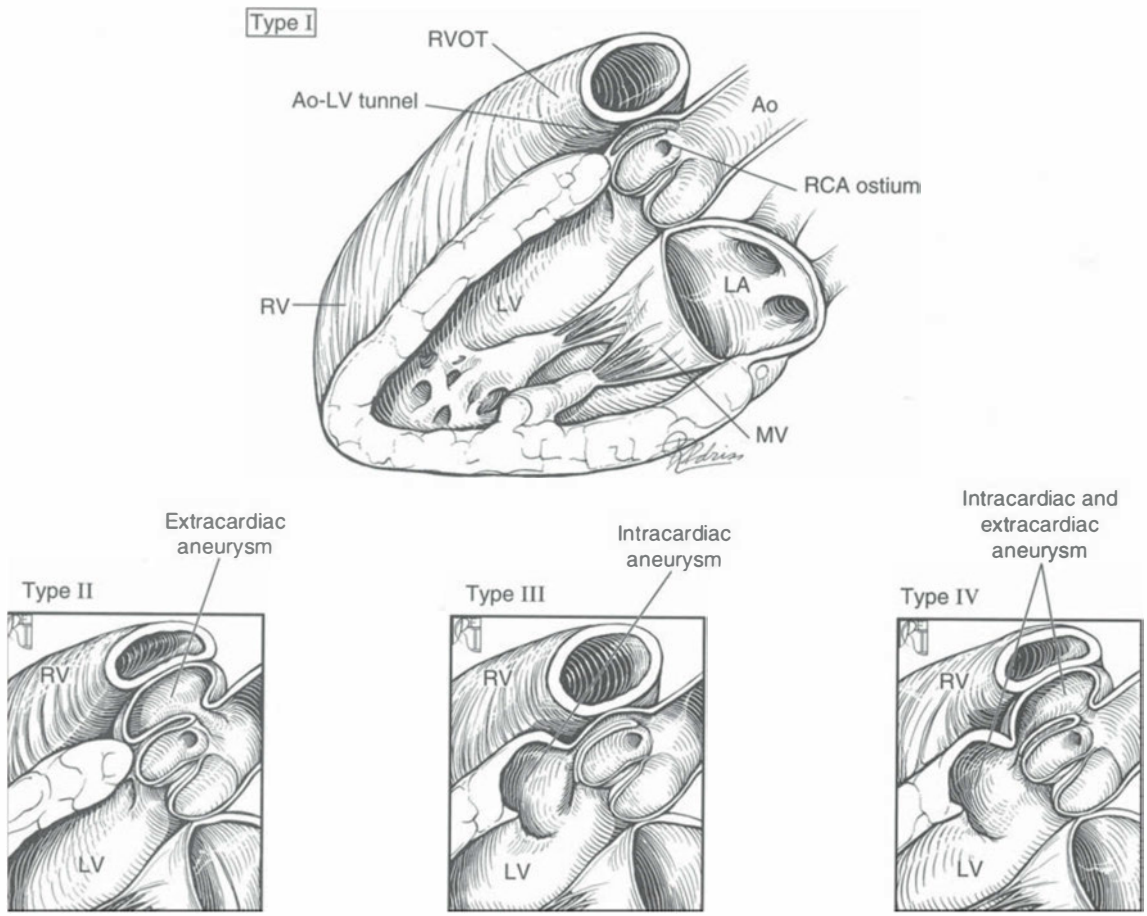


Fig. 85.4. Proposed morphologic classification of aortico-left ventricular tunnel. Type I: Simple tunnel with slit-like opening at aortic end, no aortic valve distortion. Type II: Large tunnel with oval aortic opening with or without aortic valve distortion. Type III: Intracardiac aneurysm of the ventricular portion of the tunnel, with or without right ventricular outflow tract obstruction. Type IV: Combination of types II and III. Ao, aorta; LV, left ventricle; MV, mitral valve; RCA, right coronary artery; RV, right ventricle; RVOT, right ventricular outflow tract. (Reprinted with permission from Shum-Tim D, Tchervenkov CI. Aortic-left ventricular tunnel. In Mavroudis C, Backer CL, eds. Pediatric Cardiac Surgery. 3rd ed. Philadelphia, PA: Mosby; 2003:576.)

Clinical Presentation and Diagnosis

The severity of symptoms clearly depends on the size of the aortico-LV communication and the consequent regurgitant volume. The lesion should be suspected in any neonate or infant with systolic–diastolic murmurs and congestive heart failure. Differential diagnosis includes patent arterial duct, aortopulmonary window, absent pulmonary valve syndrome, congenital coronary artery fistula, and, in older children, ruptured SoV aneurysm. Tachypnea, poor weight gain, cardiomegaly, and widened pulse pressure with bounding peripheral pulses are other typical signs and symptoms of ALVT. Rarely, cyanosis can be present, secondary to progressive right ventricular outflow tract obstruction from septal bulging beneath the pulmonary valve.

Chest X-ray typically reveals cardiomegaly, plethoric lung fields, and a tortuous ascending aorta. Transthoracic echocardiography is the diagnostic modality of choice, demonstrating the typical septal "drop-out" under the right coronary sinus. LV hypertrophy, left-axis deviation, and repolarization abnormalities are usually seen on electrocardiography. Rarely, cardiac ischemia from diastolic runoff into the fistulous tract can be present. Occasionally, it may be difficult to differentiate between aortic and paravalvular regurgitation. In such cases, aortic root angiography (with or without simultaneous occlusion of the ALVT) readily establishes the diagnosis. Magnetic resonance imaging has also been used as an alternative to angiography to define morphology.

Surgical Indications and Technique

Given the often impressive symptoms at the time of presentation and the risk of progressive aortic valve insufficiency in patients with lesser degrees of clinical acuity, operative intervention is indicated at the time of diagnosis, even in asymptomatic patients. The goal of preventing aortic valve regurgitation from progressive root distortion and secondary valvular changes is particularly relevant in small patients, in whom the alternatives to aortic valve repair are largely unsatisfactory. In case of very small ALVT diagnosed in asymptomatic patients, careful medical management and expectant follow-up can nevertheless be considered because spontaneous closure of small communications has been reported.

Intraoperatively, transesophageal echocardiography is used to assess competence of the valve and adequacy of repair after cardiopulmonary bypass.

In the absence of a septal communication, bypass is established using a single right atrial cannula and the distal ascending aorta. The left atrium is vented through the right superior pulmonary vein and systemic cooling to moderate hypothermia (24 to 28°C) is started. Because of the paravalvular regurgitation, it is important to avoid LV distension once the heart fibrillates. The aorta is, therefore, immediately cross-clamped,

and an aortotomy is performed above the sinotubular junction. Blood cardioplegia (30 cm^3/kg in pediatric patients) is then infused directly into the coronary ostia and redosed at 30-minute intervals. Alternatively, retrograde cardioplegic delivery can be used as alternative myocardial protection strategy.

As in SoV aneurysm with fistula, the defect should be approached from both the aortic root and LV, and both the inlet and outlet of the ALVT closed. This is critically important since isolated inlet closure has been associated with AVLT recurrence. Concerns have been raised about the technique of closure of the AVLT inlet, as direct closure may result in distortion of the aortic valve. We favor patch closure with Gore-Tex, bovine pericardium, or autologous pericardium in infants and small children (Fig. 85.5). The ventricular outlet of the tunnel can be exposed either through the retracted leaflets of the aortic valve or directly through the tunnel, approached from the external surface between the aorta and the pulmonary root (Fig. 85.6).

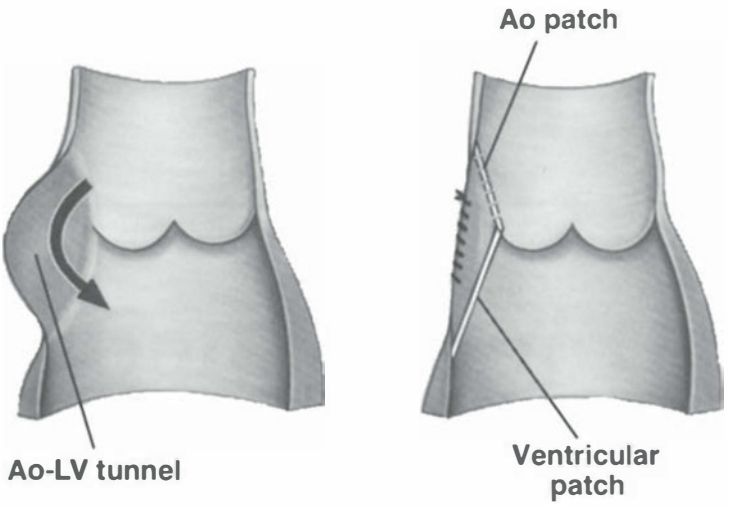


Fig. 85.6. Alternative surgical approach to repair aortico-left ventricular (Ao-LV) tunnel. Inlet and outlet orifices are exposed *through* the tunnel (*left*). Patch closure of both aortic and ventricular openings is accomplished, followed by closure of the tunnel between aorta and pulmonary artery (*right*). (Reprinted with permission from J Stark. Congenital Anomalies of the Sinuses of Valsalva and Aortico-Ventricular Tunnel. In Stark J, deLeval M (eds), Surgery for Congenital Heart Defects, 3rd ed. Wiley, 2006;607.)

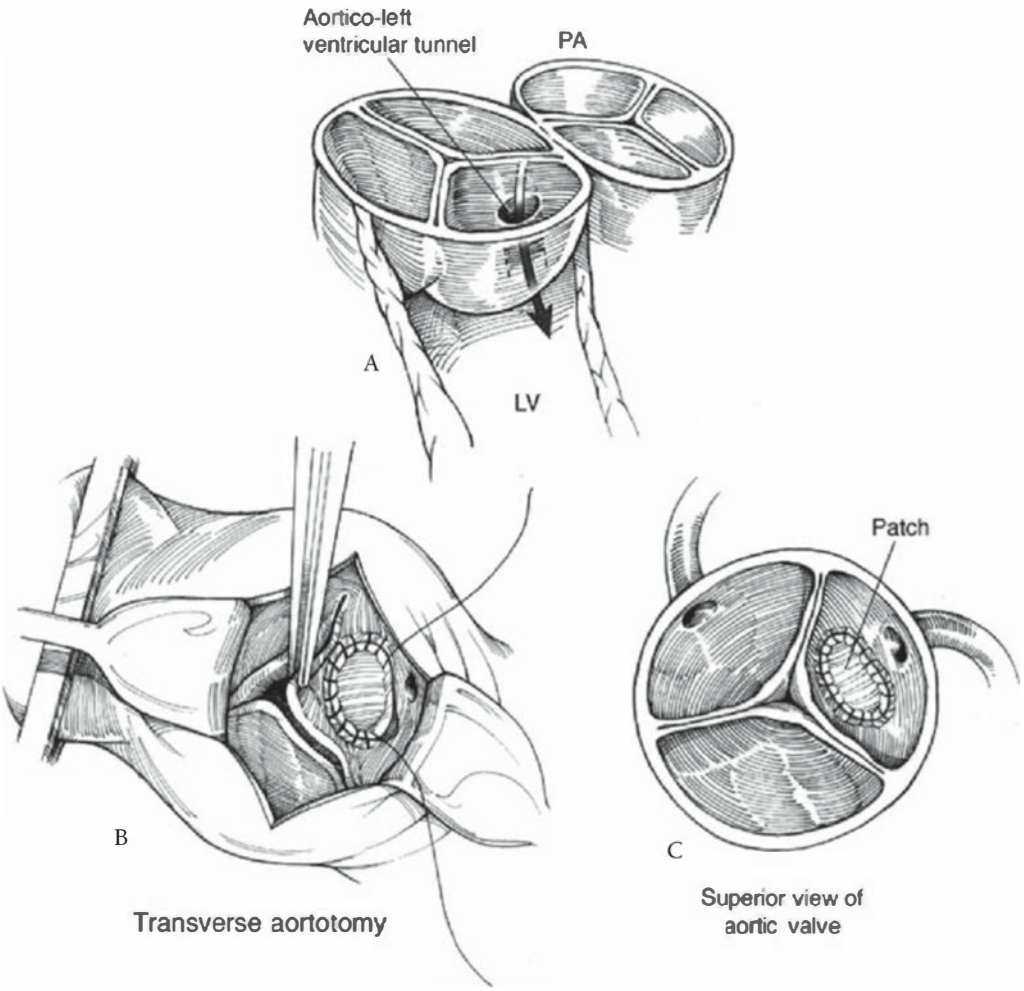


Fig. 85.5. (**A**) Aortico-left ventricular tunnel, as typically observed arising in the right sinus of Valsalva. (**B** and **C**) Through an aortotomy, the inlet orifice to the tunnel is closed with an autologous pericardial patch. The ventricular opening of the tunnel is closed through the aorta separately, either primarily or with a second patch. LV, left ventricle; PA, pulmonary artery.

Results

Reported operative mortality ranges between 0% and 16%. Reported complications include heart block and aortic valve regurgitation.

The major postoperative concern is the need for late reintervention. Up to 50% of patients may require late aortic valve replacement because of progressive valvar deformity (likely the result of postrepair turbulence) or aneurysmal dilation of the aortic root secondary to poor support of the right aortic cusp. Significant early or late aortic insufficiency is seen more frequently in children undergoing repair at an older age.

ALVT recurrence is rare, especially if closure is performed at both ends. If a residual small ALVT is observed, consideration may be given to coil embolization, but proximity to the aortic valve and right coronary artery orifice must be weighed carefully.

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EDITOR'S COMMENTS

Aneurysms of the sinus of Valsalva are often identified after rupture occurs. As noted by the authors, most sinus of Valsalva aneurysms rupture into the low-pressure right atrial or right ventricular chamber. Left coronary sinus of Valsalva aneurysms, however, can rupture into the left atrium, although even in this location rupture into the right atrium or ventricle is more common. VSD is associated with approximately one-third of sinus of Valsalva aneurysms and therefore should be carefully looked for in these patients. Although fistulous communications between the sinuses of Valsalva and cardiac chambers can be approached with a catheter technique and occluder devices, surgery remains the procedure of choice because of the direct approach to each end of the fistula and the ability to either primarily close or patch these defects without significant prosthetic material. One of the more current causes of ruptured sinus of Valsalva into the atrium is related to the use of atrial septal defect occluder devices, which, when excessively large, can erode into the base of the aortic wall and aortic sinus, resulting in an aortic to atrial fistula. Complete removal of the device and surgical closure of the aneurysm would seem to be the most appropriate treatment for these complications of device intervention. The very low morbidity and mortality of operative intervention should continue to lend favor to this approach.

A particular controversy is the patient with an asymptomatic sinus of Valsalva aneurysm identified on echocardiographic screening for other congenital conditions. In the presence of an isolated aneurysm of a single sinus of Valsalva, the indications for operation are unclear. One could argue that resection of the abnormal tissue in the sinus and replacement with homograft or other material may permit a better remodeling of the sinus and prevent late development of aortic insufficiency, but this is not clearly been demonstrated. In most cases, expectant observation of the aneurysm with operation if significant size increases are identified seems to be a preferred approach. In patients who have developed a rupture of the sinus of Valsalva aneurysm, even asymptomatic, operative intervention is recommended.

Aortico-left ventricular tunnel is an extremely rare defect; however, it is important to emphasize that when this defect is approached, both the aortic and ventricular ends of the tunnel should be closed. Simple closure of the aortic end of the defect can result in a chamber developing in the septum from systolic inflow, and the pulsatility in this chamber can create an aneurysm of the ventricular septum, which can distort the aortic valve annulus and cause progressive aortic insufficiency.

The embryologic abnormality of aortic-left ventricular tunnel bypasses the aortic valve and may affect aortic valve development. In some cases, these patients may have fusion of the commissures of the aortic valve leaflets and relative hypoplasia of the aortic valve annulus. In addition, the tunnel as it channels through the ventricular septum can affect flow into the pulmonary arteries and can also affect pulmonary valve development. Therefore, it is imperative to examine the valves carefully in these patients. In some cases, an aortic valvotomy maybe necessary or more extensive procedure such as aortic valve replacement with a Ross operation may be a preferred approach. The extreme rarity of this condition leads to a wide variety of clinical presentations and anatomy.

The optimal technique for dealing with sinus dilation in patients with Marfan syndrome or other degenerative connective tissue diseases of the aortic root remains controversial. As discussed in this chapter, the dilated sinuses of Valsalva can be addressed by graft replacement down to the level of the aortic annulus with preservation of the commissural attachments of the aortic valve and a portion of aortic wall as in the Yacoub operation, or the valve can be resuspended inside a tubular Dacron graft as in the David operation. The excellent results with the David valve-sparing root replacement for connective tissue abnormalities has made it the preferred approach in most centers for patients with dilation of the aortic root. The valve leaflets are also abnormal in these conditions, and progressive aortic insufficiency may occur even with valve-sparing procedures, as in the David operation. Thus, if there is any concern about significant aortic valve disease or aortic insufficiency, complete replacement of the aortic root with a homograft or prosthetic valve and conduit may be the best approach. With these techniques, the operative results have been excellent in the authors' series from Johns Hopkins University, and despite the frequent need for reoperation on other portions of the aorta or the mitral valve in these patients, late morbidity from valve replacement has been low.

TLS

86 Atrioventricular Canal Defects

Jeffrey P. Jacobs and Martin J. Elliott

ANATOMY

Atrioventricular (AV) canal defects have also been called endocardial cushion defects and AV septal defects (AVSDs). These defects are characterized by varying degrees of incomplete development of the septal tissue surrounding the AV valves along with varying degrees of abnormalities of the AV valves themselves. Consequently, AVSD may include defects in the inferior portion of the atrial septum, defects in the inflow portion of the ventricular septum, and defects in the tissue forming the left and right AV valves. We prefer the term AVSD because the anomaly is primarily caused by the deficiency of normal AV septal structures.

AVSDs represent a spectrum of cardiac anomalies subdivided into partial AVSDs, intermediate AVSDs, and complete AVSDs. *Partial AVSDs* (also known as incomplete AVSDs) have a crescent-shaped atrial septal defect (ASD) in the inferior portion of the atrial septum just above the AV valve. This defect may also be referred to as an ostium primum defect. The partial AVSDs also have varying degrees of malformation of the left AV valve, leading to varying degrees of left AV valve regurgitation. *Complete AVSDs* have both defects in the atrial septum just above the AV valves and defects in the ventricular septum just below the AV valves. In complete AVSD, the AV valve is the one valve that bridges both the right and left sides of the heart, creating superior and inferior bridging leaflets. Partial AVSDs and complete AVSDs represent a spectrum of cardiac pathologic conditions. An intermediate form in the middle of this spectrum has been described and termed *intermediate AVSD* (also known as transitional AVSD). This form of AVSD not only has two distinct left and right AV valve orifices but also has both an ASD just above and a ventricular septal defect (VSD) just below the AV valves. The VSD in this intermediate form of AVSD is often restrictive. Although these AV valves in the intermediate form do form two separate orifices, they remain as abnormal valves. Table 86.1 provides definitions for AV canal defects provided by the International Society for Nomenclature of Paediatric and Congenital Heart Disease (www.ipccc.net).

The AV valve apparatus in AVSD has been described as having either five or six leaflets. In partial AVSD (Fig. 86.1A), the AV valve apparatus is easily understood as having six leaflets. On the left side, the leaflets have been termed left superior, left lateral, and left inferior. On the right side, the leaflets have similarly been termed right superior, right lateral, and right inferior. In partial AVSD, both the right superior and inferior leaflets fuse with the ventricular septum to complete the structure of the right AV valve. The left superior and inferior leaflets similarly fuse with the septum to form the left AV valve. The commissure between the left superior and inferior leaflets represents the "cleft" of the left AV valve, which is found in partial AVSD. This cleft is equivalent to the line of abutment (or zone of apposition) of the superior bridging leaflet and the inferior bridging leaflet in complete AVSD.

In complete AVSD (Fig. 86.1B and 86.1C), it is more difficult to conceive of the common AV valve as a six-leaflet valve. A five-leaflet model is more realistic for complete AVSD. A superior bridging leaflet and an inferior bridging leaflet are always present. These bridging leaflets have very variable morphology in both the amount of leaflet that crosses over the ventricular septum and the degree of chordal attachment to the ventricular septum. Furthermore, scalloping of the bridging leaflets may create the illusion of extra leaflets. In addition to the superior and inferior bridging leaflets, the five-leaflet model is completed by a left lateral leaflet, a right lateral leaflet, and a right anterosuperior leaflet.

The degree of bridging and chordal attachment by the superior bridging leaflet forms the basis for the Rastelli classification of complete AVSD originally described in 1966. The Rastelli classification does not relate to the anatomy of the inferior bridging leaflet because this leaflet displays greater anatomic variation, and no consistent relationship exists between the morphology of the superior bridging leaflet and the inferior bridging leaflet. In a Rastelli type A defect (Fig. 86.1B), the superior bridging leaflet is effectively limited to the left ventricle, with its right margin being attached to the ventricular crest. The anterosuperior leaflet of the right AV valve is also attached to the ventricular septal crest, giving the appearance that the superior bridging leaflet is split at the interventricular septum. In many cases, chordal attachments pull the plane of the AV valve down into the VSD below the plane of the annulus. In Rastelli type C defects (Fig. 86.1C), there is a marked bridging of the ventricular septum by the superior bridging leaflet. The superior bridging leaflet floats freely over the ventricular septum without chordal attachment to the crest of the ventricular septum. Rastelli type B is somewhere between types A and C and is very rare in our experience; Rastelli type B involves anomalous papillary muscle attachment from the right side of the ventricular septum to the left side of the common superior (anterior) bridging leaflet.

The other important anatomic consideration when planning the repair of an AVSD is the location of the conduction system because it is very vulnerable during surgical repair (Fig. 86.2). The AV node is displaced posteriorly and inferiorly toward the coronary sinus. The AV conduction axis then runs from this node toward the ventricle through the crest of the ventricular septum. Here, the posteriorly displaced bundle of His is usually covered by the inferior bridging leaflet of the AV valve. Thus, the AV node lies in the tissue between the coronary sinus and the margin of the VSD, if present. The location of the AV node is altered in AVSD because the ostium primum ASD often pushes the coronary sinus posteriorly and inferiorly toward the left atrium. This distorts the triangle of

Table 86.1 Definitions

The International Society for Nomenclature of Paediatric and Congenital Heart Disease (www.ipccc.net) offers the following definitions:

1. **Atrioventricular septal defect (atrioventricular canal)**

   A congenital cardiac malformation with a common atrioventricular junction (both atriums connect to a common atrioventricular valve) and varying degrees of malformation of the left-sided component of the common atrioventricular valve.

2. **Partial atrioventricular septal defect with isolated atrial component (primum atrial septal defect; partial atrioventricular canal defect with isolated atrial component)**

   A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal) with an interatrial communication just above the atrioventricular valve, no interventricular communication just below the atrioventricular valve, separate right and left atrioventricular valvar orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets of the common atrioventricular valve are bound down to the crest of the scooped out ventricular septum so that the potential for shunting through the atrioventricular septal defect is possible only at the atrial level and not at the ventricular level.

3. **Atrioventricular septal defect with isolated ventricular component (atrioventricular canal defect with isolated ventricular component)**

   A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal) with an interventricular communication just below the atrioventricular valve, no interatrial communication just above the atrioventricular valve, separate right and left atrioventricular valvar orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets of the common atrioventricular valve are bound to the atrial septum so that the potential for shunting through the atrioventricular septal defect is possible only at the ventricular level and not at the atrial level.

4. **Complete atrioventricular septal defect with atrial and ventricular components (complete atrioventricular canal with atrial and ventricular components)**

   A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal) with an interatrial communication just above the atrioventricular valve, an interventricular communication just below the atrioventricular valve, and varying degrees of malformation of the left-sided component of the common atrioventricular valve. The bridging leaflets usually float to a varying extent within the atrioventricular septal defect.

5. **Intermediate atrioventricular septal defect with atrial and ventricular components and separate atrioventricular valvar orifices (intermediate atrioventricular canal defect with atrial and ventricular components and separate atrioventricular valvar orifices)**

   A congenital cardiac malformation that is a variant of an atrioventricular septal defect (atrioventricular canal) with distinct left and right atrioventricular valvar orifices but also with both an interatrial communication just above the atrioventricular valves and an interventricular communication just below the atrioventricular valves. While these atrioventricular valves in the intermediate form have two separate orifices, they remain abnormal valves. The ventricular component is usually restrictive (with a pressure gradient across the ventricular component of the atrioventricular septal defect).

6. **Atrioventricular septal defect and tetralogy of Fallot (atrioventricular canal defect and tetralogy of Fallot)**

   A congenital cardiac malformation with both atrioventricular septal defect (atrioventricular canal) and tetralogy of Fallot. An atrioventricular septal defect (atrioventricular canal) is defined as a congenital cardiac malformation with a common atrioventricular junction (both atriums connect to a common atrioventricular valve) and varying degrees of malformation of the left-sided component of the common atrioventricular valve. Tetralogy of Fallot is defined as a group of congenital cardiac malformations with biventricular atrioventricular alignments or connections characterized by anterosuperior deviation of the conal or outlet septum or its fibrous remnant, narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the malalignment type, and biventricular origin of the aorta. Tetralogy of Fallot will always have a ventricular septal defect, narrowing or atresia of the pulmonary outflow, and aortic override, and will most often have right ventricular hypertrophy.

7. **Congenital abnormality of the common atrioventricular valve**

   A congenital cardiac malformation in which there is an abnormality of the common atrioventricular valve.

8. **Common atrioventricular valvar regurgitation**

   Congenital backward flow through the common atrioventricular valve.

Koch and creates a second triangle called the nodal triangle. The nodal triangle is bounded by the coronary sinus, the posterior attachment of the inferior bridging leaflet, and the leading edge of the atrial septum at the septal defect. The ASD pushes the AV node and the corresponding conduction tissues posteriorly and inferiorly along with the coronary sinus. The AV node, therefore, lies at the apex of the nodal triangle in a more posterior and inferior location. The bundle then travels down on the crest of the ventricular septum under the inferior bridging leaflet on the rim of the VSD.



DIAGNOSIS

Patients with AVSD present in a variety of clinical conditions depending on the size of the septal defects, the direction and magnitude of the associated shunt, and the associated lesions. Patients with partial AVSD may have an asymptomatic cardiac murmur similar to those with secundum ASDs. However, when left AV valve insufficiency is more pronounced, patients may have symptoms of pulmonary congestion, cardiac failure, and dyspnea. Patients with complete AVSD are more likely to have prominent left-to-right shunting and are similarly more likely to have symptoms of congestive heart failure, fatigue, and dyspnea. Complete AVSD presents with a more

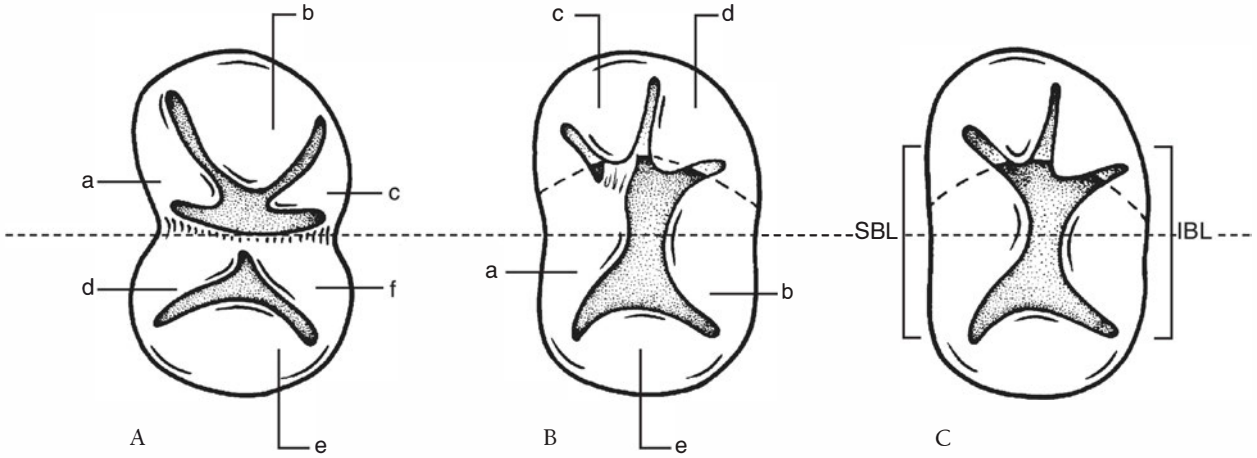


Fig. 86.1. (**A**) The atrioventricular valve apparatus in partial atrioventricular septal defect has six leaflets (a, right superior leaflet; b, right lateral leaflet; c, right inferior leaflet; d, left superior leaflet; e, left lateral leaflet; f, left inferior leaflet). The dashed line denotes the plane of the interventricular septum. (**B**) A five-leaflet model demonstrates a Rastelli A atrioventricular septal defect (a, superior bridging leaflet; b, inferior bridging leaflet; c, right anterosuperior leaflet; d, right lateral leaflet; e, left lateral leaflet). Again, the plane of the interventricular septum is demonstrated by the dashed line. (**C**) The anatomy of a Rastelli C defect. The free-floating superior bridging leaflet is demonstrated, as is the inferior bridging leaflet.

malignant course than partial AVSD. With the complete defect, severe cardiac failure is often present in infancy, and severe pulmonary hypertension will eventually develop, resulting in the death of up to 65% of infants before 1 year of age without surgical intervention. More than 50% of the patients with complete AVSD also have Down syndrome.

Physical examination often reveals a variety of cardiac murmurs. A systolic ejection murmur may be found in the pulmonary area because of increased flow across the pulmonary valve. A holosystolic apical murmur is also present when left AV valve regurgitation is significant. The ASD and VSD may also have associated cardiac murmurs.

A chest radiograph often reveals enlargement of the pulmonary artery. The film may also show right ventricular hypertrophy as symptoms of failure progress and left ventricular enlargement with significant left AV valve regurgitation. An electrocardiogram often reveals right ventricular hypertrophy and sometimes left ventricular hypertrophy as well. A vector cardiogram usually shows a counterclockwise frontal plain loop.

Echocardiography is the modality of choice for establishing a definitive diagnosis in the current era. Two-dimensional echocardiography, along with color Doppler studies, usually provides complete preoperative information for both partial AVSD and complete AVSD. Three-dimensional echocardiography is undergoing assessment in a number of centers and may ultimately help in the surgeon's understanding of AV valve morphology and in the planning of surgery. Three-dimensional echocardiography may be especially useful when reoperating for mitral regurgitation on a patient with a previously repaired AVSD because it can help define the etiology of the regurgitation.

Cardiac catheterization is required only when clinical evidence of pulmonary vascular disease exists, making operability questionable, or when additional major cardiac anomalies coexist. A left

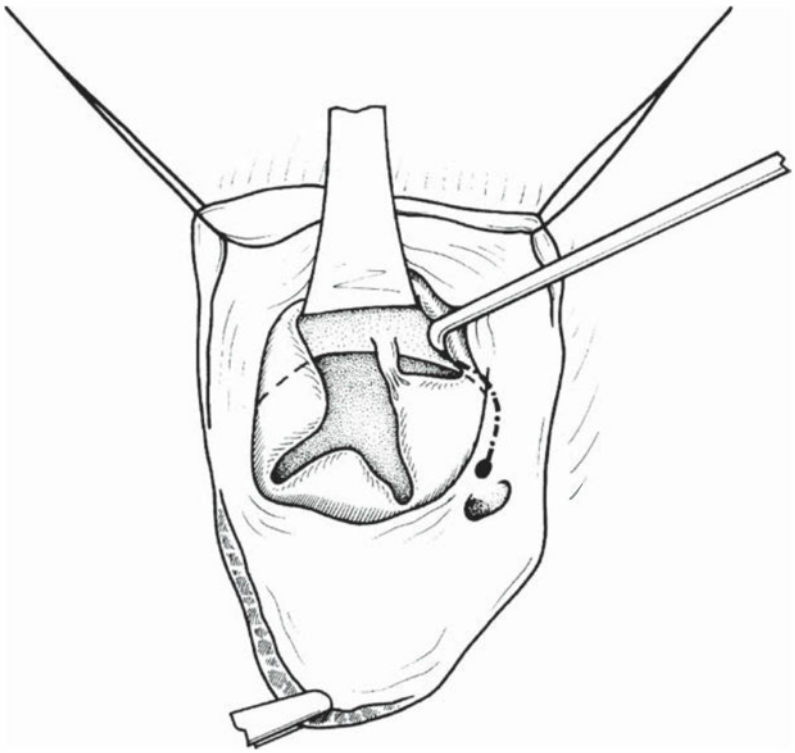


Fig. 86.2. A dashed line is used to indicate the location of the atrioventricular node and the bundle of His. The coronary sinus is also demonstrated.

ventriculogram in the anteroposterior projection shows a typical "goose-neck" deformity caused by the long, narrow left ventricular outflow tract, the lower boundary of which is made up of the superior bridging leaflet. Cardiac catheterization can also be used to measure pressures, flows, and resistances in the pulmonary and systemic circuits as well as the direction and the magnitude of shunting.

INDICATIONS

The natural history of untreated AVSD depends on the morphology of the lesion and dictates the indications and timing of surgical intervention. Partial AVSD without significant left AV valve regurgitation has a natural history similar to that of secundum ASD. Up to 15% of patients may develop high pulmonary arteriolar resistance in their adult life. The development of symptoms in adulthood often relates to the onset of atrial fibrillation. The natural history of partial AVSD with significant left AV valve regurgitation is much worse. These patients present earlier in life, and without surgical treatment, many may die in the first decade of life. Infants with complete AVSD have an even more malignant presentation, and without surgical correction, the majority die within the first year of life.

Patients with asymptomatic partial AVSD should be treated similarly to patients with secundum ASD. Elective repair is indicated before school age unless the patient develops symptoms of heart failure or failure to thrive. A minority of patients with partial AVSD and severe left AV valve regurgitation present with severe symptomatology in the first year of life and thus require earlier surgical intervention. Those few infants with severe left AV valve regurgitation who are asymptomatic should also undergo earlier surgical intervention.

Infants with complete AVSD should undergo elective correction between 3 and 6 months of life. In the past, the management of infants with complete AVSD and trisomy 21 has been somewhat controversial and has depended on the philosophy of the individual cardiologist, the cardiac surgeon, and the family. In our view, however, they should be treated exactly as those without trisomy 21. The surgical procedure of choice for both partial AVSD and complete AVSD is complete repair of the lesion as will be described. Pulmonary artery banding to palliate symptoms of congestive heart failure plays no role in the management of these lesions except in patients with complex associated cardiac anomalies or those with severely unbalanced hearts or functionally univentricular physiology with pulmonary overcirculation. Severe pulmonary infection may also be a relative indication for pulmonary artery banding.

Contraindications to surgery are based on fixed severe elevation of pulmonary vascular resistance. Pulmonary vascular resistance of >10 units/m^2 of body surface area (or a pulmonary-to-systemic resistance ratio of >0.7) represents a contraindication to repair. Elevated pulmonary vascular resistance of <10 units/m^2 (or a pulmonary-to-systemic resistance ratio of <0.7) represents an indication for more urgent surgical intervention. The assessment of elevated pulmonary vascular resistance should include cardiac catheterization under conditions of oxygen, nitric oxide, and prostacyclin to assess reversibility.

SURGICAL TECHNIQUE

After routine anesthesia, preparation, and draping, a standard median sternotomy is performed followed by a thymectomy. An eccentric pericardiotomy to the left is then performed, leaving a large piece of pericardium attached on the right for later use as a patch (Fig. 86.3). (All operative drawings presented here are viewed from the surgeon's perspective.) The aorta, ductus arteriosus, and superior vena cava (SVC) are then mobilized. Identification of the ductus is best achieved by first finding the "axilla" between the right pulmonary artery and the ductus, and then finding the "axilla" between the left pulmonary artery and the ductus using traction on the main pulmonary artery. Once the right and left pulmonary arteries have been formally identified, anything remaining inbetween must be the ductus. A silk ligature is passed around the ductus but is not yet tied, with care being taken to avoid injuring the ductus.

The lateral aspect of the SVC should be mobilized with scissors to avoid diathermy damage to the right phrenic nerve. Purse strings of 5-0 polypropylene are then placed into the aorta and the right atrial appendage. An additional, longitudinally oriented, narrow, rectangular purse string is placed on the SVC about 0.5 to 1.0 cm above the junction with the right atrium (Fig. 86.4). The aorta is then cannulated. We prefer the DLP (Medtronic, Grand Rapids, MI) aortic cannula because of its flexibility. Next, the inferior vena cava (IVC) angled metal Pacifico venous cannula (DLP) is placed temporarily into the right atrial appendage. Cardiopulmonary bypass is then established (Fig. 86.5), and the ductus arteriosus is ligated with the previously placed silk ligature. Once cardiopulmonary bypass is established, the patient is cooled. Some surgeons will cool to 24 or 25°C. Other surgeons do these cases at warmer temperatures such as 32°C. With warmer

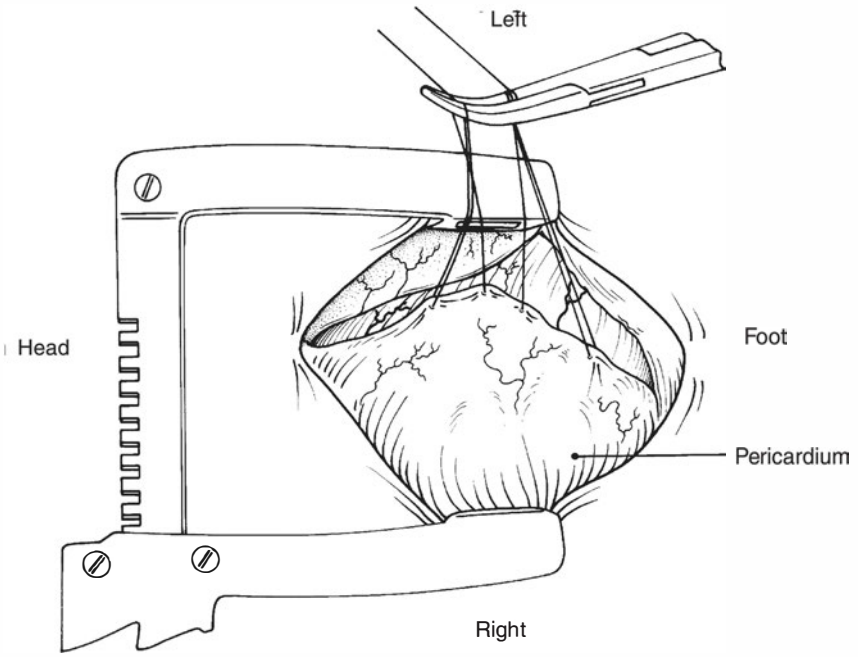


Fig. 86.3. An eccentric pericardiotomy to the left is performed leaving a large piece of pericardium attached to the right for later use as a patch. (Note: All operative drawings in this and subsequent figures are viewed from the surgeon's perspective.)

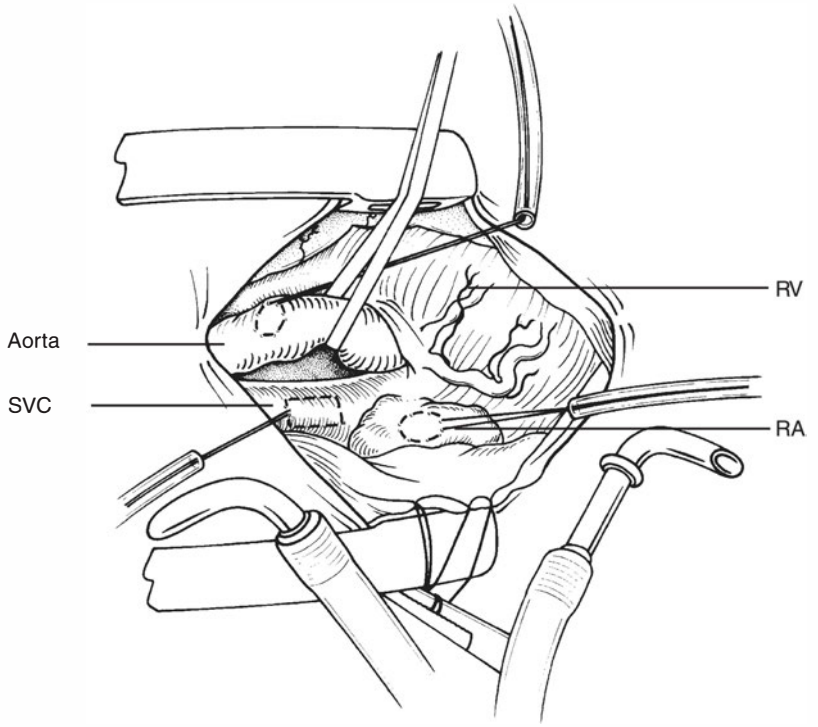


Fig. 86.4. Purse strings are demonstrated in the aorta, the right atrial appendage, and the superior vena cava (SVC). Note the position of the longitudinally oriented, narrow, rectangular purse string on the SVC about 0.5 to 1.0 cm above the junction with the right atrium (RA). RAA, right atrial appendage; RV, right ventricle; SVC, superior vena cava.

temperatures, the hematocrit is kept higher while on cardiopulmonary bypass.

The SVC is then grasped with two mosquito hemostats on each side of the previously placed purse string. The SVC is incised longitudinally with a scalpel, and the SVC is then cannulated with the second angled metal cannula (Fig. 86.6).

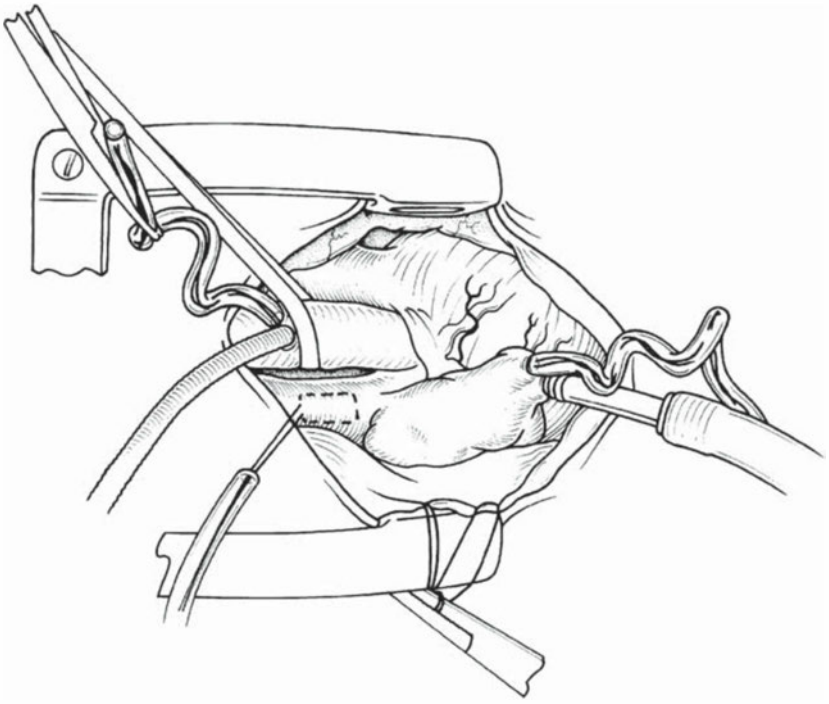


Fig. 86.5. The aorta and the right atrial appendage are both cannulated. Note that the inferior vena cava angled metal cannula is placed temporarily into the right atrial appendage. Cardiopulmonary bypass may now be established.

A vent is then inserted into the right atrial appendage after the IVC cannula is removed (Fig. 86.7). The pericardial reflection anterior to the IVC is then released, and the sub-diaphragmatic IVC is exposed down to the level of the first hepatic vein. A purse string (5-0 polypropylene) is then placed directly into the IVC below the pericardial reflection (Fig. 86.8), and the IVC is cannulated with the IVC angled metal cannula. Caval tapes are passed around the IVC and the SVC; nylon caval tapes may be used or silk suture may be used as caval tapes.

The aorta is cross-clamped, the two vena cavae are snared, the right atrium is opened parallel to the AV groove, and cardioplegic solution is instilled into the aortic root (Fig. 86.9).

A right-angled instrument is then passed through the opening of the right atrium into the left atrium and up into the right superior pulmonary vein. A No. 11 scalpel blade is then used to open the junction of the right superior pulmonary vein and the left atrium between the tips of the right-angled instrument (Fig. 86.10). The vent is then passed into the left atrium and secured into position with a 6-0 polypropylene purse-string suture.

Stay sutures are then applied to the atrial wall, and the anatomy of the AV valve is inspected (Fig. 86.11). Stay sutures of 6-0 polypropylene on an 8-mm needle are then applied to approximate the "kissing points" of the bridging leaflets (Fig. 86.12). The kissing points may be defined as the points of the superior bridging leaflet and the inferior bridging leaflet that (1) intuitively come together at the center of the superior bridging leaflet and the inferior bridging leaflets, (2) usually overlie the interventricular septum, and (3) can be identified as the midpoint between the left and right chordae on each bridging leaflet.

The Two-Patch Technique

We will first describe the following operative technique: *AVC (AVSD) repair, Complete (CAVSD), Two-patch technique*. Unwanted secondary chordae (which may limit exposure and whose function will partially be replaced by the new interventricular patch) can be divided. This permits elevation of a tethered bridging leaflet from the ventricular crest up to the plane of the annulus.

The size required for the VSD patch is then measured using black silk ligatures. Two pieces of black silk are trimmed. One silk measures the length of the patch by measuring the distance from the superior to the inferior margin of the annulus of the common AV valve in the plane of

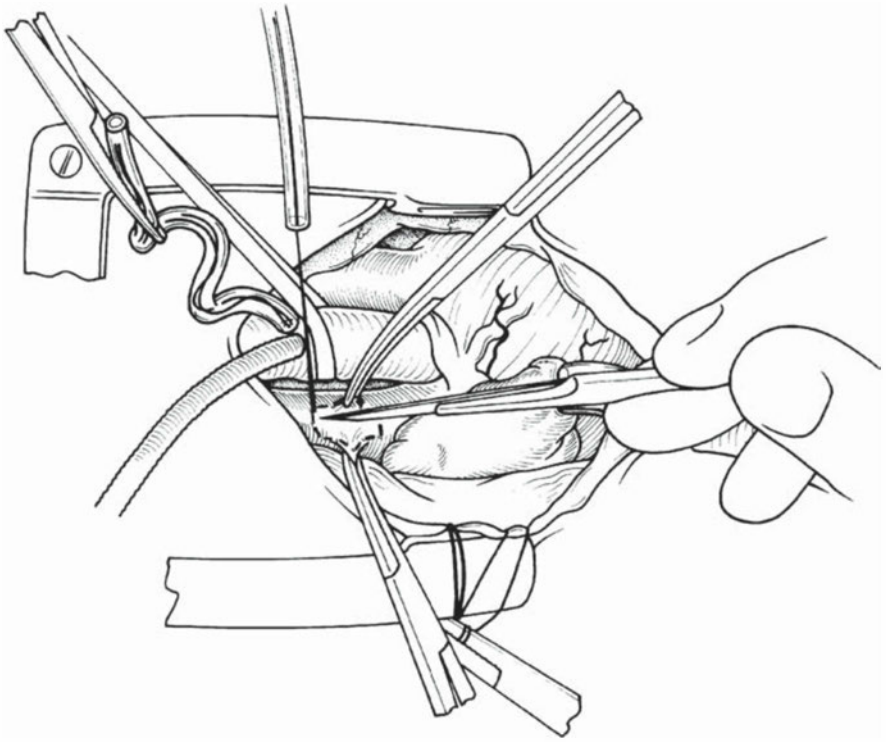


Fig. 86.6. The superior vena cava is grasped with two mosquito hemostats on each side of the previously placed purse string. The superior vena cava is incised longitudinally with a scalpel.

the interventricular septum (Fig. 86.13A). Inferiorly, extra length must be allowed to permit the patch to extend beyond the predicted position of the bundle. The second silk measures the depth of the patch by measuring the distance from the plane of the annulus of the freed common AV valve to the crest of the interventricular septum beyond the base of the VSD (Fig. 86.13B). The importance of this depth measurement is that it dictates the degree of elevation of the revised AV valve apparatus up to the plane of the annulus. However, the following operative technique that will be described later in this chapter supports the fact that the AV valve apparatus can be safely brought down to the crest of the interventricular septum at the base of the VSD: *AVC (AVSD) repair, Complete (CAVSD), VSD closed primarily (directly), and ASD closed with patch (Nunn/Wilson).*

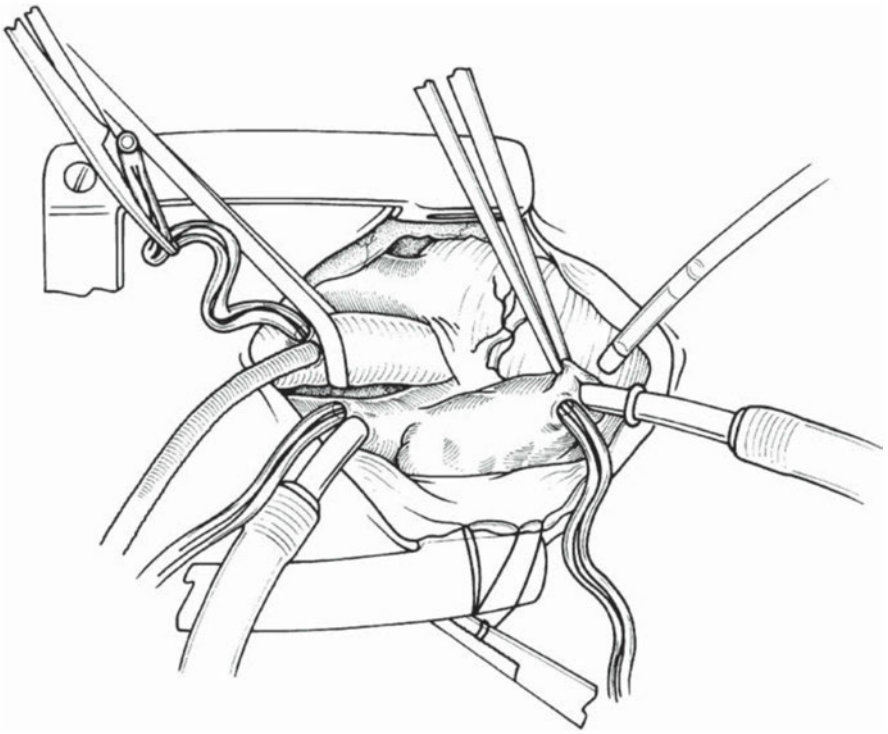


Fig. 86.7. The superior vena cava is cannulated with a second angled metal cannula. A vent is then inserted into the right atrial appendage after the inferior vena cava cannula is removed.

A patch of 0.4-mm polytetrafluoroethylene (Gore-Tex) is then trimmed to size using the previously cut silk sutures as guides (Fig. 86.13C). Alternatively, a patch of Dacron may be used. This patch is then sutured into position with a 5-0 polypropylene continuous suture. We start this suture inferiorly with the suture being brought through the right AV valve close to the annulus, well away from (inferior to) the position of the bundle (Fig. 86.14). It is easiest to place this first suture by going through the inferior bridging leaflet on the right side of the interventricular septum, passing the needle behind the right-sided chordae to the inferior bridging leaflet, and then passing the needle through the corner of the crescentic Gore-Tex patch. The needle is then taken back behind the chordae before positioning the suture deep into the muscle of the septum well away from the position of the bundle but close to the annulus. This will fix the patch in good position before commencing the running suture. This first suture is facilitated by retracting the inferior bridging leaflet inferiorly using a nerve root retractor. The running suture line is then brought superiorly along the right side of the septum, weaving behind the chordae as required. The conduction system is avoided inferiorly, and the aortic valve leaflets are carefully visualized and avoided superiorly. The 5-0 polypropylene suture is brought through the superior bridging leaflet at the annular margin and is placed on a rubber-shod clamp once the superior margin is reached (Fig. 86.15).

It is now necessary to septate the valve into a left and a right component, simultaneously preparing the atrial component of the patch. Interrupted horizontal mattress sutures of 6-0 polypropylene double-armed with 8-mm needles are then placed along the crest of the interventricular patch, with the surgeon bringing them out through the superior bridging leaflet and the inferior bridging leaflet, respectively, and then through a patch of autologous pericardium, not yet detached from the right side of the pericardium (Fig. 86.16). These

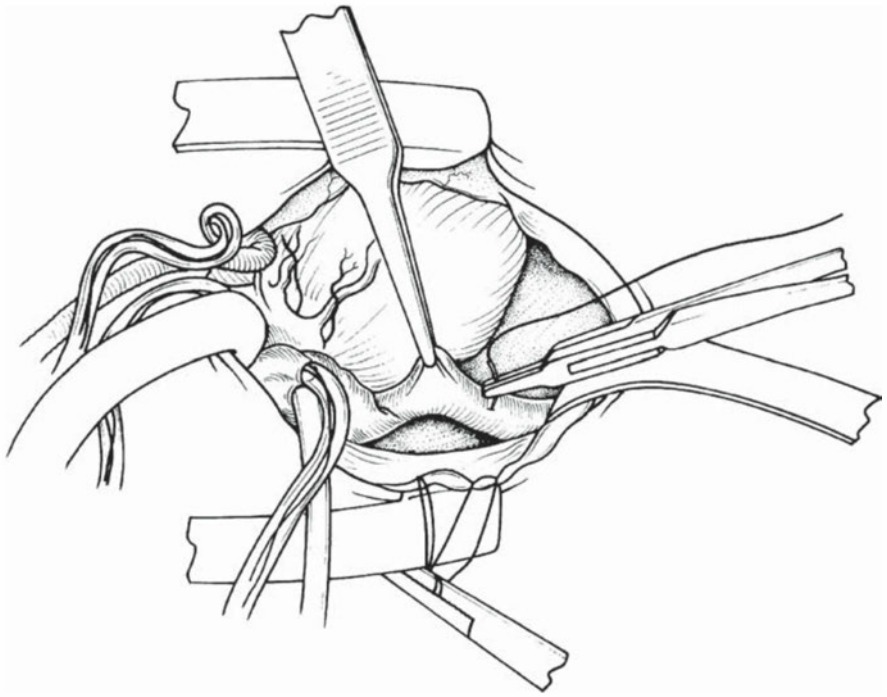


Fig. 86.8. The pericardial reflection anterior to the inferior vena cava is released and the sub-diaphragmatic inferior vena cava is exposed. A purse string (5-0 polypropylene) is then placed directly into the IVC below the pericardial reflection. The length of the IVC is exaggerated for clarity.

sutures should start inferiorly, beginning by retracting the inferior bridging leaflet once again with a nerve root retractor. The first of these sutures is placed through the VSD patch close to the inferior VSD running suture. The needle is then passed through the inferior bridging leaflet next to the inferior VSD running suture and, finally, is passed through the attached pericardial patch. The second arm of this first 6-0 polypropylene double-armed suture should then be placed a little bit farther along the crest of the Gore-Tex patch, with the surgeon bringing it in turn through the inferior bridging leaflet in a line between the inferior arm of the VSD running suture and the kissing-point stay suture. This line is important because it will represent the line of demarcation between the left and right AV valves (Fig. 86.15). The second arm of this first 6-0 polypropylene double-armed suture is then also passed through the attached pericardial patch and held in a rubber-shod clamp. The remaining interrupted horizontal mattress sutures of double-armed 6-0 polypropylene are then placed along the crest of the interventricular patch, with the surgeon bringing them out through the inferior and superior bridging leaflets, respectively, and then through the pericardial patch. We have found it to be a major advantage to keep the pericardium attached because this makes it easier to understand the relationships between the two AV valves and the atrial patch. The horizontal mattress sutures are then tied, and this brings the pericardium down to the ridge of the valve complex (Fig. 86.17). The pericardial patch is detached (Fig. 86.18A) and then swung anteriorly (Fig. 86.18B).

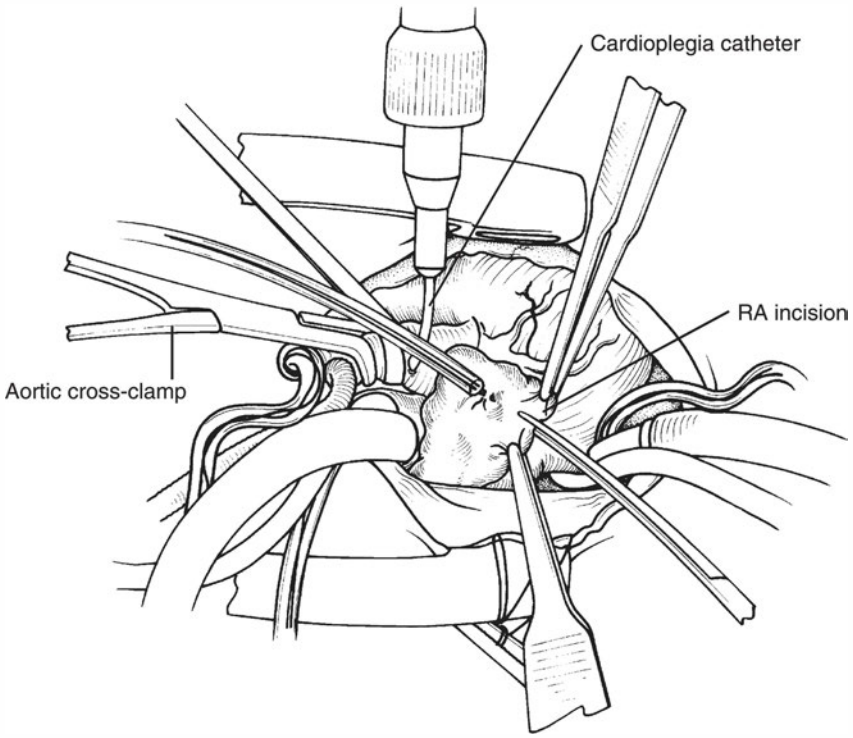


Fig. 86.9. The inferior vena cava is cannulated with the inferior vena cava angled metal cannula. Nylon caval tapes are passed around the inferior vena cava and the superior vena cava. The patient is cooled to 25°C. The aorta is cross-clamped, the two venae cavae snared, the right atrium opened parallel to the atrioventricular groove, and cardioplegic solution instilled into the aortic root.

Attaining Left Atrioventricular Valve Competence

1. The first important element in left AV valve repair is the size of the VSD patch. In prior publications, we stated: "Too big a patch and the valve will not coapt, too small a patch and the valve will become stenotic at annular level." However, the *Nunn/Wilson's* modified one-patch technique has shown us that too small a patch should not be problematic because the repair can be accomplished successfully with no VSD patch at all, as will be described below.
2. The cleft in the left-sided AV valve (or zone of abutment [or zone of apposition] between the left superior bridging leaflet and the left inferior bridging leaflet) is an unsupported commissure that needs to be closed to create a neoseptal leaflet akin to the aortic leaflet of a normal mitral valve. This closure of the "cleft" (or zone of apposition between the left superior bridging leaflet and the left inferior bridging leaflet) may be achieved using one or two horizontal mattress sutures of 6-0 polypropylene reinforced

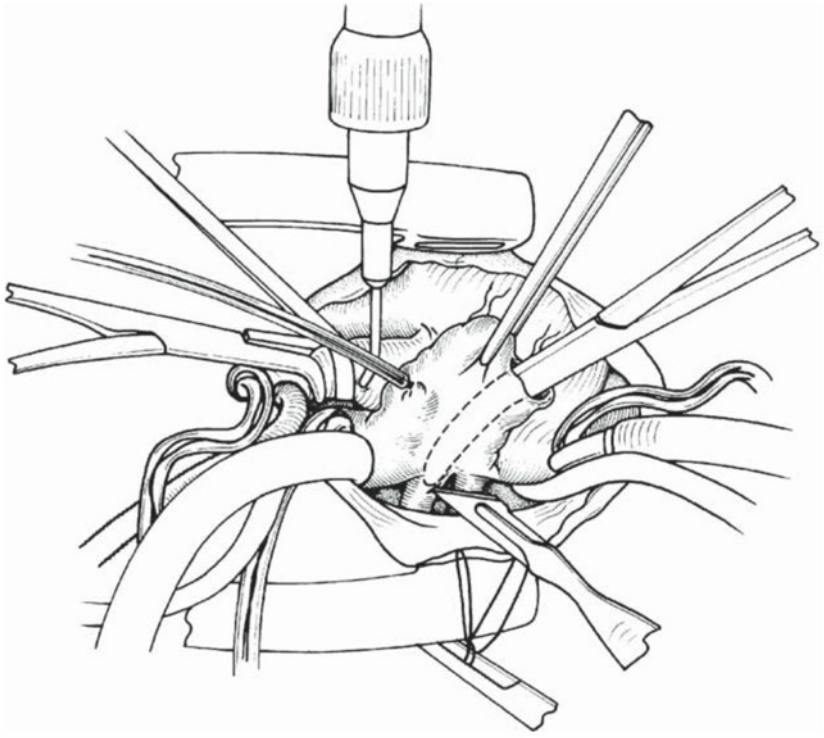


Fig. 86.10. A right-angled instrument is passed through the opening of the right atrium into the left atrium and up into the right superior pulmonary vein. A No. 11 scalpel blade then opens the junction of the right superior pulmonary vein and the left atrium between the tips of the right-angled instrument. The vent can then be passed into the left atrium and secured into position with a 6-0 polypropylene purse-string suture.

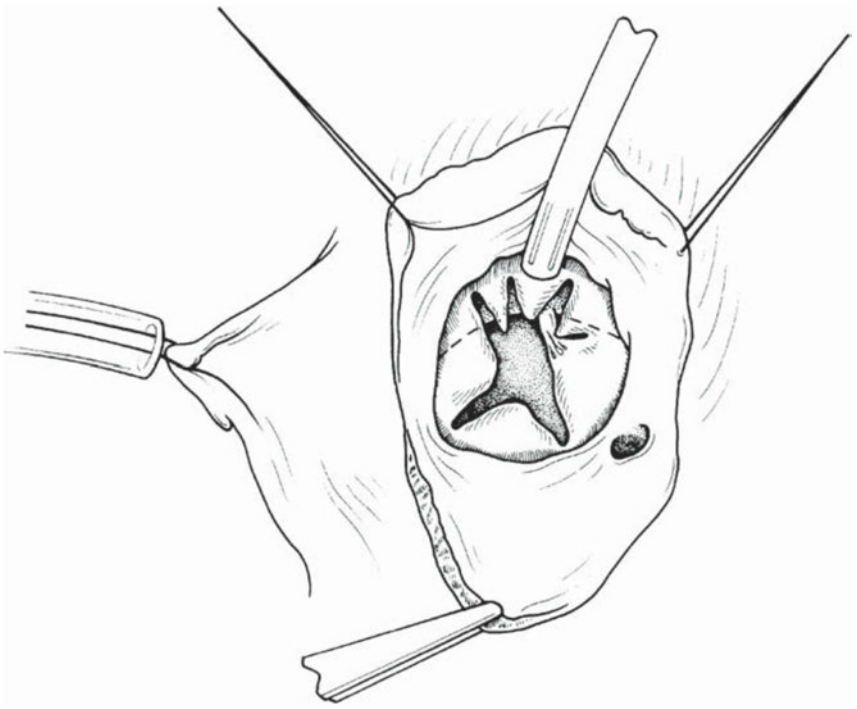


Fig. 86.11. Stay sutures are used to hold the atrial wall open to inspect the anatomy of the atrioventricular valve. The suction tip is useful as a retractor to examine the anatomy of the atrioventricular septal defect.

with autologous pericardial pledgets (Fig. 86.19). Alternatively, this closure of the "cleft" may be done with multiple non-pledgeted sutures of 6-0 polypropylene. The valve is then tested for competence by suspending the leaflets with saline (Fig. 86.20). All tension is released from all stay sutures. The autologous pericardial patch is held gently forward, and a 50-ml syringe is used to inject ice-cold saline via a size 10 nasogastric tube through the left AV valve. (Richard Jonas has described floating the valve with cold blood cardioplegia.) The valve should float up nicely and demonstrate no regurgitation (Fig. 86.21). If necessary, an additional 6-0 polypropylene suture (possibly reinforced with autologous pericardial pledgets) is placed in the cleft near the central orifice.

3. If the valve continues to leak, the problem should not be ignored. First, the location of the leak is identified: central orifice, separate orifice, or a gap between scalloped components of the bridging leaflets. If a central leak exists and the cleft is already reapproximated to the chordae, an annuloplasty, such as DeVega annuloplasty, may be performed at whatever commissures are visible. If the valve is leaking at the cleft, an annuloplasty will not help. The leaflets and the cleft must be dealt with first. Annuloplasty is supportive and cannot result in primary competence in this morphology.
4. Paradoxically, it may be better in certain circumstances not to close the cleft and to leave the superior bridging leaflet and the inferior bridging leaflet unapproximated. This is the case when the mural leaflet is tiny or absent as in the parachute arrangement of left AV valve support apparatus. There is always a balance between stenosis and regurgitation in these circumstances, and it is important to size the left AV valve using Hegar dilators to ensure good baseline information for future follow-up, and occasionally to guide the removal of an overtight stitch in the cleft, resulting in a degree of "acceptable" regurgitation.

The ASD patch is then sutured in place with continuous 5-0 polypropylene suture. The coronary sinus may be left on the left or right atrial side. If there is a large ASD or an additional secundum ASD, we often keep the coronary sinus on the left side of the heart (Fig. 86.22). It is essential to place the coronary sinus on the right side when there is a left-sided SVC. We also try to keep the coronary sinus on the right when we are concerned

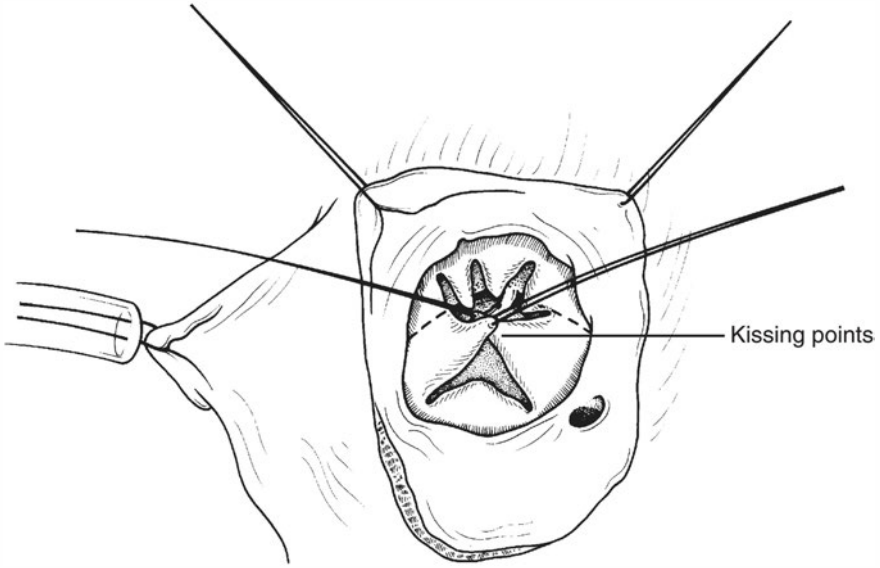


Fig. 86.12. Stay sutures of 6-0 polypropylene on an 8-mm needle are then applied to approximate the "kissing points" of the bridging leaflets.

about possible postoperative left AV valve dysfunction because a high left atrial pressure may result in high coronary sinus pressure.

During closure of the ASD, care is taken to ensure that the IVC remains on the right side and that the conduction system is carefully avoided. The first stitch of the continuous 5-0 polypropylene suture used to secure the atrial pericardial patch is placed on the inferior aspect of the ASD next to the previously placed VSD running suture currently held in a rubber-shod clamp. This first stitch is placed in the same way whether the coronary sinus will be placed on the left or the right, and this stitch is used to secure the VSD suture. (Alternatively, the VSD suture can be used to suture the ASD patch into place.) If the coronary sinus is to go on the left, this continuous polypropylene suture is then used to stitch the pericardial ASD patch along the atrial wall well inferior to the bundle and out around the coronary sinus, avoiding the Eustachian valve and keeping the IVC on the right. If the coronary sinus is to go on

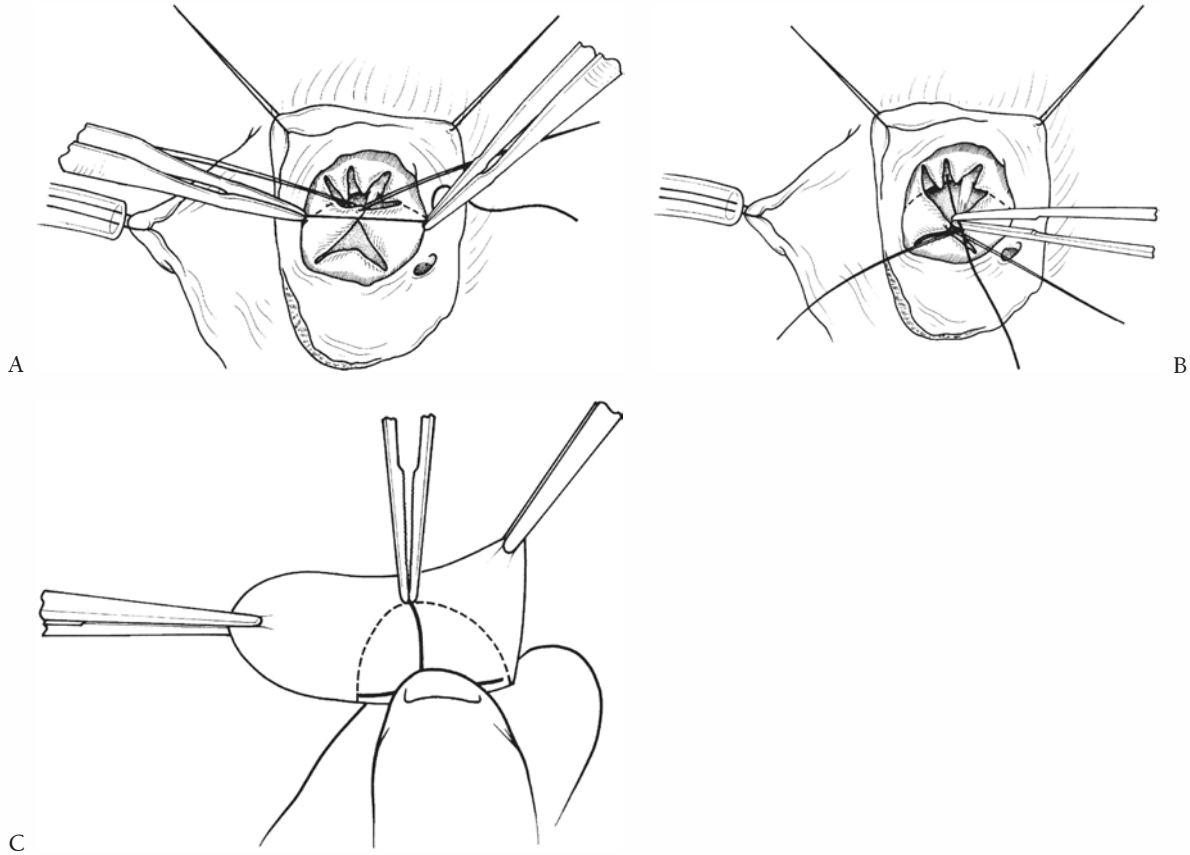


Fig. 86.13. (**A**) A piece of black silk is used to measure the length of the patch by measuring the distance from the superior to the inferior margin of the annulus of the common atrioventricular valve in the plane of the interventricular septum. Inferiorly, extra length must be allowed to permit the patch to extend below the predicted position of the bundle. (**B**) A second piece of silk measures the depth of the patch by measuring the distance from the plane of the annulus of the freed common atrioventricular valve to the crest of the interventricular septum beyond the base of the ventricular septal defect. (**C**) A patch of 0.4-mm polytetrafluoroethylene (Gore-Tex) is then trimmed to size using the previously cut silk sutures as guides.

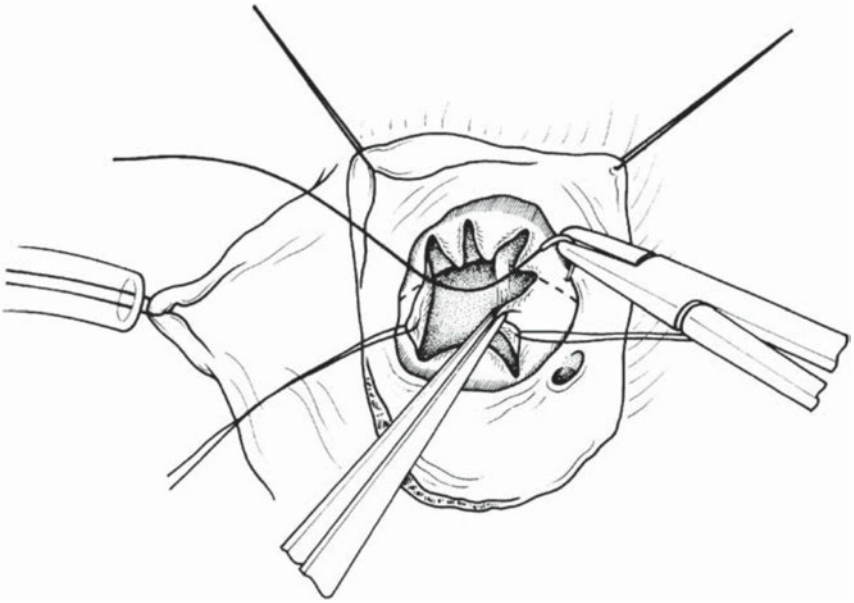


Fig. 86.14. The first suture begins inferiorly with the suture being brought through the right atrioventricular valve close to the annulus, well away from and inferior to the position of the bundle.

the right, this continuous 5-0 polypropylene suture is then used to stitch the pericardial ASD patch along the left AV valve itself at its annular margin and then onto the septum beyond the bundle. The continuous 5-0 polypropylene suture may be interrupted at the top of the ASD patch to prevent this patch from becoming constrictive. The ASD pericardial patch is trimmed as the suture line progresses and often ends up smaller than originally anticipated.

Cardioplegic solution is administered every 15 to 25 minutes throughout the cross-clamp period. After the atrial patch is sewn into position (Fig. 86.23) and all communications between the right heart and the left heart are definitely closed, the aortic cross-clamp is removed and the heart is deaired through the aortic root and the right superior pulmonary vein. The right atrium is then closed with a double layer of running 6-0 polypropylene suture on a 13-mm needle with the cross-clamp off during rewarming. Once the child is warm, ventilation is begun, deairing is completed, and the vent is removed. A left atrial pressure monitoring line may be inserted via the right superior pulmonary vein or via the left atrial appendage. The child is then weaned from cardiopulmonary bypass and modified ultrafiltration is carried out. We always use intraoperative echocardiographic assessment to define the functional status of the repair. We will redo any part of the procedure as indicated by the echocardiographic study. Protamine is then administered. Atrial and ventricular pacing wires are placed and chest drains are placed.

If the left AV valve is potentially regurgitant or dysfunctional or if any other reason exists to increase the likelihood of a redo sternotomy, a 0.1-mm-thick, low-porosity, expanded polytetrafluoroethylene pericardial membrane (Preclude Pericardial Membrane, formerly called the Gore-Tex Surgical Membrane; W. L. Gore and Associates, Flagstaff, AZ) is placed. Routine closure then follows.

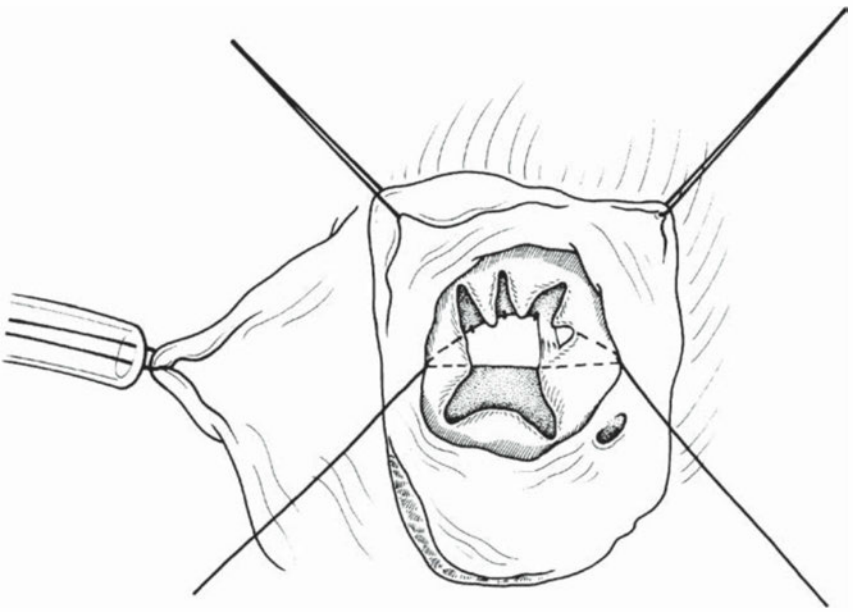


Fig. 86.15. After the running suture line attaching the Gore-Tex patch to the interventricular septum is completed, the 5-0 polypropylene suture is placed on a rubber-shod clamp at the superior margin.

An Alternative Operative Strategy: Ventricular Septal Defect Closed Primarily (Directly) and Atrial Septal Defect Closed with Patch (Nunn/Wilson)

The modified one-patch technique involves primarily closing the VSD and then using a patch to close the ASD. Cannulation, cardiopulmonary bypass, and administration of cardioplegia are the same as in the two-patch technique. Using 6-0 polypropylene double-armed suture, multiple pledgeted horizontal mattress sutures are placed from the right side of the ventricular septum just apical to the crest of the VSD, with the surgeon bringing them out through the inferior and superior bridging leaflets, respectively, and then through the pericardial patch that will close the ASD. Again, it is advantageous to keep the pericardium attached on the right side because this makes it easier to understand the relationships between the two AV valves and the atrial patch. The horizontal mattress sutures are then tied, and this closes the VSD and brings the pericardium down to the ridge of the valve complex. The pericardial patch is detached and then swung anteriorly. The "cleft" of the left AV valve is then closed as described in the two-patch technique, and the ASD is also closed as in the two-patch technique. Backer and colleagues reported that "The modified single-patch technique produced results comparable with the two-patch technique in younger patients with similarly sized VSDs. Furthermore, the modified single-patch technique was performed with significantly shorter cross-clamp and cardiopulmonary bypass times."

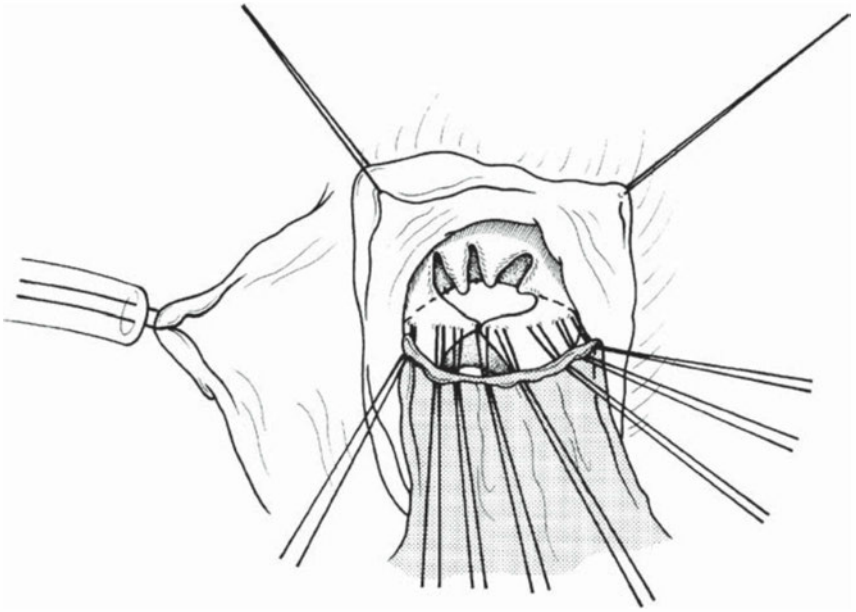


Fig. 86.16. Interrupted horizontal mattress sutures of 6-0 polypropylene double-armed with 8-mm needles are placed along the crest of the interventricular patch, with the surgeon bringing them out through the superior and inferior bridging leaflets, respectively, and then through a patch of autologous pericardium not yet detached from the right side of the pericardium. The chordae are omitted for clarity.

MANAGEMENT OF UNUSUAL VARIANTS OF COMPLETE ATRIOVENTRICULAR SEPTAL DEFECTS

Atrioventricular Septal Defect with Tetralogy of Fallot

In tetralogy of Fallot with AV canal, the common AV junction occurs in the setting of deviation of the outlet septum, producing varying degrees of aortic override. Right ventricular outlet obstruction will be present at variable levels and will have to be dealt with surgically.

The deviation of the outlet septum has consequences for the morphology of the leaflets of the AV junction and the shape of the ventricular septal crest.

In tetralogy of Fallot with AV canal, the superior bridging leaflet is always free-floating, enabling the left ventricle to access the deviated aorta. Consequently, this combination of defects does not occur in the setting of partial AVSD.

Given that such patients have a protected pulmonary vascular bed, operation is still often deferred to 2 to 3 years of age. Although the valve is larger in these older patients, valvar function and dysplasia may be paradoxically worse in the setting of chronic regurgitation with thickened dysplastic leaflets. A competent repair may, therefore, be more challenging and crucial, especially of the right AV valve, the competence of which is vital for good right ventricular function postoperatively. Right AV valve annuloplasty and/or commissuroplasty are commonly employed as a further, supportive measure for the right AV valve.

The ventricular septal patch must be tear shaped rather than elliptical, taking account of the deviated outlet septum (Fig. 86.24). The aortic margin of the patch may be difficult to access through the right atrium but can be approached through the right ventricular outflow tract that may be opened at the time of assessment of the pulmonary valve and infundibulum.

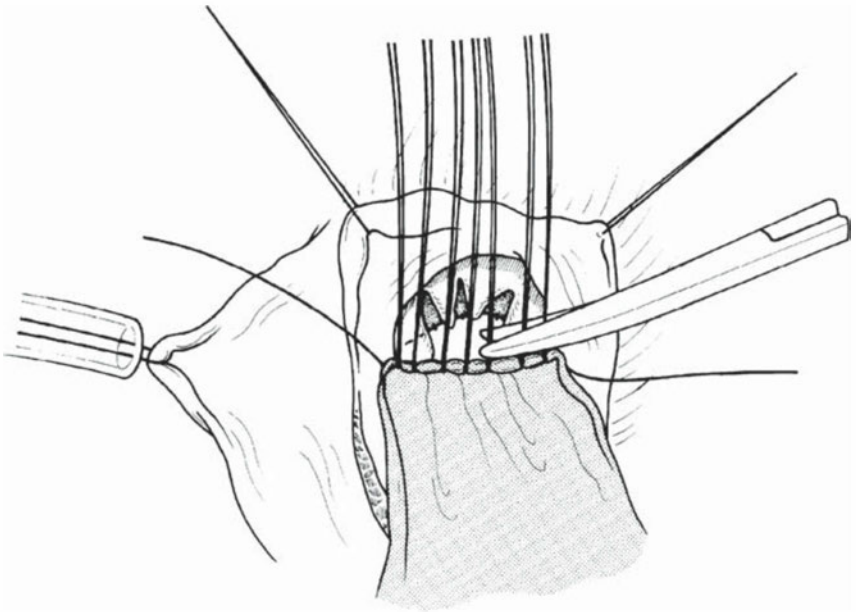


Fig. 86.17. The horizontal mattress sutures are tied, with the surgeon bringing the pericardium down to the ridge of the valve complex.

Unbalanced Atrioventricular Septal Defect

Unbalanced AVSD is difficult to define. It could be said that there is ventricular imbalance when the ventricular cavities are of unequal size or when the common AV junction is committed preferentially to one ventricle over the other. These two problems often coexist. Not only may the overall ventricular cavity be small, but it may also lack one segment of the normal tripartite ventricular morphology, such as the apical trabecular segment.

Severe imbalance of either ventricle is approached with a univentricular, Fontan, strategy, but the cut-off ratio of ventricular size that defines which strategy should be used remains undefined. This challenge is compounded by difficulty in measuring ventricular volumes. One common approach is "eye-balling" the ventricular size with two-dimensional echocardiography; however, three-dimensional echocardiography and magnetic resonance imaging might have a future role in the decision-making process.

A small left ventricle may coexist with a solitary papillary muscle arrangement (the so-called parachute valve), left ventricular outflow tract obstruction, or coarctation, as in the setting of Shone's syndrome. A small right ventricle may be seen with right

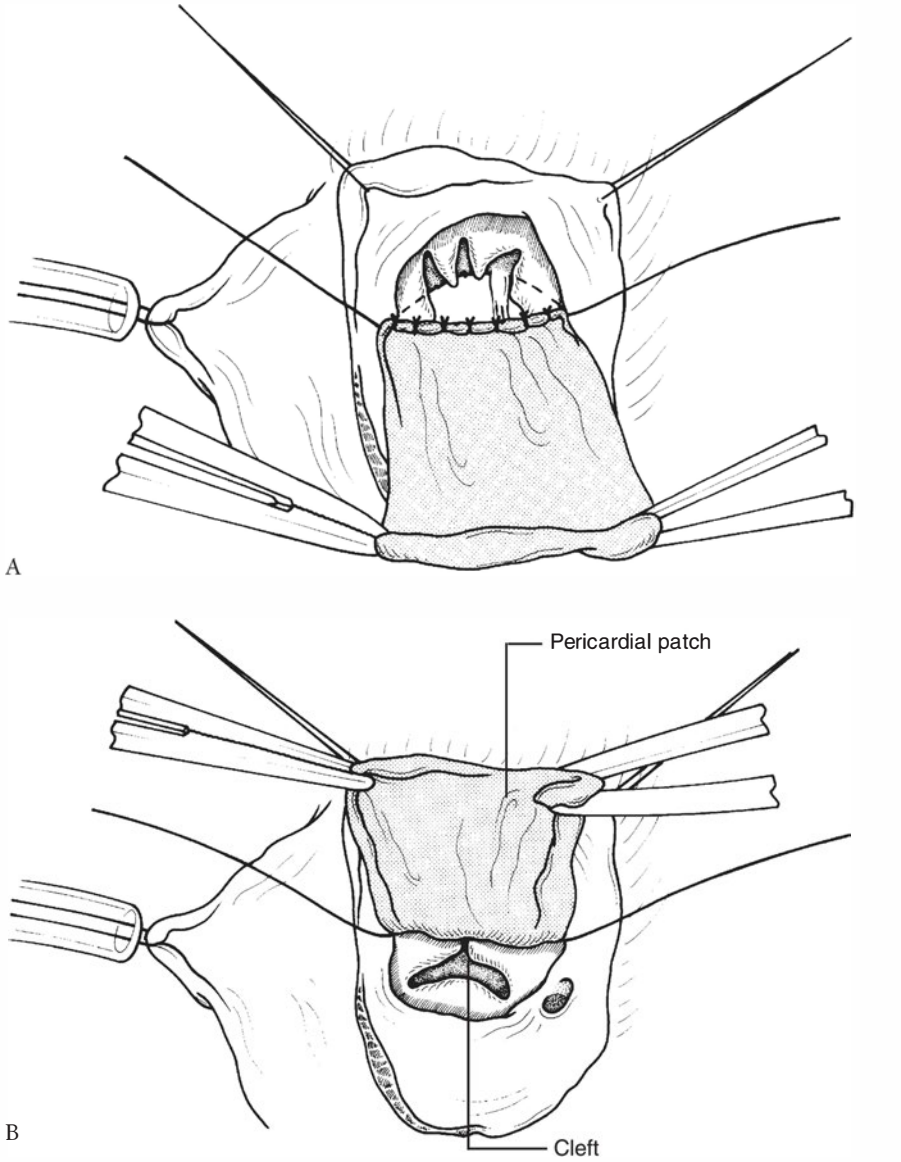


Fig. 86.18. (**A**) The pericardial patch is detached. (**B**) The pericardial patch is then swung anteriorly. This allows visualization of the "cleft" in the left atrioventricular valve. This cleft may be seen as the zone of abutment of the left superior bridging leaflet and the left inferior bridging leaflet.

ventricular outflow tract obstruction or after previous pulmonary trunk banding.

The approach to the small ventricle must be individualized according to a common morphologic algorithm. Not only must the size of the cavity be assessed at operation but so must the AV valve ring size and the state of the papillary muscles. If subjectively little of the common AV valve is committed to one ventricle, then a biventricular repair will not possible irrespective of the cavity size. Thus, the surgeon must inspect the ventricles at the annular, valvar, and subvalvar levels to determine whether a biventricular approach is feasible. If a biventricular approach is chosen, the so-called cleft must often be left open to prevent left AV valvar stenosis.

Is it possible to "recruit" cavity volume through the division of muscle bundles, in a manner similar to the ventricular overhaul utilized in patients with pulmonary atresia and intact ventricular septum? This approach is being investigated. One may closely inspect the ventricular cavity for such bundles with the view of dividing them, so long as the AV valve ring is of good size and reasonably balanced.

Meryl S. Cohen, Jeanne M. Baffa, David M. Overman and colleagues are leading an initiative by the Congenital Heart Surgeons' Society to study patients with unbalanced AVSD. They have developed echocardiographic tools to help evaluate patients with unbalanced AVSD. These tools accurately identify unbalanced AVSD and also bring into focus a zone of transition from anatomies that can support a biventricular end state and those that cannot. The atrioventricular valve index (AVVI) is expressed as the smaller AV valve area over the larger AV valve area, so that left-dominant unbalanced AVSD is expressed as right AV valve area/left AV valve area and right-dominant unbalanced AVSD the inverse. The modified AVVI is derived by dividing left AV valve area/total AV valve area. Thus, all forms of unbalance exist on a continuum from 0.0 to 1.0, with 0.0 having no left AV valve area, 1.0 having all left AV valve area, and 0.5 having exactly equal left and right AV valve areas. These tools may help determine the suitability of a given heart for univentricular or biventricular management strategies. Malalignment of the AV junction and the associated abnormalities of inflow physiology are critical in this decision-making process.

Accessory Left Atrioventricular Valvar Orifice

Accessory left AV valvar orifice is an unusual variant that is commonly cited as a risk factor for postoperative mortality and left AV valvar regurgitation.

In these cases, the larger AV valvar orifice occurs in association with a smaller orifice that is almost always located at the junction of the inferior bridging leaflet and mural leaflet. Developmentally, these two leaflets may have failed to separate or may have fused at their tips, producing a small accessory orifice along their line of closure.

At operation, this orifice usually should not be closed because it is nearly always supported by cords and therefore is competent. Closure can result in left AV valvar stenosis. For similar reasons, the zone of apposition in the main orifice must be closed carefully, ensuring that there is no resulting stenosis.

Deficiency of Atrioventricular Valvar Tissue

Deficiency of AV valvar tissue is a difficult problem and can result in "on-table mortality." Although any of the leaflets may be deficient, this pathology almost always results from deficiency of the inferior bridging leaflet over the ventricular septum. Thus, the surgical approach must be tailored according to the degree and pattern of this deficiency. Here, annuloplasty may be particularly helpful in improving the degree of central coaptation of the other leaflets. Some degree of resulting valvar stenosis has to be tolerated, given that the

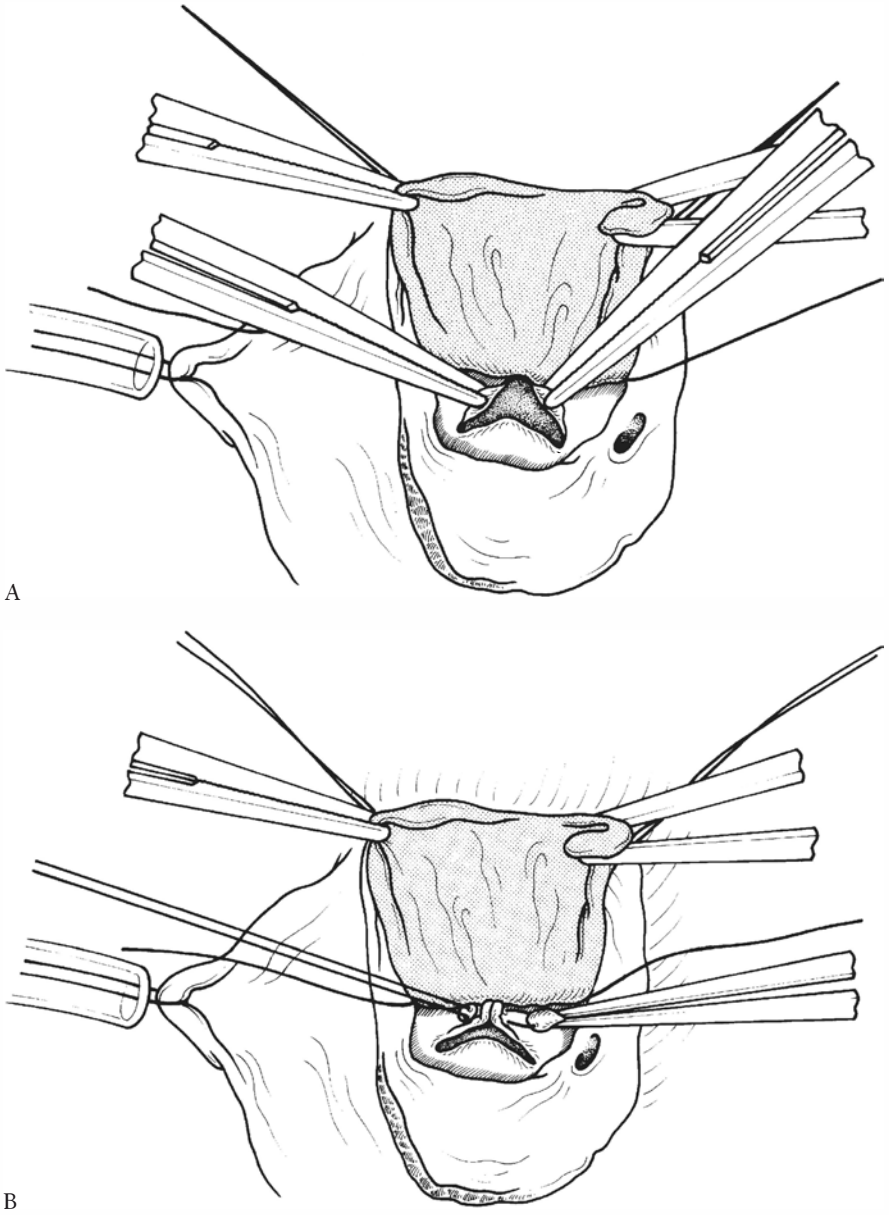


Fig. 86.19. (**A**) The cleft in the left atrioventricular valve is demonstrated. (**B**) One or two horizontal mattress sutures of 6-0 or 7-0 polypropylene reinforced with autologous pericardial pledgets are then used to close the cleft in the left atrioventricular valve.

unwanted alternative is valvar replacement at a very young age.

The Role of Atrial Septal Defect Patch Fenestration

In all of the foregoing instances, the ASD may be closed with fenestration. This technique has the advantage of acting as a "blow-off" (or "pop-off") valve when the ventricle is small, improving the postoperative hemodynamics and salvaging a difficult biventricular repair in this setting. It also gives the chance for the AV valve and ventricle to grow, redressing some of the imbalance and potentially improving AV valve function over time.

A hole is punched in the middle of a Gore-Tex patch that is sutured to the margins of the ASD or atrial septectomy. This not only prevents widening of the fenestration that can occur with an incision in autologous pericardium but also permits transcatheter closure of the hole at the appropriate postoperative time.

SURGICAL MANAGEMENT OF PARTIAL ATRIOVENTRICULAR SEPTAL DEFECT

The surgical management of partial AVSD has many similarities with that of complete AVSD, and repetition of the principles is not required. However, there are important differences that need to be highlighted.

Cardiopulmonary Bypass

Unlike complete AVSD where the patient is typically cooled to somewhere between 24 and 32°C, as described above, repair of partial AVSD is typically performed at a warmer temperature. Some will cool to 28°C, while others will repair partial AVSD at temperatures of >32°C. Some advocate normothermic repair partial AVSD and repair partial AVSD at 37°C. Cannulation is as for complete AVSD, and cardioplegia is used.

Repair

We assess the defect carefully using an approach similar to that described for repair of complete AVSD. Pericardial patch preparation is similar to that described for repair of complete AVSD. Stay sutures are not required, but attention should be paid to the quality of the valvar tissue above the crest of the interventricular septum. If the valvar tissue is judged to be of good quality (strong, thick, holding sutures well), we sometimes use a continuous suture of 5-0 polypropylene to secure the patch along the line of division between the right and left components of the AV valve. If the valvar tissue is of poor quality (friable, thin, deficient, or holding sutures poorly), we prefer an interrupted suture technique using an appropriately sized suture as horizontal mattress sutures. If the valvar tissue is particularly bad, we often take a strip of autologous pericardium from the left side of the patient and use this as a buttress to the sutures along the valvar leaflets at the crest of the septum, creating a very robust sandwich (pericardium to valve to pericardium). At the inferior margin, the bundle is avoided, and the coronary sinus is positioned exactly as for complete AVSD. After the patch has been attached to the crest of the septum, the patch is swung anteriorly and the left AV valve assessed and repaired as for complete AVSD.

SURGICAL MANAGEMENT OF TRANSITIONAL (INTERMEDIATE) ATRIOVENTRICULAR SEPTAL DEFECT

The surgical management of transitional AVSD has many similarities with that of complete AVSD and partial AVSD, and again, repetition of the principles is not required. Cannulation, cardiopulmonary bypass, and administration of cardioplegia are the same.

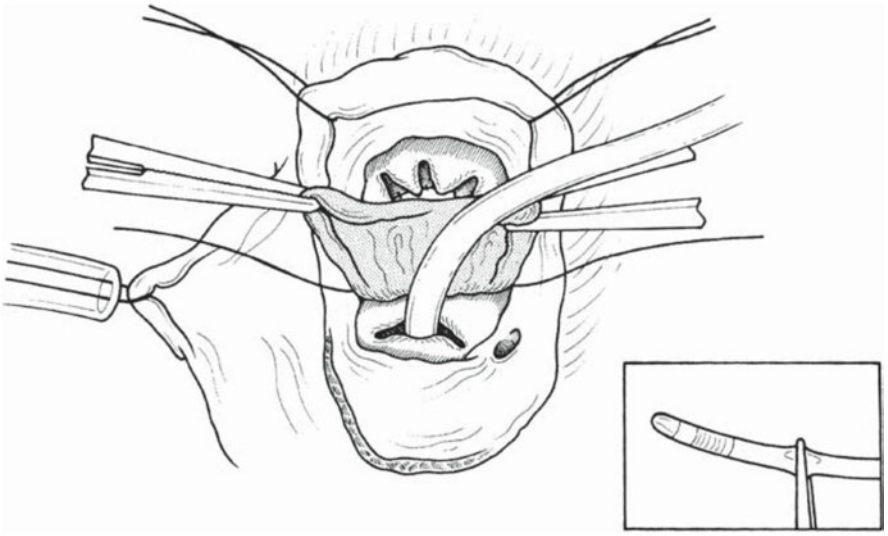


Fig. 86.20. The valve is then tested for competence by suspending the leaflets with saline. The inset demonstrates the catheter used to inject the saline to float the valve.

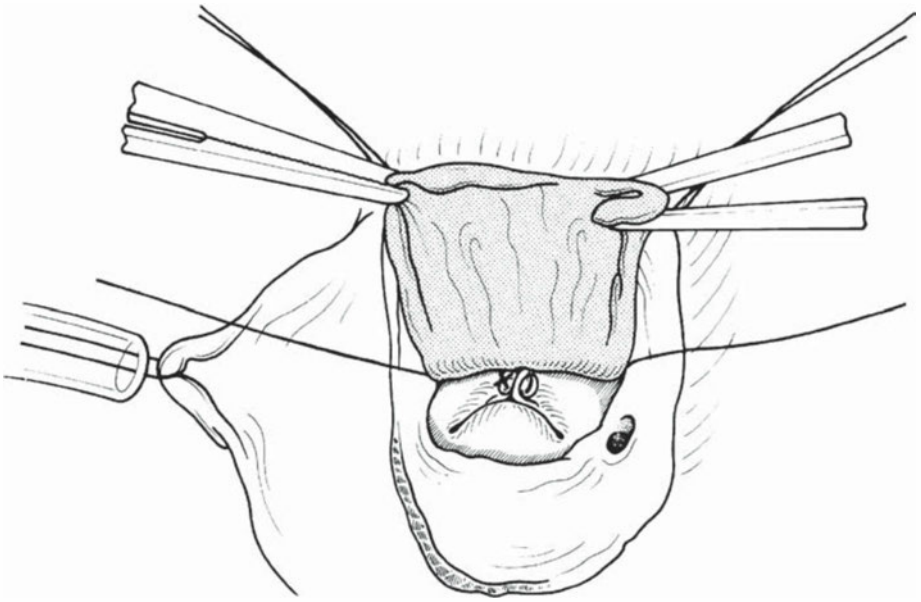


Fig. 86.21. The valve should float up nicely, demonstrating no regurgitation.

However, there are important differences that need to be highlighted. The VSD is often quite small and may be closed with one or more 6-0 polypropylene double-armed suture placed as pledgeted horizontal mattress sutures from the right side of the ventricular septum to the AV valve. The repair of the cleft of the left AV valve may be challenging because the AV valvar tissue may be scarred and dysplastic. It is important to spend the necessary time to assess this potentially dysplastic left AV valvar tissue prior to reconstruction. Closure of the primum ASD is similar to that described for partial AVSD.

SURGICAL COMPLICATIONS AND POSTOPERATIVE CARE

Improved modern surgical techniques and modern intensive care units have drastically reduced the operative mortality for these procedures. Most centers report operative mortality of <1% for surgical intervention for partial AVSD defects. This mortality is even less if the partial AVSD is not associated with significant left AV valve regurgitation. Operative mortality for complete AVSD is <3%, and most major centers report operative mortality of <2% for this problem. The operative risk does increase in several subsets of patients, including those with severe AV valvar incompetence, those with hypoplasia of one of the ventricles, and those with pulmonary vascular disease.

Potential postoperative complications include pulmonary hypertensive crisis, left AV valvar insufficiency, and heart block. Children with significant preoperative left AV valvar regurgitation and older children with complete AVSD are at risk of postoperative pulmonary hypertensive crises. The concern for pulmonary hypertension leads to certain fundamental postoperative management principles. Children believed to be at risk for postoperative pulmonary hypertensive problems, including children in the range of 6 to 9 months (or older) undergoing repair of complete AVSD, fall into a late-extubation group. These children may be managed with a pulmonary artery line and a left atrial pressure monitoring line in situ and are often kept sedated over the first 48 hours after surgery. They are monitored for pulmonary hypertension and are in a position in which pulmonary hypertensive crises can be appropriately managed. Our first-line therapy, after conventional ventilatory support, is inhaled nitric oxide (2 to 20 ppm). Phenoxybenzamine, a long-lasting α-adrenergic blocking agent, has been used in these patients prophylactically. It is given before cardiopulmonary bypass and during rewarming at doses of 1 mg/kg. It is also given postoperatively at doses of 0.5 mg/kg per dose every 8 to 12 hours until extubation. These children remain sedated and monitored. If a pulmonary hypertensive crisis occurs, appropriate interventions can be initiated. In the current era, the use of milrinone plus or minus nitric oxide is all that is usually needed to manage these patients. With milrinone, a pulmonary hypertensive crisis is less likely to occur, and when a pulmonary hypertensive crisis does occur, nitric oxide is the first therapeutic intervention and is usually effective. Additional useful interventions in these patients include increased sedation, increased supplemental oxygen, hyperventilation to a CO_2 pressure of <25 mmHg or <3.5 kPa, and possible pharmacologic paralysis. Traditional strategies for the management of pulmonary hypertensive crisis are now rarely needed; these traditional interventions include intravenous nitroglycerin or sodium nitroprusside, intravenous aminophylline, and intravenous prostacyclin administration. If a pulmonary hypertensive crisis occurs, a 24-hour period of stability is required before attempts are made to wean the patient from the ventilator. If no pulmonary hypertensive crises

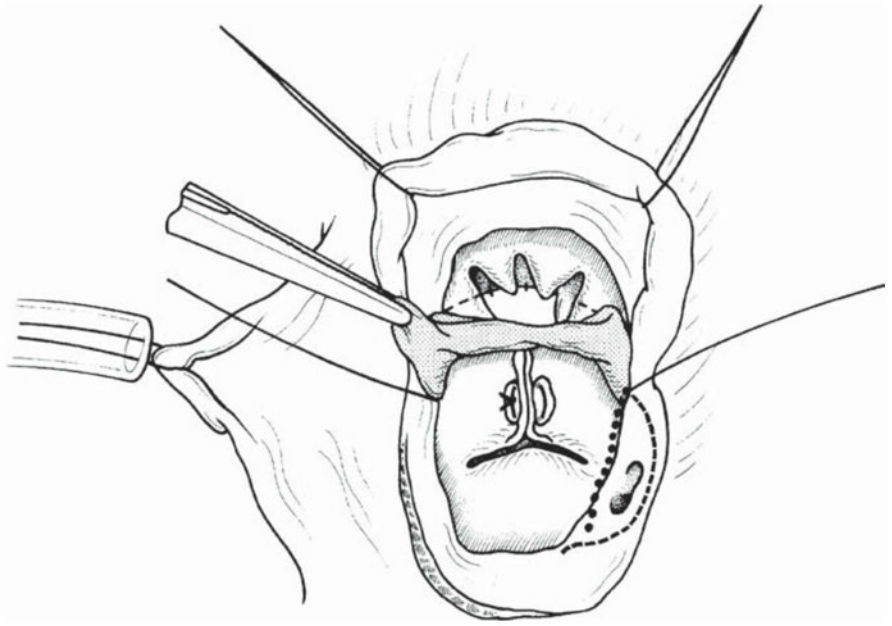


Fig. 86.22. The atrial septal defect pericardial patch is then sutured in place using continuous 5-0 polypropylene suture. The coronary sinus may be left on the left or right atrial side. The dotted line demonstrates the suture line if the coronary sinus is to be placed on the right atrial side. The dashed line demonstrates the suture line if the coronary sinus is to be placed on the left atrial side.

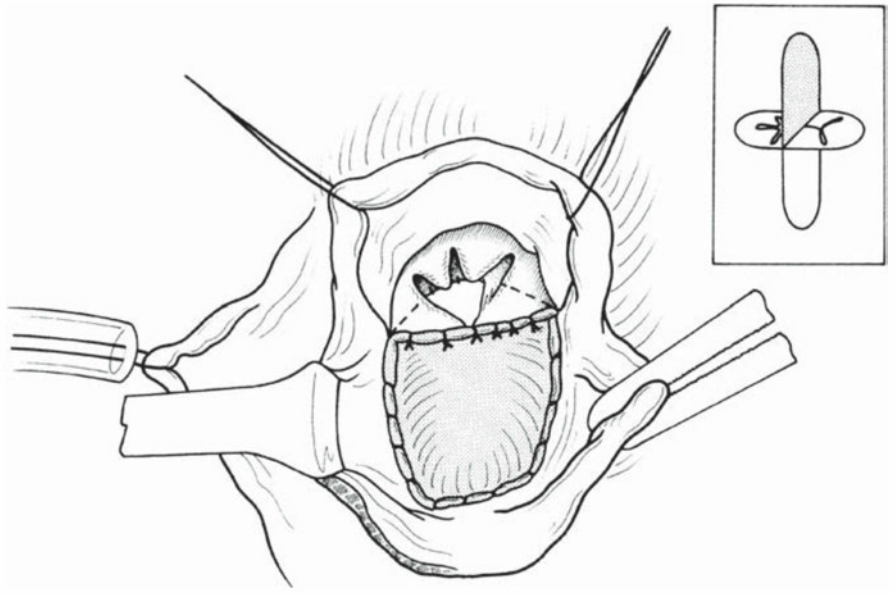


Fig. 86.23. The atrial patch is sewn into position. The inset demonstrates the relationship between the atrial patch, the ventricular patch, the right atrioventricular valve, and the left atrioventricular valve.



Fig. 86.24. In atrioventricular septal defect with tetralogy of Fallot, the shape of the ventricular septal patch must be tear shaped rather than elliptical, taking account of the deviated outlet septum.

occur, children in this group (older patients undergoing repair of complete AVSD who are at risk for the development of pulmonary hypertensive crises) are extubated at approximately 48 hours. Younger infants undergoing repair of complete AVSD in the range of 3 to 6 months, as well as children with partial AVSD and transitional AVSD, are often not at risk for pulmonary hypertensive crises; consequently, these children are often extubated sooner, often either on the operating theater table immediately after surgery or within the first 24 hours.

Postoperative left AV valvar insufficiency is present to some degree in approximately 10% of patients undergoing repair of AVSD. These children are initially managed medically with afterload reduction. This may be done intravenously in the intensive care unit with nitroglycerin or nitroprusside. Long-term afterload reduction can be done orally with angiotensin-converting enzyme inhibitors such as captopril or with other oral medications such as prazosin (Minipress). A small subgroup of these patients with left AV valve insufficiency require redo operations for either left AV valvuloplasty or left AV valve replacement in the future. Reoperation should not be delayed if the child is in the intensive care unit and failing to progress.

A third potential complication after repair of AVSD is heart block. In the early days of repair of AVSD, this problem represented a leading cause of operative and postoperative mortality. This complication is not as problematic currently because it occurs with much lower frequency with careful attention to the anatomy of the conduction system, and also because, when it does occur, it can be managed appropriately. The incidence of permanent heart block in major centers repairing AVSD is <1%. Most episodes of postoperative heart block in these patients are temporary episodes caused by edema and resolve in the first 10 days after surgery. When permanent heart block does occur, it should be treated by placement of a permanent pacemaker before discharge from the hospital.

During the 4-year time interval of 2005 through 2008, the Society of Thoracic Surgeons Congenital Heart Surgery Database documented data from 2882 operations to repair AV canal defects: partial, 623 (21.5%); intermediate, 342 (11.8%); and complete, 1917 (66.3%). Mean age at complete repair (years) was as follows: partial, 6.1; intermediate, 2.9; and complete, 0.6. Median age at complete repair (years) was as follows: partial, 2.6; intermediate, 0.9; and complete, 0.4. Down syndrome was present in 1767 patients (61.1%). Debanding of the pulmonary artery was rarely performed: partial, 1 (0.2%); intermediate, 0 (0.0%); and complete, 66 (3.4%). Deep hypothermic circulatory arrest was rarely used: partial, 6 (1.0%); intermediate, 5 (1.5%); and complete, 52 (2.7%). Discharge mortality was low: partial, 2 (0.3%); intermediate, 3 (0.9%); and complete, 38 (2.0%). AV block requiring permanent pacemaker occurred but was uncommon: partial, 6 (1.0%); intermediate, 2 (0.6%); and complete, 29 (1.5%). Unplanned reoperation

prior to hospital discharge occurred in 3.9% of complete AV canal repairs. The sternum was left open in 3.0% of complete AV canal repairs. Postoperative cardiac arrest occurred in 1.9% of complete AV canal repairs. Mean postoperative length of stay (days) was as follows: partial, 5.2; intermediate, 7; and complete, 13.1. Median postoperative length of stay (days) was as follows: partial, 4; intermediate, 4; and complete, 7. Overall, 98% to 99% of patients survive complete repair of AV canal and 96% to 97% survive complete repair of AV canal with no major complications.

Long-term prognosis is excellent after repair of AVSD. Several series have reported long-term survival over 10- to 20-year periods of >90%. Long-term freedom from reoperation is similarly high except in the subgroup of patients requiring intervention for severe left AV valvar regurgitation. The short-term and long-term prognoses in children with AVSD and Down syndrome are similar to those for children without Down syndrome.

CONTROVERSIAL ISSUES

Several issues remain controversial regarding the repair of AVSD. These include whether a single patch or two patches should be used for the repair, which type of material should be used for the patch or patches, whether the cleft in the left AV valve should be closed in partial AVSD, and where to leave the coronary sinus after repair. We prefer to use separate patches because we believe that this allows us greater ease in the reconstruction of the AV valves. By placing two patches, we believe that we are better able to avoid distorting the valves and that we have greater flexibility in creating competent AV valves. Moreover, we believe that we have greater ability to avoid conduction tissue with the use of two patches. We have also had recent success with the *Nunn/Wilson's* modified one-patch technique as described above.

We choose to use Gore-Tex or polyester (Dacron) patches for closure of the VSD. However, we prefer to use autologous pericardial patches for closure of the ASD. We also like to use pericardial pledgets to reinforce this patch closure as previously described. We believe that the pericardial closure of the ASD helps to prevent the small risk of problematic postoperative hemolysis. Otherwise, a jet of regurgitation through the left AV valve can strike a Gore-Tex or Dacron patch and subsequently lead to this hemolytic problem.

It has been proposed that the cleft leaflet in the left AV valve in partial AVSD does not need to be completely reapproximated because this valve is actually a trileaflet valve completely different in fundamental structure from a normal mitral valve. In our view, this cleft should be closed because our experience demonstrates that left AV valve cleft closure helps to prevent the development of late left AV valve regurgitation.

Finally, several options exist regarding placement of the coronary sinus on the left or right atrial side of the patch. Some advocate closing the ASD so that the coronary sinus drains to the left atrium; those who utilize this approach feel that baffling the coronary sinus to the left atrium minimizes the potential for damage to the conduction system and the development of permanent heart block. Others advocate closing the ASD so that the coronary sinus drains to right atrium; placement of the coronary sinus on the right atrial side eliminates an additional element of mixing of saturated and desaturated blood. Certainly, when a left-sided SVC drains into the coronary sinus, it is extremely important that the coronary sinus be placed on the right side of the atrial septum. It is also important to direct the coronary sinus blood flow to the right side in the setting of an unbalanced AVSD with a small left ventricle. Because the AV node typically lies at the apex of the nodal triangle, the coronary sinus can be directed to the right atrial side of the atrial patch, and the AV node can be safely avoided in the majority of circumstances. In some hearts, a sizable post-Eustachian sinus near the base of the coronary sinus allows for placement sutures that avoid conduction tissue and safely allow the coronary sinus to drain to the right atrium. In other hearts, this post-Eustachian sinus is smaller; in these hearts, it is often necessary to place the suture line within the coronary sinus ostium to keep the coronary sinus draining to the right side of the heart. By using these techniques, in the majority of circumstances one can allow coronary sinus drainage to remain in its anatomic position of the right side by paying careful attention to the anatomy of the conduction system and the nodal triangle.

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EDITOR'S COMMENTS

The progressive improvement in results of repair of complete AVSD has permitted primary repair in infancy to become the procedure of choice for this condition. The incidence of the development of pulmonary vascular occlusive disease by 1 year of age has progressively decreased the optimal age at repair to the point that now elective repair can be undertaken at any age when congestive heart failure is not controlled, and optimally between 2 and 4 months of age in most infants. We agree with the authors that pulmonary artery banding is virtually never indicated for complete AVSDs because the banding may cause increased right ventricular hypertrophy, which actually complicates the exposure of the AV valves at the time of complete repair and in our experience has resulted in less optimal exposure for the placement of the ventricular patch and reconstruction of the common AV valve. Even very tiny infants can undergo successful complete repair without resorting to banding. One subset of patients who may benefit from banding are the rare infants who have abnormal attachment of the anterior bridging leaflet that is significant enough to create left ventricular outflow tract obstruction. These patients may best be considered to have functionally single ventricle.

Partial AVSD ("ostium primum defect") is generally simple to repair, with good long-term results. However, significant left AV valve regurgitation can occur in this condition despite an adequate primary repair. We agree that closure of the cleft in the anterior mitral leaflet in this condition should be routinely performed to attempt to decrease the incidence of late AV valve regurgitation. In spite of this recommendation, some patients will nevertheless develop AV valve regurgitation despite an adequate closure and require valvuloplasty procedures at a later time. A particularly difficult subgroup of patients are those with a relatively hypoplastic left ventricle in association with primum ASD, who have a high mortality with primary repair.

Technically, our technique for repair of complete AVSDs is similar to the technique described in this chapter. We also favor a two-patch technique, which allows the best exposure of the AV valves for the assessment of competence and also permits closure of the VSD without the need to divide a common anterior bridging leaflet except in rare circumstances. If the anterior bridging leaflet is maintained undivided, the chances of dehiscence of the valve attachments to the patch become less and may decrease the need for reoperation. In addition, we have found that securing a ventricular septal patch below the common AV valve leaflets permits a suture line to which the superiorly located pericardial patch can be attached, reinforcing the suture line at the AV valves to minimize the risk of dehiscence. We have generally used Dacron for VSD closure because the patch is more flexible and small residual defects will often close spontaneously. We have elected to place the midportion of the ventricular septal patch first, and then, using gentle traction on each stitch in a running fashion, the suture line can be carried superiorly and inferiorly to complete the repair to the level of the AV valve attachments. The patch can then be trimmed again if necessary to allow the appropriate plane for attachment of the AV valve leaflets to the superior crest of the patch, which we perform in a running fashion. The common AV valve leaflets are then floated with saline solution to assess the coapting surfaces, which are then directly approximated using polytetrafluoroethylene suture, which does not cut through the tissue as readily as polypropylene suture and may avoid the need for pledget material. It is important not to evert the edges of the cleft of the mitral leaflet, which can lead to central regurgitation at the tip of the anterior mitral leaflet. In rare instances in which there is an accessory mitral valve orifice, usually posteriorly and inferiorly, the accessory orifice is not addressed in the repair. We have elected to place the coronary sinus on the right side of the atrial septal patch in essentially all patients and have not found an increased incidence of conduction defects utilizing this technique.

We use intraoperative transesophageal echocardiography at the completion of surgery in every patient who undergoes repair of partial or complete AVSDs to assess the magnitude of mitral valve competence at the end of the procedure. Patients with more than moderate residual mitral regurgitation can then undergo immediate revision of the valve repair to decrease the incidence of late reoperation. With this technique, our experience with complete AVSD repair has resulted in a mortality of ≤2.9%, with a marked reduction in reoperation rate to <10% (Canter CE, Spray TL, Huddleston CB, Mendeloff E. Intraoperative evaluation of atrioventricular septal defect repair by color flow mapping echocardiography. Ann Thorac Surg 1997;63:592).

We have not elected to use pulmonary arterial monitoring lines in most patients with AVSD repairs because with early operative intervention, the incidence of pulmonary hypertensive events has been very low.

Patients who have a restrictive VSD in unbalanced AV canal to the right may have adequate inflow into the left ventricle for a two-ventricle repair if there is antegrade flow out the aorta and across the aortic arch. Decision making for these patients can be extremely difficult, and careful attention must be given to the attachments of the common AV valve, the inflow patterns into the ventricles, and the relative size of the AV valves and ventricles.

Dr. Elliott and associates have suggested that leaving an atrial septal defect in patients with a relatively small left ventricle or compromised left AV valve may be beneficial as a "pop-off." Although this has been shown to be effective in some cases, it is not clear how this improves the hemodynamic situation, given that all of the blood decompressing from the left atrium will be represented back to the left atrium. It is thought perhaps that such atrial septal defects simply allow a larger capacitance of the left atrial chamber and may therefore limit pulmonary hypertension at the expense of a lower cardiac output and increased pulmonary blood flow. We generally believe that if an ASD is required to prevent severe left atrial hypertension, then the left-sided structures are probably too small for a two-ventricle repair.

With the advent of nitric oxide, pulmonary hypertensive events have become less of a cause of morbidity and mortality after repair of AVSD, resulting in marked improvement in operative results. Current operative survival after repair of these defects approaches 98%.

(*continued*)

EDITOR'S COMMENTS (*continued*)

In our experience, the indication for reoperation in AVSD repair has generally been left-sided AV valve regurgitation. The incidence of AV valve regurgitation after repair of complete and partially AV canal defects is higher than often believed. Review of a large number of reported series of AV canal repairs suggests that significant left AV valve regurgitation occurs in up to 30% to 35% of patients after AV canal repair, and reoperation may be necessary in 10% to 20% of these patients over time. Interestingly, despite the relatively better anatomy in partial AV canal defect, the incidence of late AV valve regurgitation requiring reoperation is as high as or higher than that for complete AV canal repair.

The patient who presents in the first few weeks of life with severe congestive heart failure with complete AV canal defect represents a difficult patient group. It is unclear why some patients do not respond to medical treatment of congestive heart failure, but in these cases pulmonary artery banding or early repair can be considered.

Rare instances of patch dehiscence have occurred that required reoperation; however, the long-term durability of the left AV valve repair remains the primary determinant of late morbidity. In many cases, the valve can be repaired at a second operative intervention either with more complete closure of the cleft of the anterior mitral leaflet or with additional annuloplasty sutures. Valve replacement should be extremely unusual after AVSD repairs.

TLS

87 Truncus Arteriosus

Michael O. Murphy and Thomas L. Spray

Truncus arteriosus represents an unusual congenital heart defect in which a single arterial trunk arises from the heart, providing the origins of the coronary arteries, the true pulmonary arteries, and the brachiocephalic vessels. The defect is usually associated with a ventricular septal defect and a single, large semilunar valve. Typically, the semilunar valve contains up to four separate leaflets, which may be dysmorphic in infancy. In situations in which more than four leaflets are present, incorporation of remnants of the pulmonary valve into the truncal valve is presumed. Very rare instances of absence of ventricular septal defect in truncus arteriosus have been reported, but usually the infundibular septum is virtually absent superiorly. Thus, the ventricular septal defect is similar to that seen in tetralogy of Fallot but with no superior rim of infundibular septum separating the pulmonary and aortic valves. This feature can be used to distinguish truncus arteriosus from pulmonary atresia with ventricular septal defect. Embryologically, the development of truncus arteriosus is related to deficiency of the aorticopulmonary septum, with the absence of the subpulmonary infundibulum and partial or complete absence of pulmonary valve tissue. These defects are associated with neural crest abnormalities, and because the neural crest also develops into thymus and parathyroids, DiGeorge syndrome is commonly associated with truncus arteriosus.

The development of aortic arches 4 and 6 varies in truncus arteriosus such that infants with hypoplasia or interruption of the aortic arch have associated large ductal connections between the truncus and the descending aorta, and infants in whom the arch is fully developed usually have absence of the ductus arteriosus.

The primary classification systems for truncus arteriosus have focused on the origins of the pulmonary arteries. The classification scheme of Collett and Edwards defines truncus arteriosus types by the presence of a main pulmonary trunk or the separation of the pulmonary arteries from the arterial trunk. From a surgical standpoint, however, the variations in the origin of the pulmonary arteries are often similar enough among the various types that separation by this classification scheme has not been as useful as the alternate scheme developed by Van Praagh. The Van Praagh classification system is described in Table 87.1. Most recently, a modified Van Praagh classification has been proposed by the Congenital Heart Surgery Nomenclature and Database project, classifying cases by the presence of confluent, or near confluent pulmonary arteries, truncus arteriosus with the absence of one pulmonary artery or truncus arteriosus in the presence of interrupted aortic arch or coarctation. The advantage of this system is that it is based on a hierarchical system and can be used interchangeably with the traditional classification systems.

Some authors have described "hemitruncus" as a situation in which the right pulmonary artery arises from the ascending aorta and the left pulmonary artery from the right ventricle. This defect is not typically associated with a ventricular septal defect. In our opinion, it is not a variation of truncus arteriosus and is best defined as aortic origin of the right pulmonary artery. Such a description distinguishes this anomaly from the Van Praagh type 3A truncus.

The truncal valve typically straddles the ventricular septum with balanced contributions from the left and right ventricles. However, in a few cases the truncal valve straddles the right ventricle, offering the potential for restriction of blood flow from the left ventricle to the truncal valve at repair. Coronary anomalies seen in truncus are typically a high origin of the left coronary artery from the truncus near the pulmonary origins and an anomalous anterior descending coronary from the right coronary artery crossing the right ventricle to supply the anterior ventricular septum.

PATHOPHYSIOLOGY OF TRUNCUS ARTERIOSUS

The defect of truncus arteriosus in the absence of arch interruption or severe hypoplasia is one of complete mixing of systemic and pulmonary venous blood with pulmonary overcirculation. As the pulmonary vascular resistance falls in the early neonatal period, the tendency is for unrestricted pulmonary blood flow from the truncus, resulting in severe congestive heart failure. Mild cyanosis may be present because of the complete mixing at the level of the atrium and ventricle. The presence of a large connection between the ascending aorta and the pulmonary arteries above the level of the semilunar valve results in pulmonary blood flow during systole and diastole, increasing the amount of pulmonary overcirculation. It is believed that this situation results in early development of pulmonary vascular obstructive disease in children with truncus arteriosus, which accounts for the high mortality of untreated patients with this condition. Infants born with truncus arteriosus and unrestricted pulmonary blood flow have a mortality of as high as 50% in the first month of life because of congestive heart failure. Pulmonary vascular obstructive disease typically develops to a significant degree by 6 months of age. Other physiologic consequences of truncus arteriosus relate to the presence or absence of truncal valve stenosis or insufficiency. Although major truncal valve stenosis is rare, truncal valve insufficiency occurs in a significant proportion of children with this condition. The presence of significant truncal valve insufficiency not only complicates the operative repair of the lesion but also may have a significant impact on morbidity and mortality after complete repair.

The presence of interrupted aortic arch in association with truncus arteriosus results in cardiovascular instability early after birth. Maintenance of ductal patency by prostaglandin E_1 infusion is critical to provide distal perfusion to the body in this condition but is associated with

Table 87.1 Van Praagh Classification of Truncus Arteriosus

1. Partially formed aorticopulmonary septum (main pulmonary artery segment present)
2. Absent aorticopulmonary septum (no main pulmonary artery segment)
3. Absence of one branch of the pulmonary artery from the trunk (ductal or aortic origin of one pulmonary artery)
4. Hypoplastic or interrupted aortic arch with a large patent ductus arteriosus

Type A: Ventricular septal defect present

Type B: Ventricular septal defect absent

progressive pulmonary overcirculation, which is not dissimilar to the situation seen in hypoplastic left heart syndrome.

DIAGNOSIS AND INDICATIONS FOR SURGICAL INTERVENTION

The diagnosis of truncus arteriosus can generally be made with accuracy from echocardiograms. Cardiac catheterization is indicated only when the patient presents late after birth and the issue of pulmonary vascular resistance is raised. Some children may have significant pulmonary arterial stenosis, which permits control of congestive heart failure early in life, and catheterization to define the level and degree of obstruction may be beneficial. In addition, abnormalities of coronary arteries and the severity of truncal valve abnormalities may occasionally prompt catheterization for greater characterization of the defects.

We have generally considered the presence of truncus arteriosus an indication for operation in a relatively urgent fashion. Because hemodynamics may be quite unstable in children with unrestricted pulmonary blood flow and because we have seen patients deteriorate rapidly while awaiting surgical intervention, we have elected to repair truncus arteriosus promptly after the diagnosis is made. This policy reflects the clear advantages of neonatal repair of truncus arteriosus. Review of series of truncus repairs reflects the trend toward earlier intervention. Whereas the first repairs of truncus arteriosus were performed in 1962 by Behrendt and associates, in 1967 McGoon first used a valved allograft for the repair. This operation was a refinement of the procedure described by Rastelli at the Mayo Clinic. The results of early repairs were quite poor, however, because of the development of pulmonary vascular obstructive disease. In most of these early repairs, the operation was undertaken after the patient reached 6 months of age, when pulmonary vascular resistance was already significantly elevated. In 1984, Ebert reported a series of 100 infants who underwent complete repair of truncus arteriosus at 6 months of age with an 11% mortality. This landmark report emphasized the improved results in early intervention in children with this lesion. In our experience, repair of truncus arteriosus has gradually improved as surgical techniques have evolved such that operative survivals of >95% are anticipated with neonatal repair in patients without associated valvar lesions. The late results of truncus arteriosus have been favorable, with little late mortality despite the need for reoperation to replace right ventricular outflow tract conduits.



SURGICAL TECHNIQUE

Repair of Truncus Arteriosus

The heart is exposed with a median sternotomy incision. The presence of thymic tissue is confirmed because many of these children have athymia associated with DiGeorge syndrome. The clear presence of thymic tissue, therefore, may have implications for later management and prognosis. The heart is suspended in a pericardial cradle and the great vessels examined. If the patient is severely overcirculated with congestive heart failure, it may be advisable to promptly encircle one of the pulmonary arteries with a snare to limit the total pulmonary blood flow. On occasion, this can stabilize the patient while dissection is performed around the arch vessels.

The operation can be performed either with the patient on continuous cardiopulmonary bypass or by utilizing circulatory arrest. We have elected to use both of these techniques, and in simple truncus arteriosus, the complete operative repair can be performed during a single period of circulatory arrest of <40 minutes in most cases. The right and left pulmonary arteries are mobilized and encircled with tourniquets for control and the origins of the pulmonary arteries examined. In addition, on opening the pericardium, it is important to determine the location of the anterior descending coronary artery and confirm that it does not cross the right ventricular wall in the area of anticipated ventriculotomy. Heparin is administered and the aorta cannulated as distally as possible. Distal cannulation is particularly important because the pulmonary bifurcation may come off relatively distally and there is little room for cross-clamping of the aorta between the anticipated division of the pulmonary bifurcation and the distal aorta. Either the venae cavae are cannulated if continuous bypass is used, or a single right atrial cannula is used with circulatory arrest. If mobilization of the pulmonary artery confluence behind the aorta is considered to be too close to the anticipated cross-clamp application site, the brachiocephalic vessels are encircled with tourniquets and circulatory arrest is used, with clamping of the aorta more distally on the arch during the reconstruction of the back of the truncus arteriosus after excision of the pulmonary bifurcation. The infant is placed on cardiopulmonary bypass and the snares on the right and left pulmonary arteries tightened as shown in Figure 87.1 to prevent pulmonary overcirculation during cooling. The child is cooled to 18°C for circulatory arrest or 34 to 37°C for continuous bypass repair. In the presence of significant truncal valve insufficiency, venting of the left ventricle is important during cooling and may be accomplished by placing a vent through the right superior pulmonary vein across the mitral valve into the ventricle, or if truncal insufficiency is severe, by compressing the heart to maintain emptying of the ventricle during cooling.

The aorta is then cross-clamped and cardioplegic solution is administered into the truncal root with the pulmonary artery snares applied to force the cardioplegic solution into the coronary arteries. Alternatively, cardioplegia can be induced in a retrograde manner or the aorta opened and the coronaries directly cannulated. After cardioplegia is achieved, the snares are removed from the pulmonary arteries, and an incision is made near the takeoff of the pulmonary bifurcation anteriorly. Through this initial incision, the truncal valve is examined and the origin of the left coronary artery carefully identified. The left coronary can arise high from above the truncal sinuses and be intimately associated with the origin of the pulmonary bifurcation. Once the coronary arteries are identified, the pulmonary bifurcation is excised from the back of the truncus arteriosus. The aorta is then repaired using a

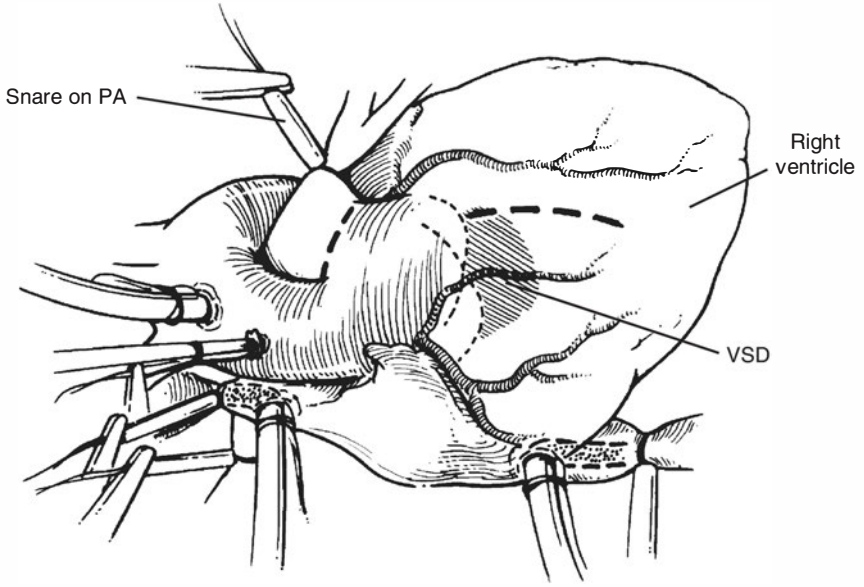


Fig. 87.1. Repair of simple truncus arteriosus. The infant is placed on cardiopulmonary bypass with either bicaval cannulation or single venous cannulation in the right atrium for circulatory arrest. The aorta is cannulated as distally as possible to allow cross-clamping proximal to the aortic cannulation site for the division of the pulmonary arteries from the truncal vessel. Snares are placed on the right and left pulmonary arteries to prevent pulmonary overcirculation during warming and to allow delivery of cardioplegic solution to the myocardium. After cardioplegia is achieved, the snares are released from the pulmonary arteries and the pulmonary bifurcation is excised from the back of the truncal artery, with care being taken to avoid the origin of the left coronary artery posteriorly, which can originate very close to the pulmonary bifurcation. An incision is made in the right ventricle, with care being taken to avoid major epicardial coronary branches and the base of the truncal valve. PA, pulmonary artery; VSD, ventricular septal defect.

patch of pulmonary homograft. Primary suture closure of the defect in the truncus may be used; however, we believe that the placement of a patch allows for less tension on the anastomosis and therefore a decreased risk of bleeding from behind the aorta. This is important after the right ventricular reconstruction has been performed because exposure of the back of the aorta is difficult after the completion of the repair. In addition, suturing bleeding sites in this area may be hazardous because the origin of the left coronary artery may be very close to the margins of the excision of the pulmonary bifurcation. Although pericardium can also be used for a patch, we have elected to use a portion of the pulmonary or aortic homograft that will be used for right ventricular outflow tract reconstruction because it results in a hemostatic suture line and is easier to work with than nontanned pericardium.

After the repair of the truncus arteriosus is complete and the pulmonary bifurcation is mobilized to bring the bifurcation to the left for reconstruction, additional cardioplegic solution is injected into the aortic root to test the suture line of the truncal patch, and additional sutures are placed if necessary. Next, an incision is made in the right ventricle, avoiding major conal branches of the coronary arteries. This incision is made at the base of the truncal valve, and care must be taken not to carry the incision too far superiorly into the truncal valve annulus. The absence of infundibular septum in this area results in the possibility of damage to the truncal valve if the incision is started too far superiorly. The orientation of the incision is made so that the takeoff of the conduit from the right ventricle will be directed toward the pulmonary bifurcation.

As shown in Figure 87.2, the ventricular septal defect is exposed through the incision in the right ventricle. The defect is closed with a patch of polyester (Dacron) material using a running technique. Superiorly, the absence of the infundibular septum mandates that the patch be more ovoid in shape, and the patch is secured to the epicardial margin of the ventriculotomy incision superiorly to avoid interference with the truncal valve (Fig. 87.3). In the majority of cases, there is a bridge of muscle between the margin of the ventricular septal defect and the tricuspid valve septal leaflet, and in these patients, suturing in this area can be performed without risk to the conduction

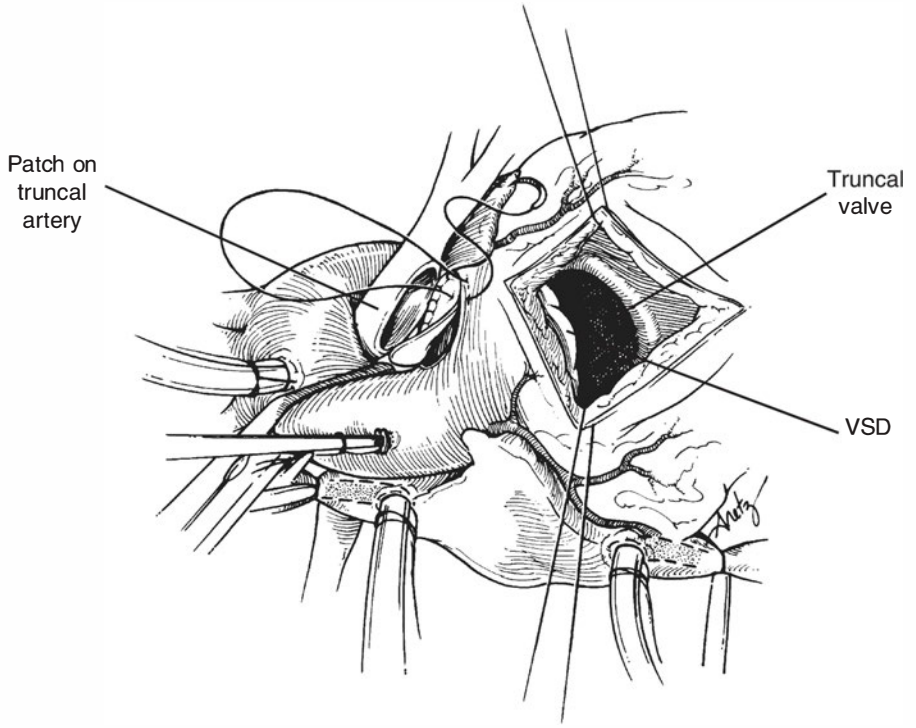


Fig. 87.2. The defect in the main trunk from excision of the pulmonary bifurcation is closed with a patch of pulmonary homograft material or pericardium with meticulous suture technique to ensure hemostasis. Cardioplegic solution is then administered to observe any leaking of the suture line because the area will be difficult to expose after complete repair. The ventricular septal defect (VSD) is exposed through the ventriculotomy incision. There is no muscle rim typically present at the superior margin of the VSD and, therefore, the patch is secured to the epicardial portion of the ventriculotomy incision superiorly.

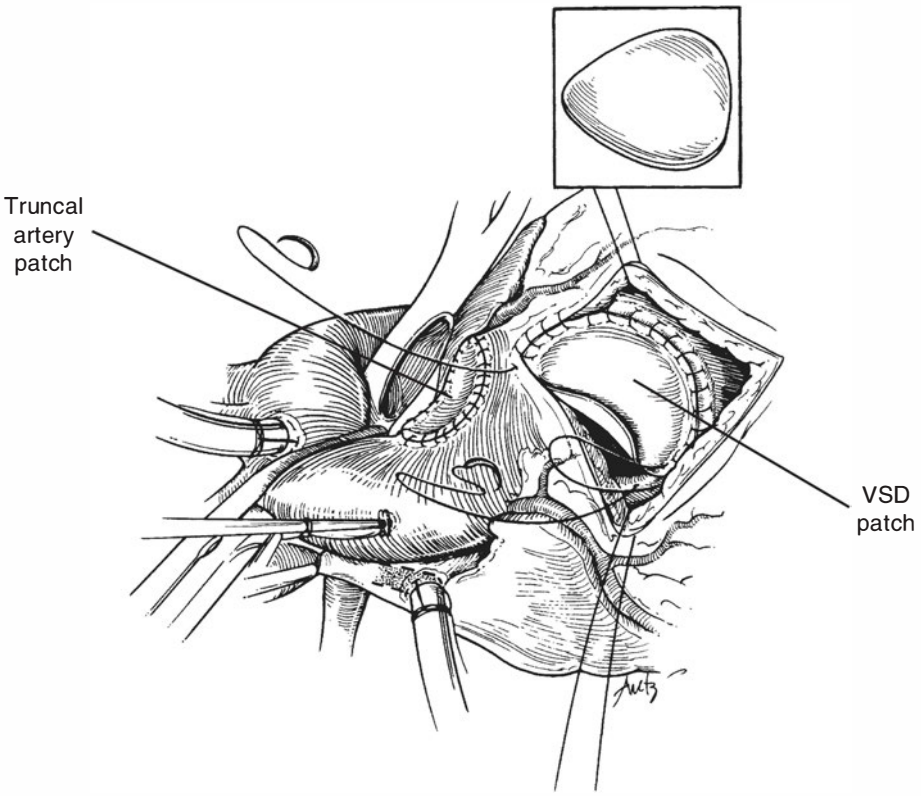


Fig. 87.3. The ventricular septal defect (VSD) is closed with an ovoid patch of polyethylene terephthalate (Dacron) material sewn to the superior margin of the ventriculotomy incision (*inset*). Typically, a rim of muscle is present inferiorly between the VSD and the tricuspid valve septal leaflet, and in these patients a running suture line can be created that avoids the conduction tissue. If the muscle bridge is not present, the inferior margin of the suture line is carried along the base of the septal tricuspid valve leaflet on the right ventricular aspect to avoid the conducting tissue.

tissue. In a minority of cases, there is absence of this muscle tissue, and suturing of the ventricular septal patch to the base of the septal leaflet of the tricuspid valve is necessary.

After closure of the ventricular septal defect is complete, the right ventricular outflow tract is reconstructed using a pulmonary or aortic homograft. Although in the past we elected to use larger pulmonary homografts to allow for the maximum possible growth of the infant before conduit change is necessary, more recently we have used aortic allografts preferentially, because the high pulmonary vascular resistance seen in the early postoperative period often causes dilation in the pulmonary allografts. Another advantage for the initial use of an aortic homograft is the curved nature of the ascending aorta that can be positioned to the left, curving around the enlarged truncal root in patients in whom the truncus is particularly enlarged.

The use of an aortic homograft in the initial repair of truncus arteriosus prevents pulmonary homograft dilation early. At the time of the conduit replacement after truncus repair which typically occurs between 3 and 6 years after the operation, a pulmonary homograft is preferentially selected since pulmonary vascular resistance is now low and placement of a large pulmonary homograft may obviate the need for additional conduit replacements in the future, and the development of pulmonary insufficiency in the homograft conduit can be treated later with a stent mounted on pulmonary valve with a percutaneous approach.

If a pulmonary homograft is used, it is important not to oversize the pulmonary homograft because even though it is desirable to have a larger homograft to allow for future growth and decrease the need for early reoperation, the capacitance of a large pulmonary homograft may equal the stroke volume of the right ventricle and limit the forward cardiac output in patients when there is severe oversizing of the homograft conduit. The conduit in such cases acts like an aneurysm of the right ventricle, decreasing overall right ventricular performance. To accommodate such a large homograft, the pulmonary bifurcation is incised, with the incision slightly into the origin of the right pulmonary artery but more into the left pulmonary artery so that the conduit will lie toward the left. An anastomosis is then created between the homograft and the pulmonary bifurcation with a running monofilament suture (Fig. 87.4).

As noted in Figure 87.5, the homograft is then anastomosed to the superior margin of the ventricular incision to which the ventricular septal defect patch has been sewn. Approximately one-third of the circumference of the ventriculotomy incision is sutured in this manner. In some cases, the homograft will have an adequate amount of right ventricular muscle attached so that it can be trimmed to allow a very small margin of muscle at the suture line to the ventricle but with an adequate

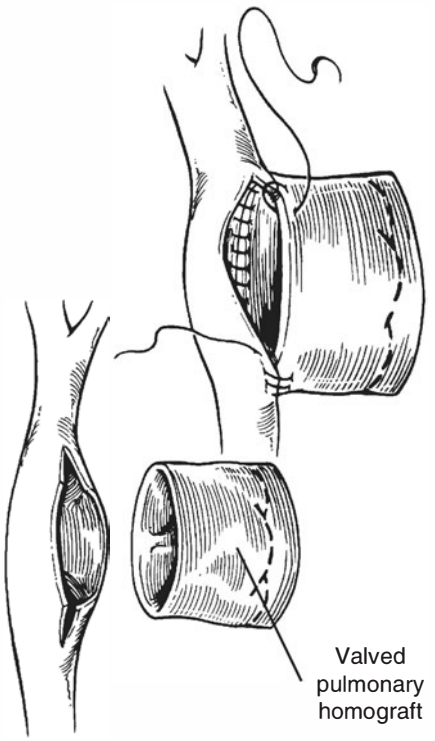


Fig. 87.4. The pulmonary arteries are reconstructed with a pulmonary homograft. To prevent kinking of the homograft with distention, it is trimmed 2 to 3 mm distal to the commissural attachments of the pulmonary valve. To provide an adequate anastomosis and size match to the pulmonary bifurcation using a large pulmonary homograft of 12 to 18 mm in diameter, an incision is carried into the origin of the right pulmonary artery for a short distance and more significantly into the left pulmonary artery if additional opening is necessary. An anastomosis is then created with a running fine monofilament suture between the pulmonary homograft and the pulmonary bifurcation.

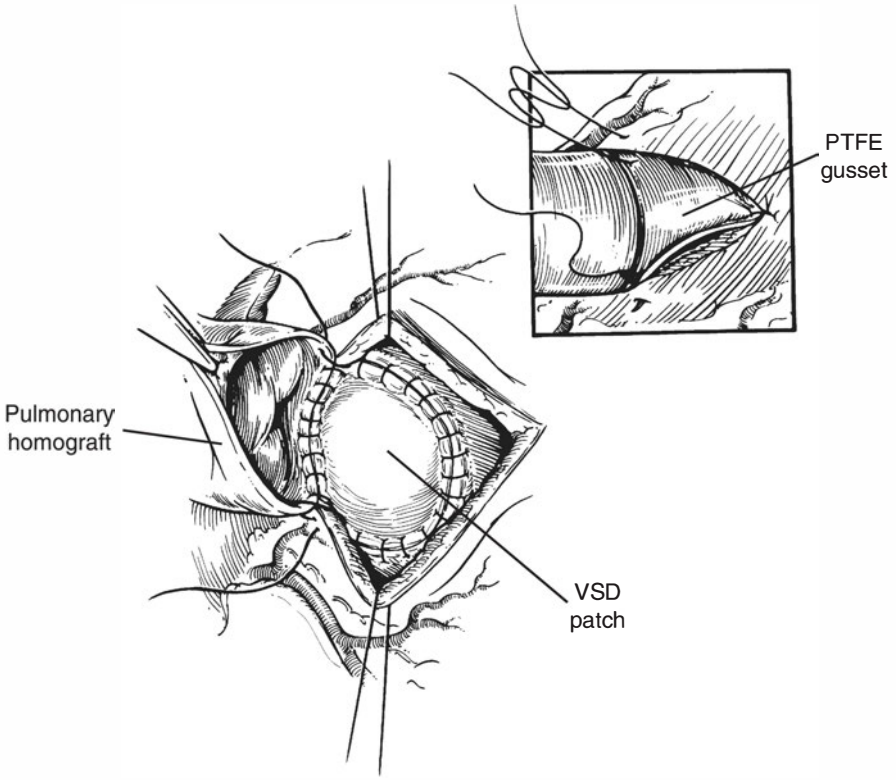


Fig. 87.5. The pulmonary homograft is then sewn proximally to the superior aspect of the ventriculotomy incision for approximately one-third of its circumference. As noted in the text, if adequate muscle is present on the pulmonary homograft, it can be sewn directly down to the right ventricular outflow tract. However, if inadequate tissue is present, the outflow tract is augmented with a gusset of polytetrafluoroethylene (PFTE) material, as shown in the inset, to create a gentle takeoff of the homograft from the right ventricular outflow tract to avoid compression. VSD, ventricular septal defect.

amount of muscle remaining anteriorly to create a complete reconstruction without augmentation. In this case, the suture line is continued around the margins of the ventricular incision until the homograft is completely sewn to the right ventricular outflow tract. Care must be taken to trim the homograft appropriately so that it is not compressed as it originates from the right ventricle and also the length of the homograft is not excessive, which can result in kinking of the posterior wall of the homograft and right ventricular outflow tract obstruction.

Typically, in a neonate or infant, division of the pulmonary homograft just above the level of the commissural attachments of the valve results in sufficient length to prevent kinking after completion of the reconstruction. With the smaller aortic allografts' use, the angle and the mitral valve of the homograft create a "gusset" that creates a gentle takeoff of the conduit from the right ventricle of the aortic arch, which is used to place the graft at the correct orientation to the pulmonary arteries. If insufficient right ventricular muscle is available on a pulmonary homograft for a direct anastomosis, a gusset of polytetrafluoroethylene (PTFE) material is used to augment the takeoff of the homograft from the right ventricle to prevent compression of its origin as noted in the inset of Figure 87.5. Whereas pericardium may be used for this gusset, we use PTFE because we have seen aneurysm formation of nonpreserved pericardium when it is used in this position if there is distal stenosis of the homograft or the pulmonary arterial anastomosis.

After the repair is completed, the atrial septum is examined, working through either an atrial purse string or direct atrial incision. In a significant proportion of patients, a secundum-type atrial septal defect is present, and this is closed partially as noted in Figure 87.6 to allow the capacity for right-to-left shunting at the atrial level if right ventricular dysfunction occurs in the early postoperative period. If a foramen ovale is present without a significant defect, the foramen is left open to permit decompression of the right atrium.

After completion of the repair, the patient is rewarmed and weaned from cardiopulmonary bypass. We have used modified ultrafiltration in our recent series of infants to decrease myocardial edema and improve early postoperative hemodynamics. If a large pulmonary homograft is chosen for construction of the outflow tract, the homograft conduit may appear to be compressed by the sternum during closure, and in these cases, opening the left pleural space widely either anterior or posterior to the phrenic nerve may permit rotation of the conduit into the left pleural space and allow the chest to be closed without compression.

Alternative Repairs of Truncus Arteriosus

Several variations on repair of truncus arteriosus have been described, primarily to avoid the need for an allograft conduit

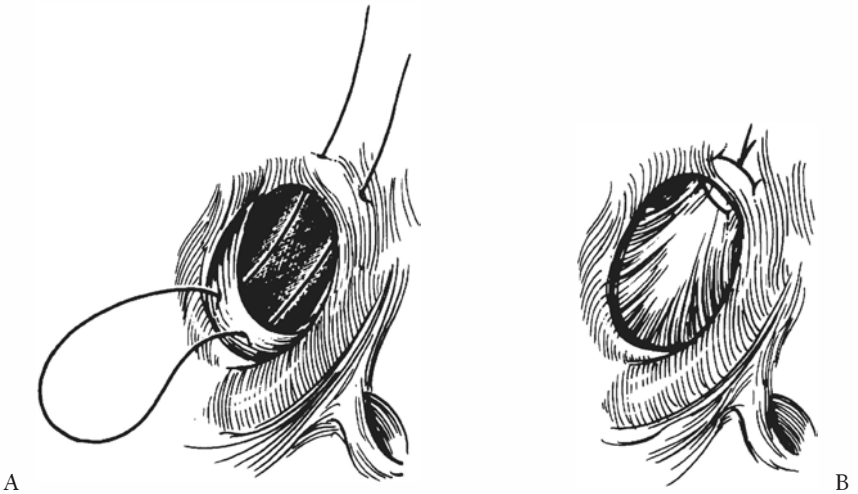


Fig. 87.6. If a secundum atrial septal defect is present (**A**), a partial closure is accomplished (**B**) to allow for the capacity for right-to-left shunting at the atrial level if ventricular dysfunction occurs postoperatively.

from the right ventricle to the pulmonary arteries, which will commit most children to eventual conduit replacement. We have used conduit reconstruction in the majority of infants because the early morbidity and mortality after truncus arteriosus repair are commonly related to the occurrence of pulmonary vascular hypertensive crises and right ventricular dysfunction, and in this setting, the absence of a pulmonary valve exacerbates the hemodynamic instability. The presence of a competent valve in the right ventricular outflow tract seems to minimize the early hemodynamic instability in these patients, and replacement of the right ventricular outflow allograft is associated with a low morbidity and mortality.

In the interest of completeness, several alternative techniques are described here. The first alternative repair involves incision of the truncus arteriosus above the semilunar valve on the pulmonary arterial side of the common trunk (Fig. 87.7). Working through this incision, the surgeon can patch the origin of the pulmonary bifurcation from the arterial trunk with a piece of homologous pericardium or homograft material, with care being taken to avoid interference with the coronary ostium of the left coronary artery and allowing unobstructed flow from the truncal valve to the ascending aorta. An advantage of this technique is the lack of external bleeding with this suture line, which can be troublesome after patching of this area after division of the pulmonary bifurcation. A potential disadvantage, however, is that any dehiscence of this suture line will result in a significant left-to-right shunt. If this type of repair is performed, an incision is made in the right ventricle, and the ventricular septal defect is closed. The pulmonary bifurcation is then mobilized freely and either brought anterior to the ascending aorta or mobilized sufficiently to allow direct anastomosis to the superior margin of the right ventriculotomy. In situations in which the pulmonary bifurcation cannot be anastomosed directly to the right ventriculotomy without compression of the pulmonary bifurcation by the ascending aorta or if an anomalous anterior descending coronary artery crosses the outflow tract, it is possible to create a floor for the outflow tract reconstruction by the use of autologous tissue, such as the left atrial appendage. In Figure 87.8, the left atrial appendage is opened, creating a flap of tissue that is then brought over the outflow tract and secured to the superior margin of the ventricular incision and the pulmonary bifurcation. The base of the atrial appendage is oversewn. This creates an autologous connection between the right ventricle and the pulmonary bifurcation that has the potential for growth. The outflow tract is then augmented by an anteriorly placed patch of homologous pericardium or pulmonary homograft material. In this manner, a valveless connection is created between the right ventricle and the pulmonary bifurcation.

Another alternative is the use of a monocusp reconstruction of the right ventricular outflow tract, which has the potential advantage of early pulmonary valve competence during the initial period of hemodynamic instability postoperatively and the potential for later growth to decrease the need for conduit reconstruction later in life. In this technique (Fig. 87.9), a piece of homologous pericardium is used to create a single cusp, which is sewn to the wall of an outflow patch of pulmonary homograft, homologous pericardium, or PTFE material. This pericardial monocusp is created in a generous manner to allow it to close against the right ventricle (Fig. 87.10). The posterior floor of the reconstruction is performed as noted in Figure 87.8; however, the anterior reconstruction is completed with this monocusp outflow patch. The monocusp valve appears to function adequately for as long as several months and may decrease the incidence of late conduit changes, but it is more difficult to accomplish without at least some early postoperative pulmonary insufficiency.

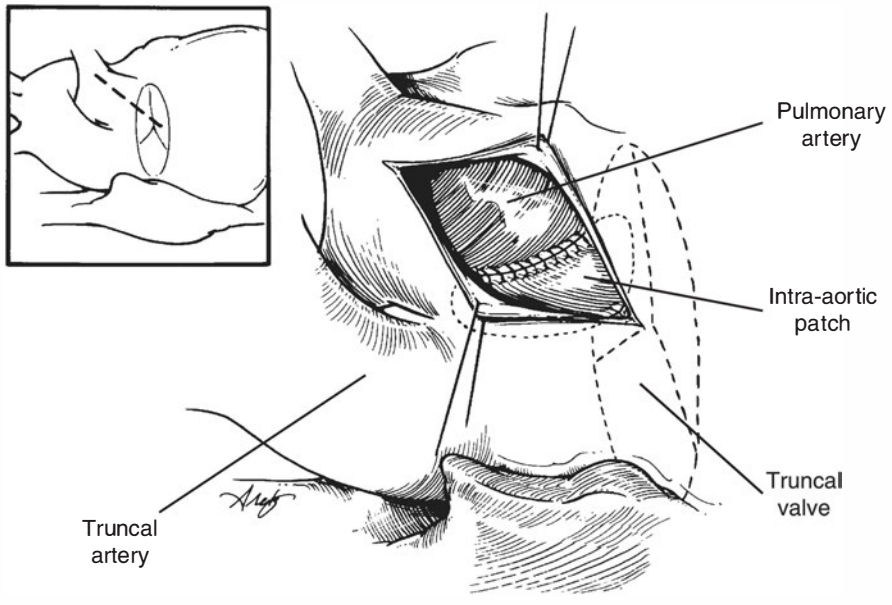


Fig. 87.7. Truncal repair can be accomplished without division of the pulmonary bifurcation by incising the truncus anteriorly toward the pulmonary bifurcation (*inset*) and patching the origin of the pulmonary arteries from the inside of the truncus using a piece of pulmonary homograft or autologous pericardium. Care must be taken to avoid suturing near the origin of the left coronary artery and to avoid obstruction to outflow from the truncal valve. Again, meticulous suture technique is imperative to prevent leakage of this patch and resultant left-to-right shunt into the pulmonary artery after complete repair. In addition, the patch cannot be made too redundant or it will bow into the main pulmonary artery, potentially compromising the right ventricular outflow reconstruction. The pulmonary outflow tract is then reconstructed with a conduit or by alternative techniques.

Repair of Truncus Arteriosus with Interrupted Aortic Arch

When interrupted aortic arch is associated with truncus arteriosus, the operation is usually performed with the patient under circulatory arrest. Because of the instability of these infants, surgery in the first week of life is typical. After mobilization and snaring of the pulmonary arteries to prevent pulmonary overcirculation on bypass, cannulation of the main trunk and the right atrial appendage is performed. Because perfusion of both the brachiocephalic vessels and the distal aorta across the ductus arteriosus will be accomplished by a single cannulation, the arterial cannulation can be placed proximally in the main portion of the truncal vessel. Snares

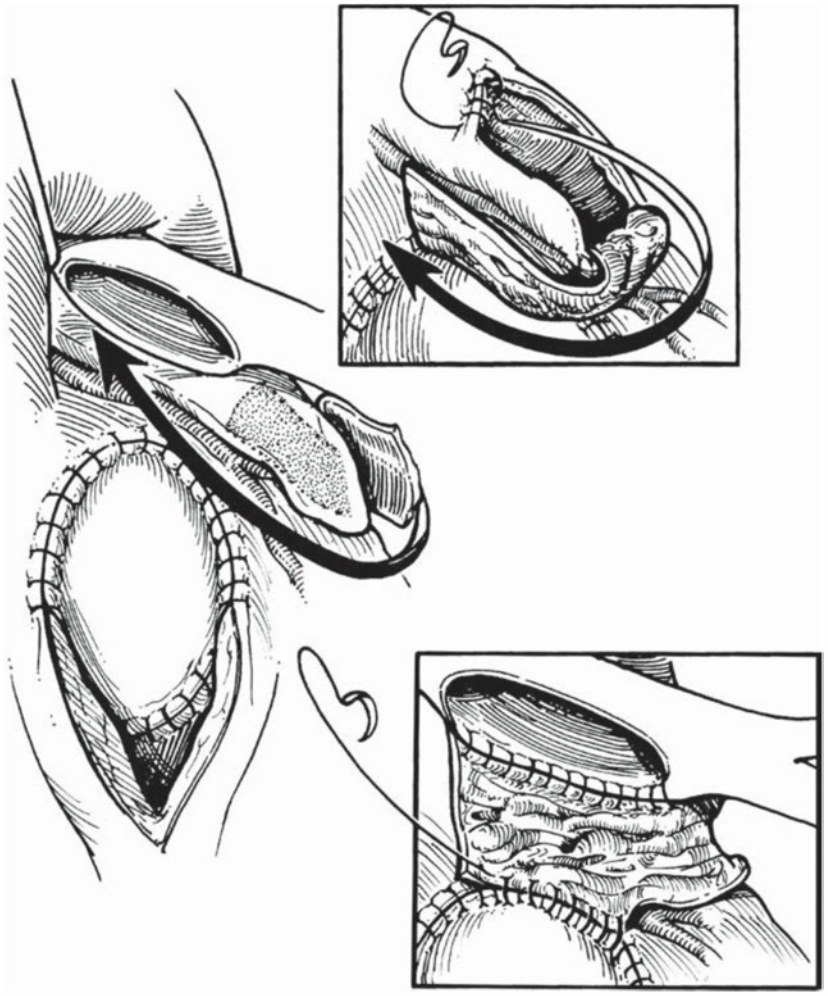


Fig. 87.8. In situations in which a pulmonary homograft is not used for outflow reconstruction, autologous tissue may be used. In this alternative technique, the left atrial appendage is incised at its base and a flap of atrial appendage anastomosed to the pulmonary bifurcation superiorly and to the ventriculotomy incision inferiorly to bridge the ventriculotomy incision and the pulmonary bifurcation with autologous tissue. The base of the atrial appendage is then oversewn. It is important if this technique is used to open the atrial appendage to create a long bridge of tissue. The use of the unopened atrial appendage has been suggested; however, in our experience, sufficient tension may be present on this atrial appendage that with cardiac distention and contraction of the atrium, the bridge of tissue may compress the left coronary artery and cause myocardial ischemia. The repair is then completed with augmentation of the autologous floor of the outflow tract reconstruction with an anteriorly positioned patch of pericardium or homograft material.

are placed also on the brachiocephalic vessels originating from the ascending aorta. The infant is placed on bypass and cooled to a nasopharyngeal temperature of 18°C while snares on the right and left pulmonary arteries prevent pulmonary overcirculation and distal perfusion is provided across the ductus arteriosus. During cooling, the descending aorta and the left subclavian artery may be mobilized and a snare placed on the left subclavian artery if necessary. In some cases, aberrant origin of the right subclavian artery is present in this condition. In rare cases, it may be necessary to divide the right subclavian artery to gain adequate mobilization for repair, as in repair of interrupted aortic arch, but this is usually not necessary. Although several techniques are available for reconstruction of the aortic arch, we prefer to use a technique in which the arch is augmented with pulmonary homograft material to prevent tension on the anastomosis. After circulatory arrest is established and cardioplegia induced, which can be accomplished by snaring of the ductus arteriosus and the brachiocephalic vessels and infusion of cardioplegic solution through the aortic cannula in the proximal arterial trunk, the cannulae are removed from the heart. With the arch vessels snared, an incision is made on the lateral aspect of the ascending aorta beyond the origins of the pulmonary arteries and then carried inferiorly and across the pulmonary bifurcation (Fig. 87.11). Ductal tissue is excised and the ductus ligated on the pulmonary arterial end. In this manner, the pulmonary bifurcation is excised from the truncus. The pulmonary arteries are mobilized freely; then, all ductal tissue is excised from the descending aorta and an incision is made superiorly into the origin of the left subclavian artery. A side-to-side anastomosis is then created from the origin of the left carotid to the origin of the

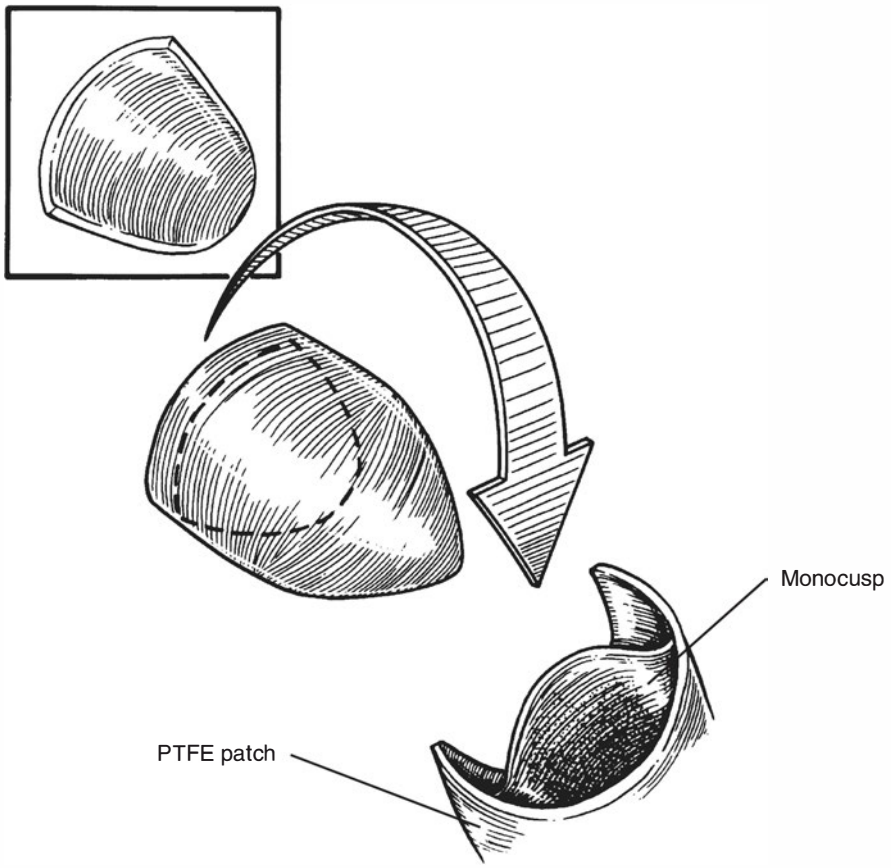


Fig. 87.9. An additional variation of outflow reconstruction uses a monocusp of homologous pericardium positioned in a polytetrafluoroethylene (PTFE) or homograft outflow patch, which can provide temporary pulmonary competence in the early postoperative period. We have elected to create a generous cusp of autologous pericardium and secure it to the undersurface of a PTFE patch fashioned to the size of the outflow tract and then suture the PTFE reconstruction with the monocusp already in place. An alternative technique involves sewing a patch of pericardium that contacts the ventricular septum to the right ventricular incision and then placing an onlay PTFE patch over this area.

left subclavian artery, providing a length of autologous tissue on the superior surface of the arch to allow for growth of the arch (Fig. 87.12). The ventriculotomy and closure of the ventricular septal defect are then performed in the usual manner, and the arch of the aorta is reconstructed with a patch of pulmonary homograft to provide unobstructed flow through the arch of the aorta and to decrease tension on the anastomosis to decrease the risk of bleeding or distal coarctation (Fig. 87.13). The pulmonary artery reconstruction is then performed as in simple truncus arteriosus.

Repair of Truncus Arteriosus with Truncal Valve Repair or Replacement

Newborns who have significant truncal valve stenosis and insufficiency represent a higher risk subset for neonatal repair. Fortunately, significant truncal

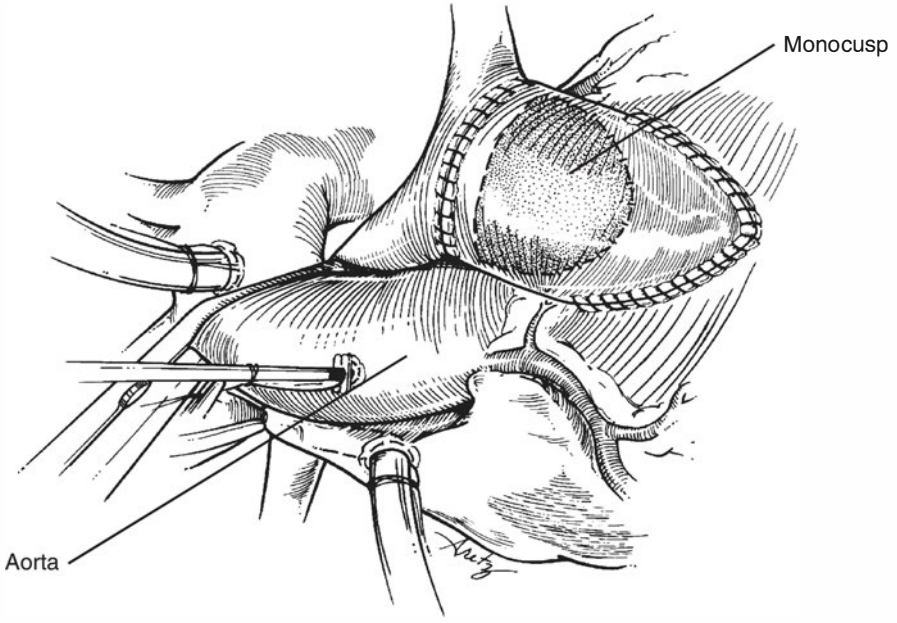


Fig. 87.10. After reconstruction with a monocusp, the pericardial patch closes against the superior margin of the ventricular incision or the base of the autologous reconstruction of the right ventricular outflow tract and can limit pulmonary insufficiency.

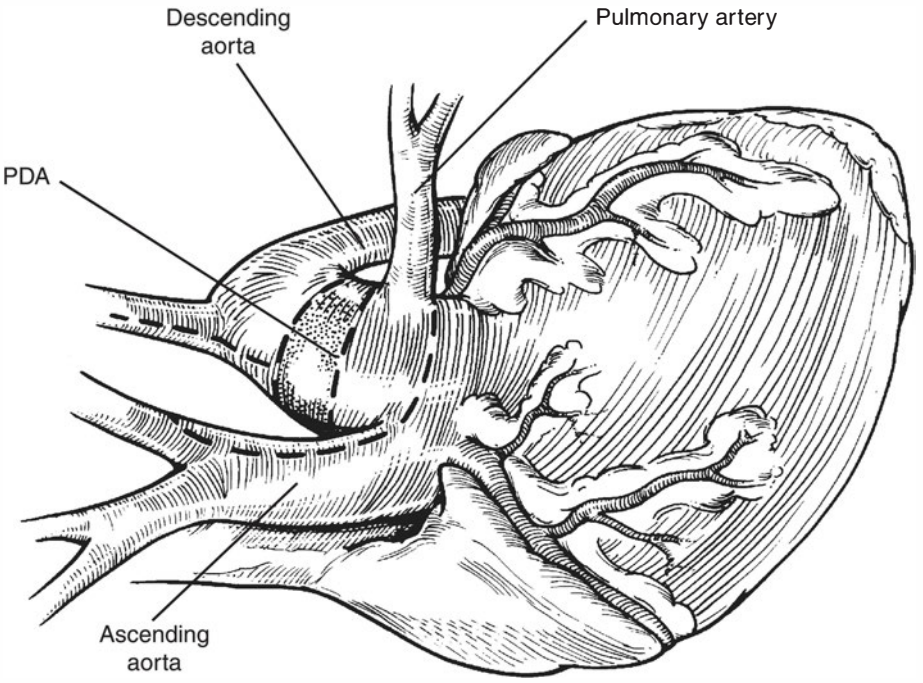


Fig. 87.11. Repair of interrupted aortic arch with truncus arteriosus. A vertical incision is made in the ascending aorta and brachiocephalic vessels onto the origin of the left carotid artery. This incision is placed medially and then carried transversely across the truncus to the base of the left pulmonary artery origin. The pulmonary bifurcation is then excised from the back of the truncus. The ductus arteriosus is ligated distal to the pulmonary bifurcation, and ductal tissue is excised from the descending aorta. An incision is then made vertically in the left subclavian artery and onto the descending aorta beyond any ductal tissue remnants. PDA, patent ductus arteriosus.

valve stenosis is rare, and only in the most severe forms of truncal stenosis valve replacement is necessary at primary operation. Often the elimination of the large left-to-right shunt seen in these patients decreases the flow across the truncal valve, and what appears to be a significant level of stenosis preoperatively may be acceptable after the ventricular septal defect is closed and the truncal repair has been completed. Thus, primary valve replacement for repair of truncus arteriosus should be extremely rare.

A more common situation is the presence of significant truncal valve insufficiency in association with unrepaired truncus arteriosus with or without interrupted aortic arch. Over the last several years, an increasing experience has been reported of valve repair for truncal valve regurgitation at the initial repair of truncus arteriosus or as a secondary procedure. In patients in whom the truncal valve is quadricuspid and there is significant prolapse or abnormality of a single cusp, resection of the cusp and recreation of a trileaflet truncal valve may result in significant improvement in truncal valve insufficiency. In this repair, after division of the truncal root, an incision is made in the aortic wall immediately adjacent to the commissural attachment of the valve leaflet that is to be excised and the incision carried across the valve annulus obliquely. A second incision is made in the more anterior commissural attachment of this same valve cusp, and the cusp is excised with a portion of the truncal wall. Primary repair of the defect then results in a trileaflet valve, which may have less central insufficiency and a more normal distribution of forces on the valve sinuses, aiding closure of the valve centrally (Fig. 87.14A–C). In other cases, if the truncal valve leaflets are not obviously abnormal, or if there is an obvious area of truncal regurgitation at a commissural attachment, closure of the commissure between adjacent truncal valve leaflets with or without leaflet augmentation and annuloplasty may result in significant improvement in truncal insufficiency (Fig. 87.14D).

Often the valve leaflet that needs to be excised in patients with truncal regurgitation and quadraleaflet truncal valve is the leaflet adjacent to the left coronary ostium. In these cases, excision of the leaflet can be performed as described in Figure 87.14E and 87.14F. However, the left coronary artery needs to be mobilized with a button on the aortic wall prior to excision of the valve leaflet and then reimplanted into a suitable site in the truncal root as is often performed with the arterial switch procedure. With these techniques, any of the leaflets of the truncal valve can be excised, although excision of leaflets adjacent to the conduction tissue in the anterolateral aspect of the aorta can be associated with the development of heart block, and therefore, it is preferable to excise noncoronary leaflets or left-sided leaflets if possible.

In the situation of truncus repair with truncal valve replacement, the operation is typically performed with the patient on

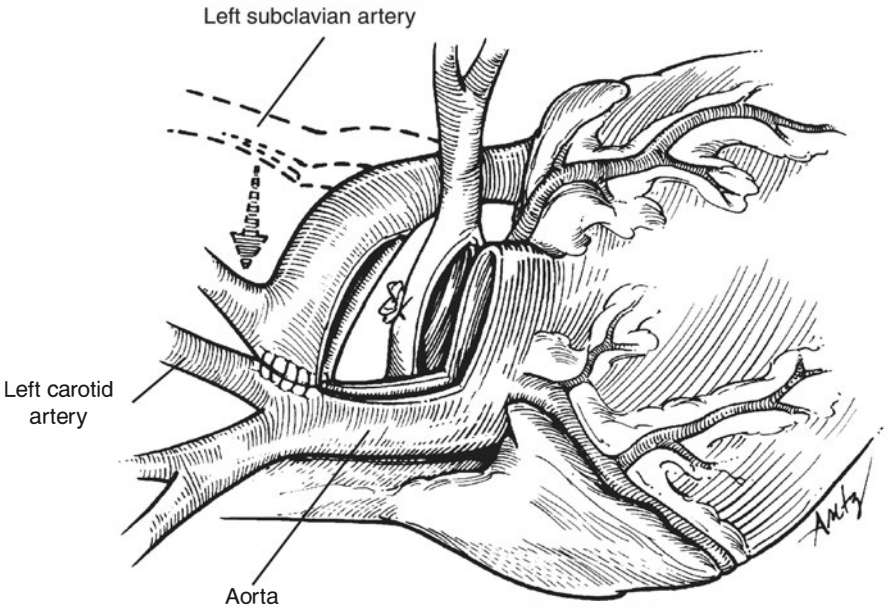


Fig. 87.12. The arch is reconstructed by direct anastomosis of the origins of the subclavian and carotid vessels with absorbable suture to create an autologous superior aspect to the arch reconstruction.

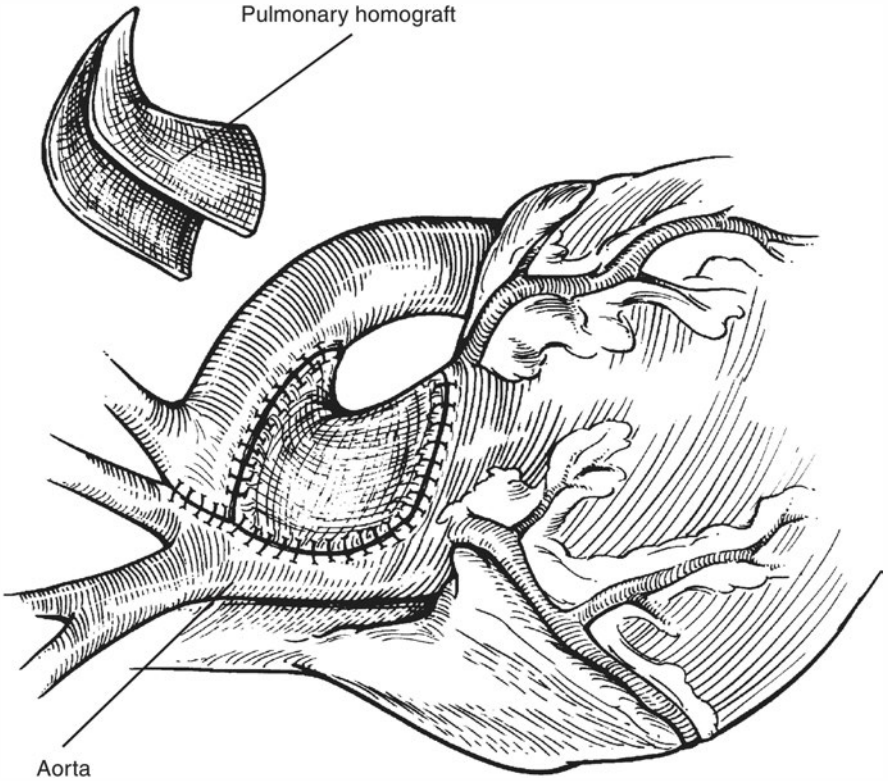


Fig. 87.13. A patch of pulmonary homograft material is then used to create arch augmentation beyond all ductal tissue to the proximal trunk. In this manner, an unobstructed arch is created with the least amount of tension on the suture lines, which prevents compression of the pulmonary bifurcation from the descending aorta and decreases the risk of coarctation of the arch anastomosis.

continuous cardiopulmonary bypass or with intermittent circulatory arrest. Bicaval cannulation is used and the aortic cannulation performed as distally as possible. The left heart is vented through the right superior pulmonary vein across the mitral valve to maintain decompression during cooling and rewarming. The aorta is clamped and cardioplegic solution administered directly into the truncal root if there is only modest truncal insufficiency or directly into the coronary arteries after the truncal vessel is opened if severe insufficiency is present. Alternatively, cardioplegia can be induced in a retrograde manner. In our experience, however, direct cannulation of the coronary ostia is possible in even very small neonates, permitting adequate distribution of cardioplegic solution. The truncus is divided transversely at the level of the takeoff of the pulmonary bifurcation and the bifurcation mobilized freely. An incision is then made vertically across the truncal valve annulus into the right ventricle, widely opening the right ventricular outflow tract (Fig. 87.15). The right and left coronary ostia are mobilized with a button of aortic wall for anastomosis to the reconstruction. The truncal valve is then excised as in Figure 87.16. The ventricular septal defect is readily apparent. We have generally preferred aortic allografts for truncal root replacement as in the early postoperative period high systemic pressures can lead to pulmonary homograft dilation and valvar insufficiency. When root replacement is required, the proximal anastomosis of the allograft is performed to the annulus of the truncal valve posteriorly. This suture line is created for approximately one-half to two-thirds of the circumference of the truncal valve annulus (Fig. 87.17). The ventricular septal defect is then closed with a patch of Dacron or PTFE material, baffling the left ventricular flow to the homograft valve. Thus, the superior margin of the ventricular septal defect patch is sewn to the base of the valve allograft anteriorly, reconstructing the left ventricular outflow tract. At suitable sites on the allograft, buttons of tissue are excised and the coronary arteries reimplanted as for the arterial switch operation for transposition of the great arteries (see Chapter 79). The distal anastomosis of the allograft to the ascending aorta is then completed. At this point, additional cardioplegic solution is injected into the aortic root to ensure that the suture lines are hemostatic and that no insufficiency of the allograft valve is present. The right ventricular reconstruction is then performed (Fig. 87.18), with an allograft valve from the right ventricular incision to the pulmonary bifurcation augmented with a gusset of PTFE material if necessary or by one of the other alternative techniques described previously. If truncal valve replacement is required later after initial repair, it is performed as in homograft root replacement (see Chapter 92).

A recent review of the Society of Thoracic Surgeons (STS) congenital heart surgery database showed that patients undergoing truncal valve surgery had a significantly higher mortality, particularly in the subset of patients who had associated interrupted aortic arch compared with patients undergoing isolated truncus arteriosus repair.

POSTOPERATIVE MANAGEMENT

The management of infants after repair of truncus arteriosus is primarily directed at minimizing pulmonary vascular resistance and right heart dysfunction. Sedation and occasionally paralysis with hyperventilation are standards of postoperative management. Low-dose inotropic support with phosphodiesterase inhibitors may be necessary to improve the right ventricular function in the early postoperative period. With these techniques, pulmonary hypertensive crises and acute right heart dysfunction have been relatively uncommon in our more recent series. However, significant

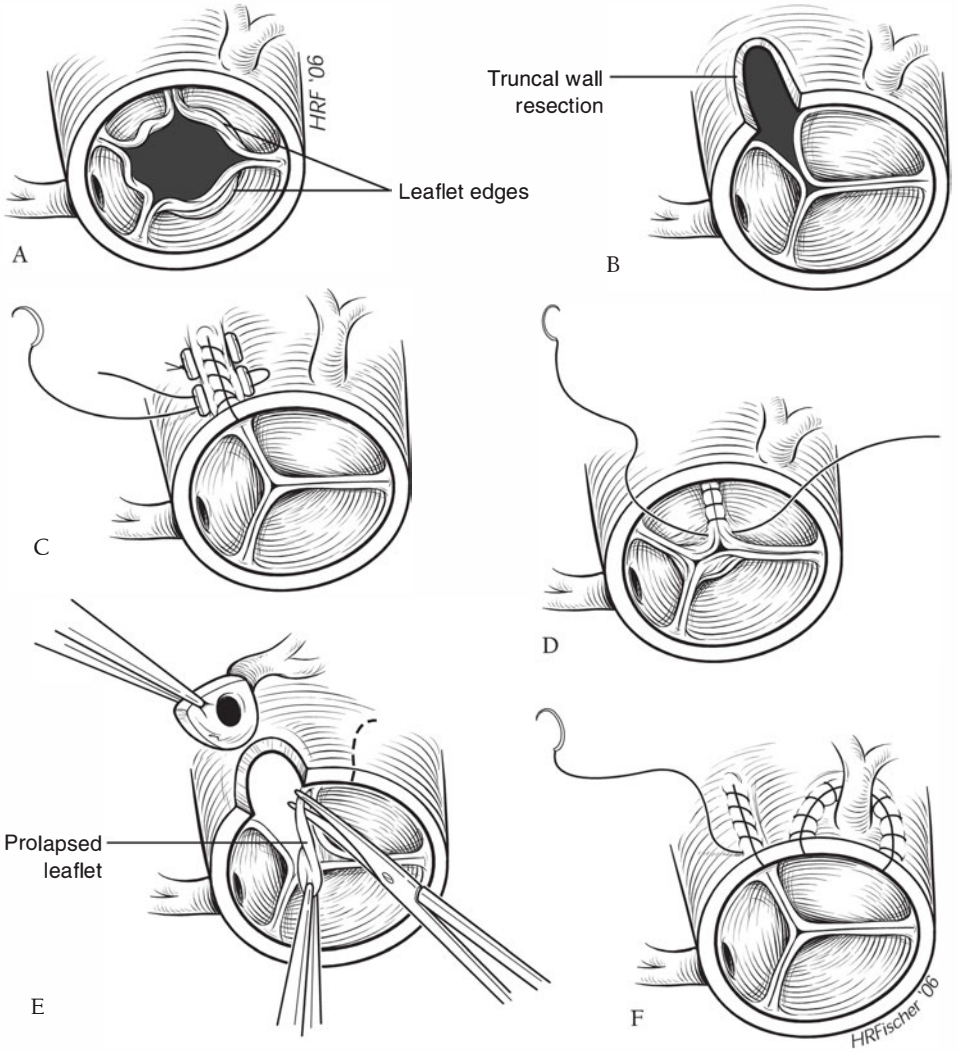


Fig. 87.14. Repair of truncal valve insufficiency. (**A**) The truncus is transected, and the leaflet to be excised is identified. Typically, a quadricuspid valve is present, and there may be a significant prolapse of one leaflet. An incision is made adjacent to the commissural attachment of the leaflet to be excised and is carried down across the truncal valve annulus obliquely. (**B**) A second incision is made anteriorly in a similar manner, and the valve leaflet along with a 1- to 2-mm section of the valve annulus is excised. (**C**) Primary closure of the valve annulus and the aortic wall then results in coaptation of the commissural attachments of the remaining truncal leaflets and decreases the magnitude of central insufficiency, improving the central coaptation between the remaining truncal valve leaflets. (**D**) In patients with truncal insufficiency in whom there is prolapse or dysplasia of a truncal valve leaflet resulting in localized regurgitation, closure of the commissural attachments between valve leaflets may be effective. The truncus is divided, and the truncal valve is examined from above. If areas of deficiency between valve leaflets at the commissures are identified, the commissure can be closed with interrupted or running sutures of monofilament PTFE material. We prefer PTFE suture, which seems to cut through the very delicate valve leaflet tissues less than other types of suture material. (**E**) In patients in whom the valve leaflet to be excised is associated with an adjacent coronary ostium, mobilization of the coronary ostium with excision of a button of aortic wall allows the valve leaflet and annulus to be excised as in panel B, permitting primary reconstruction of a trileaflet valve. (**F**) The coronary button can then be reimplanted at a suitable site on the aortic wall either to a punch opening or to a medially based flap incision in the aortic wall.

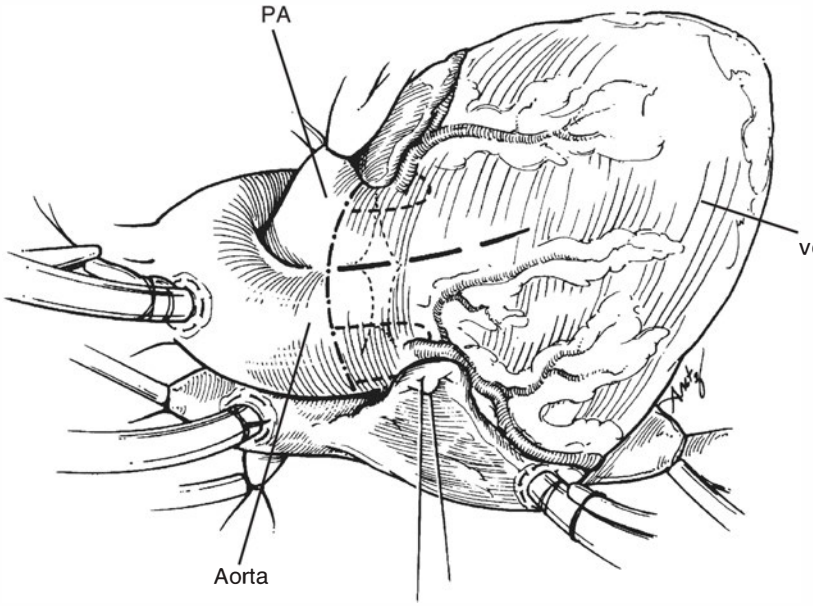


Fig. 87.15. Repair of truncus arteriosus with truncal valve replacement. After circulatory arrest or with continuous bypass, the aorta is cross-clamped and cardioplegic solution is injected into the truncal root. The pulmonary arteries are snared to allow flow of cardioplegic solution into the coronary arteries if possible. If truncal insufficiency is severe, the truncus is opened superiorly as noted in this diagram and then cardioplegic solution is injected into the left and right coronary ostia under direct vision. The right and left coronary ostia are excised with a button of aortic wall, and a vertical incision is made down across the truncal valve annulus into the right ventricle. PA, pulmonary artery.

right ventricular dysfunction or unmanageable pulmonary hypertension with low cardiac output may result in severe hemodynamic instability and on rare occasion has been successfully managed with extracorporeal membrane oxygenation support for several days.

RESULTS

Although mortality rates for truncus arteriosus repair were as high as 60% to 70% until the early 1980s, the trend toward earlier repair and improvements in surgical and postoperative management techniques have resulted in a progressive improvement in the operative results such that survival rates of >95% are anticipated in simple truncus arteriosus. The association of severe truncal insufficiency or interrupted aortic arch is a relative risk factor for early mortality; however, good results have been achieved in these subsets of patients also. Patients who survive the perioperative interval have surprisingly good long-term results after repair of truncus arteriosus. Recent evaluation of the long-term results from San Francisco has shown a significant need for reoperation for conduit revision; however, the rates of late morbidity and mortality with this lesion are very low, and the long-term functional results are excellent. Durability of truncal valve repair has improved with contemporary techniques, although truncal valve surgery remains a significant risk factor for death.

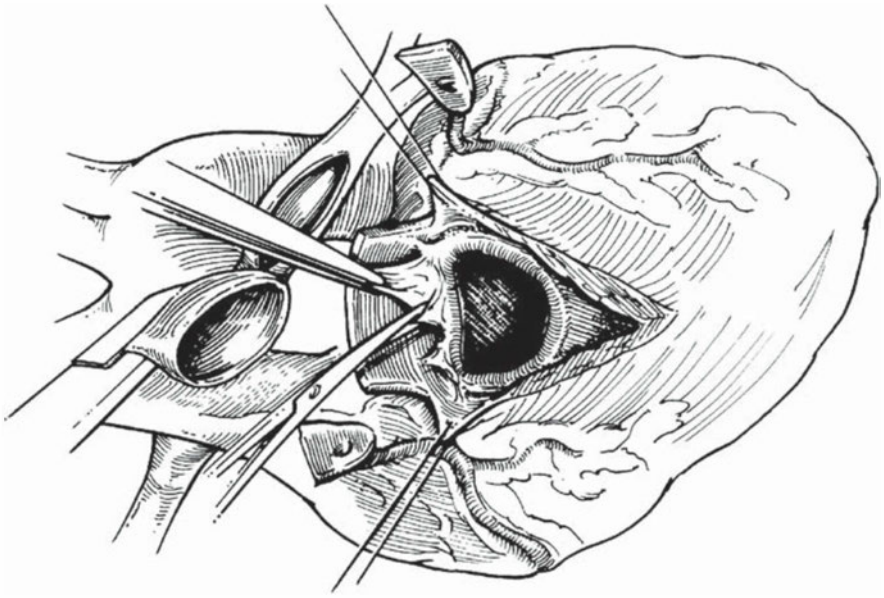


Fig. 87.16. The truncal valve is excised, and then a pulmonary homograft is used to replace the truncal valve sewn to the truncal valve annulus over the posterior one-half to two-thirds of its circumference.

CONCLUSION

Whereas truncus arteriosus has been associated with a very significant early and late mortality in the past, the repair of truncus arteriosus in the neonatal period has been associated with a progressive improvement in results. Although several innovative techniques of repair have been developed to obviate the need for use of allograft conduits for outflow tract reconstruction of the right ventricle, we believe that the presence of a competent pulmonary valve early after repair leads to improved postoperative hemodynamic stability. The low morbidity and mortality from late conduit revision have supported this approach. Nevertheless, the use of an allograft conduit in a neonate will lead to a virtually certain need for operative reintervention for conduit change; therefore, the use of autologous tissue in right ventricular outflow tract reconstruction has inherent appeal. Improvements in postoperative management of neonates after complex reconstructive surgery have resulted in significant improvement in the prognosis for children with truncus arteriosus complex, even those with significant additional abnormalities including truncal valve stenosis and regurgitation or interrupted aortic arch.

SUGGESTED READINGS

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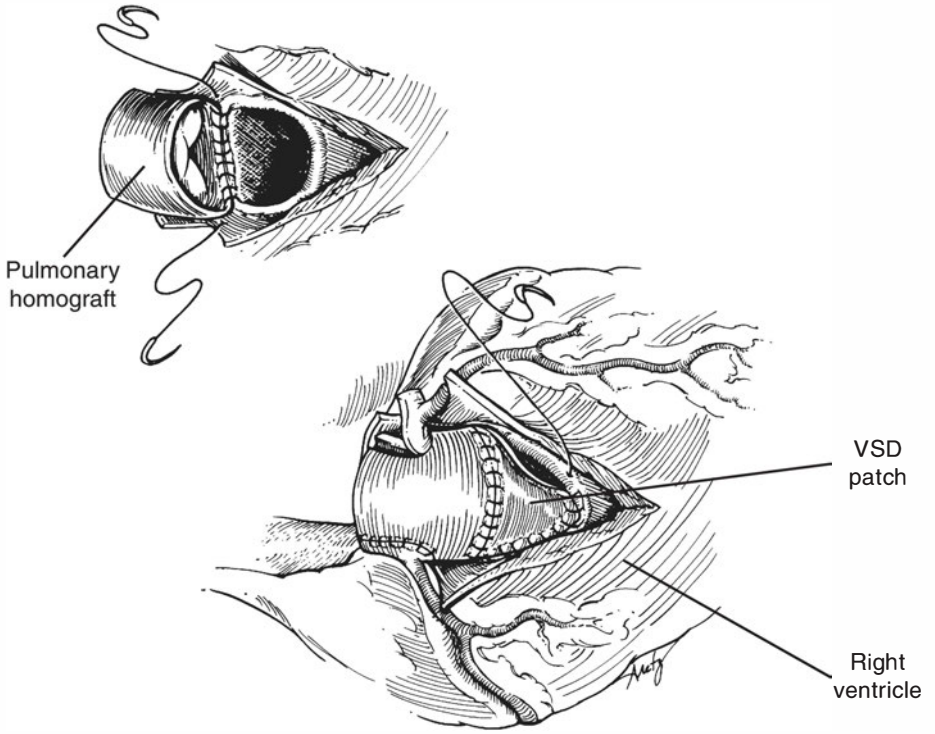


Fig. 87.17. The anterior circumference of the pulmonary homograft is secured with a polytetrafluoroethylene (Gore-Tex) or a polyethylene terephthalate (Dacron) patch sewn to the margins of the ventricular septal defect (VSD) up to the pulmonary homograft, reconstructing the left ventricular outflow tract. The coronary ostia are then reimplanted into the pulmonary homograft in the usual manner and the distal anastomosis to the ascending aorta created.

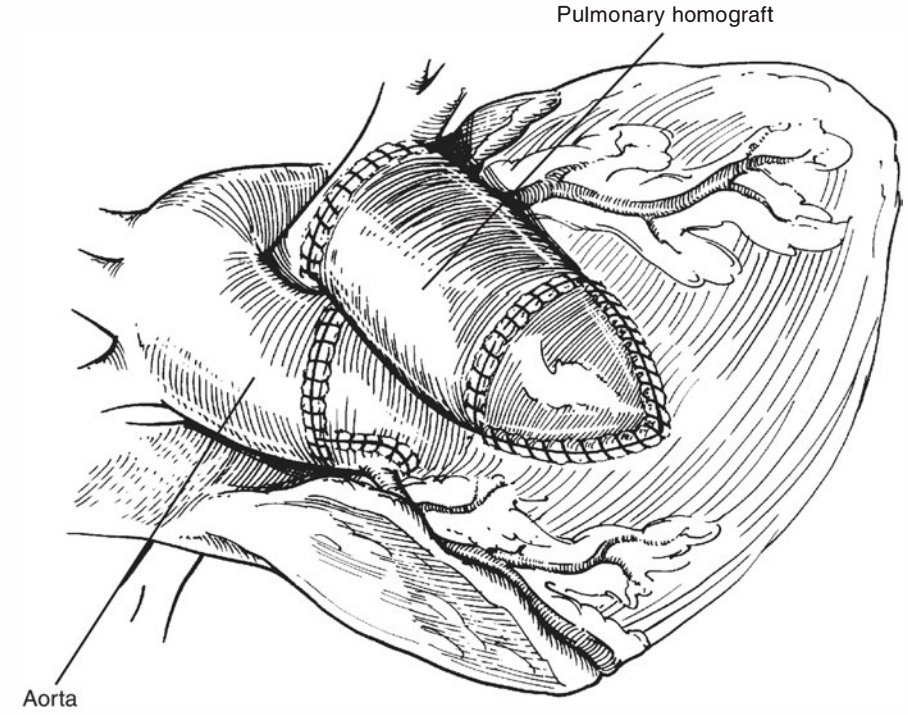


Fig. 87.18. The right ventricular outflow tract is reconstructed.

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EDITOR'S COMMENTS

Truncus is a highly lethal defect that must be repaired in infancy. As these authors have described, Dr. Ebert presented the first large series of these patients who were operated on with an acceptable surgical mortality.

The best parts of this description are the emphases on safety during the initial repair. The importance of snaring the pulmonary artery if the patient is overcirculated and the possibilities of pulmonary overcirculation doing cardiopulmonary bypass are well described. In addition, the issues with the coronaries are also described. The left main coronary is often quite high and one must be careful to look for this when transecting the aorta and removing the pulmonary artery bifurcation.

We have differed from Dr. Spray and used pulmonary homograft early on. He has suggested aortic homograft because of the proper curve. He makes a good point but the early calcifications of these conduits are worrisome to us. Finally, I think there is a place for the percutaneous pulmonary valve in the reoperative situation if the conduit is big enough.

ILK

88 Double Outlet Ventricles

Kirk R. Kanter

DOUBLE-OUTLET RIGHT VENTRICLE

Definition

Double-outlet right ventricle (DORV) refers to a heterogeneous group of cardiac malformations characterized by an abnormal ventriculoarterial connection in which both great arteries are related to the right ventricle. Although the term *double-outlet right ventricle* can be correctly applied to hearts with atrioventricular discordance (e.g., congenitally corrected transposition of the great arteries) or to hearts with univentricular atrioventricular connections (e.g., double-inlet left ventricle), for simplicity of discussion, only hearts with atrioventricular concordance and two adequate ventricles are discussed in this chapter.

The actual definition of what constitutes a DORV has been the source of controversy in the literature. Although some have required the presence of bilateral infundibula or atrioventricular valve–semilunar valve discontinuity (most commonly mitral-aortic discontinuity), these criteria are not essential in establishing the diagnosis of DORV. From a surgical perspective, it is most useful to adopt the "50% rule" in defining DORV. With this rule, a heart is termed DORV if >50% of both great arteries arise from the right ventricle. Usually, all of one great artery and ≥50% of the other great artery arise from the right ventricle in DORV.

Classification

A ventricular septal defect (VSD) is almost always present with DORV. Based on the work of Lev and colleagues, DORV is classified into four groups based on the relationship of the VSD to the great arteries (Table 88.1): subaortic, subpulmonary, doubly committed, and noncommitted (remote).

Subaortic Ventricular Septal Defect

DORV with subaortic VSD (Fig. 88.1A) is the most common group of DORV. It may or may not be associated with pulmonary stenosis. The presentation without pulmonary stenosis is similar to that of a child with a large VSD (heart failure). If there is pulmonary stenosis, it is usually infundibular, so the clinical presentation in this subgroup is similar to that of tetralogy of Fallot (cyanosis, hypercyanotic episodes). Accordingly, if a patient is not a candidate for complete repair at the time of presentation because of size, clinical condition, or other variables, palliative procedures in DORV with subaortic VSD without pulmonary stenosis would be pulmonary artery banding; in children with pulmonary stenosis, a systemic-to-pulmonary artery shunt would be appropriate.

Subpulmonary Ventricular Septal Defect

DORV with subpulmonary VSD (the Taussig–Bing anomaly) is the second most common group of DORV (Fig. 88.1B). Because of the location of the VSD, oxygenated left ventricular blood preferentially streams through the VSD into the pulmonary artery and desaturated right ventricular blood streams into the aorta as in transposition of the great arteries with VSD. These children present with cyanosis and heart failure. Associated coarctation of the aorta occurs commonly; aortic arch obstruction and subaortic stenosis can also be associated with the Taussig–Bing anomaly. Because DORV with subpulmonary VSD is prone to early development of pulmonary vascular obstructive disease, intervention in infancy is usually necessary. In addition to repair of the aortic coarctation and arch hypoplasia, if present, balloon atrial septostomy is commonly needed to improve mixing of oxygenated blood at the atrial level as in transposition of the great arteries. Although palliation can be accomplished with pulmonary artery banding, it is preferable to proceed with complete repair in infancy.

Doubly Committed Ventricular Septal Defect

In DORV with doubly committed VSD, the VSD is immediately beneath both the pulmonary artery and the aorta (Fig. 88.1C). Usually, the infundibular septum is absent or hypoplastic. As in DORV with subaortic VSD, there may be associated pulmonary stenosis. Thus, clinical presentation and surgical approach would be similar to that for DORV with subaortic VSD with or without pulmonary stenosis.

Noncommitted Ventricular Septal Defect

In the final group of DORV, the VSD is committed neither to the aorta nor to the pulmonary artery (Fig. 88.1D). The remote location of the VSD may be in the inlet septum as with atrioventricular septal defects or a muscular VSD in the trabecular septum. Pulmonary stenosis may be present. The presentation and surgical palliation of DORV with noncommitted VSD would be similar to that for DORV with subaortic VSD with or without pulmonary stenosis.

Surgical Techniques

The goal of surgical repair in DORV is to achieve a biventricular repair utilizing the left ventricle as the systemic ventricle with unobstructed right and left ventricular outflow tracts. Usually, this can be accomplished within the first 6 to 12 months of life, thus obviating the need for palliative procedures. If it is anticipated that the final repair will require an extracardiac valved conduit or a complicated intraventricular tunnel, it is reasonable to delay correction to allow for growth by palliating, if necessary, with a pulmonary artery band or a systemic-to-pulmonary artery shunt. Also, in children who eventually will need a Fontan procedure, it is critical to protect the pulmonary vascular bed early on with appropriate palliation.

Table 88.1 Classification of Double-Outlet Right Ventricle

VSD relationship	Pulmonary stenosis	Clinical mimicry
Subaortic	Absent	VSD
Subaortic	Present	TOF
Subpulmonary	Absent	TGA/VSD
Doubly committed	Absent	VSD
Doubly committed	Present	TOF
Noncommitted	Absent	VSD
Noncommitted	Present	TOF

TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

Roughly 10% of children with DORV will have a restrictive VSD. By definition, this means that outflow from the left ventricle is obstructed, and therefore, early repair with enlargement of the VSD is mandated since spontaneous closure of the VSD, rather than being curative as in isolated VSD, would be fatal in DORV. Pulmonary artery banding is certainly not appropriate in patients with DORV and a restrictive VSD.

Intraventricular Tunnel Repair of Double-Outlet Right Ventricle with Subaortic Ventricular Septal Defect without Pulmonary Stenosis

Repair of DORV with a subaortic VSD is accomplished by creating an intraventricular tunnel channeling left ventricular blood through the VSD to the aorta (Fig. 88.2). This is facilitated by the use of a polyester (Dacron) or collagen-impregnated polyester tube graft corresponding to the size of the aorta. This is opened longitudinally so that about two-thirds of its circumference is available (Fig. 88.2A). The advantage of using a tube graft for the intraventricular tunnel is that the corrugations keep the required curve in the baffle to allow unobstructed left ventricular outflow. A flat Dacron or polytetrafluoroethylene (Gore-Tex) patch that one would use to close a simple VSD would be prone to kinking

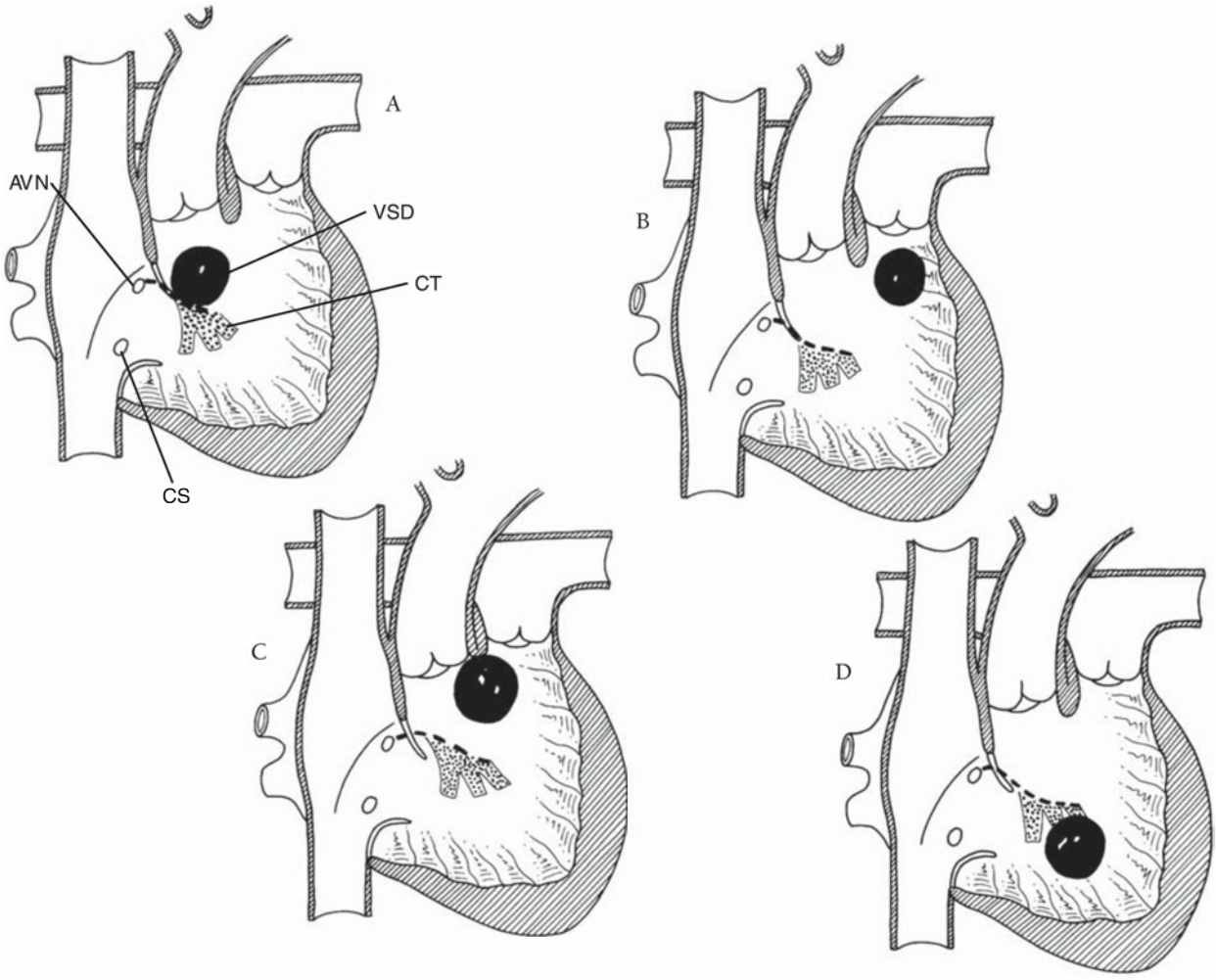


Fig. 88.1. Types of double-outlet right ventricle classified by the relationship of the ventricular septal defect (VSD) to the great arteries. The location of the atrioventricular node and conduction tissue is depicted. (**A**) Subaortic VSD. (**B**) Subpulmonary VSD. (**C**) Doubly committed VSD. (**D**) Noncommitted VSD. AVN, atrioventricular node; CS, coronary sinus; CT, conduction tissue.

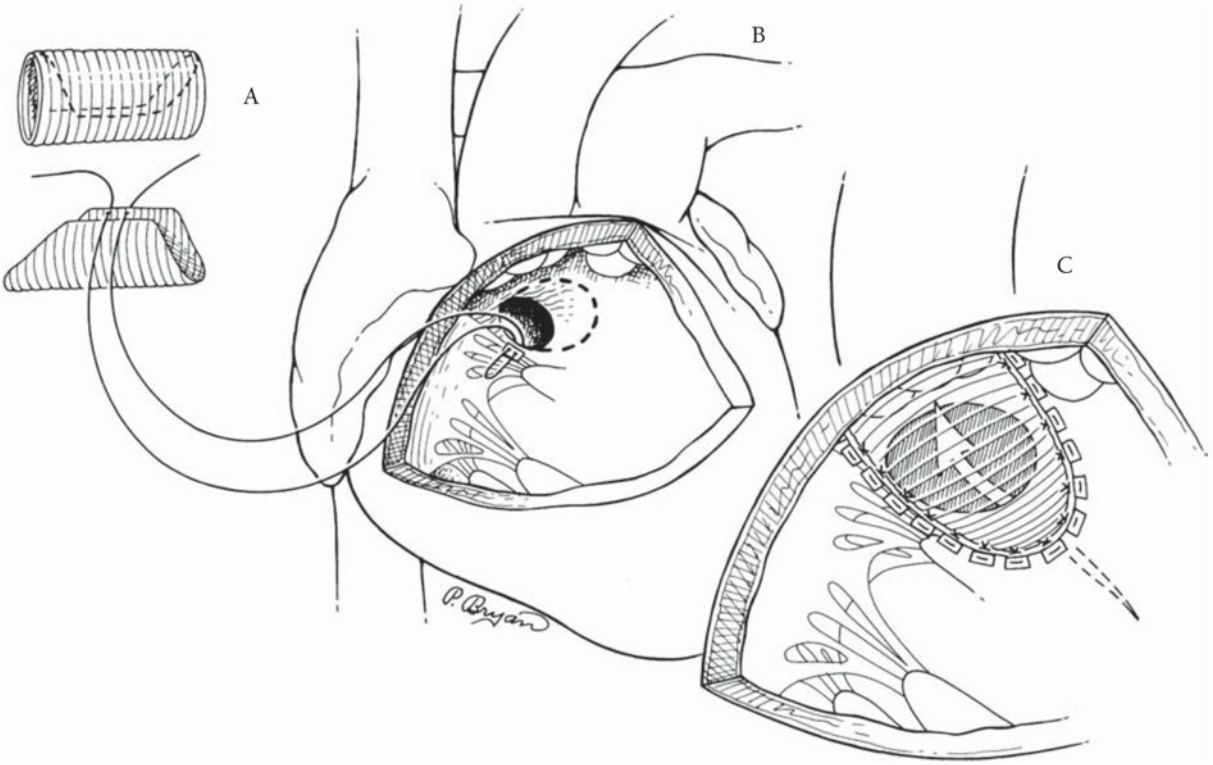


Fig. 88.2. Intraventricular tunnel repair of double-outlet right ventricle with subaortic ventricular septal defect (VSD) without pulmonary stenosis. (**A**) A (polyester) Dacron or Hemashield tube graft the size of the aorta is opened longitudinally for use as the patch. Suturing is started through the midportion of the graft and the base of the anteroseptal commissure of the tricuspid valve. (**B**) The VSD is exposed through a right ventriculotomy. The safe area for the enlargement of the VSD is shown with a dashed line. (**C**) The intraventricular tunnel is completed with pledgetted or continuous sutures. Care is taken to maintain orientation of the tube graft so one end is at the anterior-inferior margin of the VSD and the other end is anterior to the aortic valve. The arrow indicates flow of blood from the left ventricle through the enlarged VSD and tunnel to the aorta.

and would create obstruction unless the geometry and the size of the patch were exactly perfect.

After routine establishment of cardiopulmonary bypass with bicaval cannulation and cardioplegic arrest, the intracardiac anatomy is carefully inspected through a right atriotomy. The VSD is visualized through the tricuspid valve and its relationship to the aorta is confirmed. If there is any suspicion preoperatively or intraoperatively that the VSD is smaller than the aorta, it should be enlarged. This can be accomplished through the tricuspid valve or the right ventricle with either a transverse or a longitudinal right ventriculotomy. The VSD is enlarged superiorly and anteriorly (Fig. 88.2B), thus resecting some of the infundibular septum. Enlarging the VSD posteriorly and superiorly through the ventriculoinfundibular fold runs the risk of going outside the heart. The conduction tissue runs inferiorly and of course should be avoided (Fig. 88.1A).

Once the VSD is enlarged (if necessary), the Dacron tube graft is oriented so that the longitudinal axis of the graft corresponds to an imaginary line from the anterior-most portion of the aorta to the anterior-inferior limit of the VSD (Fig. 88.2C). It is helpful to place the first suture through the base of the tricuspid valve leaflet at the anteroseptal commissure, and then through the midportion of the tube graft (Fig. 88.2B). About one-third of the VSD sutures are placed along the posterior and inferior rim of the defect through the tricuspid valve, with care taken to avoid the conduction tissue, with several of these pledgetted sutures on the atrial side of the septal leaflet of the tricuspid valve. These sutures are passed through the Dacron tube graft, which is seated down and the sutures tied. The remainder of the circumference of the VSD is closed through the right ventriculotomy, with care being taken to maintain proper orientation of the patch (Fig. 88.2C). Anteriorly and superiorly, it may be advantageous to anchor some of the pledgetted sutures through the anterior wall of the right ventricle above the aortic valve, as is done in a Rastelli procedure. Rather than using interrupted pledgetted sutures, the VSD can be closed with a continuous suture technique completely through the right ventriculotomy.

If it appears that the intraventricular tunnel is bulging into the right ventricular outflow tract, the right ventriculotomy is closed with a patch of autologous pericardium to avoid right ventricular outflow tract obstruction.

Repair of Double-Outlet Right Ventricle with Subaortic Ventricular Septal Defect and Pulmonary Stenosis

In the group of patients with DORV with subaortic VSD and pulmonary stenosis, it is useful to mark the planned right ventriculotomy incision with stay sutures before cardioplegic arrest after carefully identifying the course of the epicardial coronary arteries (Fig. 88.3A). The intraventricular tunnel repair of the VSD is identical to that used for patients with DORV and subaortic VSD without pulmonary stenosis (Fig. 88.3B). If a major coronary artery crosses the right ventricular outflow tract or if the pulmonary vascular resistance is elevated or if there is distal pulmonary arterial obstruction, a valved conduit should be used to establish right ventricle–pulmonary artery continuity. The pulmonary trunk is divided, with the proximal main pulmonary artery oversewn (Fig. 88.3B). The proximal portion

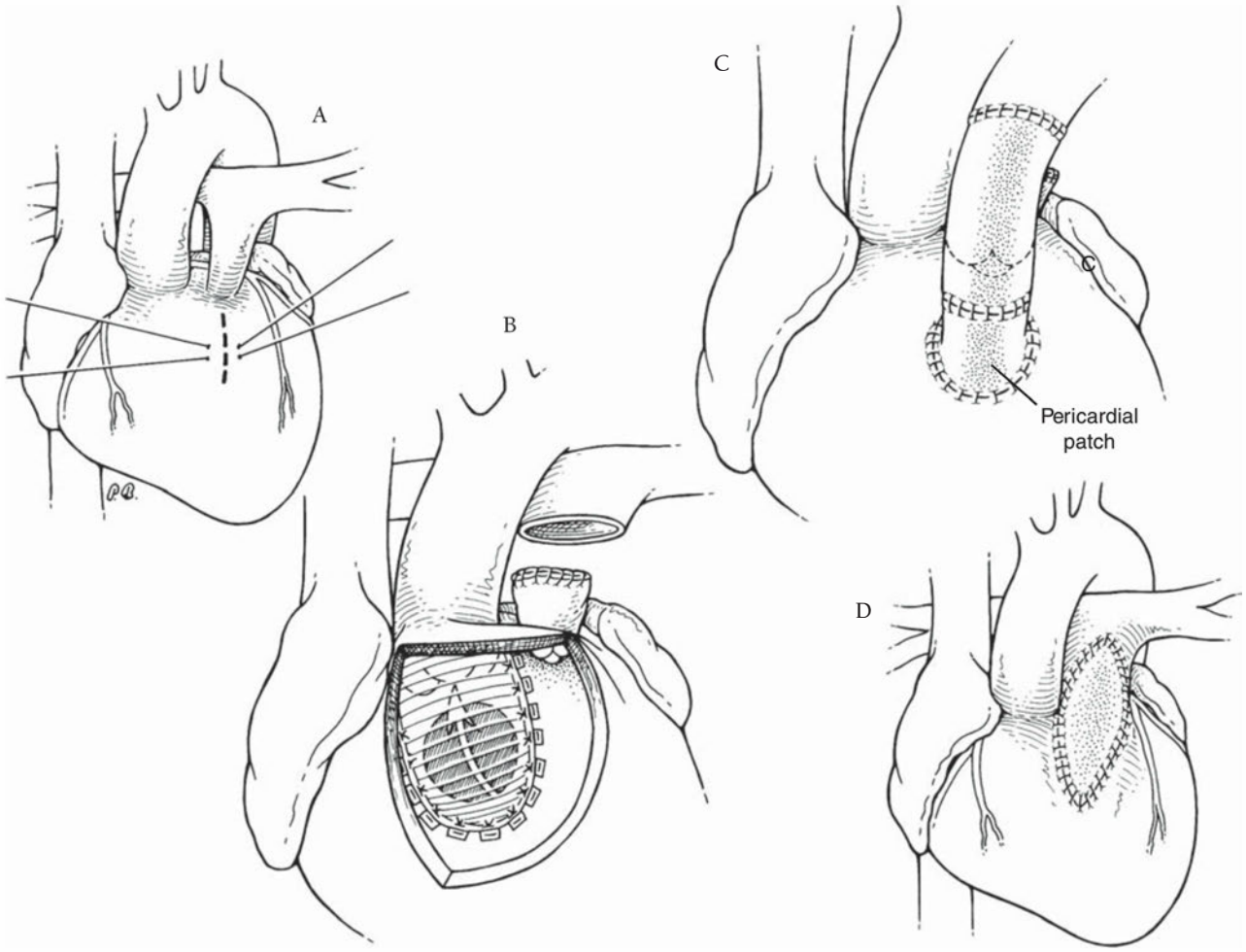


Fig. 88.3. Repair of double-outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis. (**A**) The site of the proposed right ventriculotomy is shown by a dashed line. Marking sutures are placed before cardioplegic arrest to aid with retraction and to properly orient the ventricular incision. (**B**) An intraventricular tunnel is fashioned as in Figure 88.2. The main pulmonary artery is divided and the cardiac end oversewn. (**C**) Right ventricle-to-pulmonary artery continuity is reestablished with a valved homograft conduit. About one-half of the circumference of the proximal homograft is sutured to the superior aspect of the right ventriculotomy. A piece of pericardium or homograft material is used to roof the gap between the right ventricle and the homograft. (**D**) Alternatively, a nonvalved transannular right ventricular outflow tract patch can be used to relieve the pulmonary stenosis.

of a valved homograft is sewn to the superior aspect of the right ventriculotomy. The homograft is trimmed to the proper length, and an end-to-end anastomosis between the distal homograft and the distal main pulmonary artery (or pulmonary bifurcation if the main pulmonary artery is small) is performed. Finally, the gap between the proximal right ventriculotomy and the proximal homograft is roofed with leftover homograft material or autologous pericardium (Fig. 88.3C). Alternatively, the native pulmonary trunk can be left intact, and the distal homograft conduit is sewn end-to-side to the junction of the native pulmonary trunk and pulmonary bifurcation allowing blood flow through both the stenotic native pulmonary valve and the new conduit.

Often it is possible to establish an adequate right ventricular outflow tract without a valved conduit as is done with repair of tetralogy of Fallot. In this case, after the division of obstructing right ventricular muscle bundles, a transannular outflow tract patch is created using autologous pericardium (Fig. 88.3D). If the pulmonary valve annulus is of good size, a nontransannular patch is adequate after a pulmonary valvotomy is performed. Otherwise, a transannular patch is necessary.

Sakamoto and colleagues have described an interesting transaortic approach for enlarging the VSD and tunneling left ventricular outflow through the VSD to the aorta in two patients with DORV with subaortic VSD and pulmonary stenosis.

Anatomic Repair of Double-Outlet Right Ventricle with Subpulmonary Ventricular Septal Defect

Currently, the preferred surgical repair of DORV with subpulmonary VSD (the Taussig–Bing anomaly) is the anatomic repair (the arterial switch operation). Since coarctation of the aorta is commonly seen in this group, these patients may have had prior coarctation repair with a pulmonary artery band, although simultaneous repair of the coarctation at the time of anatomic repair is now the accepted standard.

The VSD is closed either through a right atriotomy or through a right ventriculotomy channeling left ventricular blood into the pulmonary artery (Fig. 88.4A). The aorta is transected slightly higher than the pulmonary trunk (Fig. 88.4A). If present, the coarctation and associated aortic arch hypoplasia can be corrected at this time with a patch of homograft material using a brief period of profound hypothermia with circulatory arrest or regional low flow perfusion.

Particularly with side-by-side great arterial relationships, the circumflex coronary artery often arises with the right

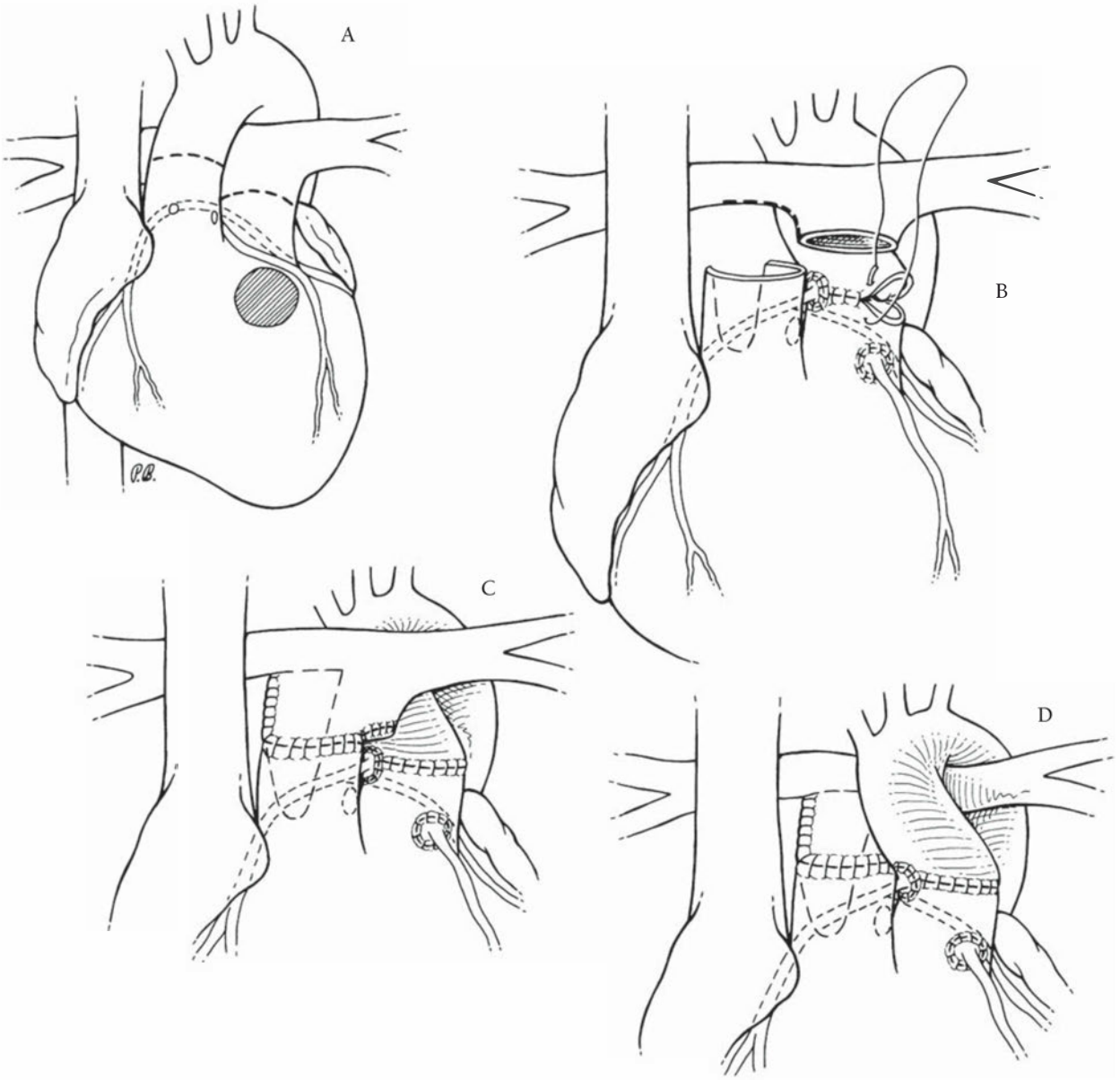


Fig. 88.4. Anatomic repair of double-outlet right ventricle with subpulmonary ventricular septal defect (arterial switch operation). (**A**) The level of transection of the great arteries is shown with a dashed line. With a side-by-side relationship of the great arteries, the aorta should be divided higher than the main pulmonary artery. The ventricular septal defect is closed through a right ventriculotomy or right atriotomy, directing blood to the pulmonary artery. (**B**) The anterior descending coronary ostial button is transferred to an oval defect in the neoaorta. The right coronary–circumflex coronary ostial patch extends to the level of aortic transection as a U-shaped button. It is incorporated into the aortic suture line. The pulmonary bifurcation has been brought anterior to the distal aorta (the LeCompte maneuver). The incision along the undersurface of the right pulmonary artery (RPA) is indicated with a dashed line. (**C**) A large pericardial patch closes the U-shaped defect in the proximal neopulmonary artery and extends onto the undersurface of the RPA. Separate patches close the harvest site of the anterior descending coronary ostial button and the left lateral aspect of the pulmonary trunk. (**D**) With the same technique as in Figure 88.4C, the LeCompte maneuver can be avoided, leaving the pulmonary bifurcation posterior to the ascending aorta.

coronary artery from the right posterior-facing sinus, whereas the anterior descending coronary artery originates from the left posterior-facing sinus. In this situation, the anterior descending coronary artery is excised as a button and transferred to a defect created in the left anterior-facing sinus of the proximal pulmonary trunk (neoaorta; Fig. 88.4B). The right and circumflex coronary ostium is excised as a large U-shaped button. It is transferred above the right anterior-facing sinus of the proximal pulmonary trunk (neoaorta) and incorporated into the suture line between the proximal pulmonary trunk (neoaorta) and the distal ascending aorta (Fig. 88.4B). Positioning this coronary button more superiorly as it is transferred to the left avoids potential kinking of the circumflex coronary artery.

Reconstruction of the right ventricular outflow tract when the great arteries are side by side can cause compression of one of the coronary arteries or predispose to right ventricular outflow tract obstruction if performed exactly as one would with typical transposition of the great arteries, since there is so much offset between the "old" proximal ascending aorta (neopulmonary artery) and the distal pulmonary trunk. This potential problem can be addressed by opening the right lateral aspect of the distal pulmonary trunk onto the undersurface

of the right pulmonary artery (Fig. 88.4B) after performing the LeCompte maneuver (bringing the pulmonary bifurcation anterior to the distal ascending aorta). The U-shaped defect in the "old" proximal ascending aorta at the site of the right and circumflex coronary ostial button is repaired with a patch of autologous pericardium or homograft material that is much wider distally. This increases the circumference of the proximal neopulmonary artery to accommodate for the typical size discrepancy between the smaller proximal aorta and the larger distal pulmonary trunk. This same patch is incorporated into the incision on the undersurface of the right pulmonary artery (Fig. 88.4C) to allow for a generous tension-free pulmonary arterial anastomosis and has the benefit of pulling the new pulmonary trunk right away from the right coronary artery. Both the old site of the anterior descending coronary artery button and the slight overhang on the left lateral aspect of the distal pulmonary trunk are closed with small pericardial or homograft patches. If the U-shaped patch filling the defect in the right posterior-facing sinus is broad enough distally, it accomplishes the same effect as a pantaloon patch for repairing both coronary button defects and is somewhat easier to do.

Alternatively, with side-by-side great arteries, pulmonary artery reconstruction can be performed using the same techniques outlined above without incorporating the LeCompte maneuver (Fig. 88.4D). To do this, it is very important to transect the aorta higher than the pulmonary trunk at the beginning of the repair (Fig. 88.4A).

Intraventricular Repair of Double-Outlet Right Ventricle with Subpulmonary Ventricular Septal Defect

In patients with DORV and subpulmonary VSD with side-by-side great arteries, an intraventricular tunnel repair is feasible. The subpulmonary VSD is exposed through a transverse right ventriculotomy (Fig. 88.5A). Often it is necessary to enlarge the VSD anteriorly and superiorly. The infundibular septum between the aorta and pulmonary artery is usually quite prominent and in fact can cause subaortic obstruction preoperatively. This is excised to provide an unimpeded channel from the left ventricle through the VSD to the aorta (Fig. 88.5B). Usual care must be taken to avoid injury to the conduction tissue (Fig. 88.1B). The intraventricular tunnel is now created using an opened Dacron or Hemashield tube graft as employed with DORV and subaortic VSD (Fig. 88.5C). Although the illustration in Figure 88.5C depicts interrupted pledgetted sutures, a continuous suture technique can also be used.

If the relationship of the great arteries is more anteroposterior than side by side, there is inadequate distance between the tricuspid valve and the pulmonary valve. The intraventricular tunnel will obstruct the subpulmonary region. Therefore, patients with DORV and subpulmonary VSD with anteroposterior great arteries should not undergo an intraventricular tunnel repair; anatomic repair with closure of the VSD to the pulmonary artery and an arterial switch operation are preferable.

Damus–Kaye–Stansel Repair of Double-Outlet Right Ventricle with Subpulmonary Ventricular Septal Defect

There is a subset of patients with DORV and subpulmonary VSD who have significant subaortic stenosis that may not lend itself to adequately successful resection. This would preclude an arterial switch operation, since postoperative right ventricular outflow tract obstruction would result. There are also a very small number of patients with coronary artery anatomy that makes it too risky to perform an arterial switch operation depending on the experience of the individual surgeon (e.g., intramural coronary artery or single coronary artery), although these situations have become less common as techniques dealing with these more challenging coronary abnormalities have evolved. In these situations, an arterial switch operation without coronary translocation (Damus–Kaye–Stansel procedure) is a feasible alternative.

With the Damus–Kaye–Stansel procedure, the subpulmonary VSD is closed either through the right atrium or through a right ventriculotomy incision to direct left ventricular blood through the pulmonary valve (Fig. 88.6A). The pulmonary trunk is transected just proximal to the pulmonary bifurcation. It is critical to avoid distortion of the proximal pulmonary trunk or ascending aorta, which may result in semilunar valve insufficiency. Before cardioplegic arrest, it is helpful to place a marking suture on the right medial aspect of the proximal pulmonary trunk at the planned site of transection. A corresponding marking suture is placed on the left medial aspect of the ascending aorta to define the proximal extent of the aortic incision and to ensure proper orientation of both great arteries. The aorta is opened along its left medial aspect from the previously placed marking suture distally for a length corresponding to the diameter of the pulmonary trunk. The incision on the aorta should start just above the commissural posts of the aortic valve. Attention should be given to carefully identify the coronary arteries and avoid any distortion. An end-to-side anastomosis between the proximal pulmonary trunk and the left medial aspect of the ascending aorta is performed using a continuous suture technique. It is surprising how often this anastomosis can be performed primarily without additional patch augmentation (Fig. 88.6A). However, in children with previous pulmonary artery banding, the level of transection of the pulmonary trunk is at the level of the band. After the scar tissue is excised from the band site, it is usually necessary to augment the end-to-side pulmonary aortic anastomosis with autologous pericardium or homograft material. Right ventricle-to-pulmonary artery continuity is now established with a homograft valved conduit (Fig. 88.6B).

Alternatively, the ascending aorta can be transected at the same level as the pulmonary trunk. A partial side-to-side anastomosis is constructed between the proximal ascending aorta and the proximal pulmonary trunk. The distal ascending aorta is anastomosed to this double-barreled arterial connection, commonly after patch enlargement of the ascending aorta and arch. The advantage of this double-barreled technique compared with the end-to-side anastomosis depicted in Figure 88.6 is less risk of semilunar valve distortion with subsequent insufficiency as well as the ease of enlarging the distal aorta.

With the Damus–Kaye–Stansel procedure, the aortic valve is still connected to the right ventricle. Since aortic pressure is always higher than right ventricular pressure throughout the cardiac cycle, the aortic valve remains closed. Right ventricular output is through the valved conduit into the lower pressure pulmonary arteries rather than through the aortic valve into the higher pressure systemic circulation. Obviously, any significant aortic valve insufficiency would be poorly tolerated, since the regurgitant blood would flow into the right ventricle, causing a left-to-right shunt that could be misdiagnosed as a residual VSD. In the case of significant aortic valve insufficiency, the aortic valve itself can be oversewn primarily without much difficulty. With current advances in the arterial switch procedure and the creation of intraventricular tunnels, it is uncommon to resort to a classical Damus–Kaye–Stansel procedure.

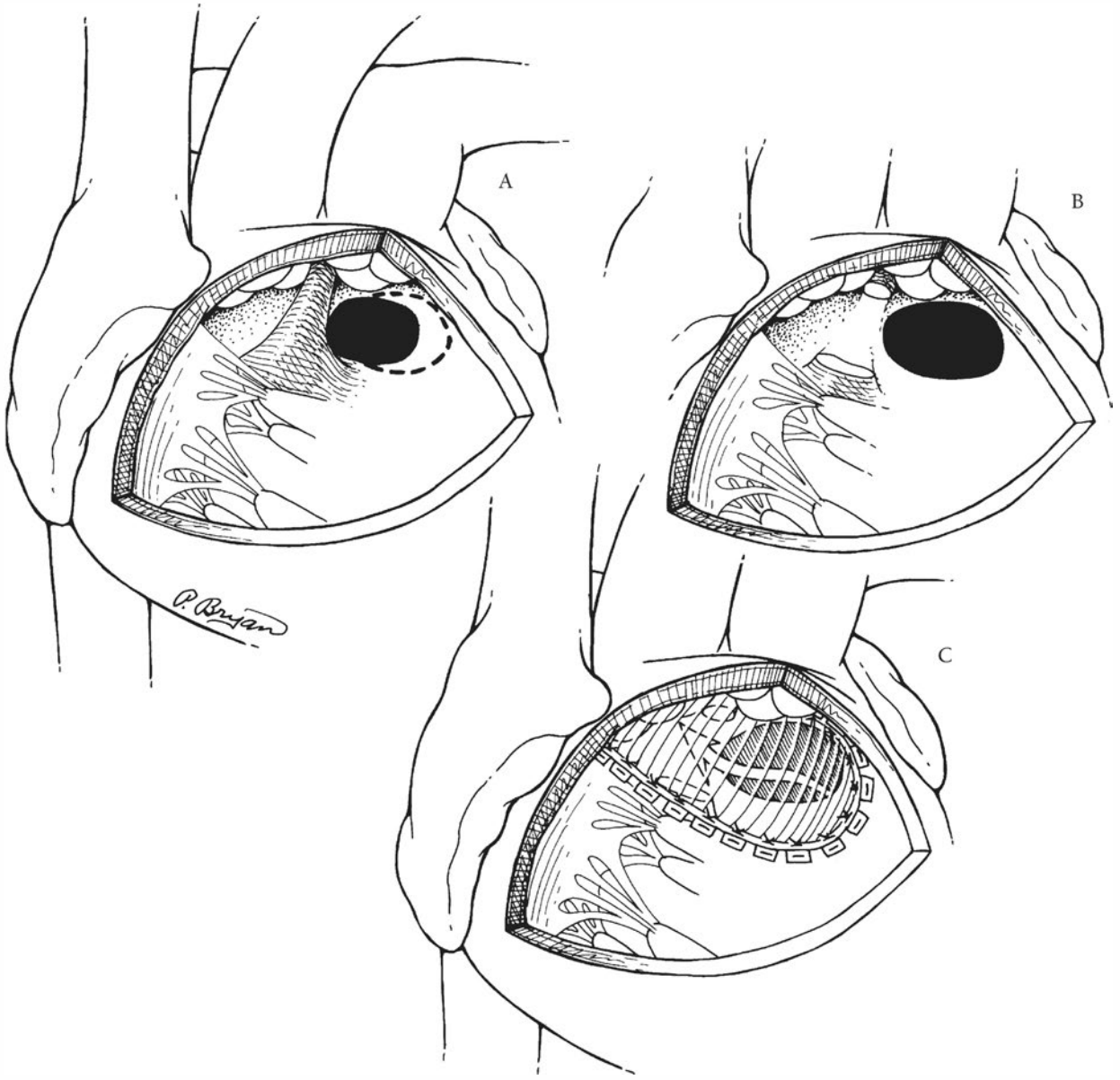


Fig. 88.5. Intraventricular repair of double-outlet right ventricle with subpulmonary ventricular septal defect (VSD). (**A**) The VSD is exposed through a transverse right ventriculotomy. The safe area for the enlargement of the VSD is marked with a dashed line. The prominent infundibular septum is well seen. (**B**) The infundibular septum is partially excised and the VSD is enlarged. (**C**) The intraventricular tunnel is created with an opened polyester (Dacron) tube graft, channeling left ventricular blood through the enlarged VSD to the aorta (*arrow*).

Repair of Double-Outlet Right Ventricle with Doubly Committed Ventricular Septal Defect

Surgical repair of the uncommon variant of DORV with doubly committed VSD is managed in much the same manner as described for DORV with subaortic VSD. The VSD is usually large, so channeling left ventricular blood to the aorta with an intraventricular tunnel does not present much difficulty. If there is pulmonary stenosis or if the VSD patch obstructs flow into the pulmonary artery, it may be necessary to perform a right ventricular outflow tract patch or to use a right ventricle-to-pulmonary artery valved conduit.

Repair of Double-Outlet Right Ventricle with Noncommitted Ventricular Septal Defect

A satisfactory biventricular repair of DORV with noncommitted VSD is more difficult because the remoteness of the VSD necessitates a complex intraventricular tunnel to direct blood from the left ventricle through the VSD to the aorta. If the VSD is in the inlet perimembranous location, the VSD must be carefully inspected for straddling tricuspid valve or even mitral valve tissue, which would preclude biventricular repair. If the VSD is small, it can be safely enlarged superiorly and anteriorly (Fig. 88.7A), since the conduction tissue in a perimembranous inlet VSD courses along the posterior and inferior rim of the defect on the left ventricular side of the septum similar to DORV with subaortic VSD (Fig. 88.1A). The intraventricular tunnel is then fashioned with a patch tailored from a Dacron or Hemashield tube graft (Figs 88.2A and 88.7B). The techniques and precautions described in the section on intraventricular tunnel repair of DORV with subaortic VSD are very much applicable to this repair of

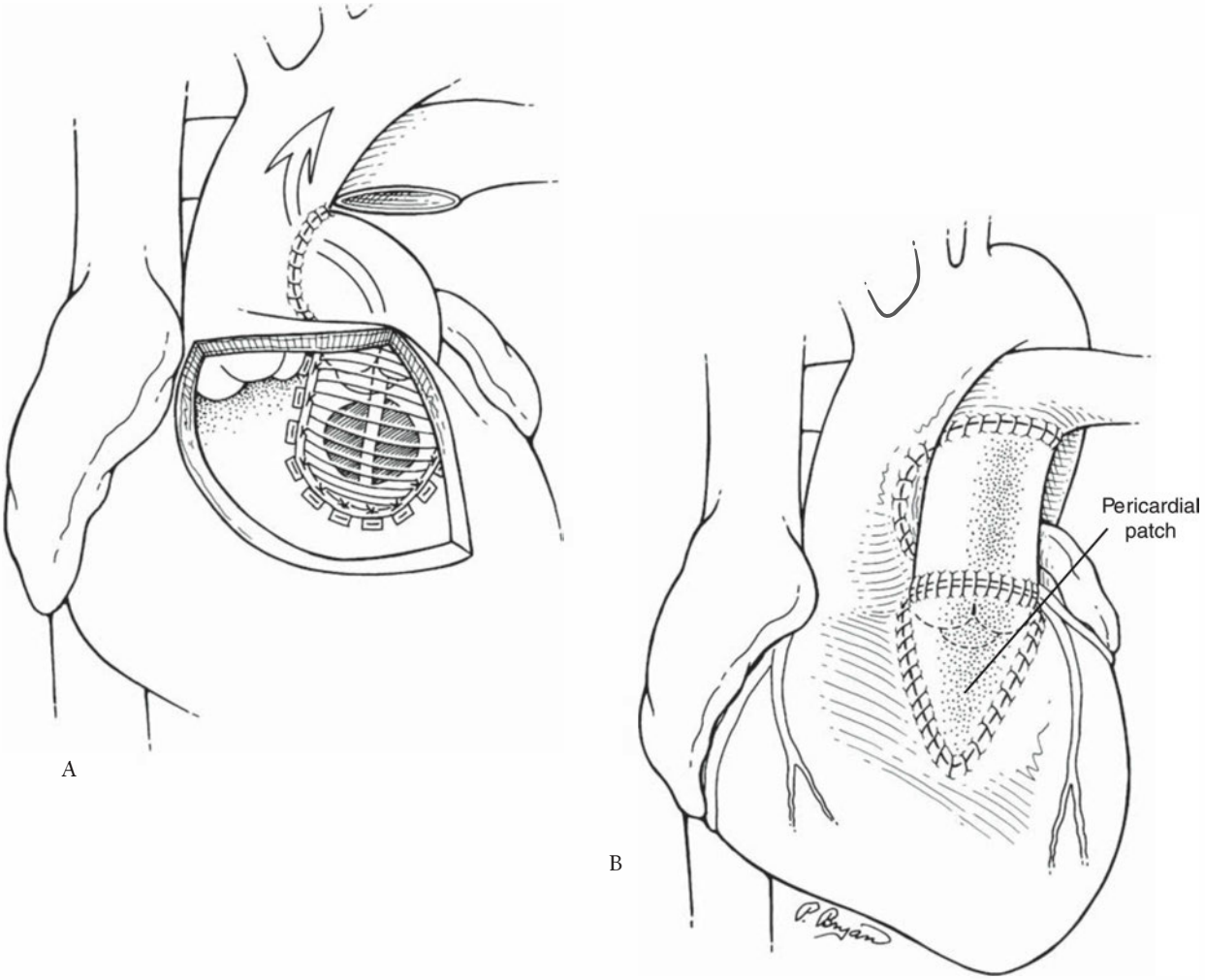


Fig. 88.6. Damus–Kaye–Stansel repair of double-outlet right ventricle with subpulmonary ventricular septal defect (VSD). (**A**) The VSD is closed, channeling left ventricular blood to the pulmonary artery (*arrow*). The main pulmonary artery is transected, and an end-to-side anastomosis is created with the proximal ascending aorta. The aortic valve remains connected to the right ventricle. (**B**) Right ventricle-to-pulmonary artery continuity is re-established with a valved homograft; about one-half the circumference of the proximal homograft is sutured to the superior aspect of the right ventricle. A piece of pericardium or homograft material is used to roof the gap between the right ventricle and the homograft.

DORV with noncommitted perimembranous inlet VSD. Usually, even if there is no preexisting pulmonary stenosis, the large and bulky intraventricular patch creates some obstruction to the pulmonary valve so that a right ventricular outflow tract patch or a right ventricle-to-pulmonary artery valved conduit is necessary.

Sometimes, the noncommitted VSD is a muscular defect in the trabecular septum. If the VSD is small, it is enlarged anteriorly and inferiorly (Fig. 88.8A), since the conduction tissue courses on the superior posterior aspect of the defect (Fig. 88.1D). A suture line with the intraventricular tunnel repair as described above would not only place the conduction tissue at jeopardy as the patch is brought up to the aorta but also risks distortion of the tricuspid valve apparatus. For these reasons, in this situation the Dacron or Hemashield tube graft is left intact, with one end sutured around the VSD and the other end around the aortic valve (Fig. 88.8B). This large intraventricular tube is quite bulky and may obstruct right ventricular outflow, mandating the use of right ventricular outflow tract patch or right ventricle-to-pulmonary artery valved conduit. An obvious disadvantage to this intraventricular tube graft repair is the lack of potential for growth in the channel from the VSD to the aorta. Therefore, the tube graft must be of adult size at the time of the original repair or one must be resigned to reoperation to enlarge the tube as the child outgrows it.

An alternative approach to correction of DORV with noncommitted VSD has been described by Lacour-Gayet and colleagues wherein an intraventricular baffle is constructed to route the left ventricular outflow through the VSD (after surgical enlargement) to the pulmonary artery and then an arterial switch procedure is performed. An even more complex biventricular repair strategy is the use of multiple intraventricular patches to direct left ventricular outflow through the noncommitted VSD to the aorta.

There are situations in which a satisfactory biventricular repair cannot be safely accomplished with DORV and noncommitted VSD. Examples are multiple muscular VSDs, an inability to reliably channel the remote VSD to the aorta, or straddling atrioventricular valve tissue (although Serraf has described the innovative techniques to establish a biventricular repair with straddling atrioventricular valves). In these patients who are unsuitable for a biventricular repair, a modified Fontan procedure can be used effectively. Even though this will result in a physiologic univentricular repair, it is reasonable to enlarge a restrictive VSD

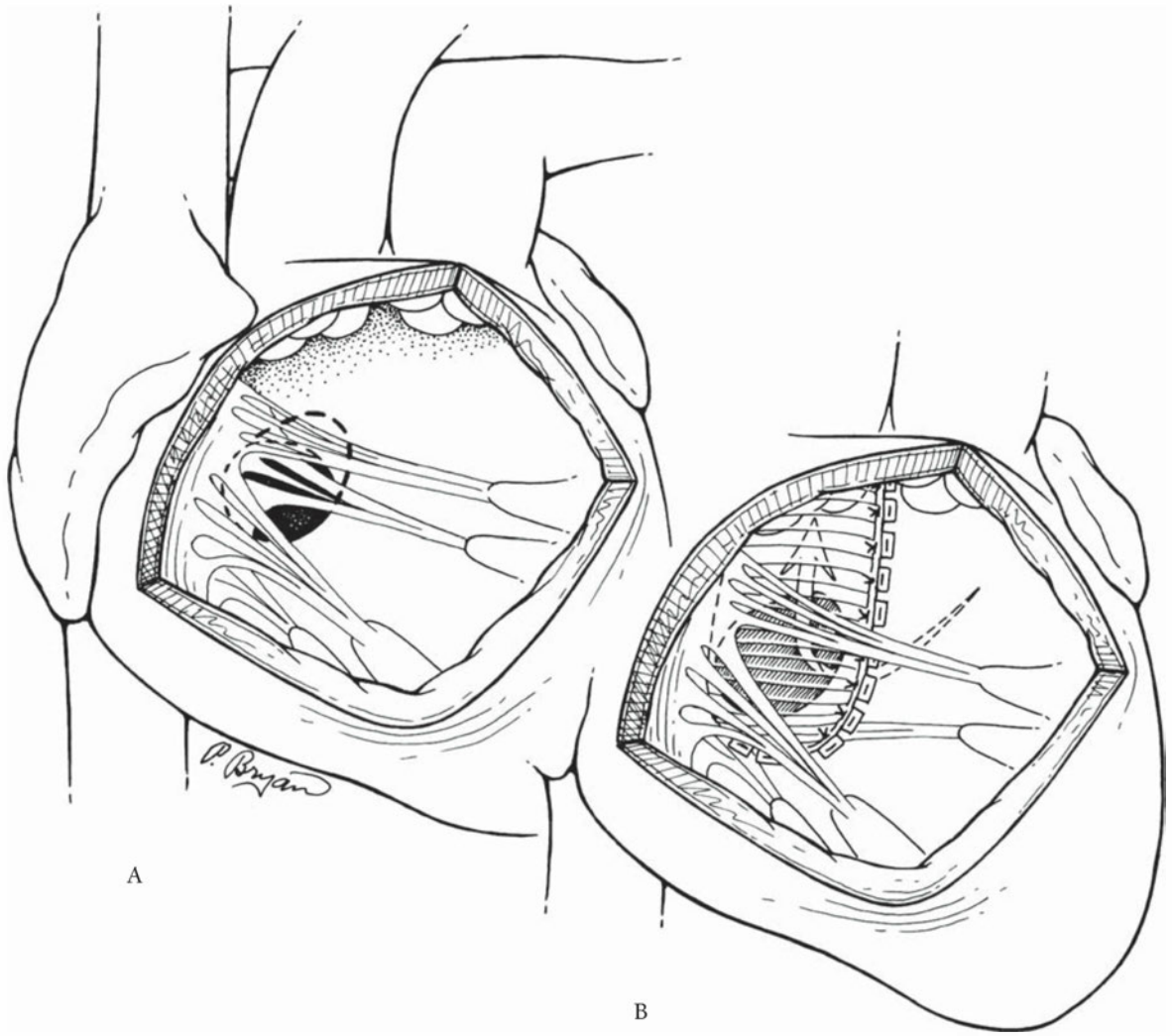


Fig. 88.7. Intraventricular tunnel repair of double-outlet right ventricle with noncommitted perimembranous inlet ventricular septal defect (VSD). (**A**) The VSD is exposed through a right ventriculotomy. The safe area for superior and anterior enlargement of the VSD is marked with a dashed line. (**B**) The intraventricular tunnel is created with an opened polyester (Dacron) tube graft, channeling left ventricular blood through the VSD to the aorta (*arrow*).

when present to allow unobstructed left ventricular contribution to the systemic output. In these children whose eventual repair will be a Fontan procedure, careful consideration must be given to protecting the pulmonary vascular band with a pulmonary artery band early, particularly if there is no associated pulmonary stenosis.

Complications and Postoperative Considerations

Many of the important complications after surgical repair of DORV are mechanical in nature and should be routinely sought out in the operating room after the patient is separated from cardiopulmonary bypass and addressed at that time if possible. With the routine use of intraoperative transesophageal echocardiography, many of these problems are recognized early allowing expeditious correction.

The same possible complications seen with closure of straightforward VSDs can occur during repair of DORV with VSD, particularly if it is necessary to enlarge the VSD during the correction. Surgically induced heart block can occur and underscores the importance of assiduously avoiding the area of the conduction tissue during repair (Fig. 88.1). Residual left-to-right shunting across the VSD patch or intraventricular baffle can severely compromise the patient's hemodynamic status. In addition to intraoperative transesophageal echocardiography which will help precisely locate any residual leak, measurements of oxygen saturations across the right heart will help quantify the magnitude of the shunt. Certainly, any residual calculated Q_p/Q_s ratio of 2:1 should be addressed in the operating room and probably a ratio of $>$1.5:1 should also be corrected.

The use of an intraventricular tunnel in the repair of most forms of DORV raises the possibility of left ventricular outflow tract obstruction, from either inadequate enlargement of a restrictive VSD or poor configuration of the intraventricular tunnel patch with resultant obstruction. Again, intraoperative echocardiography is helpful in identifying and localizing any narrowings. Careful simultaneous measurements in the operating room of left ventricular and aortic pressures will identify any residual left ventricular outflow tract gradient. If there is a significant residual gradient, it is helpful to inspect the left ventricular outflow tract through an aortotomy. Residual obstructing muscle often can be resected through the aortic valve. If the patch is the cause of obstruction, rather than replacing the entire intraventricular tunnel patch, it is possible to visualize the narrowed area through the

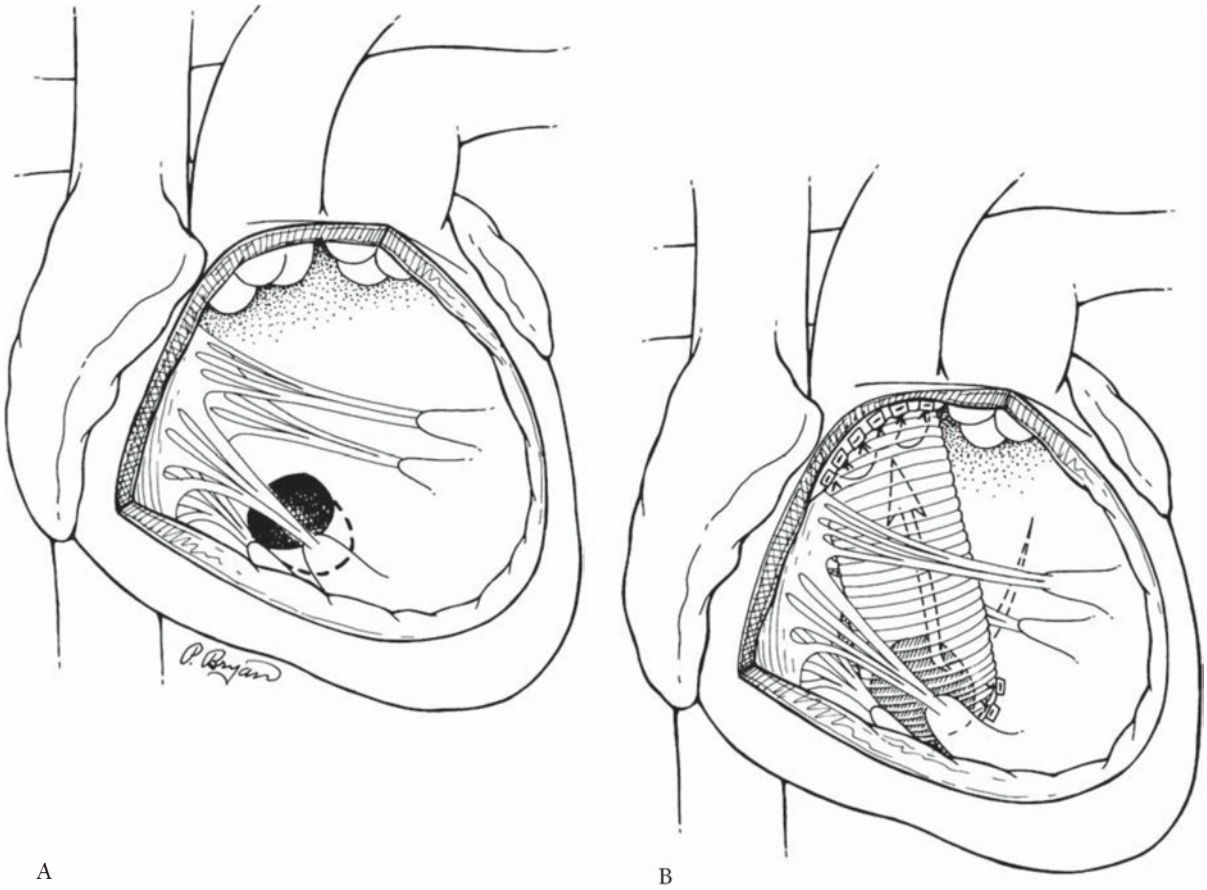


Fig. 88.8. Repair of double-outlet right ventricle with noncommitted muscular trabecular ventricular septal defect (VSD). (**A**) The VSD is exposed through a right ventriculotomy. The safe area for inferior and anterior enlargement of the VSD is marked with a *dashed line*. (**B**) An intact polyester (Dacron) tube graft is used with one end sutured around the VSD and the other end up to the aortic valve, creating an intraventricular conduit for blood from the left ventricle through the VSD to the aorta (*arrow*).

aorta and relieve the obstruction through the right ventricle by incising the intraventricular tunnel patch at the site of narrowing and enlarging it with a separate patch of Dacron or pericardium. Although perhaps not aesthetically pleasing, this technique can be an easy solution to a potentially life-threatening complication.

Similarly, significant right ventricular outflow tract obstruction can occur after repair as a result of inadequate relief of preexisting pulmonary stenosis, obstructing muscle bundles, or obstruction from the intraventricular tunnel patch. Again, intraoperative pressure measurements and echocardiography help identify and locate the site of obstruction, which should be dealt with at the time of recognition if significant.

Myocardial dysfunction can be a significant problem postoperatively because of the complex and sometimes lengthy intraoperative repairs as well as the uniform presence of significant right ventricular hypertrophy and possibly even left ventricular hypertrophy in patients with a restrictive VSD. Naturally, diligent intraoperative myocardial protection is crucial. Postoperatively, in addition to inotropic support, fairly high right-sided filling pressures are often necessary to maintain adequate cardiac output because of the stiff, poorly compliant, hypertrophied right ventricle. In patients in whom it is anticipated that there will be postoperative right ventricular dysfunction due to existing right ventricular hypertrophy, extensive right ventricular incisions, or residual right ventricular outflow tract obstruction, it can be helpful to leave an interatrial communication (typically by not closing the patent foramen ovale) to allow right-to-left shunting. This strategy maintains systemic ventricular output at the expense of usually well-tolerated systemic arterial desaturation.

Particularly in children without pulmonary stenosis, pulmonary hypertension can be problematic in the early postoperative period and should be identified and treated with the usual measures of sedation, paralysis, pulmonary vasodilatation, and hyperventilation. In the past, when complete repair was delayed until later in life, pulmonary vascular obstructive disease was an important cause of postoperative mortality. Occasionally, delaying sternal closure until after the improvement of myocardial dysfunction and reduction of tissue edema can be life saving.

Finally, children undergoing the arterial switch repair of DORV with subpulmonary VSD can develop coronary ischemia even with technically adequate coronary transfer. Avoidance of ventricular distention and systemic hypertension in this situation is key.

DOUBLE-OUTLET LEFT VENTRICLE

Definition

Double-outlet left ventricle (DOLV) is a rare form of congenital heart disease in which >50% of both great arteries arise from the left ventricle. As with DORV, DOLV can be associated with atrioventricular

discordance or with univentricular atrioventricular connections. For the purposes of this chapter, only DOLV with concordant atrioventricular connections and two adequate ventricles is discussed.

Anatomy and Presentation

A VSD is almost always present with DOLV and is usually subaortic in location, although it can be subpulmonic. There are no reports of DOLV with two well-developed ventricles in which the VSD is remote or absent. Pulmonary stenosis is present in most cases of DOLV and can be either valvular or subvalvular.

Children with DOLV with pulmonary stenosis will present with a clinical picture similar to that for tetralogy of Fallot with cyanosis. Palliation with a systemic-to-pulmonary shunt is very satisfactory. The uncommon patient with DOLV without pulmonary stenosis will present with symptoms of excessive pulmonary blood flow with heart failure, much like a patient with an unrestricted VSD. Interestingly, there may be some systemic desaturation due to the mixing of systemic and pulmonary venous return in the left ventricle. These children may be palliated early in life with a pulmonary artery band.

Surgical Technique

Complete surgical repair of DOLV is usually simpler than repair of DORV, since the aorta arises completely from the left ventricle, thus eliminating the need for a complex intraventricular tunnel. Furthermore, the VSD is unrestrictive, so there is no need to enlarge the VSD.

In patients with DOLV with pulmonary stenosis, the VSD is exposed through a right ventriculotomy (Fig. 88.9A). It is closed with a Dacron patch, with standard precautions taken to avoid injury to the conduction tissue, which lies in the usual location on the left ventricular side of the posterior-inferior portion of the defect (Fig. 88.9B). The proximal main pulmonary artery is divided and the cardiac end oversewn (Fig. 88.9A). Right ventricle-to-pulmonary artery continuity is established using a homograft valved conduit (Fig. 88.9C).

There are reports of total intracardiac repairs of DOLV without pulmonary stenosis in which the VSD is patched to direct right ventricular outflow through the VSD into the pulmonary artery. However, except in the most ideal situations, it is still probably easier to repair DOLV without pulmonary stenosis in the same manner as DOLV with pulmonary stenosis (i.e., simple patch closure of the VSD, oversew the proximal pulmonary artery, and establish right ventricle-to-pulmonary artery continuity with a homograft valved conduit; Fig. 88.9). In many cases with subaortic VSD and pulmonary outflow tract obstruction, the anatomic substrate for the pulmonary obstruction is typically subvalvular with only mild or no pulmonary valve stenosis. In these cases, after closing the subaortic VSD channeling left ventricular outflow to the aorta, the pulmonary root can be harvested with the valve intact (similar to the technique for a Ross procedure) and then translocated to the right ventricle. This obviates the need for a homograft valved conduit with its attendant requirement for eventual replacement.

Complications and Postoperative Considerations

As with surgical closure of a routine VSD, in repair of DOLV the risks of surgically induced heart block or residual left-to-right shunts exist, but there is nothing about DOLV that makes these complications more or less likely when the usual precautions and careful surgical techniques are used. Since all patients with DOLV have hypertrophied right ventricles, postoperative right ventricular dysfunction with a stiff, poorly compliant right ventricle

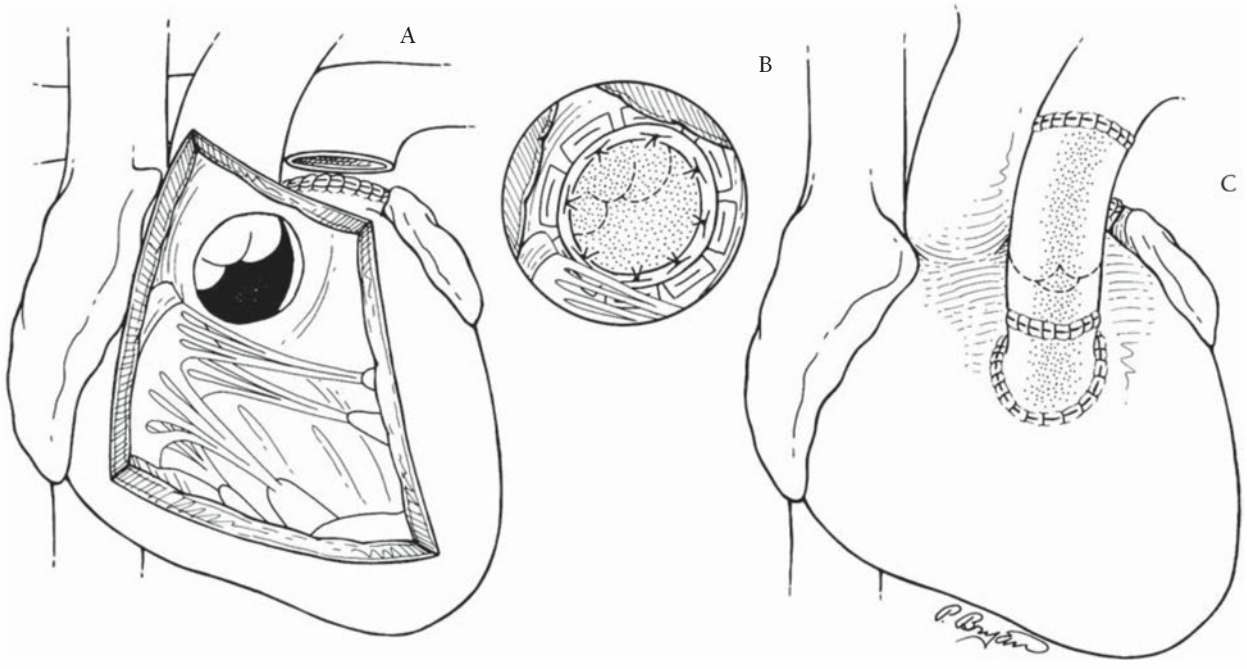


Fig. 88.9. Repair of double-outlet left ventricle. (**A**) The ventricular septal defect (VSD) is exposed through a right ventriculotomy. The aortic valve can be visualized through the subaortic VSD. The main pulmonary artery is divided and the cardiac end oversewn. (**B**) The VSD is closed with a polyester (**Dacron**) patch using interrupted pledgetted sutures or a continuous suture technique, with care taken to avoid the conduction tissue. (**C**) Right ventricle-to-pulmonary artery continuity is established with a valved homograft; about one-half of the circumference of the proximal homograft is sutured to the superior aspect of the right ventricle. A piece of pericardium or homograft material is used to roof the gap between the right ventricle and the homograft.

can occur but is usually not problematic. Finally, as with any extracardiac valved conduit, improper technique can result in right ventricular outflow tract obstruction, which should be identified and corrected in the operating room.

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EDITOR'S COMMENTS

The anatomic spectrum of DORV continues to require multiple operative approaches. In complex forms of these defects, surgical decision making can be difficult. The use of intraventricular tunnel repairs, often with the need for right ventricular outflow tract reconstruction with valved conduits, has led to a significant incidence of reoperation in these patients for conduit exchange. In addition, inadequate resection of septal muscle and enlargement of the VSD can result in progressive obstruction of the intraventricular tunnel and subaortic stenosis. The problems with these late defects have led some authors to suggest that in complex forms of DORV, conversion to a single-ventricle type of repair is preferable and may actually decrease the risk of late reoperation and complications.

A patient with subpulmonary VSD and DORV (the Taussig–Bing heart) commonly has associated coarctation of the aorta. Although repair of the coarctation and pulmonary artery banding can be performed for this condition, we perform primary complete repair in these infants. The division of the aorta at the time of the arterial switch procedure allows reconstruction of the arch by patch enlargement without difficulty and a low risk of recurrence of stenosis.

Even when complete correction requires the use of an extracardiac conduit, we primarily repair most patients with nonrestrictive pulmonary outflow tracts to prevent the complications of pulmonary artery banding, which may exacerbate myocardial hypertrophy and, in some cases with relatively restrictive VSDs, cause progressive subaortic obstruction. The progression of subaortic obstruction then drives more blood through the pulmonary artery band. These patients represent a very difficult subgroup at second-stage operation. Often they have been followed with a pulmonary artery band with good oxygen saturations and yet have had progressive subaortic obstruction. Thus, the actual Q_p/Q_s ratio may not be restricted, and a significant left-to-right shunt may predispose the patient to develop pulmonary vascular disease. This is a particularly difficult situation in those patients who are being staged to single-ventricle correction. Bidirectional Glenn shunting or hemi-Fontan procedures should be performed early even if pulmonary artery banding is undertaken in infancy. Subjecting these patients to significant left-to-right shunts as subaortic stenosis develops in the face of a pulmonary band results in a very high risk of inadequate pulmonary circulation at the time of reoperation for Damus–Kaye–Stansel connection and bidirectional Glenn shunting. Therefore, a close follow-up is necessary after pulmonary artery banding to avoid this complication.

When a Damus–Kaye–Stansel operation is necessary because of subaortic obstruction, we augment the arch of the aorta as in the Norwood operation in most cases. Patients with significant subaortic

(*continued*)

EDITOR'S COMMENTS (*continued*)

obstruction in infancy often have hypoplasia of the ascending aorta, and there is a significant size discrepancy between the ascending aorta and the proximal pulmonary artery. Therefore, direct connection of the pulmonary artery to the aorta, either primarily or with an augmentation hood, can result in kinking of the distalmost portion of the superior anastomosis with obstruction of the aorta, requiring additional augmentation or stenting. Creation of a long patch underneath the arch of the aorta prevents this kinking and augments the entire ascending aorta. A brief period of circulatory arrest is used for this portion of the operation.

Late restriction of the VSD when an intracardiac conduit is used is common enough that one should consider enlargement of virtually all VSDs in DORV at the time of complete repair. The exception may be isolated subaortic defects where there is a 50% override of the aorta and where the VSD is clearly as large as or larger than the aortic annulus.

Although intracardiac conduits can obstruct right ventricular outflow, it is also possible that the conduit attachments to the septal tricuspid leaflet can limit leaflet mobility and cause functional tricuspid stenosis.

Patients with DORV and uncommitted VSDs remain a difficult surgical challenge. Although resection of the anterior septum with inlet defects can occasionally permit an adequate left ventricular outflow without interfering with tricuspid valve function, in many cases complex intraventricular tunnels may be necessary, creating a virtual right-angled tunnel along the ventricular septum to the aorta.

In some cases with noncommitted VSD, the VSD is more directly related to the pulmonary artery than to the aorta. In these cases, in the absence of significant pulmonary stenosis, closure of the VSD to the pulmonary artery and arterial switch procedure is the best approach as is done in Taussig–Bing anomaly. In each case with noncommitted VSD, it is imperative to identify the relationship of the VSD to the great arteries and to conceptually craft a pathway from the VSD to the great vessels to decide when a two-ventricle repair can be accomplished. Very complex baffles with significant angulation are most likely to result in late stenosis; in such cases, the potential for obstruction is significant and, therefore in many cases, a single-ventricle-type repair might be the best approach.

There has been increasing interest in the use of three-dimensional (3-D) modeling systems using MRI data to help aid surgical planning in patients with DORV and noncommitted VSD. Although these modeling techniques do not account for the issue of straddling valves, which are best determined by echocardiography, creating 3-D models including creation of the 3-D printed casts of the cardiac anatomy can aid in designing the patches to prevent a pathway obstruction when an intraventricular tunnel is created.

The REV procedure described by LeCompte is a good alternative in patients who have significant amounts of infundibular muscle, which can interfere with the pathway from the VSD to aorta. Extensive resection of this muscle can create a more direct pathway from the left ventricle to aorta and prevent recurrent LV outflow tract obstruction following repair. DaSilva in Brazil has also described the technique of pulmonary artery translocation which can be used in some complex forms of DORV, moving the pulmonary root anterior to the aorta to the right ventricular outflow tract. Removing the pulmonary valve and root resects the subaortic and subpulmonary conus, which makes the VSD pathway to the aorta less obstructive.

DOLV remains an unusual condition. Although patch closure of the VSD is simple, oversewing of the pulmonary outflow and creation of a right ventricle-to-pulmonary artery conduit exposes the child to recurrent operation for conduit changes. As noted by Dr. Kanter, in some cases with unrestricted right ventricular outflow, the entire pulmonary valve and annulus can be excised from the ventricle and relocated onto the right ventricle with patch closure of the defect in the left ventricle. In this manner, a native valve can be reconstructed to the right ventricular outflow tract and potentially decrease the risk of late reoperation. Patients with significant pulmonary stenosis, however, are best treated with outflow reconstruction, either with a valved conduit or by direct reconstruction of the pulmonary confluence onto the right ventricle with the LeCompte maneuver and an anterior patch of pericardium or homograft material to create a nonvalved reconstruction. Coronary location often precludes primary outflow tract reconstruction with a transannular patch.

TLS

89 Transposition of the Great Arteries

Aaron W. Eckhauser and Thomas L. Spray

Complete transposition of the great arteries (TGA) is a congenital cardiac defect in which there is anatomic reversal of the relationship of the great arteries. The aorta arises entirely or largely from the right ventricle, and the pulmonary artery arises entirely or largely from the left ventricle (ventriculoarterial discordant connection). The lesion is incompatible with life without surgical intervention because in the resulting physiologic abnormality the pulmonary and systemic circulations exist in parallel instead of in series. Survival, therefore, depends on mixing between the pulmonary and systemic circulations. In spite of the severity of this cardiac defect, surgical therapy has become standardized such that anatomic and physiologic repair can be accomplished in the first few days of life in the majority of affected children.

TGA may coexist with other cardiac lesions, including coarctation of the aorta and patent ductus arteriosus. Transposition is associated with intact ventricular septum in approximately 50% of patients, ventricular septal defect (VSD) in 25%, and VSD with functional or anatomic left ventricular outflow tract obstruction (pulmonary stenosis) in 25%. Transposition with interruption of the aortic arch is a rare association.

ANATOMY

Complete TGA is characterized by atrioventricular concordance and ventriculoarterial discordance ({S,D,D} according to the Van Praagh classification system). There have been many interpretations and applications of the term transposition, but in this chapter, *complete TGA* is defined as atrial situs solitus, atrioventricular concordance, and ventriculoarterial discordance {S,D,D}.

The morphogenesis of the abnormal relationship between the great arteries and the ventricles in TGA is controversial. In normal cardiac development, the subaortic conus is static, and dominant growth of the pulmonary conus forces the pulmonary valve anterior, superior, and to the left. Van Praagh suggested that in TGA the subaortic conus persists during normal looping of the ventricles, whereas the subpulmonary conus undergoes absorption. Differential growth of the subaortic conus in transposition, therefore, pushes the aorta anteriorly and disrupts the aortic-mitral valve continuity. If the subpulmonary conus fails to develop, the pulmonary artery will maintain a posterior location and pulmonary-mitral valve continuity will occur. Consequently, the aortic valve becomes anterior to the pulmonary valve, permitting both semilunar valves to connect with the distal great vessels without the rotation that is hypothesized to occur in normal cardiac development. Because conal development determines the rotation of the truncus arteriosus, the great arteries are similar in relationship at the semilunar valves as they are at the arch. Anatomic variations are often encountered, although the heart is left-sided with atrial situs solitus in 95% of patients. Left-to-right juxtaposition of the atrial appendages is a sign of other intracardiac anomalies. A true ostium secundum atrial septal defect is present in 10% to 20% of cases, but the majority of atrial communications are via a patent foramen ovale. Right aortic arch is present in 4% of patients with intact ventricular septum and up to 16% of those with VSD. Up to 50% of patients with TGA will have an associated VSD, many of which will spontaneously close. The VSDs are commonly perimembranous (conoventricular) in location, although they may be found anywhere in the ventricular septum. Pulmonary stenosis or atresia, overriding or straddling atrioventricular valves, coarctation of the aorta, and interruption of the aortic arch have all been noted in association with transposition and VSD.

The spatial relationship of the great vessels is quite variable, but the aorta is most frequently to the right and anterior to the pulmonary artery {S,D,D}. Less commonly, the aorta can be anterior and to the left with mirror-image branching {I,L,L} or anterior and to the left with normal arch branching {S,D,L}. The sinuses of Valsalva and coronary artery ostia typically face the corresponding pulmonary arterial sinuses of Valsalva, permitting transfer of the coronary arteries in the arterial switch operation. Only a small number of patients who have a coronary artery that originates from a nonfacing coronary sinus pose a problem for arterial switch reconstruction.

The coronary anatomy can be described using a modified Leiden convention. In this description, the surgeon places themselves in the nonadjacent aortic sinus looking toward the pulmonary valve. The sinus to the surgeon's right hand is labeled "1" and the sinus to the surgeon's left hand is labeled "2" (Fig. 89.1). Coronary branching patterns can be described as normal, looping, or intramural. In the "normal" pattern, which accounts for 60% to 70% of cases, sinus 1 gives rise to the anterior descending and circumflex arteries. Sinus 2 gives rise to the right coronary artery. In this pattern, none of the major arterial branches crosses in front of or behind either of the major arterial trunks. The "looping" pattern, found in one-third of cases, results when one or more of the three major coronary arteries run in front of or behind the major arterial trunks. Posterior looping results when a coronary artery runs posterior to the pulmonary artery, most commonly the circumflex arising from sinus 2. Anterior looping results when one of the branches runs anterior to the aorta. This can occur when the left anterior descending artery arises from the right coronary artery in sinus 2 or a single coronary artery arises from either sinus 1 (right coronary artery crosses) or 2 (left main equivalent crosses). Branching patterns giving rise to double loops are not uncommon and account for 15% to 20% of cases. Finally, the "intramural" course occurs when a coronary artery appears to arise from one sinus, crosses through the media of the aortic wall behind the valve commissure, and

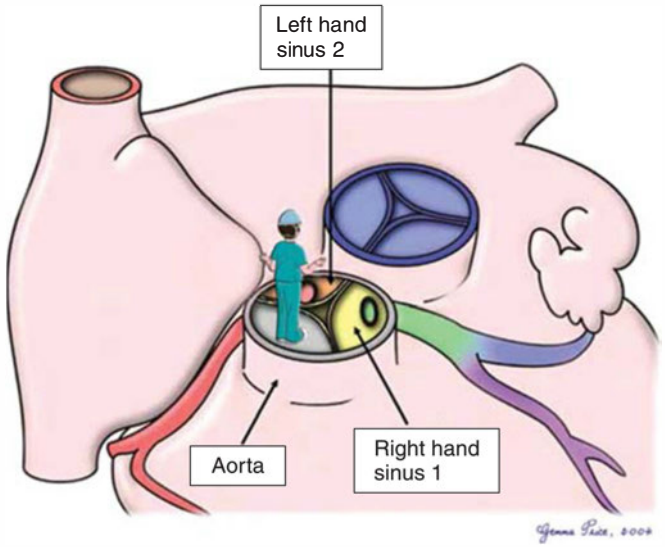


Fig. 89.1. Leiden convention for coronary sinus description in transposition. The surgeon is standing in the nonadjacent aortic sinus facing the pulmonary artery. The surgeon's right hand represents sinus 1 and the left hand represents sinus 2.

truly arises from the other adjacent aortic sinus. Occasionally, there is no true circumflex coronary artery, but separate branches arise from the left coronary to supply the corresponding portion of the left ventricle. Abnormal coronary branching patterns are more commonly seen in transposition with VSD than when an intact ventricular septum is present.

Important left ventricular outflow tract obstruction is unusual in association with TGA but has significant implications for management strategies. The most common type of left ventricular outflow tract obstruction is dynamic, resulting from leftward displacement of the hypertrophied muscular ventricular septum secondary to the development of higher right systemic ventricular pressure. The septum may then narrow the outflow tract, resulting in abnormal systolic anterior mitral leaflet motion and a situation similar to that noted in hypertrophic obstructive cardiomyopathy. Posterior malalignment of the ventricular septum may create a tunnel-like obstruction, and fibrous tags arising from the mitral apparatus or membranous septum can result in significant subvalvar obstruction, more commonly when a VSD is present. True valvar stenosis is uncommon, but a nonobstructive bicuspid pulmonary valve is not infrequently present. In rare cases, aortic arch obstruction with coarctation or true interruption of the aortic arch has been observed in patients with TGA, left ventricular outflow tract obstruction, and VSD.

PATHOPHYSIOLOGY

TGA {S,D,D} is a relatively common form of congenital heart disease, accounting for 9.9% of infants with congenital heart disease in a New England study and representing a frequency of 0.206 per 1000 live births. A distinct male predominance is noted, with a male-to-female ratio of 2:1, which increases to 3.3:1 when the ventricular septum is intact. In complex forms of transposition, a sexual predominance has not been noted. Untreated, 90% of children with D-TGA and an intact ventricular septum will die by 1 year of age.

The parallel relationship of the pulmonary and systemic circulations in TGA results in nonoxygenated venous blood passing through the right ventricle to the aorta, whereas the oxygenated pulmonary venous blood passes through the left ventricle back to the pulmonary artery. Mixing between the pulmonary and systemic circulations at the atrial, ventricular, or great vessel level through a patent foramen ovale or atrial septal defect, a VSD, or a patent ductus arteriosus is mandatory for survival. Patients with TGA and an intact ventricular septum survive initially because of aortopulmonary flow through a patent ductus arteriosus. After birth, both ventricles are relatively noncompliant, and infants with transposition often have increased pulmonary blood flow. This results in left atrial enlargement and functional incompetence of the foramen ovale with atrial level mixing of oxygenated and nonoxygenated blood. Inadequacy of mixing, however, will result in marginal tissue oxygenation that does not improve with oxygen administration. Atrial balloon septostomy results in improved admixture and improved tissue oxygen delivery.

Patients with TGA and significant VSD often have higher oxygen saturations by virtue of greater pulmonary blood flow and greater mixing at both the atrial and ventricular levels. In children with high pulmonary blood flow, pulmonary resistance may progressively increase during infancy. The early development of severe pulmonary vascular disease in children with TGA is exacerbated in patients with associated VSD. The rapid development of pulmonary obstructive disease may be related to hypoxemia, increased sympathetic activity, and excessive pulmonary blood flow.

Although neonatal pulmonary vascular resistance is elevated in infants with TGA, the resistance falls progressively during the neonatal period, coinciding with changes in the pulmonary and systemic ventricular compliance. In a normal infant, there is an increase in the left ventricular volume and pressure load and a decrease in the right ventricular volume and pressure load shortly after birth, resulting in a rapid increase in the left ventricular myocardial mass. Normal development of the left ventricle is lost in infants with D-TGA because the left ventricle ejects to the low-resistance pulmonary vascular bed. Thus, the left ventricle does not increase muscle mass relative to the right ventricle and within a few weeks loses the ability to maintain adequate cardiac output against significant afterload. This change occurs despite the fact that the left ventricle maintains a volume load in patients with transposition and intact ventricular septum. However, when a VSD or large patent ductus arteriosus is present, both volume and pressure overload of the left ventricle are maintained. In D-TGA with left ventricular outflow tract obstruction without a VSD, a ventricular pressure load is imposed without a significant volume load. These physiologic changes in the neonatal heart are important for the consideration of surgical approaches because after a few weeks of postuterine life, the left ventricle in D-TGA with intact ventricular septum takes on the characteristics and wall thickness of a pulmonary ventricle and may not be adequate to support the systemic circulation.

CLINICAL FEATURES

The most common clinical finding in an infant with TGA is cyanosis (arterial partial pressure of oxygen 25 to 40 mmHg), which varies in degree depending on associated anomalies. Typically, the cyanosis is more pronounced when the ventricular septum is intact and is often present at birth. The development of cyanosis later in infancy is usually associated with the presence of a significant VSD or left ventricular outflow tract obstruction. Congestive heart failure (CHF) may be the predominant clinical finding in patients with a large VSD or patent ductus arteriosus. Symptoms of cardiac failure, however, are rarely present in the first week of life but commonly appear by 1 month of age as pulmonary vascular resistance decreases and pulmonary blood flow becomes excessive, even in a patient with an intact ventricular septum.

MANAGEMENT

The widespread use of fetal ultrasound techniques has resulted in the common antenatal diagnosis of TGA. In fetuses with TGA and intact ventricular septum, the four-chamber view is normal. The parallel course of the aorta and pulmonary artery differentiates the fetus with TGA from those in which the arterial trunks are normally crossing. Color and pulsed wave Doppler showing inflow and outflow patterns of the left ventricular outflow tract coupled with a typical ejection pattern from the pulmonary artery further confirm the diagnosis of transposition. Postnatally, echocardiography has now generally supplanted cardiac catheterization in the majority of patients with simple complete transposition. Cardiac catheterization is now indicated only in infants in whom inadequate shunting is noted or if associated intracardiac or extracardiac abnormalities require clarification. Echocardiographic views confirming a posterior great vessel that divides into right and left pulmonary arteries and arises from the left ventricle in association with an anterior aorta arising from a right ventricle confirm the diagnosis of TGA. The intracavitary shunts can be determined by Doppler echocardiography, and several echo views can determine the size and location of VSDs with reference to the infundibular septum, the nature and size of atrial communications, the anatomy of the atrioventricular valves, and the presence and location of significant degrees of subpulmonary stenosis. In the majority of cases, the origins and anatomic distributions of the coronary arteries can be adequately visualized by echocardiography without the need for catheterization. Since virtually all coronary patterns can be dealt with surgically, preoperative identification of coronary anatomy is not necessary.

Cardiac catheterization is reserved for patients with significant clinical instability in order to improve the degree of intracavitary shunting by enlarging the interatrial septal communication. At catheterization, the left atrial pressure is usually greater than the right atrial pressure, and the pressure in the pulmonary (left) ventricle depends on the presence or absence of a VSD, the valvar or subvalvar stenosis, the age of the patient, and the magnitude of elevation of pulmonary vascular resistance.

The most important aspect of preoperative management of TGA is that oxygenated left atrial blood gets to the head. A PDA only helps to fill the left atrium, so atrial or ventricular level mixing is critical. If inadequate interatrial shunting results in clinical instability (acidemia, severe hypoxemia), the mainstay of management has been a Rashkind balloon atrial septostomy. A balloon-tipped catheter is passed through the systemic veins, through the right atrium and foramen ovale, and into the left atrium. The balloon is inflated and pulled vigorously across the atrial septum to tear the foramen ovale, improving admixture of pulmonary and systemic venous blood. This procedure may be performed in the intensive care unit with echocardiographic guidance in children who are clinically unstable. In patients with TGA and intact ventricular septum, atrial level mixing results in moderate hypoxemia but rarely CHF. Since an atrial septostomy decompresses the left ventricle, delayed repair may result in poor left ventricular performance. Prostaglandin E_1 is often administered to maintain ductal patency and increase pulmonary blood flow helping to stabilize patients before early operative repair. In patients with atrial and ductal level shunting, mixing is improved with only mild hypoxemia but can be complicated by CHF and low diastolic pressures. Patients with PDA and restrictive atrial level shunting often develop pulmonary edema and metabolic acidosis. In patients with TGA and VSD, atrial, ventricular, and ductal shunting results in minimal hypoxemia but severe CHF once the pulmonary resistance falls. Therefore, patients who are good anatomic candidates for an arterial switch procedure who have acceptable arterial oxygen saturations are generally referred for early arterial switch without intervening atrial septostomy.

HISTORY OF SURGICAL REPAIR

Initial surgical therapy for TGA involved creation of an atrial septal defect using a closed technique to increase the mixing between the systemic venous and pulmonary venous circulations. This was first performed by Blalock and Hanlon in 1950. Although early mortality was high with this operative approach, successful creation of an atrial septal defect resulted in significant palliation in many of these children. Initial attempts to reverse the transposed vessels by both Mustard and Bailey were frustrated by an inability to maintain coronary perfusion and poor function of the anatomic left ventricle. Thus, initial surgical therapy was directed toward atrial transposition of the pulmonary and systemic venous returns. In 1952, Lillehei and Varco transferred the right-sided pulmonary veins to the right atrium and connected the inferior vena cava to the left atrium. A successful modification of this technique using an allograft to connect the inferior vena cava to the left atrium was described by Baffes in 1956.

Multiple attempts were made at both atrial and arterial repairs of TGA during the 1950s, and in 1954 Albert suggested the concept of switching the atrial septum so that caval return was directed to the left ventricle and the pulmonary venous return to the right. The atrial switch concept was first successfully accomplished by Senning in 1959 using an ingenious technique for relocating the walls of the right atrium and the atrial septum. Many of the early attempts at atrial repair were frustrated by the fact that the operation was performed on patients between 1 and 2 years of age and significant pulmonary vascular obstructive disease had already developed in many of these children. In 1964, Mustard described an alternate procedure for intra-atrial repair: excising the atrial septum and creating a large interatrial baffle of pericardium to redirect pulmonary and systemic venous blood resulting in a larger atrium than the Senning operation. Early results with the Mustard operation were markedly improved over the previously reported Senning repairs and reflected the significant population of patients in Toronto who had undergone successful Blalock–Hanlon atrial septectomies early in life.

A major development in the surgical treatment of TGA occurred in 1966 when Rashkind and Miller reported the use of a

balloon catheter technique to enlarge the atrial septal defect in patients with TGA, resulting in improved early physiologic stability and decreasing the need for operative atrial septectomy. During the 1960s, the Mustard operation became the most commonly performed procedure for transposition. As surgical techniques improved, it became clear that repair in the first few months of life could be accomplished with a low operative mortality and improved results compared with repair at a later age. In 1970, the Senning procedure reemerged as persistent problems with baffle obstruction and arrhythmias after the Mustard operation became well defined and the ability to use autologous tissue for the atrial reconstruction became a preferred approach.

The success with the atrial switch operations for TGA with intact septum did not translate to repair of transposition with a large VSD. Disappointing results with VSD closure and atrial repair in this group of patients continued to be a stimulus for the development of an arterial switch procedure, which was first successfully performed by Jatene and colleagues in 1975. Yacoub shortly thereafter reported additional successful cases. The success of the arterial switch procedure with reimplantation of the coronary arteries in some patients with transposition and VSD led to reintroduction of this technique for patients with intact ventricular septum. Yacoub's initial attempts in patients with TGA and an intact ventricular septum were unsuccessful in 1972. However, additional reports by 1976 suggested that such repair was possible in infancy. Early mortality with the arterial switch in infancy was related to the fact that the pulmonary left ventricle was not prepared to sustain systemic pressure. Therefore, initial approaches included pulmonary arterial banding (with or without a systemic-to-pulmonary shunt) as a first stage followed by a later arterial switch. However, Yacoub, Quaegebeur, Brawn, and Castaneda subsequently demonstrated the ability to repair simple transposition in the first few days of life by an arterial switch operation while the pulmonary ventricle has relatively high pressure. The rapidly decreasing mortality for arterial switch operations has resulted in this surgical approach becoming the standard corrective surgical procedure, which results in both an anatomic and physiologic reconstruction of this cardiac defect.

Operative approaches to TGA, VSD, and significant left ventricular outflow tract obstruction were less successful in the early period of cardiac surgery. In 1969, Rastelli suggested the combination of an intraventricular tunnel repair: closing the VSD and baffling the pulmonary venous blood from the left ventricle to the aorta, closing the pulmonary artery exit from the left ventricle, and creating an extracardiac conduit from the anatomic right ventricle to the pulmonary bifurcation, resulting in an anatomic repair for TGA/VSD and left ventricular outflow tract obstruction. Obstruction of the reconstructed left ventricular outflow tract remains a common complication of the Rastelli-type repair and has led to recommendation of a variation of intraventricular repair by Lecompte (the "réparation à l'étage ventriculaire" [REV] procedure) and the aortic root translocation (Nikaidoh procedure).

OPERATIVE INTERVENTION: PALLIATIVE OPERATIONS

The advent of the balloon atrial septostomy by Rashkind and Miller has essentially eliminated the need for the Blalock–Hanlon atrial septectomy. Infants with associated cardiac abnormalities and a thick atrial septum who are considered for later atrial baffle repair may benefit from the Blalock–Hanlon technique, although the safety of cardiopulmonary bypass has resulted in the common use of open atrial septectomy in such patients. Pulmonary arterial banding has been used for palliation with TGA and VSD in young infants who have intractable CHF until operative repair at 3 to 6 months of age. As the results with the arterial switch procedure and VSD closure in infancy have improved, banding in most instances is unnecessary because complete repair can be performed safely. Therefore, banding of the pulmonary artery has now been limited to very small neonates who might benefit from a delay in corrective surgery and those patients with transposition and an intact ventricular septum who present late for arterial switch repair and require "training" of the left ventricle to work at a higher pressure in order to become the systemic ventricle. For similar reasons, pulmonary arterial banding is used in patients with TGA who develop right ventricular dysfunction and failure after atrial baffle operations as a component of staged conversion to an arterial switch repair. Pulmonary arterial banding in TGA is a delicate procedure because limitation of pulmonary blood flow results in significant hypoxia and metabolic acidosis, and loose banding results in inadequate protection of the pulmonary vascular bed and poor development of the left (pulmonary) ventricle. Thus, in most situations in which preparation of the left ventricle is undertaken for conversion to an arterial switch procedure, banding must be associated with creation of an aortopulmonary shunt to maintain adequate pulmonary blood flow to prevent hypoxemia and ventricular dysfunction.

SURGICAL CORRECTION

Successful correction of TGA results in the rerouting of systemic venous blood to the pulmonary circulation and the pulmonary venous blood into the systemic arterial circulation. This may be accomplished at the atrial, ventricular, or great arterial level. The earliest repairs of TGA involve rerouting of the systemic and pulmonary venous returns at the atrial level, resulting in an adequate physiologic repair but not an anatomic repair because the morphologic right ventricle continues to be the systemic ventricle. Ventricular (Rastelli) and great arterial (arterial switch) repairs are more anatomic corrections resulting in the morphologic left ventricle as the systemic ventricle.

Atrial Repair

Despite the excellent results with atrial switch operations for TGA, anatomic correction has resulted in the arterial switch procedure becoming the standard surgical repair for TGA. Therefore, the Senning and Mustard operations will not be described in detail in this chapter.

Arterial Repair

The technique of the arterial switch operation involves transection of the great arteries, transfer of the coronary arteries, and repositioning of the great vessels. The procedure is performed through a median sternotomy with the use of cardiopulmonary bypass with mild or no hypothermia. In rare cases, the entire operation can be performed with a single period of circulatory arrest, or circulatory arrest can be used for a brief period for atrial septal defect exposure and coronary transfer. After sternotomy, a portion of the anterior pericardium can be excised for use as an autologous patch for reconstruction of the anterior great vessel. The patch can be used either fresh or fixed in a glutaraldehyde solution to make for easier handling. Currently, we use pulmonary homograft patch material for pulmonary artery reconstruction. The ligamentum arteriosum or patent ductus arteriosus is dissected out, and the branches of the right and left pulmonary arteries are mobilized well out into the hilum of the lungs bilaterally. It is important to freely mobilize the

pulmonary arterial branches out beyond the bifurcation to the lobar branches to permit safe anterior relocation of the pulmonary artery during the Lecompte maneuver without causing compression of the pulmonary arteries by the aorta. As noted in Figure 89.2, the aorta is cannulated as distally as possible to allow room for manipulation of the proximal aorta during the reconstruction. A cardioplegia needle is inserted into the aortic root, and bicaval cannulation is performed in most cases. In D-TGA, the aorta is anterior and to the right of the pulmonary artery, and the pulmonary artery is typically larger than the ascending aorta. After bypass is established, the ductus arteriosus is ligated but not divided. The aorta is clamped as close to the aortic cannula as possible, and cardioplegic solution is administered into the aortic root.

If a VSD is present, it can be approached either through the right atrium across the tricuspid valve or occasionally through the anterior great vessel or pulmonary artery (anterior muscular or conoseptal hypoplasia defects). In the majority of cases, however, we approach the VSD across the tricuspid valve. We prefer to address the VSD and atrial septal defect after the initial cross-clamping of the aorta so that additional doses of cardioplegic solution can be administered before aortic reconstruction. The VSD is closed in the usual manner across the tricuspid valve as described in Chapter 76. If an atrial septal defect is present, it is closed partially to allow for right-to-left shunting if necessary in the early postoperative period; if a patent foramen ovale is present, it is left unclosed. After the VSD is closed, if present, additional cardioplegic solution is injected into the aortic root, and then the aorta is transected above the level of the commissural attachments of the aortic valve and the pulmonary artery transected at the level of the bifurcation as noted in Figure 89.3. The ductus arteriosus is divided on the pulmonary arterial end, and the pulmonary arteries are freely mobilized from adjacent tissue. At this point, we perform the LeCompte maneuver, relocating the pulmonary bifurcation anterior to the aorta and then recross-clamp the aorta, keeping the pulmonary bifurcation cephalad and out of the operative field while coronary reconstruction is performed. The coronary ostia are examined carefully and excised from the anterior great vessel with a button of aortic wall extending down to the base of the sinuses of Valsalva, as noted in Figure 89.4 (inset A). The epicardial courses of the coronary arteries are then mobilized adequately to permit translocation of the coronary ostia to the posterior great vessel without kinking of the vessels, as noted in Figure 89-4. On occasion, small conal branches of the coronary arteries may need to be sacrificed to permit adequate mobilization. At a suitable site on the posterior great artery, a vertical incision is made with a medially based flap to allow takeoff of the coronary ostia without tension or kinking. Whereas generally the

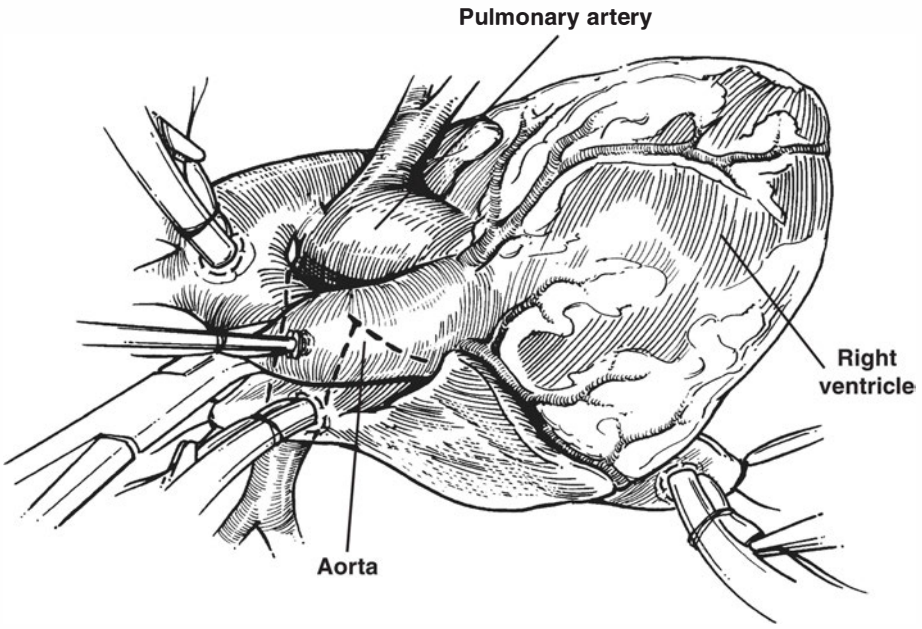


Fig. 89.2. The anatomy in transposition of the great arteries with the aorta anterior and to the right of the pulmonary artery. The aorta is cannulated distally, and bicaval cannulation with snares is placed.

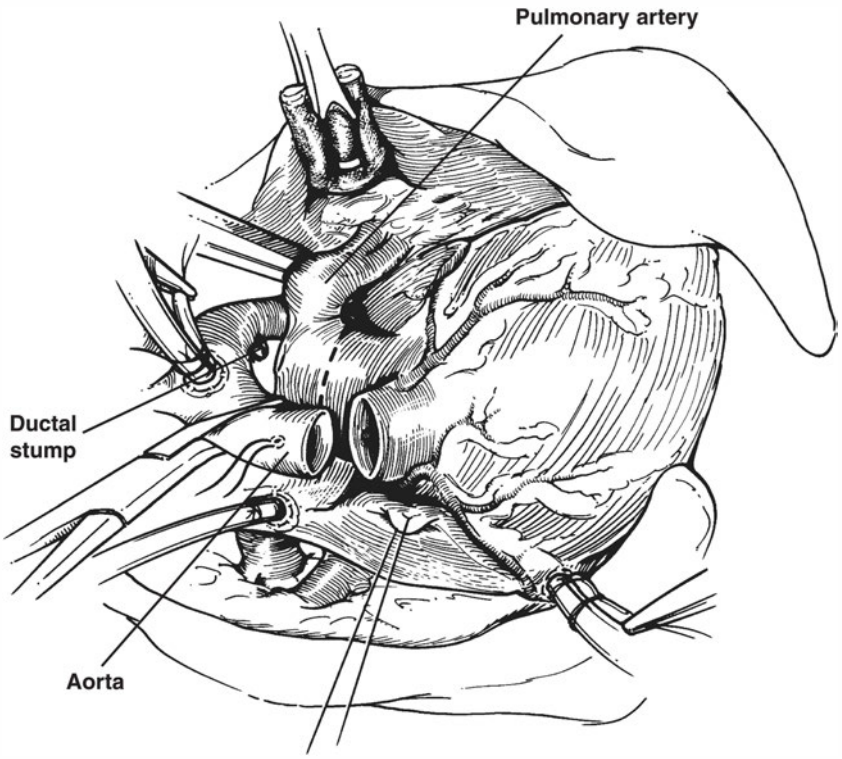


Fig. 89.3. The aorta is transected just above the level of the commissural attachments of the aortic valve, and the pulmonary artery bifurcation is transected at the dashed line at the bifurcation. Care must be taken not to cut into the origin of the right pulmonary artery. The pulmonary bifurcation is mobilized freely bilaterally out to the hilar branches to allow for translocation anteriorly.

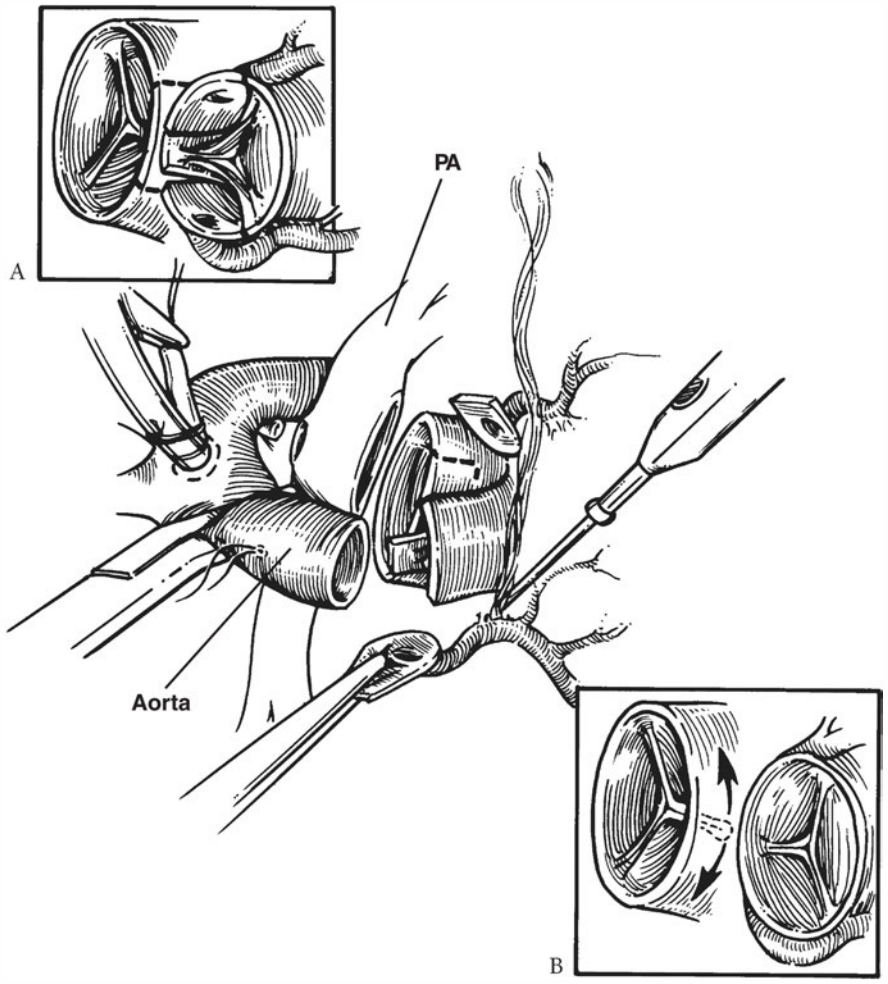


Fig. 89.4. The coronary ostia are excised with a button of aortic wall, and the proximal coronary arteries are mobilized with the cautery, sacrificing small conal branches if necessary to allow unrestricted translocation to the posterior great vessel. Medially based flap incisions are made posteriorly for reimplantation of the coronary arteries as noted by the dashed lines. (**Inset A**) The excision of the coronary ostia down to the sinuses of the aorta is noted, and the relationship of the vertical incisions in the posterior great artery is shown. (**Inset B**) The fact that the commissural attachment of the pulmonary valve may vary in location and relationship to the anterior great artery is shown. Division of the pulmonary artery (PA) at the bifurcation allows a 5- or 6-mm distance of the PA above the commissural attachments so that the coronary arteries can be reimplanted above the sinuses if necessary.

sinuses of the aortic and pulmonary valves face each other, the commissural attachment of the pulmonary valve may be dislocated superiorly or inferiorly and does not necessarily align directly with the commissural attachment of the aortic valve, as noted in Figure 89-4 (inset B). This fact must be taken into account when deciding about relocation of the coronary ostia. It is not uncommon for both coronary ostia to end up arising superior to the commissural attachments of the pulmonary valve or for both coronary ostia to be reimplanted to the same sinus of the pulmonary valve because of the variations in anatomy of the commissural attachments. It is generally preferable to reimplant the coronary arteries somewhat higher in the pulmonary artery than down into the sinuses of the pulmonary valve. This permits the coronary to come off of the neoaorta with less risk of kinking at the origin during distention of the aorta. As shown in Figure 89.5, the coronary ostia are then reimplanted into the medially based flap incisions. We prefer to use polypropylene suture for these anastomoses. On occasion, the placement of a vertically oriented suture line can result in kinking of the origin of the coronary artery, as noted in Figure 89.5D; in these cases, the rotation of the coronary buttons medially (Fig. 89.5E) can resolve the distortion.

Common variations in coronary translocation techniques are illustrated in Figures 89.6 and 89.7. As noted in Figure 89.6, when both coronaries come off a single sinus with the origin of the left coronary artery from an orifice near the commissural attachment of the aortic valve, it is possible to mobilize the commissure and excise the coronary arteries in the usual manner before translocation to the posterior great vessel. It is particularly important to note that the coronary artery may run intramurally in the aortic wall before exiting to the epicardial surface, and care must be taken not to cut across the coronary (Fig. 89.6B). An alternative technique for translocation that does not require separate coronary transfer is noted in Figure 89.7. The common coronary orifice or adjacent coronary orifices can be excised from the aortic wall as a single patch and then sewn side to side to the posterior great artery. The distal aorta can then be fashioned to create a flap over the origin of the coronary arteries, allowing unobstructed flow into the vessels (Fig. 89.7C). In the majority of cases, however, it is possible to transfer the coronary arteries directly to the posterior great vessel even when there is a single coronary from the right posterior-facing sinus, as noted in Figure 89.7D and 90.7E. Care must be taken, however, to position the coronary artery in such a manner that it does not kink one of the branches (Fig. 89.7E). Rotation of the coronary flap or translocation to a higher level on the pulmonary artery can adjust for many of the variations and possible kinking of the origins of the coronary arteries and allow coronary transfer in virtually all anatomic variations of TGA. In situations of single coronary ostium or side-by-side origin of the coronaries from the same sinus with very little distance between the orifices and intramural course, take down of the commissure, division of the coronary

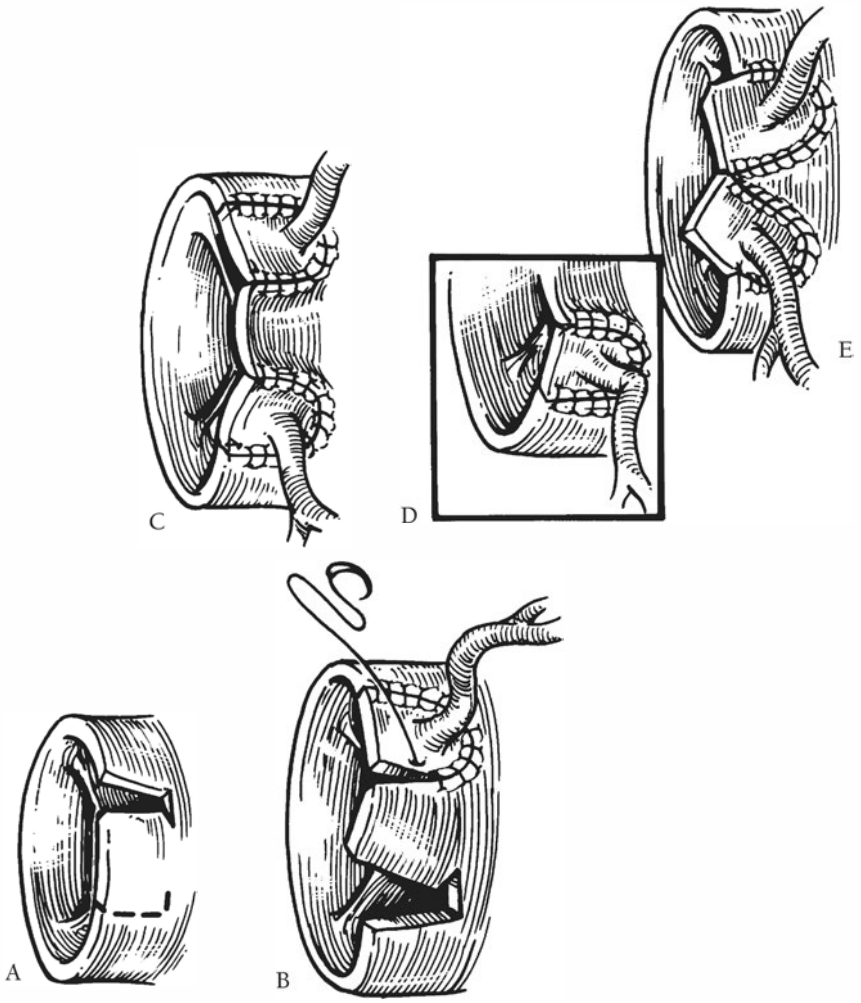


Fig. 89.5. (**A**) The medially based flap incisions in the posterior great artery are shown. (**B**) The coronary arteries are translocated to these incisions and anastomosed with absorbable running suture. (**C**) The transfer of the coronary arteries is complete and the takeoff of the vessels noted. (**D**) If the coronary artery is implanted vertically, in some cases the vessel is too low in the posterior great artery, and the origin of the vessel can kink during distention of the neoaorta. (**E**) In these cases, translocation of the coronary artery at an angle that is more superior can result in an unobstructed takeoff of the coronary artery from the aorta.

arteries, and rotation and reimplantation into the posterior great vessel may still leave an intramural course of the coronary that is prone to kinking and occlusion. The flap technique described earlier leaving the coronaries in situ may sometimes solve this potential problem; however, the Lecompte maneuver, bringing the pulmonary bifurcation anterior to the aorta, can compress the anteriorly located flap and limit the flow into the coronaries causing ischemia and potential late arrhythmias during exercise. One technique that has worked well for us in this situation is dividing the coronary arteries with reimplantation to the posterior great vessel and then performing an ostial coronary arterioplasty, cutting into the orifice of the intramural course of the coronary vessel and augmenting the origin with a small triangular patch of pulmonary homograft material or pericardium. This widely opens the orifice to the coronary artery and prevents kinking at the end proximal portion of the intramural course of the vessel (Fig. 89.7F and 90.7G).

Once coronary artery transfer has been completed, the distal aorta is then anastomosed to the neoaorta using polypropylene suture. Because there is frequently a size discrepancy between the neoaorta (to which the coronaries have been transferred) and the distal aorta, a vertical incision is used to make up the size discrepancy (Fig. 89.8). As noted in Figure 89.9, this incision can allow for adequate primary anastomosis in most circumstances even if there is a significant size discrepancy. The anteriorly placed vertical incision in the aorta has the additional advantage of pulling the aorta posteriorly, which helps to prevent compression of the pulmonary bifurcation that has now been anteriorly translocated. At this point, additional cardioplegic solution is injected into the aortic root (Fig. 89.10) to check for hemostasis of the suture lines, to ensure free perfusion of the coronary ostia without kinking, and to provide additional cardioplegia. Next, the defects in the anterior great vessel from which the coronaries have been excised are repaired with a generous portion of glutaraldehyde-fixed pericardium or pulmonary homograft material (Fig. 89.11). The patch is fashioned in a pantaloon shape and posteriorly is extended for 3 to 5 mm to allow for some extension and length of the pulmonary artery to the bifurcation. Anteriorly, native tissue is left in place and the patch trimmed appropriately to allow for an anterior portion of native tissue to be anastomosed to the pulmonary bifurcation in the hope of encouraging growth at this portion of the suture line. As noted in Figure 89.12, the anterior great vessel is then sutured to the pulmonary artery bifurcation, with care being taken to avoid tension on the pulmonary arteries, which may interfere with symmetric pulmonary blood flow. If necessary, an incision is carried out onto the origin of the pulmonary arteries to allow for an adequate size match and to prevent distortion of the origins of the pulmonary arteries. This anastomosis is occasionally completed with the aortic cross-clamp

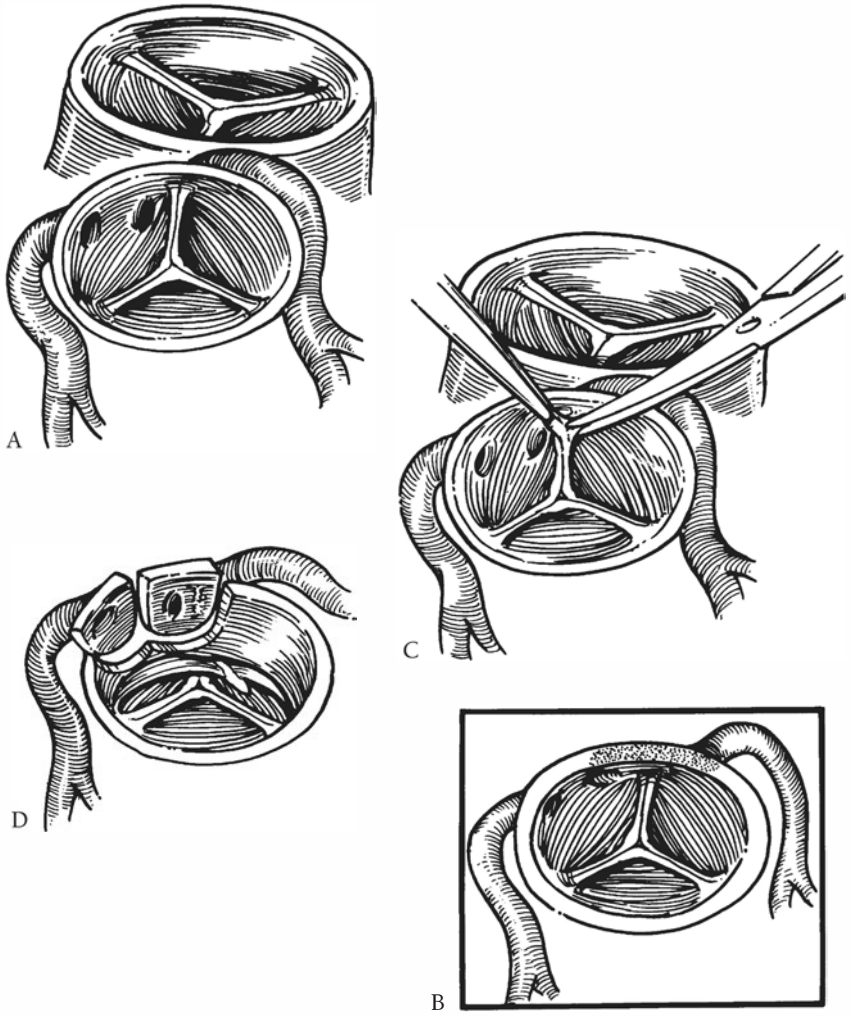


Fig. 89.6. (**A**) In variations in which both coronary arteries come off a single sinus, the coronary ostia may have an intramural course. (**B**) Care must be taken not to transect the coronary artery during excision of the coronary button from the aortic wall (*stippled area*). (**C**) The aortic valve leaflet may need to be detached from the commissure to allow access to the coronary ostia for excision. (**D**) Separate excision of the two coronary ostia can be effective in most cases.

released and the heart reperfusing. After pulmonary artery reconstruction, the right atrium is closed, and the pulmonary origin of the ductus is oversewn, with care being taken not to interfere with the flow into the origin of the left pulmonary artery (Fig. 89.13).

When the great vessels are located side to side rather than anteroposteriorly, it is occasionally advisable to leave the pulmonary confluence posterior to the aorta, close the pulmonary bifurcation on the left, and incise the right pulmonary artery more rightward. This may facilitate reconstruction to the right ventricle after the arterial switch has been performed.

After completion of the repair, right atrial lines are placed for postoperative monitoring and the volume infusions and temporary pacemaker wires applied to the right atrium and the right ventricle. After the patient is weaned from cardiopulmonary bypass, a systemic pressure of no more than 60 to 70 mmHg is preferable to prevent distention of the newly systemic left ventricle.

The Rastelli Operation

Infants with significant left ventricular outflow tract obstruction represent a small proportion of children with TGA. Left ventricular outflow tract obstruction in TGA is often dynamic in nature, so the relative contributions of the dynamic components of obstruction and the fixed components such as subvalvar fibrous rings and mitral valve leaflet tags are difficult to determine. At the time of arterial switch operation, some of the fixed obstruction can be resected across the pulmonary valve, but complete relief is not often possible. In the majority of cases, however, moderate relief of left ventricular outflow tract obstruction can be well tolerated because the pulmonary ventricle has been preconditioned to elevated intracavitary pressure.

When significant left ventricular outflow tract obstruction in association with TGA presents in infancy, creation of an interatrial communication and a systemic-to-pulmonary arterial shunt is often the best early approach, followed later by the Rastelli operation. In this situation, early complete repair is preferred, and we have generally elected to complete the operative procedure by the time the patient is 4 to 6 months of age even if a systemic-to-pulmonary artery shunt is initially performed.

In the Rastelli operation (Figs. 89.14 to 89.19), pulmonary venous blood is directed across the VSD to the aorta with an intracardiac baffle, and a valved conduit is used to reconstruct the right ventricular outflow tract to the distal pulmonary arteries. Figure 89.14 shows the transposed great arteries with the ascending aorta larger than the posteriorly located pulmonary artery because of significant pulmonary stenosis. A right ventriculotomy is created through an oblique incision heading leftward toward the pulmonary bifurcation. The VSD is easily exposed through this incision. As noted in Figure 89.15, the VSD is often somewhat smaller than the ascending aorta, and enlargement of the VSD is recommended to prevent late development of

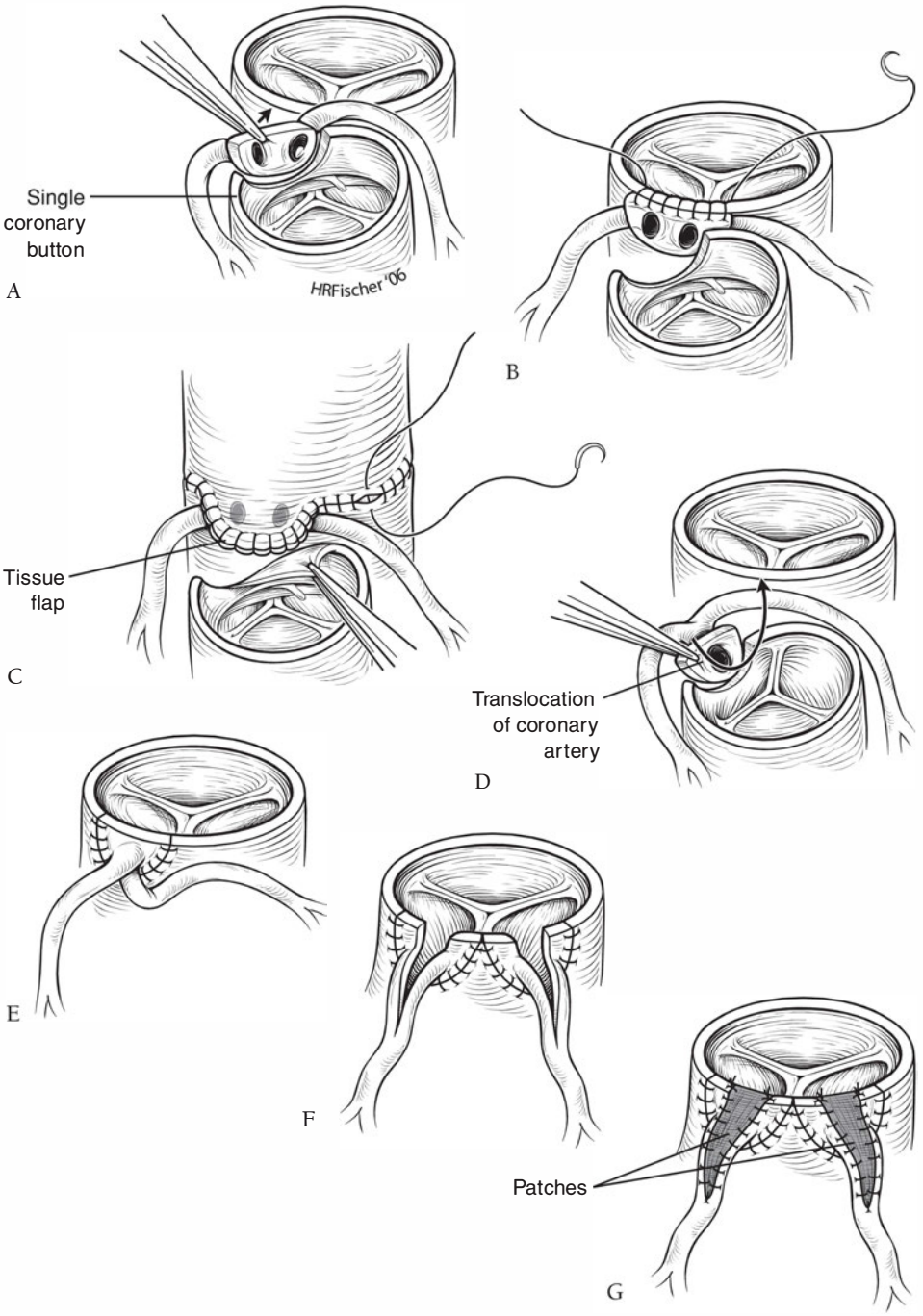


Fig. 89.7. (**A**) In some cases, excision of the single coronary ostium supplying both right and left circulations or side-by-side ostia from the same sinus are best translocated with a single coronary button excised from the sinus of the aortic valve. (**B**) The superior margin of the coronary button is anastomosed to the superior margin of the posterior great vessel. In this manner, there is no translocation of the coronary ostia posteriorly, but they are left in their normal anatomic relationships to prevent kinking of the arteries. (**C**) The aorta is then mobilized to create a flap of tissue sewn down over the coronary ostia, allowing unobstructed flow into the coronary vessels. (**D**) Translocation of a single coronary artery to the posterior great vessel can be effective in most cases. Care must be taken, however, to orient the artery such that kinking of the left coronary branch does not occur. (**E**) A more superior location of the coronary artery on the posterior great vessel generally relieves such kinking. (**F**) In situations with intramural coronaries with ostia that arise adjacent posteriorly from the aorta, the coronary buttons can be excised and the intramural course opened inside the aortic wall. The coronaries are then rotated and then reimplanted into the posterior great artery in the usual manner. (**G**) The narrow nature of the coronary arteries despite opening of the intramural course still often leaves stenosis, and therefore, incision of the coronary with patch augmentation with a small triangular patch of pulmonary homograft or pericardium material can ensure that there is no coronary inflow problem. This patch coronary ostioplasty must be done in a meticulous manner to prevent distortion and kinking of coronary flow.

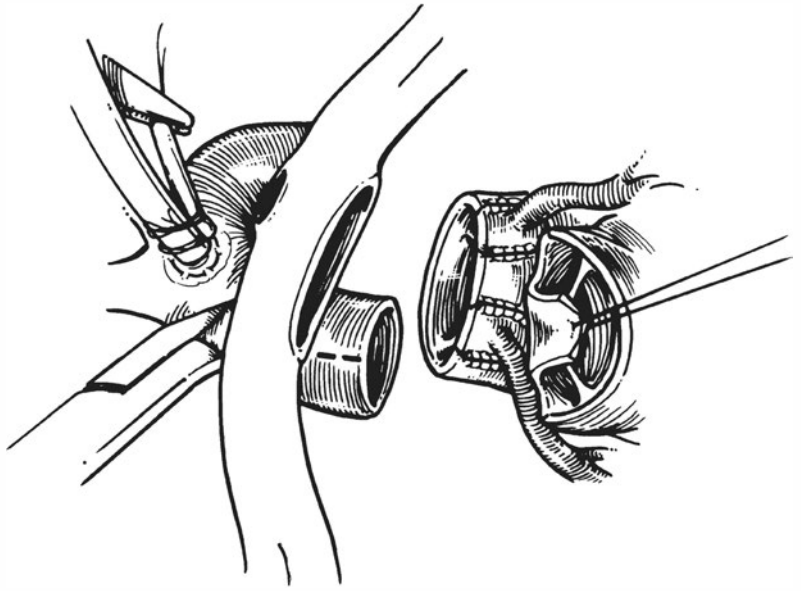


Fig. 89.8. The pulmonary bifurcation is translocated anterior to the great artery in the Lecompte maneuver. To obtain an adequate size match between the distal ascending aorta and the proximal great artery to which the coronaries have been transferred, a vertical incision is used.

obstruction across the intracardiac baffle, which becomes the outlet to the ascending aorta. Restriction of the VSD is one of the most common late problems after the Rastelli operation. In Figure 89-15 (inset A), the VSD is enlarged anteriorly and superiorly away from the area of the conducting tissue. The pathway of the conducting tissue along the right ventricular septum is noted in Figure 89.15B (stippled line), and the suture line for placement of the baffle from the left ventricle to the aorta is noted by the dashed line. In Figure 89.16, the creation of a conduit-like patch from either a polyester (Dacron) tube graft or a polytetrafluoroethylene (Gore-Tex) tube graft is described.

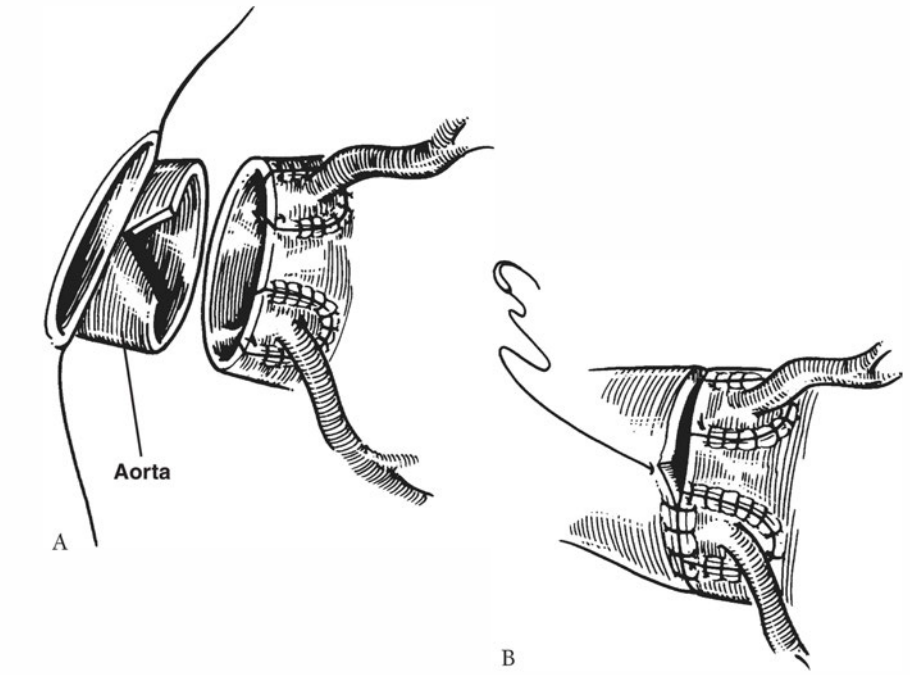


Fig. 89.9. The vertical incision in the distal aorta is shown, creating an adequate size match for the anastomosis (**A**), which is completed with a running absorbable suture (**B**).

Alternately, a patulous rectangular patch of Dacron material can be used, allowing for a large amount of the material to bow into the right ventricle to permit an unobstructed connection from the VSD at the ascending aorta. We prefer to use Gore-Tex material for these connections because the rough surface of the Dacron patch material may cause hemolysis early postoperatively.

After the baffle is completed, the left ventricular output is directed across to the ascending aorta anteriorly (Fig. 89.17). The pulmonary artery is then divided and the pulmonary bifurcation incised for connection to a pulmonary homograft conduit of the largest size that will satisfactorily fit into the chest cavity (Fig. 89.18). Homograft material is preferred for reconstruction of the right ventricular outflow tract because it is compressible and typically will allow the conduit to sit in the left chest underneath the sternum without significant distortion. The pulmonary valve is oversewn to prevent antegrade flow out of the ventricle into a blind pouch, which could be a source of potential thrombi and thromboembolism. The stump of the pulmonary artery is also oversewn. The right ventricular outflow tract is then reconstructed with a pulmonary valved homograft with or without the use of a gusset, allowing a gentle takeoff from the right ventricle to the pulmonary outflow tract (Fig. 89.19).

Unusual cases of children with transposition and intact ventricular septum with significant left ventricular outflow tract obstruction may be treated by atrial baffle repair and connection of a conduit from the left ventricular cavity to the pulmonary bifurcation on the left.

Lecompte modified the Rastelli operation to decrease the risk of obstruction of the conduit from the left ventricle to the aorta by creation of the REV procedure. In this operation, resection of the subaortic conus is more extensive than in the Rastelli operation, and resection of muscle is carried to just below the pulmonary valve annulus superiorly (Fig. 89.20A and 89.20B). In addition, when tricuspid valve attachments to the ventricular septum are abnormal, a septal flap containing the important tricuspid valve attachments can be created and repositioned onto the ventricular septal patch after it is sewn in place. These more extensive resections of the ventricular septum create a more direct pathway from the left ventricle to the ascending aorta with a lower risk of recurrent stenosis. In addition, Lecompte suggested translocation of the pulmonary

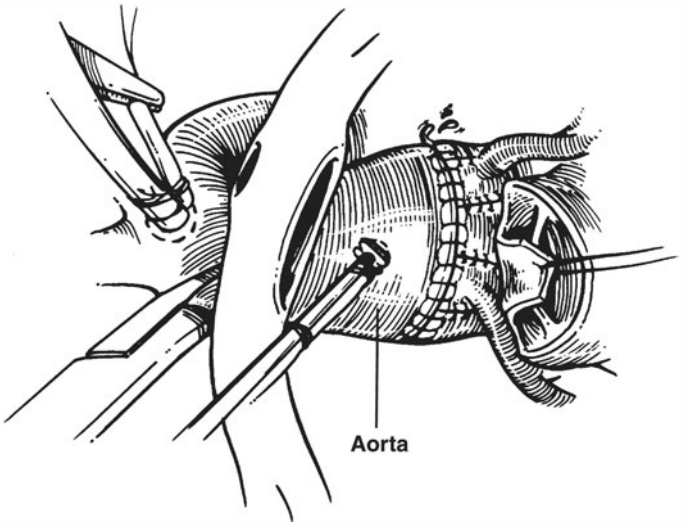


Fig. 89.10. After the coronary transfer and ascending aortic reconstruction are completed, cardioplegic solution is injected into the aortic root to ensure free flow into the coronary arteries and hemostasis at the suture lines.

bifurcation anteriorly in these patients with resection of a short portion of the ascending aorta to reposition the aorta more posteriorly and allow direct connection of the pulmonary bifurcation to the right ventricular outflow tract with a monocusp patch or nonvalved direct connection. In this manner, complete repair can be performed in patients with TGA, VSD, and left ventricular outflow tract obstruction without the need for a homograft conduit, which would require later revision. A modification of the approach described by Lecompte resects a short segment of the aorta decreasing the length of the aorta and bringing it into a more posterior position. This short segment of aorta that is excised can then be used to reconstruct the pulmonary outflow tract using autologous tissue to allow for potential growth.

A variation on the REV procedure has been described by Nikaidoh and is useful for patients who have transposition-like forms of double-outlet right ventricle with VSD and small pulmonary valve annulus. In this approach, the pulmonary annulus can be incised across the conal muscle into the VSD and the aorta excised with mobilization of the coronary arteries as in the pulmonary autograft procedure for aortic valve replacement. The entire aortic root with the coronaries attached can then be positioned more posteriorly into the left ventricular outflow tract and secured to the pulmonary valve annulus posteriorly (Fig. 89.21A). The VSD is then closed anteriorly with a patch secured to the inferior margin of the translocated aorta. In this manner, the outflow from the left ventricle becomes more directly related to the aorta without a complex intracardiac baffle and often without requiring enlargement of the VSD. The right ventricular outflow tract can then be reconstructed either with direct connection to the pulmonary bifurcation or with the use of a homograft valved conduit (Fig. 89.21B and 89.21C). In some cases, it is simpler to excise the coronary artery buttons and mobilize them to gain access to the subaortic area for excision of the aortic root before translocation of the aortic root posteriorly. The coronary buttons can then be reimplanted in a suitable orientation to the mobilized aorta, decreasing the potential risk of kinking along the epicardial course of the vessels.

An alternate approach to some of these patients may be pulmonary artery translocation. This operation has been described by DaSilva and takes the pulmonary valve from the left ventricle and transposes it to the right ventricle with patch closure of the pulmonary outflow tract. This removes the subaortic obstruction and can position the right ventricular outflow tract in a more anatomic location and provide more direct connection from the left ventricle to the ascending aorta.

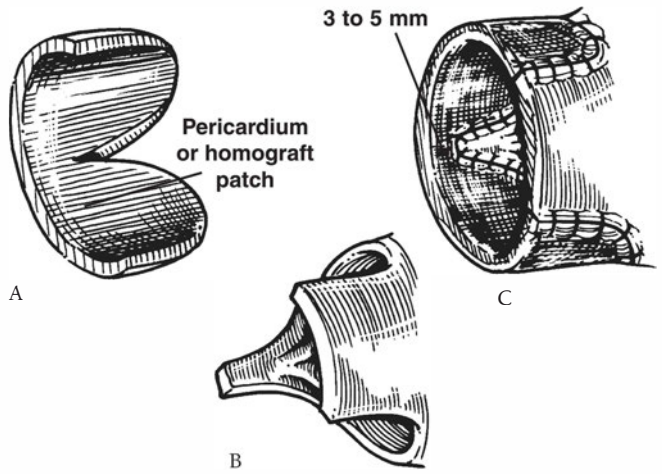


Fig. 89.11. The anterior pulmonary artery is then reconstructed with a pantaloon-shaped patch of pulmonary homograft or autologous pericardium with an extension approximately 3 to 5 mm above the posterior commissure to elongate the pulmonary artery before the bifurcation.

SURGICAL RESULTS OF REPAIRS OF TRANSPOSITION OF THE GREAT ARTERIES

The results of either a Mustard or a Senning atrial switch procedure have generally been good, with survival rates of 80% to 95% reported for complete repair in infancy in patients with intact ventricular septum. Atrial switch operations have been complicated by a high incidence of sinus node dysfunction or other atrial dysrhythmias, and the development of dynamic left ventricular outflow tract obstruction from systemic right ventricular pressure loading. The Senning operation has been associated with improved rates of sinus rhythm compared with the Mustard operation, although atrial arrhythmias are not uncommon with this approach. The incidence of late

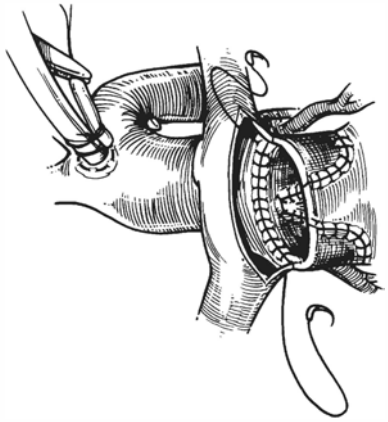


Fig. 89.12. The pulmonary bifurcation is then reconstructed onto the anterior great artery using running nonabsorbable suture. Incisions onto the right and left pulmonary arteries are made as necessary to create an adequate size match.

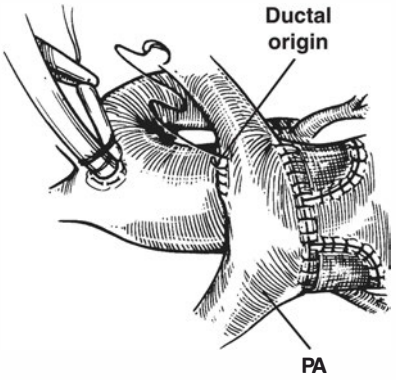


Fig. 89.13. After reconstruction, the origin of the ductus arteriosus is oversewn with absorbable suture, with care being taken not to narrow the takeoff of the left pulmonary artery (PA).

deterioration in systemic right ventricular function is unknown but may become clinically significant in 10% of patients. Results with atrial correction appear to be improved with early repair before the development of significant pulmonary vascular obstructive disease, and age alone does not appear to be a significant independent predictor of operative risk.

Arterial switch operations have been associated with progressive improvement in operative and hospital mortality. Perhaps, the largest series of arterial repairs for transposition of the great vessels has been reported by the group at the Boston Children's Hospital, in which the 1-, 5-, and 8-year survival rates were 93%, 92%, and 91%, respectively. Additionally, they have shown that left ventricular size, mass, functional status, and contractility are normal on follow-up with no evidence of late deterioration. Follow-up studies of atrial and ventricular arrhythmias after the arterial switch operation have shown a 96% incidence of sinus rhythm on electrocardiography and 99% during Holter monitoring at a mean of 2.1 years postoperatively, which represents a marked improvement over the results with the atrial operations. The results of these careful studies have confirmed the superiority of the anatomic arterial switch procedure over atrial repairs. In spite of the excellent intermediate-term results with the arterial switch operation, late problems do occur in some of these children. Supravalvar pulmonary stenosis is the most frequent anatomic complication following the arterial switch, with a peak incidence during the first year in 2% to 30% of patients. Dilation of the aortic root and ascending aorta with progressive aortic insufficiency has been seen with increasing frequency in older patients after the arterial switch operation, perhaps reflecting the abnormal pulmonary arterial wall of the aortic root. In a large series out of Boston Children's Hospital, over half of patients developed aortic root dilation beyond three standard deviations after 10 years. Coronary arterial obstruction following arterial switch has also been reported in 2% to 7% of patients. Symptomatic patients with significant obstruction clearly require intervention, but there are little data regarding the rate of progression and timing of intervention on asymptomatic patients with mild-to-moderate obstruction. Overall though, most patients currently undergoing arterial switch operations can expect near normal developmental, neurologic, and cardiac function.

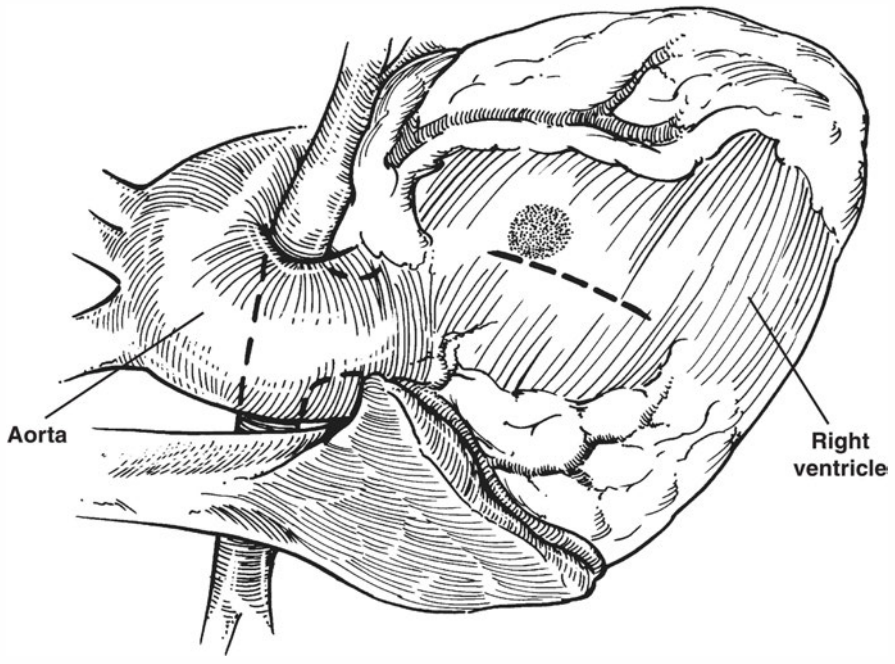


Fig. 89.14. The Rastelli operation. In transposition of the great arteries with significant left ventricular outflow tract obstruction, the aorta is larger than the posteriorly located pulmonary artery. The dashed lines show the direction for the ventriculotomy incision in the right ventricle avoiding major epicardial coronaries, and the location of the ventricular septal defect is noted.

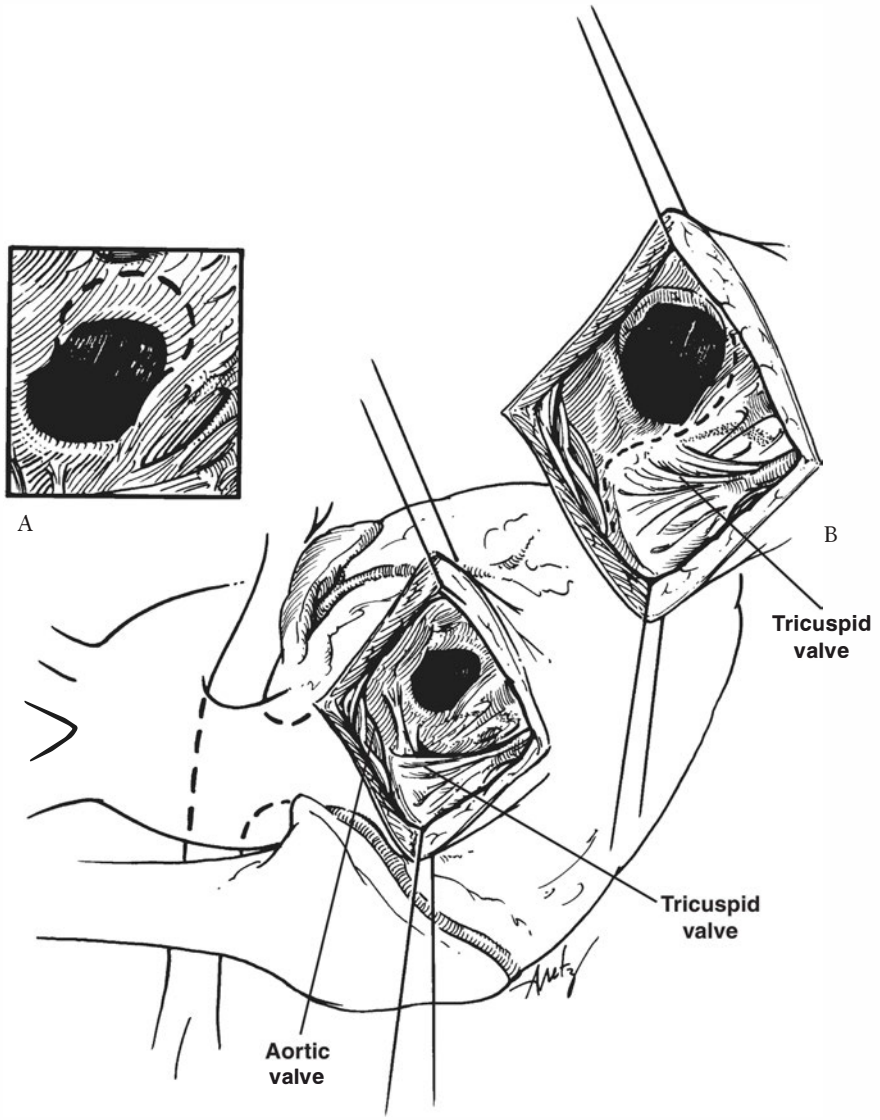


Fig. 89.15. The ventricular septal defect and tricuspid valve attachments. **(Inset A)** Excision of the anterior superior margin of the ventricular septal defect away from the conducting tissue allows an unobstructed connection from the left ventricle to the aorta. **(Inset B)** In the "réparation à l'étage ventriculaire" (REV) procedure, the conal muscle beneath the aorta separating the aorta and pulmonary valves is also resected at the anterior left lateral margin of the ventricular septal defect away from the conducting tissue (*stippled lines*). The direction of the suture line for the ventricular septal defect patch to the aorta is shown by the dashed lines, avoiding the conduction tissue.

As experience has been accumulated with patients after the arterial switch operation, it is apparent that the overall ventricular performance and the incidence of arrhythmias are significantly improved over atrial switch-type operations. Nevertheless, the neoaortic root continues to initially have been a pulmonary root and root dilation over time has been seen as in patients after the Ross operation or after repair of hypoplastic left heart syndrome. While the need for reoperation for aortic insufficiency and root dilation appears to be quite low, continued monitoring of the patients over time will be necessary to determine whether the aortic valve will require reintervention in the future. In addition, there is a significant incidence of coronary artery stenosis and even asymptomatic coronary occlusion after the arterial switch operation, which may require additional interventions in the future. While it is clear that the arterial switch operation results in a significant improvement over the atrial switch-type repairs, continued longitudinal follow-up of patients will be necessary over a lifetime to determine the need for reintervention for various complications of this congenital anomaly.

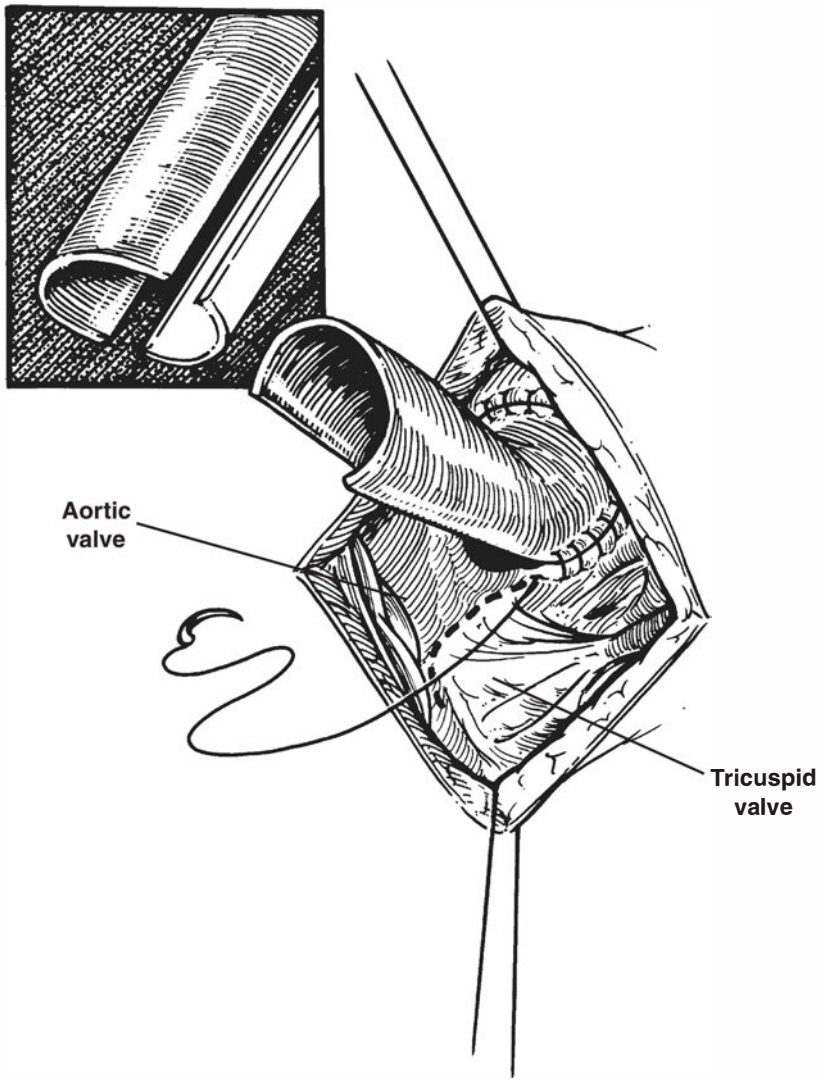


Fig. 89.16. A baffle created from a polytetrafluoroethylene (Gore-Tex) tube graft **(inset)** is used to create the connection from the left ventricle to the aorta.

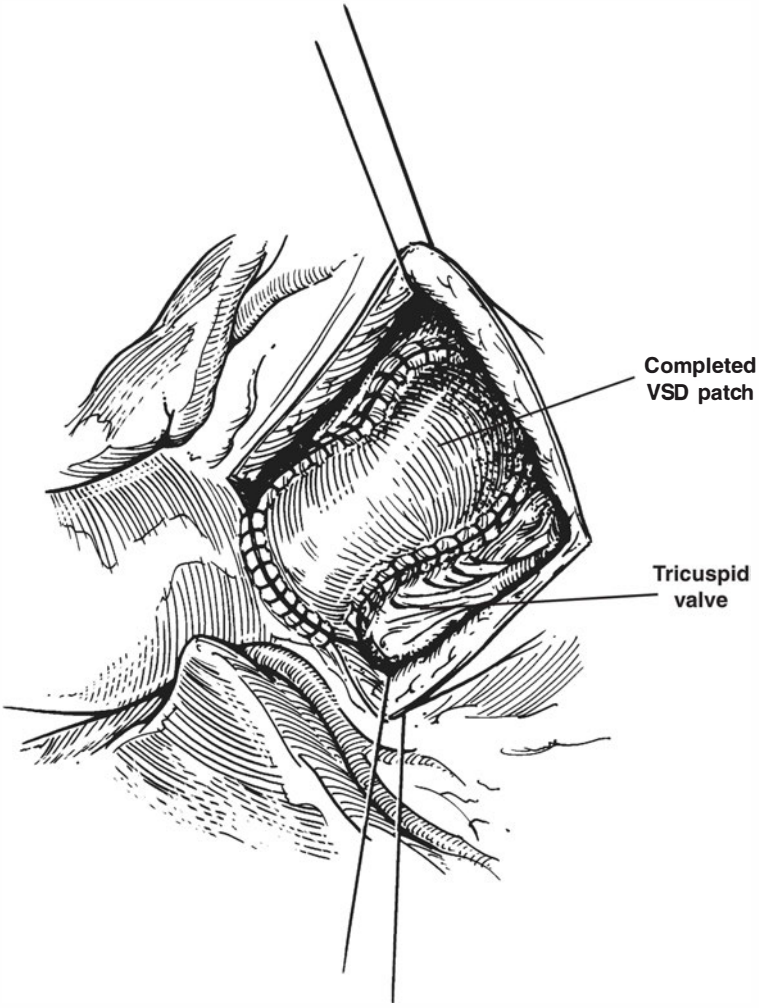


Fig. 89.17. Completion of the baffle creates a conduit from the left ventricle to the aorta sewn to the base of the septal tricuspid valve leaflet inferiorly. VSD, ventricular septal defect.

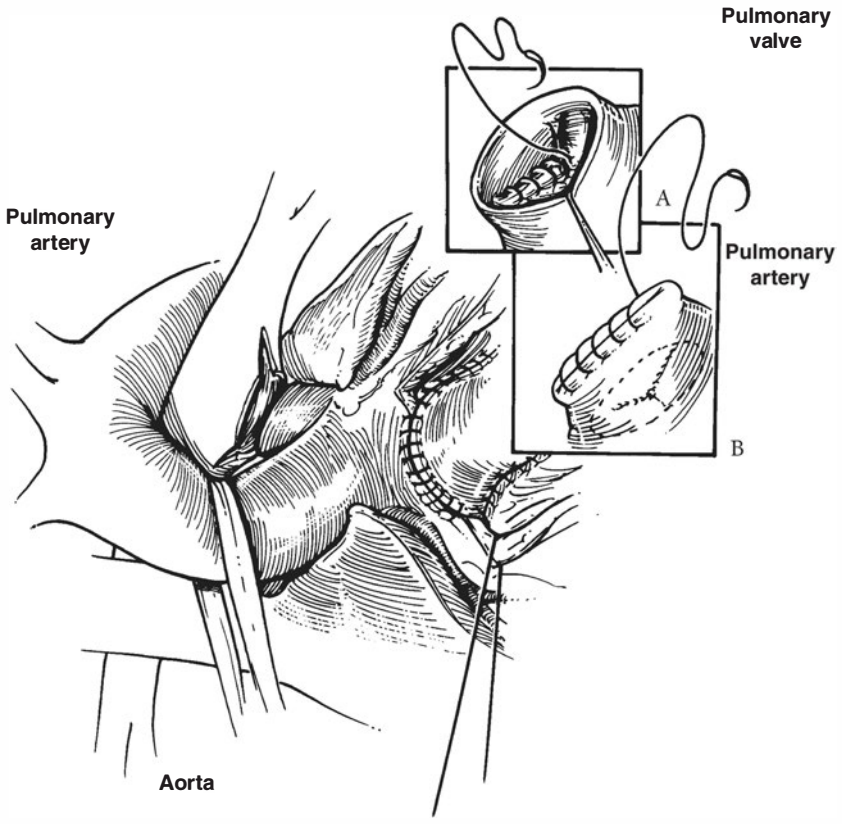


Fig. 89.18. After the intracardiac portion of the repair is completed, the pulmonary artery is transected and the pulmonary valve (**inset A**) and the main pulmonary artery (**inset B**) are oversewn to prevent stasis and bleeding. Alternatively, the pulmonary valve can be excised. The distal pulmonary artery is opened adequately for reconstruction with a pulmonary homograft.

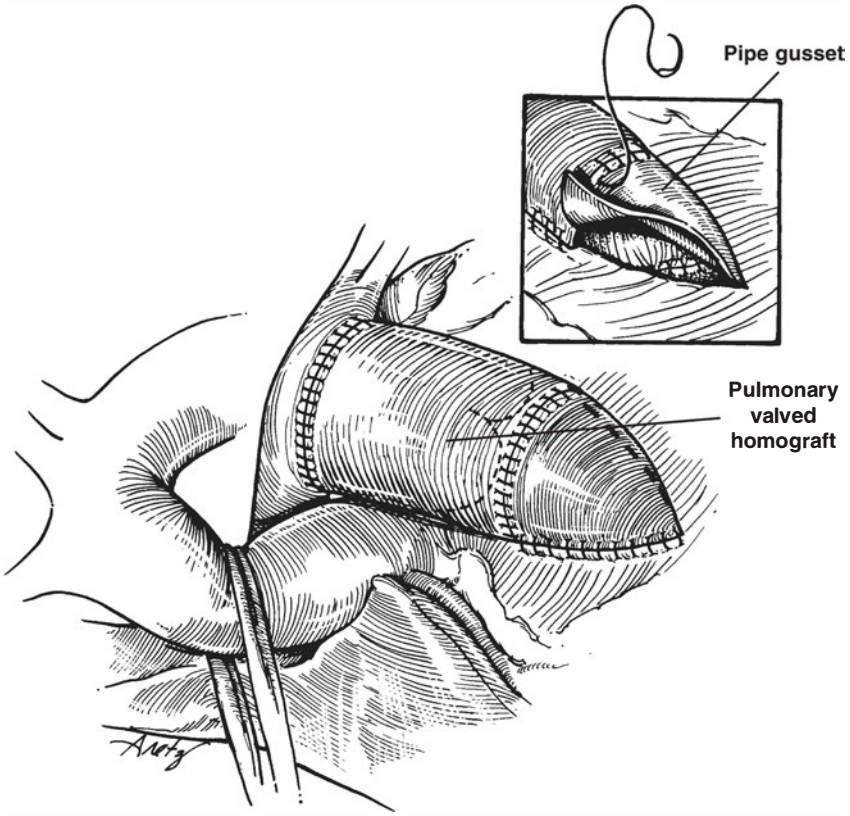
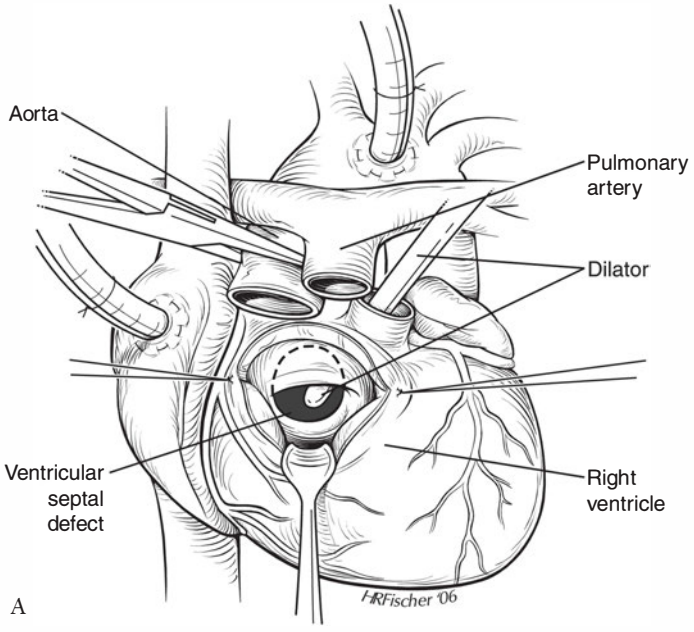


Fig. 89.19. The right ventricular outflow tract is reconstructed using a pulmonary valved homograft to the pulmonary bifurcation augmented by a polytetrafluoroethylene (PTFE) gusset from the right ventricle (**inset**).



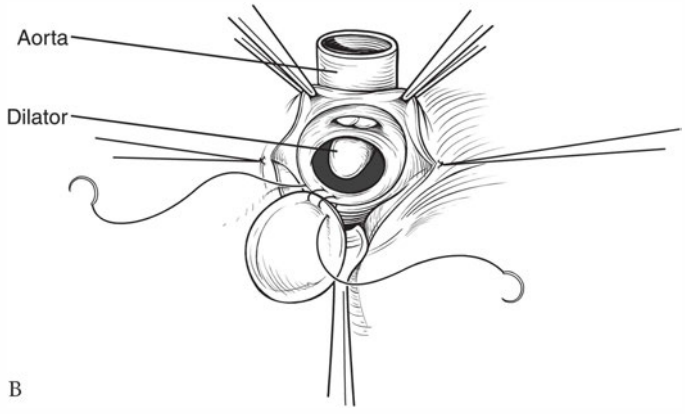


Fig. 89.20. (**A**) In the "réparation à l'étage ventriculaire" (REV) procedure, the aorta and the pulmonary artery are transected, and through an incision in the right ventricular outflow tract the infundibulum is widely excised to create an unobstructed pathway from the ventricular septal defect (VSD) to the aorta. A dilator can be passed across the outflow to guide resection of the VSD to avoid damage to the semilunar valve. (**B**) A generous patch of Dacron is used to close the VSD, directing the flow to the aorta. The resection of conal muscle provides a more direct connection of the ventricle to the aorta than in the standard Rastelli operation. The pulmonary artery is then reconnected to the right ventricular outflow tract after a Lecompte maneuver is performed, bringing the pulmonary arteries anterior to the aorta.

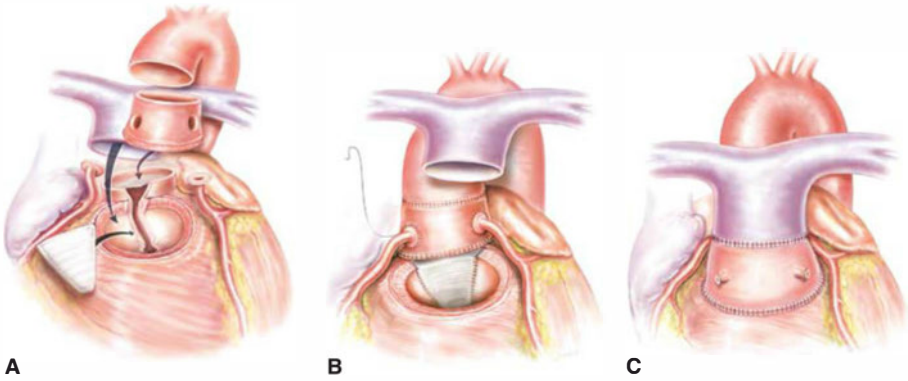


Fig. 89.21. (**A**) The Nikaidoh procedure involves excising the aortic root including the aortic valve from the ventricle by making an incision below the aortic valve annulus and mobilizing the entire root along with the attached coronary arteries. If exposure is difficult, the coronaries can be excised for reimplantation, much as in an arterial switch operation. Once the aortic root has been mobilized, the muscle between the pulmonary annulus and the ventricular septal defect (VSD) is divided, widely opening up the outflow from the left ventricle to the pulmonary valve annulus, and additional muscle resected if necessary. After infundibular muscle is resected, the aortic root is reimplanted into the left ventricular outflow tract at what was the previous pulmonary valve annulus. After the aortic root is translocated posteriorly, the anterior VSD is closed with a patch of Dacron or polytetrafluoroethylene material, which is secured anteriorly to the anterior portion of the aortic root, thus baffling the blood from the left ventricle to the aorta. (**B** and **C**) The right ventricular outflow tract reconstruction is created with an aortic or pulmonary homograft conduit.

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EDITOR'S COMMENTS

Dr. Eckhauser and Spray have written a thorough description of the physiology and surgery for transposition of the great arteries. There has been a huge transformation of the procedure for this disease. I was involved very early on with the mustard and senning procedures which left the anatomic right ventricle as a systemic left ventricle. We learned the hard way how these ventricles often failed in the long run. The change to this procedure basically made these infants normal.

There is also no procedure that is technically more exacting in congenital heart disease. It is absolutely critical to get the coronaries right. There needs to be adequate mobilization and we have used marking ahead of time to determine the proper place to put these buttons while the heart is still full and beating. The differences are huge. This is a procedure that if done correctly should have virtually no mortality as long as there are no issues that causes kinking or compression of the coronary arteries. These authors have demonstrated different approaches for different coronary anatomy variations. The bottom line is that the vast majority of patients can have a switch successfully done.

ILK

90 Congenitally Corrected Transposition of the Great Arteries

Ergin Kocyildirim and Victor O. Morell

INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a complex cardiac lesion involving discordant atrioventricular (AV) and ventriculoarterial connections. The term congenitally corrected transposition was introduced by von Rokitansky in 1875. He described two patients in whom; the abnormal relationship of the great arteries was corrected functionally by the position of the ventricular septum. Cardell, Anderson et al., Lev and Rowlatt, and Schiebler et al. reported extensive anatomical descriptions and clinical studies. Anderson et al. described the anatomy of the conduction system. Alternative terms used to describe ccTGA such as L-TGA, for situs solitus (S), L-loop (L), and the aorta left to the pulmonary artery, and {I,D,D} for situs inversus with mirror image anatomy. D-loop is for D-transposed great arteries, where L means left and D means right. ccTGA have become a universal termination according to the Society of Thoracic Surgeons Congenital Heart Surgery Nomenclature and Database Project.

In this complex cardiac lesion, the right atrium is connected to the morphologic left ventricle, which is connected to the pulmonary artery and the morphologic left atrium is connected to a morphologic right ventricle, which is connected to the aorta (Fig. 90.1). As a result of the discordant connections at both levels, the blood flows in normal physiology. ccTGA represents the 1% of all congenital cardiac anomalies and about 94% of all cases are associated with other cardiac lesions, with the most common abnormalities involving the tricuspid valve (up to 91% of patients). Other associated defects are ventricular septal defects (VSDs), and pulmonary valve or subpulmonary stenosis resulting in left ventricular outflow tract obstruction (LVOTO). Also, dextrocardia is frequently associated with this cardiac defect. Most commonly, the associated lesions determine the severity of the symptoms and the surgical management strategy.

EMBRYOLOGY AND ANATOMY

During embryonic development, the primitive heart tube develops several curvatures. Corrected transposition in visceroatrial situs solitus develops when the primitive heart tube loops to the left instead of the right, resulting in the absence of spiral rotation of the conotruncal septum. This causes the aorta to connect to the morphologic right ventricle and the pulmonary artery to connect to the morphologic left ventricle. The normal process of septation and valve formation is also affected and the ventricular morphology is maintained consistent within the ventricular chamber. The AV valve and conduction tissue correspond to the overall morphology of each ventricle. The process of malseptation leads to a VSD, which is a frequent associated anomaly (50% to 80% of all cases). The VSD is usually perimembranous and large but can be located anywhere in the septum.

When atrial situs inversus is present, a mirror-image relation exists and the aorta is almost always to the right of the pulmonary artery with dextrocardia. Malalignment of the atrial and ventricular septa is present, except where the pulmonary, mitral, and tricuspid valves meet and are joined by the right fibrous trigone. The atrial septal attachment to the fibrous skeleton lies to the right of the ventricular septal attachment. These changes result in the abnormally positioned AV node and His bundle. Abnormalities of the AV node, including dual AV node with the abnormal His bundle, are quite common and many of those ccTGA patients develop complete heart block spontaneously during intra- or extra uterine life. Tricuspid valve and VSD surgery may also precipitate the complete heart block.

The left ventricular outflow tract in this cardiac defect is located in the region between the septal leaflet of the mitral valve and the muscular ventricular septum. Anatomic obstruction is significant and hemodynamically important in about 50% of the patients.

Tricuspid valve anomalies are noted frequently. When Ebstein anomaly occurs in the presence of ccTGA, the tricuspid valve and the right atrial morphology are different. Unlike single Ebstein anomaly, the anterior leaflet is not sail-like and the atrialized part of the right ventricle is small.

In atrial situs solitus, the right-sided coronary artery, which feeds left ventricle, usually gives rise to the anterior descending and circumflex branches. The left-sided coronary artery supplies the right ventricle and becomes the posterior descending artery. The most common variation of the coronary artery distribution is the existence of a single coronary artery that arises from the right-facing sinus and divides into right and left main branches. In ccTGA, there is also a tendency toward early branching of the left main.

DIAGNOSIS

Patients with ccTGA may seek medical advice at any age, and their timing of presentation is usually dependent on the presence and severity of associated cardiac anomalies. Cyanosis could be the initial clinical presentation of a neonate or infant with severe pulmonary stenosis or pulmonary atresia requiring palliation with a systemic to pulmonary artery shunt. Not infrequently patients with pulmonary stenosis and a VSD have a nicely balanced circulation allowing for delayed surgical management during late infancy or early childhood.

Infants may experience congestive heart failure due to a large VSD, severe tricuspid regurgitation, aortic coarctation, or aortic arch abnormalities. Bradycardia can be the initial presenting symptom and it is most often caused by the spontaneous

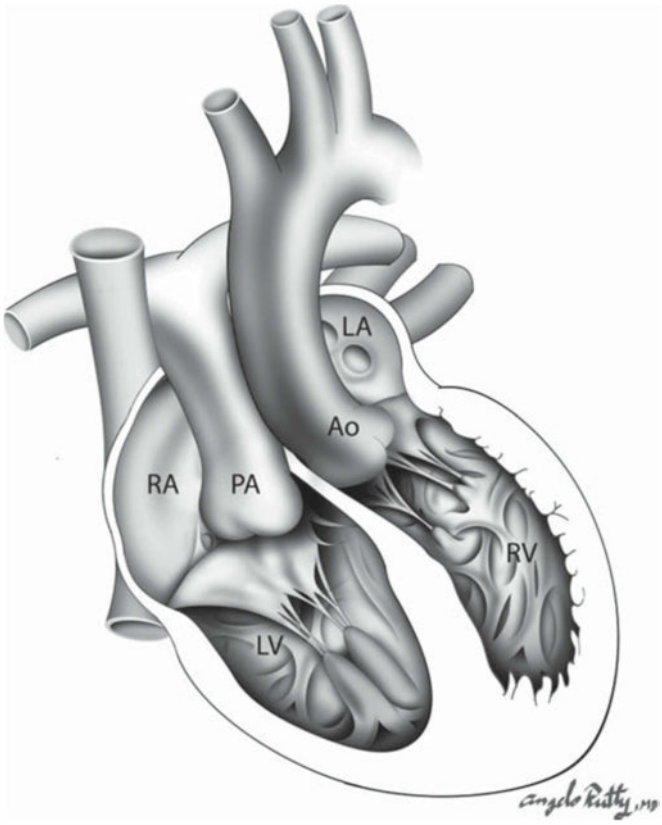


Fig. 90.1. Congenitally corrected transposition of the great arteries. Note the presence of atrioventricular and ventriculoarterial discordance. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.

development of complete heart block. The proportion of patients with heart block increases 2% per year to reach 30% by adult life. In the absence of associated cardiac anomalies, most often the defect manifests itself in older childhood or in the second decade, with the symptoms of exercise intolerance and cyanosis. In rare conditions, patients may go undiagnosed until late adulthood, when they present with symptoms of atypical chest pain and RV failure.

The physical findings generally do not provide sufficient information to confirm the diagnosis of ccTGA. If tricuspid valve regurgitation is present, left parasternal lift and a systolic murmur akin to mitral regurgitation can be heard at the left sternal border. A leftward aorta and rightward posterior pulmonary artery can be present on the chest X-ray. The presence of AV block and a Q wave in the right precordial leads suggests the diagnosis.

The diagnosis of ccTGA is most often made by echocardiography. This study is useful to assess the ventricular morphology, mitral-pulmonary fibrous continuity, ventriculoarterial discordance, great vessels, and the associated cardiac lesions. Transesophageal echocardiography is also a useful tool in making diagnosis in adults.

Cardiac catheterization is useful in assessing ventricular morphology, systemic AV valve regurgitation, systemic ventricular function, and the presence and significance of any associated cardiac anomalies. Coronary angiography is indicated and important in older patients.

SURGICAL TREATMENT

Palliative Operations

Patients in need of palliative surgery usually have either significant cyanosis or pulmonary overcirculation. In neonates and infants, significant pulmonary valve and/or subvalvar obstruction (LVOTO) in the presence of a VSD results in cyanosis. These patients can be initially managed with the placement of a modified Blalock–Taussig shunt.

Pulmonary artery banding could be utilized in neonates and infants with pulmonary overcirculation and failure to thrive secondary to the presence of a VSD. In these patients, the aim of the procedure is to reduce the distal pulmonary arterial pressure to $<50\%$ of the systemic pressure.

In patients who develop right ventricular failure after a classic repair pulmonary artery banding may be considered for LV retraining when a double-switch strategy is being considered. The ideal morphologic LV pressure after the banding should be approximately 75% of systemic. The development of morphologic LV dysfunction after the procedure is well recognized and could require postoperative inotropic support and/or loosening of the band.

Definitive Operations

The approaches to the surgical management of patients with ccTGA can be grouped into two options: physiologic (classic) or anatomic repairs. With a *physiologic repair*, the right ventricle remains the systemic ventricle and all other associated cardiac lesions are repaired. This approach is simpler but results in a RV-dependent systemic circulation, which has negative long-term implications, mainly RV failure and tricuspid regurgitation. After *anatomic repair*, the left ventricle becomes the systemic ventricle and the long-term outcomes are better. Nevertheless, the anatomic repair involves some form of a technically challenging double-switch procedure.

Closure of Ventricular Septal Defects

The majority of the VSDs in ccTGA are located in the perimembranous region. The presence of an overriding or straddling mitral valve usually signifies the presence of inlet extension. Most commonly, the surgical approach is via the right atrium with exposure of the VSD through the mitral valve. In selected cases, partial detachment of the septal leaflet of the mitral valve allows for better visualization of the defect. Transatrial approach is applicable to both physiologic and anatomic repairs. In patients requiring placement of a conduit between the morphologic LV and pulmonary arteries, the VSD could be easily closed through the ventriculotomy.

In the SLL type of L-TGA, it is important to realize that the conduction tissue runs in the subpulmonary infundibulum and along the anterior border of the defect. During VSD closure sutures must be placed on the left (systemic) side of the defect in order to avoid injury to the conduction system (Fig. 90.2). Frequently, the left-sided tricuspid valve has attachments to the posterior edge of the VSD and, therefore, the sutures need to be placed on the morphologic left ventricular side of the VSD.

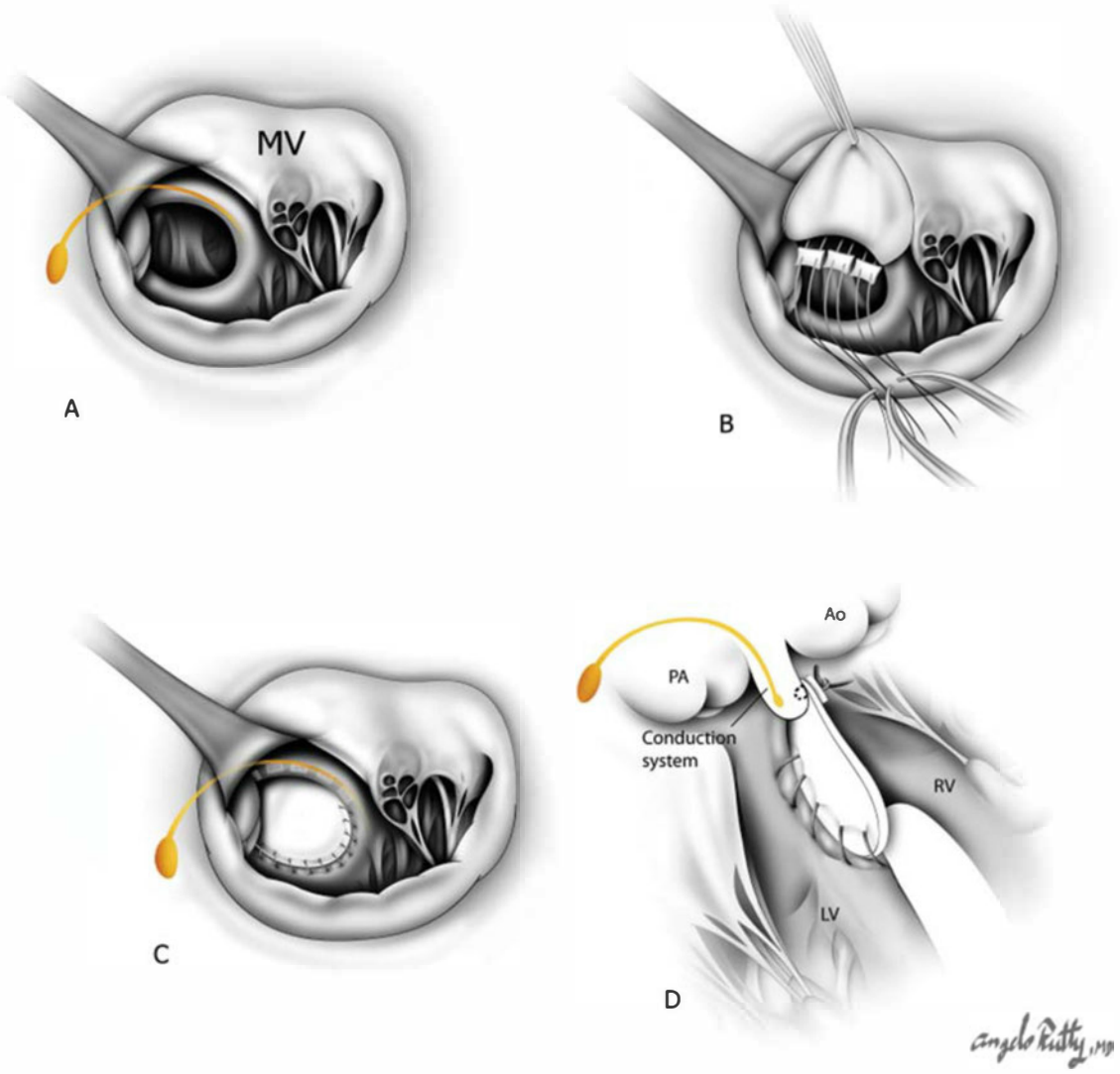


Fig. 90.2. Ventricular septal defect (VSD) closure via a right atriotomy. (**A**) The VSD is visualized through the right-sided mitral valve. The conduction tissue (*yellow*) is anterior and cephalad to the pulmonary valve and descends along the anterior margin of the VSD. (**B–D**) The VSD is closed with a prosthetic patch. Note that the sutures along the superior and anterior VSD border are placed on the morphologic right ventricular side of the septum to avoid the conduction tissue. Ao, aorta; LV, left ventricle; MV, mitral valve; PA, pulmonary artery; RV, right ventricle.

The conduction system in patients with IDD type of L-TGA runs in the posterior edge of the defect. Therefore, VSD closure would require placement of all sutures along the pulmonary (left side) edge of the defect in order to avoid surgically induced complete heart block

Also, a VSD could be closed through the aortic valve if the aortic root is large. Sutures are placed on the left (systemic) side of the VSD. Extra care must be taken not to interfere with the chordae or the tricuspid valve leaflets, which may cause AV valve insufficiency. In patients with L-TGA, VSD closure is associated with a significant risk of complete heart block, either from suture damage to the conducting bundle or from retraction on the anteriorly placed AV node and bundle.

Repair of Left Ventricular Outflow Obstruction (Subpulmonary Obstruction)

Thirty to 50% of patients with L-TGA present with some form of pulmonary or subpulmonary obstruction. Direct relief of the LVOTO is usually not satisfactory because of the posterior position of the narrowed pulmonary outflow tract, which is wedged between the two AV valves. Accessory mitral valve chordal attachments to the subpulmonic area commonly contribute to the obstruction. Isolated valvular stenosis or discrete subvalvar membranes can be relieved by valvotomy and/or membrane resection. Another complicating factor is the presence of the abnormal conduction system in close proximity to the pulmonary valve in this patient population.

During a "physiologic repair," the surgical management of LVOTO commonly involves the placement of an extra-anatomic conduit between the morphologic left ventricle and the pulmonary arteries (Fig. 90.3). The conduit takeoff from the LV is in a more apical position due to the location of the papillary muscles and coronary arteries. These conduits commonly sit retrosternally, an important fact to remember at the time of reoperation. Valved conduits are most

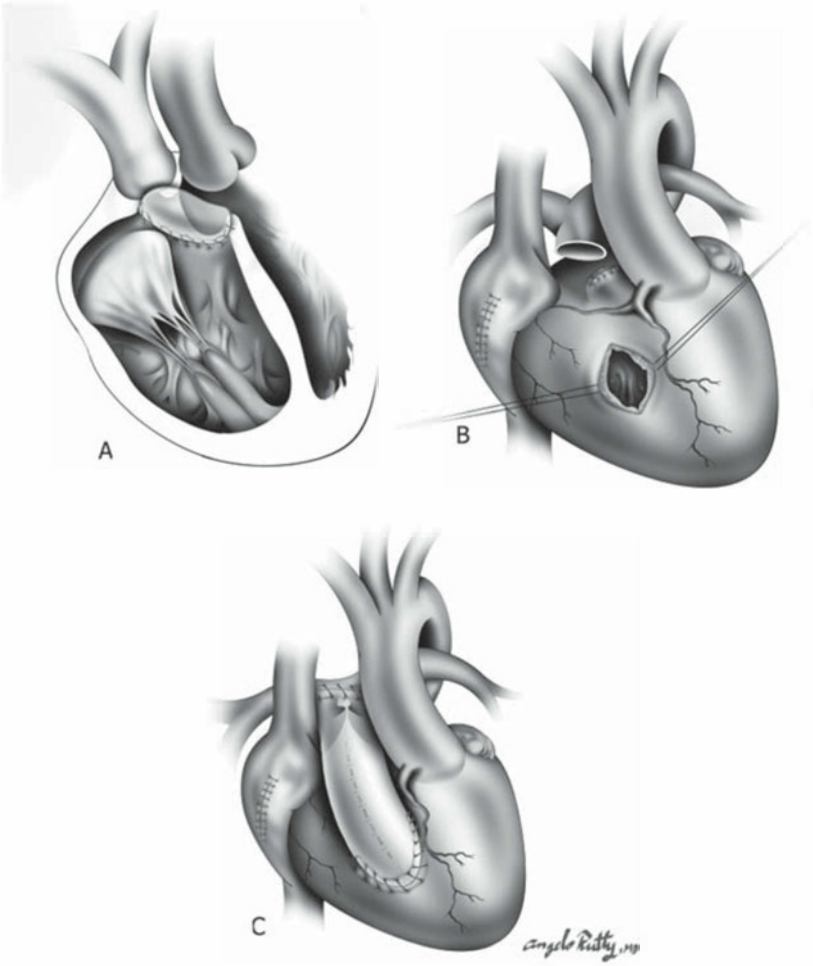


Fig. 90.3. "Classic repair" in congenitally corrected transposition of the great arteries (ccTGA) with ventricular septal defect (VSD) and pulmonary stenosis (PS). (**A**) The VSD is closed leaving the right ventricle (RV) as the systemic ventricle. (**B**) A left ventriculotomy is performed avoiding injury to the coronary arteries and the left ventricle (LV) papillary muscles. (**C**) An LV to pulmonary artery (PA) conduit is placed.

frequently used but a nonvalved synthetic tube can also be used when the pulmonary resistance is low and the distal pulmonary tree is adequate in size.

SURGICAL MANAGEMENT OF TRICUSPID VALVE INSUFFICIENCY

The tricuspid valve could be normal, but frequently there is Ebsteinoid-like displacement of the septal leaflet down into the body of the RV without atrialization of the RV free wall. Also, dilatation of the RV and tricuspid annulus contributes to the development of regurgitation, even in morphologically normal valves. Surgical exposure of the tricuspid valve is often challenging because of the abnormal location of the valve within the ventricle often requiring a trans-septal approach. Exposure of the valve via a left atriotomy through a left thoracotomy incision has also been advocated. Although we have successfully repaired a few of these valves, the more "standard" management remains valve replacement.

ANATOMIC REPAIR

Anatomic repairs reestablish the LV as the systemic ventricle. To achieve a complete anatomic repair, the systemic and pulmonary venous returns also need to be redirected or switch, thus the term "double-switch operation." Three surgical options exist as follows:

1. Rastelli procedure + atrial switch (most common).
2. Arterial switch procedure + atrial switch.
3. Aortic translocation procedure + atrial switch.

It is important to realize that these procedures are technically demanding and are usually associated with a prolonged cross-clamp time. We perform "double-switch" operations under standard bicaval cardiopulmonary bypass with moderate hypothermia and cold blood cardioplegia.

The Rastelli procedure with an atrial switch can be performed in children with significant LVOTO or pulmonary atresia. Kawata et al. described this approach which aims to divert the morphologic left ventricle outflow through the VSD to the aorta in combination with an atrial level switch procedure. After an infundibular incision, the interventricular tunnel is created with a prosthetic patch, funneling the left ventricular blood into the aorta. A conduit is used to establish right ventricle to pulmonary artery continuity (Fig. 90.4). Another alternative is the REV (*Réparation à l'Étage Ventriculaire*) procedure, which involves the enlargement of the VSD to create a more direct communication between the left ventricle and the aorta. A direct anastomosis between the right ventricle and the pulmonary artery is created.

The concept of combining the *arterial and atrial switch* procedures for the management cc-TGA was introduced by Ilbawi in 1990. Currently, it is most frequently utilized in the presence of an unrestrictive VSD without significant pulmonary or subpulmonary obstruction. Patients with mild-to-moderate subpulmonary obstruction secondary to resectable accessory AV valve tissue are also candidates. A significant number of these patients might have undergone pulmonary artery banding to manage pulmonary overcirculation or for LV retraining. Also, this approach should be considered in children with important tricuspid insufficiency and/or RV dysfunction.

The technical aspects of the arterial switch in ccTGA are quite similar to the "usual" d-TGA technique (Fig. 90.5). Both great vessels are transected and the coronary buttons harvested from the native aorta. After the LeCompte maneuver, the distal aorta is anastomosed to the neoaortic root and the coronary arteries are reimplanted. The neopulmonary root is reconstructed with native pericardium and the pulmonary artery anastomosis is performed. VSD closure is performed through the right atrium or via the transected "native" aorta .

The aortic translocation procedure in combination with an atrial switch, first described in 2005 by Morell and colleagues, is applicable to patients with LVOTO and a VSD. This technique consists of harvesting the aortic root from the right ventricle,

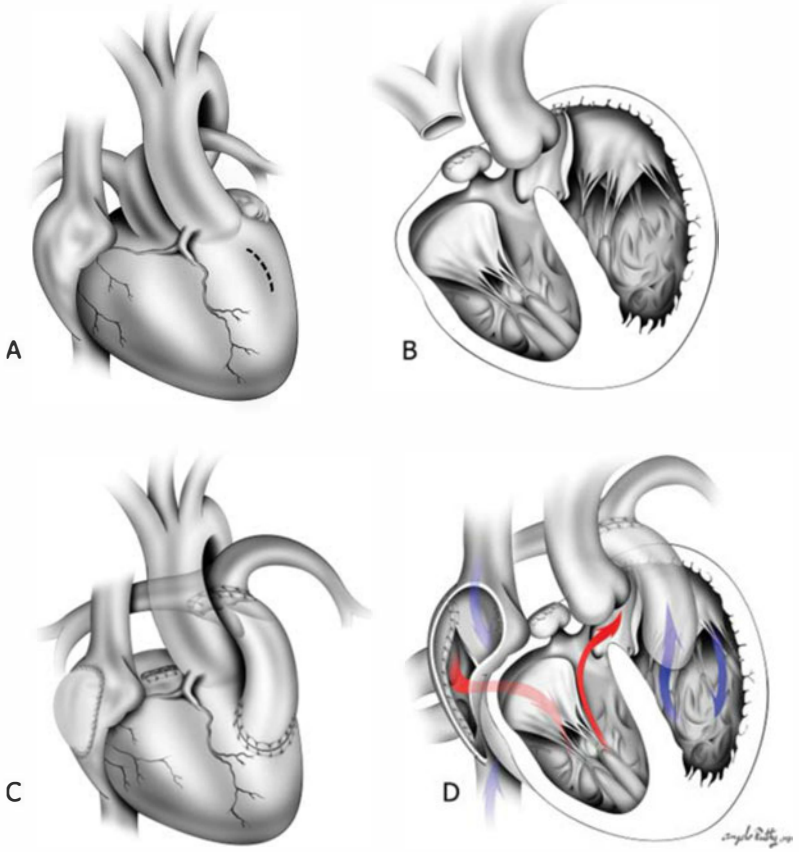


Fig. 90.4. Rastelli procedure in congenitally corrected transposition of the great arteries (ccTGA). (**A** and **B**) Via a distal right ventriculotomy, an intraventricular tunnel is created with a prosthetic patch, leaving the left ventricle (LV) as the systemic ventricle. (**C** and **D**) A right ventricle (RV) to pulmonary artery (PA) conduit is then placed and an atrial switch procedure is performed. (*Blue arrow* means venous blood; *red arrow* means Arterial blood.)

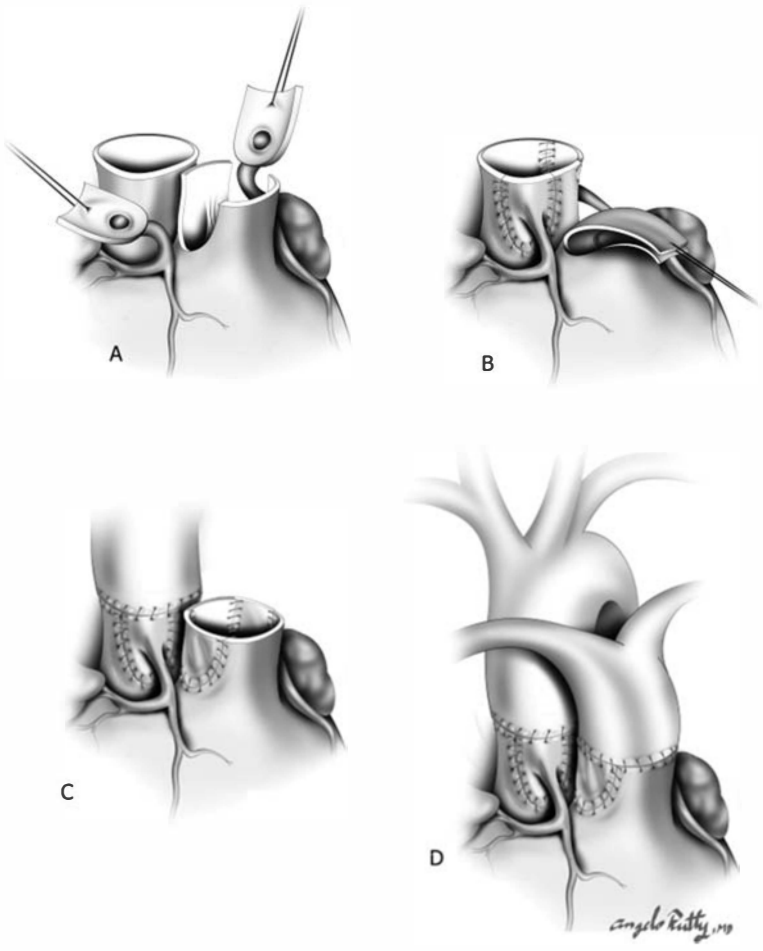


Fig. 90.5. Arterial switch procedure in congenitally corrected transposition of the great arteries (ccTGA). (**A**) The ascending aorta and main pulmonary artery are transected and the coronary buttons are harvested. The proximal coronary arteries are extensively mobilized to achieve tension-free coronary artery reimplantation. (**B**) The coronary buttons are reimplanted to the neoaorta. (**C**) The neopulmonary artery is reconstructed with autologous pericardium. (**D**) The pulmonary artery anastomosis is performed; note that the LeCompte maneuver was utilized.

usually removing one (left main coronary button most commonly) or both coronary buttons from the aorta because of the side-by-side relationship of the great vessels. Then, the main pulmonary artery is transected proximally and the LVOTO is relieved by dividing the outlet septum and the hypoplastic pulmonary valve annulus. The aortic root is translocated and sutured to the pulmonary annulus, positioning the aorta closer to the LV, the VSD is then closed. The coronary buttons are reimplanted and the RVOT is reconstructed with or without a conduit (Fig. 90.6). This technique creates better alignment of the right and left ventricular outflow tracts and is especially useful for patients with an inlet VSD, hypoplastic RV, or straddling AV valves.

There are several options available for the "atrial switch" component of the operation. We have utilized three techniques, namely, (1) the Mustard, (2) the Senning, and (3) the hemi-Mustard/Bidirectional Glenn. The Senning procedure, with pericardial patch augmentation of the pulmonary venous baffle, is our preferred option (Fig. 90.7). In the presence of dextrocardia, which is associated with a smaller right atrial mass, we tend to favor the Mustard or hemi-Mustard.

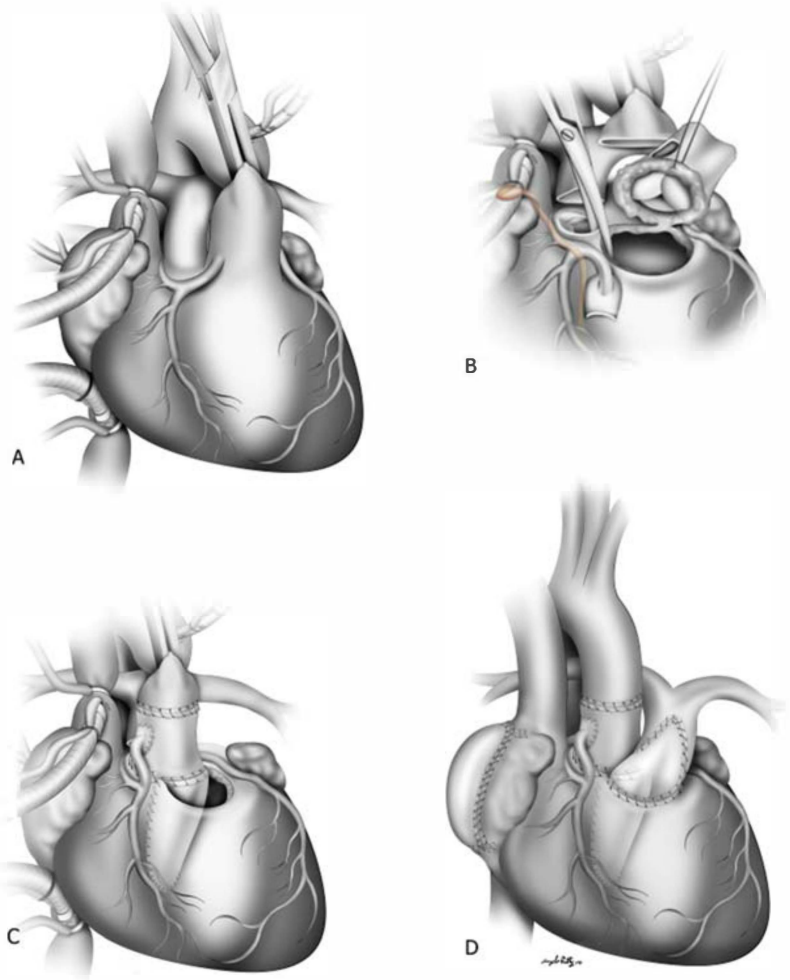


Fig. 90.6. Aortic translocation in congenitally corrected transposition of the great arteries (ccTGA). (**A** and **B**) The aortic root is harvested from the right ventricle (RV), removing the left main coronary button. Then the proximal pulmonary artery is transected and the outlet septum divided. (**C**) The aortic root is translocated and the coronary button reimplanted. The left ventricular outflow tract (LVOT) is reconstructed with the ventricular septal defect (VSD) patch. (**D**) A direct RV to pulmonary artery (PA) connection is performed with an anterior pericardial patch to enlarge the hypoplastic main PA. An atrial switch procedure is finally performed.

RESULTS

Physiologic (Classic) Repair

The long-term outcomes of patients with ccTGA who have undergone a physiologic surgical repair are unsatisfactory. The published reports reveal 20-year-survival and surgical reintervention rates that approximate 50%. As expected, systemic right ventricular dysfunction and/or tricuspid valve insufficiency are predictors of poor outcomes. In these patients, the incidence of right ventricular dysfunction starts to become noticeable during their second decade of life; by 45 years of age, 32% to 56% of patients have developed ventricular dysfunction.

Systemic AV valve (tricuspid) regurgitation tends to be universally present in patients with systemic RV dysfunction and congestive heart failure. Do these patients develop TR because of systemic ventricular dysfunction, or does the TR hasten the development of it? To date, there is no clear answer. Hraska and associates found that in patients with ccTGA undergoing "classic repairs," the poorest outcome was seen in those undergoing tricuspid valve replacement. In this study, Ebsteinoid malformation of the TV was not only a risk factor for late development of TR but was also an independent preoperative predictor of mortality. These findings are consistent with other published reports.

Anatomic Repair

The poor outcomes associated with the "classic repair" have led us to explore anatomic repairs. It is fair to say that "double-switch" procedure is considered the preferred surgical therapy for the management of ccTGA, especially in the presence of tricuspid valve anomalies or insufficiency and right ventricular dysfunction. In 1997, Karl et al. showed that the initial risk of the double-switch repair was similar to that of classical repairs. Duncan et al. showed that the double-switch operation could be performed with excellent hospital and long-term survival.

In 2009 Ly et al. achieved successful double-switch operation in 20 patients with ccTGA, 17 of whom had pulmonary artery banding procedure. The mean time elapsing from first banding to final repair was 14 months. Actuarial survival at 10 years was 100%, and the freedom from reoperation at 5 and 10 years was 93% and 77%, respectively.

In 2012, Hiramatsu and associates published the largest surgical series on the double-switch operation in 90 patients (72 "Rastelli/REV" and 18 arterial switches). The overall in-hospital mortality was 10%, with a 20-year-survival rate of approximately 80%. The freedom from reoperation was 77.6% in the Rastelli/REV group versus 94.1% in the arterial switch group. The freedom from arrhythmia was statistically higher in the arterial switch group (78.6%

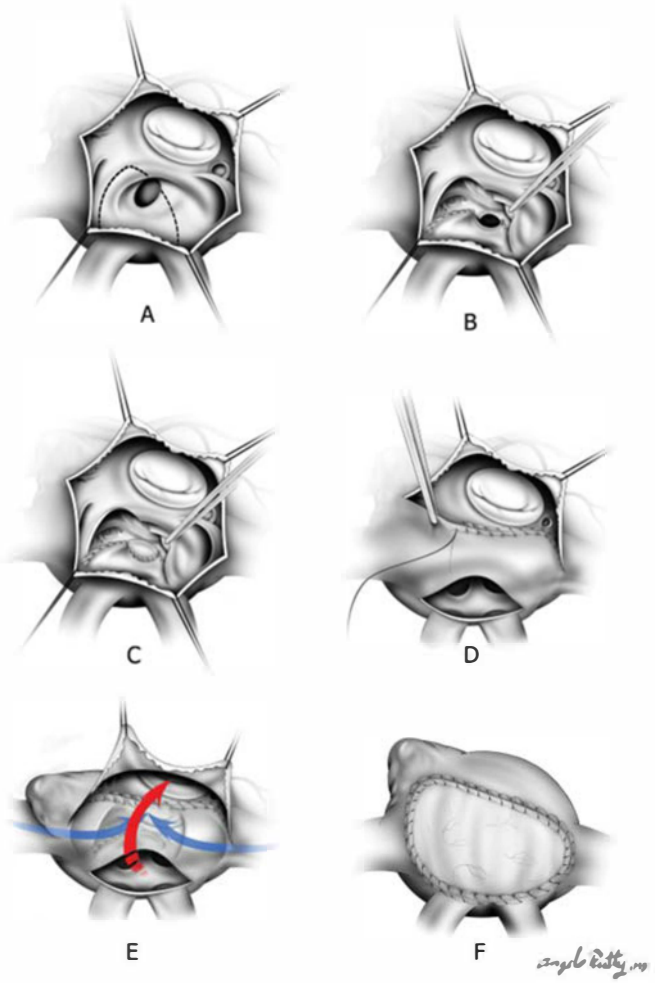


Fig. 90.7. Senning procedure. (**A**) A right atriotomy is carried out parallel to the atrioventricular groove and the interatrial septum is exposed. The septum primum is widely resected to facilitate baffling of the systemic venous return to the posterior tricuspid valve. (**B**) A laterally based flap of atrial septum is created and moved posteriorly, over the orifices of the pulmonary veins, becoming the pulmonary venous baffle. (**C**) Frequently, a segment of pericardium is needed to augment the flap. (**D**) A separate incision is made in the left atrium, just anterior to the right-sided pulmonary veins. The systemic venous baffle is created by suturing the posterior edge of the right atrial incision to the medial rim of the atrial septum. (**E** and **F**) The pulmonary venous channel is completed with a pericardial patch. (*Blue arrow* means venous blood; *red arrow* means Arterial blood.)

vs. 57.1%). The ratio of NYHA class I to II at outpatient clinic was exactly the same in both groups (86%).

The overall experience with the aortic translocation/atrial switch procedure is limited; the hospital mortality appears to be similar to the other surgical alternatives, but there are truly no long-term data available. Aortic translocation should be considered an alternative approach for patients with intracardiac anatomy not suitable for the Rastelli.

Fontan Procedure

Shin'oka et al. reported on 38 patients with ccTGA that were managed with a Fontan procedure. The actuarial survival at 22 years was 79%, which was no different than patients managed with anatomic repairs. Furthermore, there were fewer reoperations in patients who underwent a Fontan procedure than those undergoing a biventricular repair. The Fontan procedure is certainly an acceptable option for patients with unfavorable anatomy for a biventricular repair.

Left Ventricular Training

The success of the double-switch procedure depends on the ability of the left ventricle to support the systemic circulation. In patients with ccTGA, this usually requires the placement of a pulmonary artery band in order to retrain the LV, by inducing left ventricular hypertrophy. Left ventricular retraining frequently requires a minimum period of 6 to 12 months. Bove et al. noted that older patients are less likely to achieve satisfactory left ventricular retraining. Brawn and associates do not recommend retraining for patients older than 15 years of age. However, the upper age limit is not yet defined. Pulmonary artery banding has been identified as a risk factor for the development of pulmonary root dilatation and neoaortic valve regurgitation.

CONCLUSION

Patients with ccTGA represent a rare but complex group for whom the surgical correction remains a challenge. "Overall" patient outcomes appear to be improved with surgical management that reestablishes a left ventricular-dependent systemic circulation.

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EDITOR'S COMMENTS

ccTGA, or {S,L,L} transposition, continues to present a challenge for surgical intervention. The tenuous nature of the conducting tissue in this condition has resulted in a significant incidence of late complete AV block with or without surgical intervention. Despite attention to the surgical details of VSD closure, AV block continues to be a late problem requiring pacemaker insertion in up to one-third of cases. In addition, even with direct intracardiac repair, patients are left with a systemic morphologic right ventricle with often an abnormal AV valve and, in addition, frequently a conduit from the morphologic left ventricle to the pulmonary arteries that will require replacement. Thus, the incidence of late reoperation is high and the problem of late systemic ventricular dysfunction significant. Valve reparative procedures on the left side of the heart to the abnormal anatomic tricuspid valve have not been particularly successful, and valve replacement has been associated with late ventricular function deterioration and the complications of prosthetic heart valve insertion. Even conversion to the single-ventricle type of morphology in patients with significant pulmonary stenosis has not been associated with improved long-term results because left AV valve regurgitation may compromise ventricular performance after conversion to single-ventricle physiology. The suboptimal late results have led some authors to suggest the use of a double-switch procedure for selected patients with {S,L,L} anatomy. As described in this chapter, the double-switch procedure creates a Senning or Mustard baffle at the atrial level associated with closure of the VSD to the pulmonary artery and an arterial switch procedure at the level of the great vessels. In this case, the anatomic left ventricle becomes the systemic ventricle, and ventricular inflow is switched at the atrial level. The abnormal tricuspid valve is then associated with the pulmonary ventricle, which is presumably at a lower pressure, and therefore the regurgitation is better tolerated. The use of this technique has been increasing, and the follow-up results for these patients suggest improved outcomes compared with alternative "physiologic" repairs. As nicely summarized by the authors of this chapter, there are now several series with significant numbers of patients being reported with intermediate short-term follow-ups of up to 5 years with a low incidence of atrial arrhythmias but with the need for reoperation in a significant number of the patients. Overall mortality, however, has been quite low. The known

(*continued*)

EDITOR'S COMMENTS (*continued*)

problems of atrial arrhythmias from atrial baffles and the need for conduit revision with these repairs may very well limit the long-term durability of the operation even though ventricular function may be preserved. The operation may also be used for patients with right ventricular dysfunction and tricuspid regurgitation who present later in life either without prior palliative procedures or with prior palliative shunts or bands. Some patients who have had late ventricular deterioration after complete intracardiac repairs despite good anatomic results have had sufficient cardiac dysfunction to require cardiac transplantation for corrected transposition.

The authors of this chapter have described the use of pulmonary artery banding in patients with congenitally corrected transposition of the great arteries to prepare the left ventricle for a double-switch operation. This approach has been used in patients with tricuspid regurgitation to shift the ventricular septum to the right ventricular side, which can significantly decrease the amount of tricuspid regurgitation and to even preserve ventricular function. In patients with corrected transposition and unrestricted pulmonary blood flow, pulmonary banding early in life may be a long-term palliative approach. The technical aspects of pulmonary artery banding are not complex; but in this patient population, the procedure can be associated with extreme ventricular dysfunction with even minor changes in band tightness. Pulmonary banding in this patient population becomes one of the more challenging operations in the armamentarium of the congenital heart surgeon. Generally, pulmonary banding has to be done with continuous monitoring by transesophageal echocardiography to watch the shift of the septum. Even when the band appears to be in perfect position, the ventricles can rapidly deteriorate in function resulting in hypotension and cyanosis. Generally, the band has to be tightened and then released somewhat to allow the ventricle to slowly adapt to increasing pressure. These patients may be extremely ill in the postoperative Cardiac Intensive Care Unit requiring inotropic support and ventilation and sedation for several days to adjust to the acute afterload on the ventricle. Some of these patients may benefit from remotely adjustable pulmonary arterial bands, which can be gradually tightened using remote devices. The use of these adjustable bands may allow the ventricle to more gradually adapt to the increasing pressure. The primary difficulty in banding the pulmonary artery and preparing the left ventricle for the double-switch procedure as noted by the authors is the fact that there is no clear cut-off of age range when this is likely to be successful. The reported series of ventricular retraining and subsequent switch procedures have been associated with a significant operative mortality and morbidity, but in some cases can result in a good anatomic repair and preclude the need for cardiac transplantation.

TLS

91 Pulmonary Stenosis and Pulmonary Atresia with Intact Septum

Deborah J. Kozik and Mark D. Plunkett

PULMONARY STENOSIS

Pulmonary stenosis at the valvar level has been reported to account for 8% to 10% of all congenital heart defects. Although critical pulmonary stenosis may present in the newborn period and require early intervention, most of these lesions are less severe and present after the neonatal period. The pulmonary valve is usually stenotic and dome shaped with fused leaflet tissue and a small central orifice. The right ventricle is usually normal in size, but secondary hypertrophy of the ventricle and infundibulum may occur. The etiology is not known and is probably multifactorial. There is a reported increased incidence of 2% to 4% in siblings of patients with this defect. Treatment of this congenital defect with either balloon valvotomy or surgical valvotomy is associated with low morbidity and mortality and excellent long-term outcomes.

Diagnostic Considerations

Infants with critical pulmonary stenosis may present in the newborn period with severe cyanosis and heart failure. The clinical manifestations are relative to the severity of the stenosis and the patency of a foramen ovale or an atrial septal defect (ASD). In most children with pulmonary valve stenosis and intact ventricular septum, symptoms develop more slowly. Most patients are initially identified by a harsh systolic ejection murmur and a thrill over the pulmonic region on physical examination. An electrocardiogram usually reveals right-axis deviation, prominent P waves, and right ventricular (RV) hypertrophy. A chest radiograph may show prominent pulmonary artery shadows secondary to poststenotic dilation. The heart shadow is normal except in severe cases with congestive failure. Subsequent studies should include echocardiography to establish the severity of the lesion and identify associated abnormalities. Doppler evaluation allows estimation of the gradient across the valve and RV outflow tract. Finally, cardiac catheterization may be performed for additional diagnostic information, hemodynamic data, and for possible therapeutic intervention with balloon valvotomy.

Surgical Treatment and Techniques

Patients with pulmonary valve stenosis and intact ventricular septum require intervention for symptomatic lesions and for significant transvalvular pressure gradients. Historically, surgery was the mainstay of therapy for isolated pulmonary valvar stenosis. Currently, however, catheterization with balloon valvotomy has replaced surgery as the cornerstone of initial treatment. In cases of recurrent stenosis, repeat balloon valvotomy may be attempted before surgical therapy. The incidence of pulmonary insufficiency after balloon valvotomy is high (80%), but it is often clinically mild and well tolerated in most patients. Failed balloon valvotomy can be an indication for urgent surgical intervention. Surgical pulmonary valvotomy may be performed as an open technique using cardiopulmonary bypass or more rarely through a closed transventricular approach. Operative mortality is minimal except in cases of critical stenosis with associated RV hypoplasia and congestive heart failure. The results of treatment are directly related to the size of the RV chamber and the age of the patient at presentation.

Open Pulmonary Valvotomy Using Cardiopulmonary Bypass

Exposure for open pulmonary valvotomy is obtained through a median sternotomy. After heparinization, an aortic cannula is placed in the ascending aorta, and bicaval cannulation is performed. Snares are placed around the inferior and superior venae cavae. A catheter is placed in the ascending aorta for the antegrade delivery of cold blood cardioplegia. A patent ductus arteriosus must be ligated or snared at the initiation of cardiopulmonary bypass. An aortic cross-clamp is applied, and cardioplegia is delivered antegrade to achieve myocardial arrest. The snares are secured around the venous cannulae. The procedure may also be performed without a cross-clamp and cardioplegic arrest if there is no atrial or ventricular communication present.

A longitudinal pulmonary arteriotomy is performed above the level of the valve commissures. The stenotic valve is inspected, and the fused commissures are identified and carefully incised with a no. 11 scalpel blade. The incisions should extend to the annulus (Fig. 91.1A, and 91.1B). Any valvar adhesions to the pulmonary arterial wall are sharply incised. A partial valvectomy may be performed to remove thickened valve tissue or dense scarring on the leaflets. Dysplastic portions of the valve may require excision. The infundibulum is then inspected through the valve for any residual subvalvular stenosis. Sharp infundibular resection through the valve should be performed if necessary. Rarely, a ventriculotomy is required with an infundibular or transannular patch closure. If a small annulus is present, a Hegar dilator may be used to size the valve annulus. The need for a transannular patch can be determined based on this measurement. If there is supravalvular hypoplasia, a pericardial patch is used to close the incision in the pulmonary artery from the annulus to the base of the left pulmonary artery. The arteriotomy is otherwise closed with a running polypropylene (prolene) suture.

If there is evidence of an ASD or patent foramen ovale, a right atriotomy is performed and the atrial septum is inspected. Closure of a patent foramen ovale or ASD is performed using primary closure or pericardial patch closure with a running prolene suture technique. The tricuspid valve (TV) leaflets are retracted to expose the infundibular outflow tract. Infundibular resection may be performed through the TV to relieve any remaining stenosis. The TV is tested for competence before closure. The atriotomy is then closed

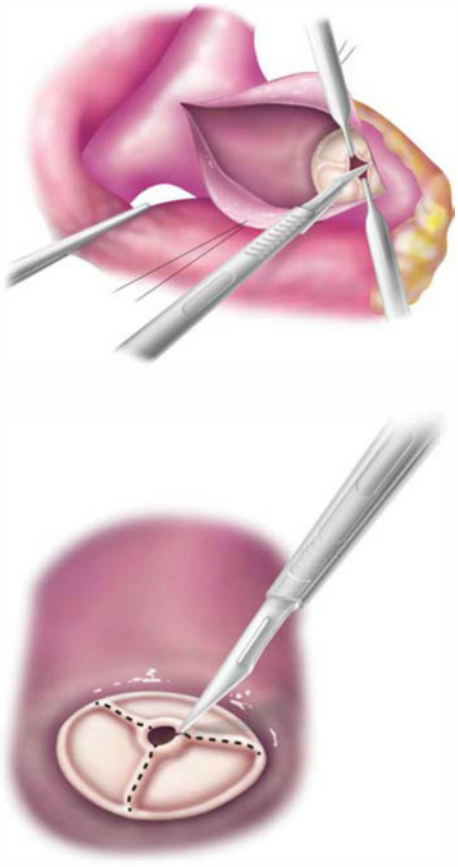


Fig. 91.1. (**A**) Open pulmonary valvotomy with division of the fused valve leaflets with a no. 11 blade scalpel or fine scissors. (**B**) The incision in the valve leaflet tissue should extend to the annulus.

in a two-layer manner with a running prolene suture.

The patient is weaned from cardiopulmonary bypass. If the right ventricle is hypoplastic, a patent foramen ovale is left partially open. Measurements of pressure in the right ventricle and the main pulmonary artery will document any residual gradient and should be performed before decannulation. Intraoperative transesophageal echocardiography is used to assess any residual gradient or pulmonary valve insufficiency. There is often a small residual gradient across the valve postoperatively, which may regress with time.

Off-Pump Transventricular Pulmonary Valvotomy

If a restrictive patent foramen ovale is present or the atrial septum is intact, pulmonary valvotomy may be performed using a closed technique that avoids the use of cardiopulmonary bypass. A wet pump should be available on standby for possible use if necessary. A median sternotomy is used, and a purse string of 4-0 prolene suture is placed on the anterior aspect of the right ventricle just below the infundibulum. A 14-gauge angiocatheter with a pressure transducer is first introduced through the right ventricle into the pulmonary artery. Hegar dilators of progressively larger sizes (up to 7 or 8 mm) are then introduced across the valve. If the membrane does not dilate easily, a long vascular clamp can be passed through the purse string and right ventricle to disrupt the valve membrane. We now prefer the use of a balloon dilation catheter with a balloon, 1 mm larger in diameter than the annulus of the pulmonary artery. The balloon can be positioned by measurement and by palpation. A needle pressure transducer is then used to assess the remaining gradient across the valve. After adequate dilation is achieved, the purse string is tied and an additional reinforcing suture is placed in the ventricular epicardium.

Perioperative Management

Most patients with pulmonary stenosis are operated on electively with routine preoperative and postoperative care. Neonates with critical stenosis should be stabilized in the intensive care unit and operated on as soon as possible. Acidosis, electrolyte abnormalities, and congestive heart failure are corrected preoperatively.

These patients may also have stenosis in the infundibular region secondary to RV myocardial hypertrophy. Inotropes must be used with caution in the preoperative and postoperative periods because increased contractility may cause increased dynamic obstruction across the pulmonary outflow tract and further compromise pulmonary blood flow.

Postoperative Care and Surgical Complications

Postoperative care for patients after surgical pulmonary valvotomy should focus on adequate RV filling pressures and reduced pulmonary artery pressures. Pulmonary vasodilators may be used in the early postoperative period to increase pulmonary flow and reduce RV afterload. Inotropic support may be useful for the first few days.

Transthoracic monitoring catheters may be placed in the right atrium or through the right ventricle into the pulmonary artery to continuously monitor hemodynamic data postoperatively. These are generally discontinued within the first 48 hours.

Pulmonary insufficiency may result from either open or closed valvotomy. Most patients will tolerate this residual valve incompetence with little difficulty. RV dysfunction may be present in the early postoperative period. This is usually transient, but may require moderate inotropic support during the early postoperative recovery.

Infants with critical pulmonary stenosis and an adequate-sized pulmonary valve annulus are often treated with balloon valvuloplasty rather than surgical intervention. However, patients with severely dysplastic pulmonary valves or significant RV outflow tract obstruction with dynamic infundibular obstruction may still come to surgical intervention and require open valvotomy or a transannular patch. If the obstruction is adequately relieved, any residual infundibular hypertrophy will regress over time. Most patients recover without complications, and long-term outcomes are excellent for both balloon valvotomy and surgical valvotomy.

PULMONARY ATRESIA WITH INTACT SEPTUM

Pulmonary atresia with intact septum (PA/IVS) is a rare defect that represents between 1% and 3% of all congenital heart defects. The exact cause or event that leads to pulmonary atresia is unknown. The lesion is sporadic, and no significant familial pattern has been identified. By definition, there is no communication between the right ventricle and the pulmonary arteries. Consequently, a patent ductus arteriosus is essential for early survival with this defect. The confluence of the branch pulmonary arteries with the main pulmonary artery is usually normal and the main pulmonary artery then tapers to an atretic pulmonary valve. The atresia of the pulmonary valve may vary significantly from a thick muscular obstruction without significant valve tissue to a thin plate-like fused trileaflet valve with well-developed sinuses and commissures. There are usually varying degrees of hypoplasia of the right ventricle and the TV, and RV-to-coronary artery fistulas may be present. Morphologically and functionally, the hypoplasia of the TV usually varies directly with the hypoplasia of the right ventricle. The TV is almost always abnormal with dysplastic leaflets, shortened chordae, and restricted leaflet motion. An Ebstein-like malformation may be present in up to 10% of patients and is usually associated with severe tricuspid

regurgitation. Coronary artery fistulas are present in 45% of cases and are more common in patients with severely hypoplastic right ventricles and small competent TVs. Of note, aortopulmonary collaterals are rarely found in patients with this defect. Without early surgical intervention, children with PA/IVS have an extremely high mortality rate. The natural history of this defect is associated with 50% mortality at 2 weeks and 85% mortality at 6 months.

Diagnostic Considerations

Diagnosis of this lesion is usually made in neonates and is prompted by the presence of hypoxia in varying degrees in the newborn period. Physical examination findings are often remarkable for prominent cervical venous pulsations and hepatic enlargement. A significant murmur is indicative of tricuspid regurgitation or may be related to the patent ductus arteriosus. An electrocardiogram shows progressive evidence of right atrial enlargement with peaked right atrial P waves. A chest radiograph is unremarkable at birth but may later reveal an increased cardiac shadow secondary to right atrial and left ventricular enlargement. The diagnosis of pulmonary atresia with intact ventricular septum is usually made by two-dimensional echocardiography. Ventricular cavity sizes, valve dimensions and function, and the nature of the pulmonary artery obstruction can be determined by echocardiography. Cardiac catheterization is used for a definitive diagnosis and further evaluation. Information determined during right and left heart catheterization should include the size and competency of the TV, the functional status of the ventricles, the degree of RV hypoplasia, the degree of infundibular hypoplasia, the presence or absence of coronary sinusoids and their communications with the anatomy of the coronary arteries, and the size of the pulmonary arteries. In addition to an injection of contrast medium into the right ventricle, selective coronary angiograms are required to assess the native coronary anatomy, particularly in the severely hypoplastic RV group. Newborns with hypoxemia and poor perfusion in spite of medical management should be assessed for the presence of a restrictive ASD, which can be enlarged by balloon septostomy at the time of catheterization. If the ductus arteriosus is closed or restrictive, prostaglandin E_1 (PGE_1) is infused to open the ductus. In the current era, a role is developing for stenting of the ductus arteriosus at the time of catheterization as an initial palliative intervention.

Anatomic Considerations

Our initial management of patients with PA/IVS is based largely on an anatomic classification, which specifically defines the degree of RV hypoplasia and the TV annular size measured as a *Z*-score. This classification not only allows initial management strategies but also has predictive value for the possible use of the right ventricle in subsequent definitive biventricular repair. The *Z*-score is determined by comparing the diameter of the TV (as measured by echocardiography) to the "expected" size and calculating the difference in standard deviations. This allows for a quantitative assessment of the degree of hypoplasia of the TV, which in most patients varies directly with the degree of hypoplasia of the RV. We and others have found both the degree of RV hypoplasia and the TV *Z*-score correlate with short- and long-term outcomes of surgical management of PA/IVS.

In this classification, neonates with PA/IVS are initially separated into three groups of mild, moderate, and severe RV hypoplasia. In patients with mild RV hypoplasia, the TV and RV cavity are approximately two-thirds or greater of calculated normal size and the RV outflow tract is well developed. This usually correlates with a *Z*-score for the TV of 0 to −2. In patients with moderate RV hypoplasia, the TV and the RV cavity are approximately one-half of calculated normal size (with a range of one-third to two-thirds normal), and the outflow tract is developed enough to allow an effective pulmonary valvotomy. This usually correlates with a *Z*-score for the TV of −2 to −4. In patients with severe RV hypoplasia, the TV and RV cavity are one-third or less of calculated normal size and the outflow tract is severely hypoplastic or obliterated and is often not amenable to an effective pulmonary valvotomy. This usually correlates with a *Z*-score for the TV of −4 to −6. Our approach is not based on a single anatomic finding but instead assesses the overall RV morphology and the degree of both TV and RV hypoplasia.

The TV is often anatomically and functionally abnormal in patients with PA/IVS. Therefore, a TV *Z*-score or valve diameter alone may not adequately predict the likelihood of achieving a biventricular repair. Similarly, some patients with only moderate RV hypoplasia may have a severe tricuspid malformation and ultimately undergo a single-ventricle approach with a Glenn or Fontan procedure. In addition, the compliance of the abnormal right ventricles with significant ventricular hypertrophy and small cavity size may limit the possibility of a complete two-ventricle repair even in those with only moderate hypoplasia.

A fourth subgroup of patients may present with marked cardiomegaly caused by right atrial enlargement, severe tricuspid regurgitation, and an Ebstein's anomaly of the TV. Dilation and dysfunction of the right atrium (RA), RV, and ventricular septal wall in these patients often compromise left ventricular function and lead to biventricular failure. These patients may have significant compromise of the left ventricle by the dilated right ventricle, and the inefficient flow of blood in and out of the right ventricle compromises overall systemic blood flow. The creation of an aortopulmonary shunt may stabilize pulmonary blood flow in these patients, but the effect of the dilated right ventricle on systemic cardiac output may remain. Patch closure of the TV is usually not suitable for these patients, because there will be no outflow of the coronary sinus return, and often sinusoidal connections and Thebesian vessels into the right ventricle cannot then be decompressed. Surgical intervention in these patients is associated with >50% mortality. In these patients, the best approach may be early cardiac transplantation.

During the initial evaluation of patients with PA/IVS, particular attention must be paid to the anatomy of the coronary circulation. Abnormalities of the coronary circulation are often found in the severely hypoplastic group and may dictate which surgical management option is indicated. During fetal development, RV hypertension may cause intramyocardial sinusoids to develop. These sinusoids often communicate by fistulas with the coronary artery circulation. The morphology of these sinusoids and their specific communications are extremely variable. Proximal coronary artery stenosis or obstruction may develop in a native coronary artery supplied by these intramyocardial sinusoids. If the distal coronary artery flow is dependent on these sinusoids for adequate myocardial perfusion, they are termed RV-dependent coronary circulations (RVDCCs). Decompression of the right ventricle in these patients is contraindicated as it may lead to acute myocardial ischemia and death. A limited experience has shown that a shunt from the aorta to the right ventricle may be beneficial in these patients to augment myocardial perfusion and coronary blood flow.

Initial Surgical Treatment and Techniques

Most infants with PA/IVS will require surgical management early in life to survive.

Treatment with intravenous PGE_1 will maintain pulmonary flow through the patent ductus arteriosus and allows time for evaluation and interventional decision making. Recently, some neonates have been successfully palliated with stenting of the ductus arteriosus at the time of catheterization. This has allowed delay of surgical intervention in those neonates who would otherwise be treated with a central shunt as an initial palliation. The efficacy of this catheter-based therapy appears promising for some neonates with PA/IVS, but the indications remain to be defined. Once the anatomy and morphology of these lesions have been identified at cardiac catheterization, classification is determined, and an appropriate operative strategy is planned. Delay in surgical treatment is often hazardous and may reduce survival. The primary goals of initial therapy should be to minimize early mortality and maximize the potential for a biventricular repair later in life. The choice of surgical approach is based on the anatomic classification previously described (Table 91.1).

Initial Procedures for Neonates with Mild Right Ventricular Hypoplasia

Neonates with mild RV hypoplasia (the right ventricle approximates two-thirds of normal or greater) and a tricuspid *Z*-score of 0 to −2 are best treated with a pulmonary valvotomy, ligation of the patent ductus arteriosus, and creation of a central systemic-to-pulmonary artery shunt. Occasionally, there are patients in which a pulmonary valvotomy alone will be adequate to restore pulmonary blood flow. These are rare and this surgical strategy should be performed with caution. Most of these patients will require a shunt as an additional source of pulmonary blood flow. Neonates with moderate RV hypoplasia and a tricuspid *Z*-score of −2 to −4 are treated with a pulmonary valvotomy, patch augmentation of the pulmonary outflow tract, insertion of a central shunt, and ligation of the patent ductus arteriosus. Pulmonary valvotomy with patch augmentation of the pulmonary outflow tract relieves RV hypertension, reduces tricuspid regurgitation, and promotes growth of both the RV cavity and TV. Neonates with severe RV hypoplasia and a tricuspid *Z*-score of −4 to −6 are usually treated with a shunt only. If there is no RVDCC and no pulmonary valvotomy is performed, then the TV can be made incompetent to decompress the RV using an open or closed tricuspid valvotomy technique. Catheter balloon disruption of the TV has also been described.

Off-Pump Transventricular Pulmonary Valvotomy

Closed pulmonary valvotomy may be performed on patients with mild-to-moderate RV hypoplasia who have an adequately developed RV outflow tract below the obstructed pulmonary valve. This should be performed with wet-pump standby in case there is hemodynamic instability during the procedure. The technique is performed as previously described. Many of these patients can now be effectively treated with perforation of the membrane using radiofrequency ablation catheters and subsequent balloon catheter dilation valvotomy. This approach may obviate the need for initial surgical intervention in this group of patients.

Table 91.1 Initial Surgical Management of Pulmonary Atresia with Intact Septum According to the Degree of Right Ventricular and Tricuspid Valve Hypoplasia

Classification of RV hypoplasia	RV and TV morphology	Initial treatment
Mild	Tricuspid valve *Z*-score 0 to −2 and RV cavity more than two-thirds of normal size, well-developed RV outflow tract	Pulmonary valvotomy with transannular patch and shunt or valvotomy only
Moderate	Tricuspid valve *Z*-score −2 to −4 and RV cavity one-third to two-thirds of normal size, moderately hypoplastic RV outflow tract	Pulmonary valvotomy with transannular patch and shunt
Severe[a]	Tricuspid valve *Z*-score −4 to −6 and RV cavity less than one-third of normal size, severely hypoplastic or absent RV outflow tract	Shunt only; possible open or closed tricuspid valvotomy

[a]Patients with Ebstein malformation of the TV may be considered for heart transplant.
RV, right ventricular; TV, tricuspid valve.

Aorta-to-Pulmonary Artery Shunt

A median sternotomy is used in performing a central shunt. The shunt insertion is performed by connecting either the innominate artery to the right pulmonary artery or the ascending aorta to the main pulmonary artery using a polytetrafluoroethylene (Gore-Tex) graft. A 3.0-mm shunt is used for neonates ≤3.0 kg, a 3.5-mm shunt in those >3.0 kg, and 4.0-mm shunt in those >4.0 kg. Cardiopulmonary bypass is usually not necessary but immediately available during the procedure. Partial occluding thin-bladed vascular C-clamps of appropriate sizes are applied to either the innominate artery or the aorta and to the pulmonary artery. Care must be taken to avoid occluding or compromising flow in the patent ductus. The arteriotomies are performed with a no. 11 scalpel blade and fine vascular scissors. The proximal anastomosis is performed on the anterior aspect of the aorta or the innominate artery in an end-to-side manner using a running 7-0 prolene suture technique. A similar anastomosis is performed to the anterior aspect of the main pulmonary artery or the right pulmonary artery. We prefer to leave the proximal clamp on during the distal anastomosis and avoid clamping the graft or allowing blood in it before flow is established. This reduces the risk of thrombus formation in the graft prior to release of the clamps. The graft is flushed with heparinized saline prior to completion of the distal anastomosis. The clamps are released slowly to deair the graft, and flow is established through the shunt. The ductus arteriosus is ligated after completion of the central shunt procedure.

Initial Procedures for Neonates with Moderate Right Ventricular Hypoplasia

Neonates with moderate RV hypoplasia are best treated with ligation of the patent ductus arteriosus, insertion of a central shunt, a pulmonary valvotomy, and patch augmentation of the pulmonary outflow tract. The transannular patch and pulmonary valvotomy may be performed with cardiopulmonary bypass and cardioplegic arrest. This allows a motionless bloodless field to perform an effective valvotomy, resect obstructing infundibular myocardium, and ensure an adequate patch augmentation. Incision of the RV outflow tract in pulmonary atresia must be performed carefully to avoid injury to the base of the aortic valve leaflet. Thus, the use of cardioplegia and cardiopulmonary bypass may allow more accurate placement of the valvotomy incision.

In select patients, we have also successfully used a closed off-pump technique to perform a pulmonary valvotomy and place

a transannular outflow tract patch of pericardium without the use of either cardioplegic arrest or cardiopulmonary bypass.

Off-Pump Pulmonary Valvotomy with Transannular Right Ventricular Outflow Tract Patch

In neonates with moderate RV hypoplasia, the infundibulum may be long and narrow but reaches the pulmonary valve membrane. In these patients, a pulmonary valvotomy and a pericardial transannular patch may be performed without using cardiopulmonary bypass.

A median sternotomy incision is used. Wet-pump bypass is available with the lines on the field in case bypass is necessary. A pediatric cross-clamp is placed immediately beneath the bifurcation of the pulmonary artery. The ductus is kept patent to provide pulmonary blood flow. A vertical incision is made in the main pulmonary artery down to the atretic pulmonary valve and RV junction. A partial-thickness myocardial incision is made over the right ventricle to bring the incision over the RV cavity. Part of the muscle is resected to a depth of 1 to 2 mm to thin out the right ventricle. An elliptical pericardial patch is now sutured to the pulmonary artery incision with a running 5-0 or 6-0 prolene suture down to the RV junction. The suture line is continued onto the myocardium to the edges of the RV incision, leaving the sutures loose inferiorly. A no. 12 blade scalpel is then used to incise the pulmonary valve membrane and to cut through the remaining myocardium into the RV cavity under the pericardial patch. The sutures are pulled up to control bleeding and the cross-clamp on the pulmonary artery is removed. If the RV pressure is not adequately reduced to between one-half and one-third of systemic pressure, a rhizotomy knife is introduced through a purse string in the pericardial patch, and the RV muscle is further incised until an adequate outflow tract has been created to adequately reduce the RV pressure (Fig. 91.2A–91.2C).

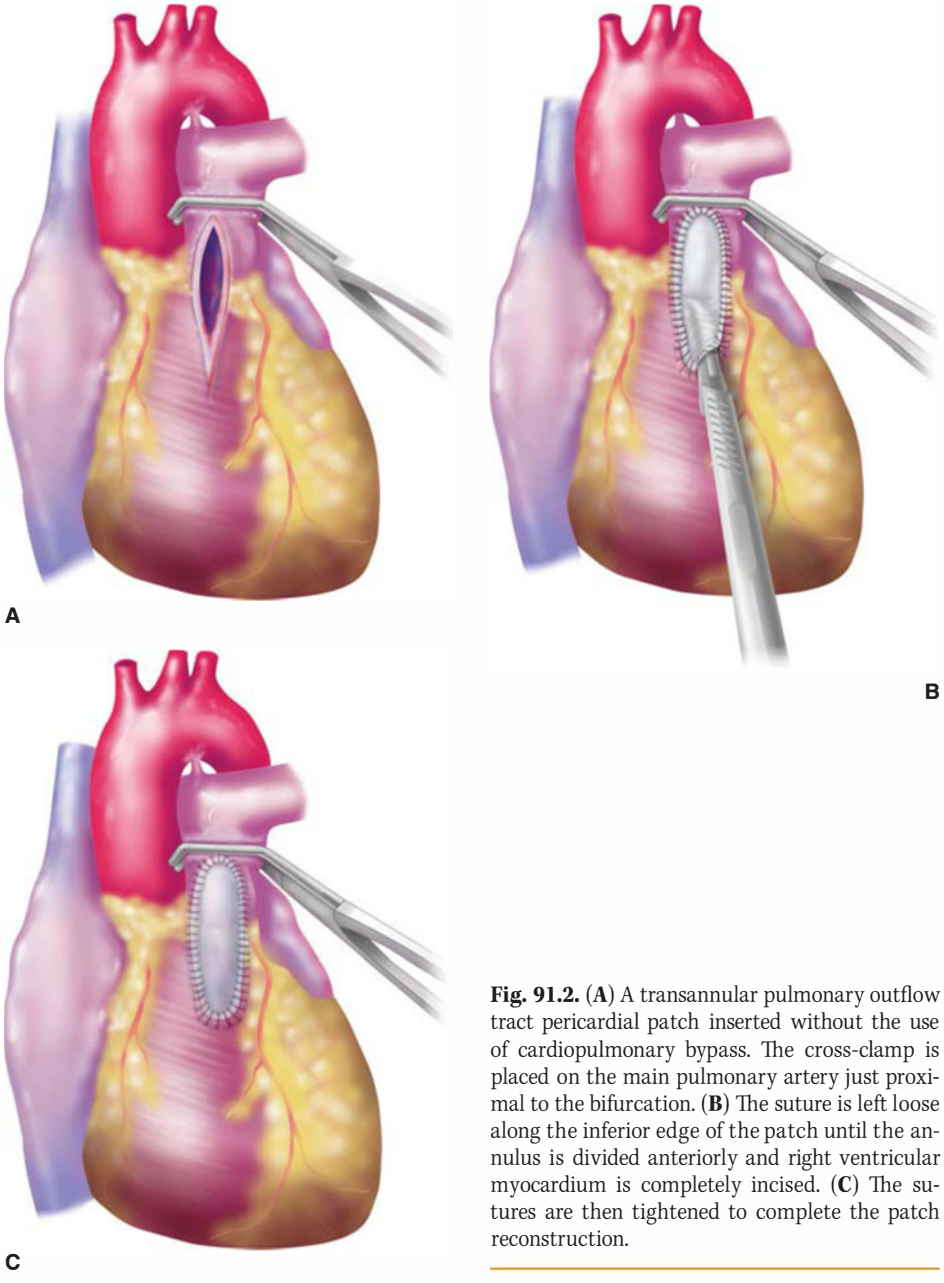


Fig. 91.2. (**A**) A transannular pulmonary outflow tract pericardial patch inserted without the use of cardiopulmonary bypass. The cross-clamp is placed on the main pulmonary artery just proximal to the bifurcation. (**B**) The suture is left loose along the inferior edge of the patch until the annulus is divided anteriorly and right ventricular myocardium is completely incised. (**C**) The sutures are then tightened to complete the patch reconstruction.

Initial Procedures for Neonates with Severe Right Ventricular Hypoplasia

Neonates with severe RV hypoplasia (less than one-third normal) can be difficult to manage. If there is a restrictive atrial communication, then balloon atrial septostomy may be performed at the time of catheterization. The ductus is kept patent with PGE_1 and may also be stented at the time of initial catheterization. At the time of surgery, simple pulmonary valvotomy is usually not effective in relieving RV hypertension and establishing antegrade flow to the pulmonary arteries. An extensive myomectomy and pericardial outflow tract patch may be performed using cardiopulmonary bypass. A Gore-Tex central shunt is inserted from the aorta to the main pulmonary artery, and the ductus arteriosus is ligated. If there are no sinusoids or if the sinusoids are tortuous and narrow and the native coronary circulation is normal, the right ventricle may be decompressed by incising the TV under direct vision using cardiopulmonary bypass and cardioplegic arrest or using a closed technique as described. It should be noted that patients who undergo tricuspid valvotomy decompression of the right ventricle should not have a concomitant pulmonary valvotomy with associated pulmonary insufficiency. This will allow the shunt flow to be directed retrograde across the outflow tract to the right ventricle and then into the right atrium, resulting in a significant steal of blood away from the pulmonary vascular bed. Repeat cardiac catheterization within 3 months is performed in this group of patients.

Closed Tricuspid Valvotomy

Closed tricuspid valvotomy is used in patients with severe RV hypoplasia where the possibility of a subsequent biventricular repair is minimal. Preoperative catheterization must determine the absence of sinusoids or an RVDCC before decompression of the right ventricle. As noted previously, RVDCC is one in which the native coronary circulation has proximal stenoses and the sinusoids

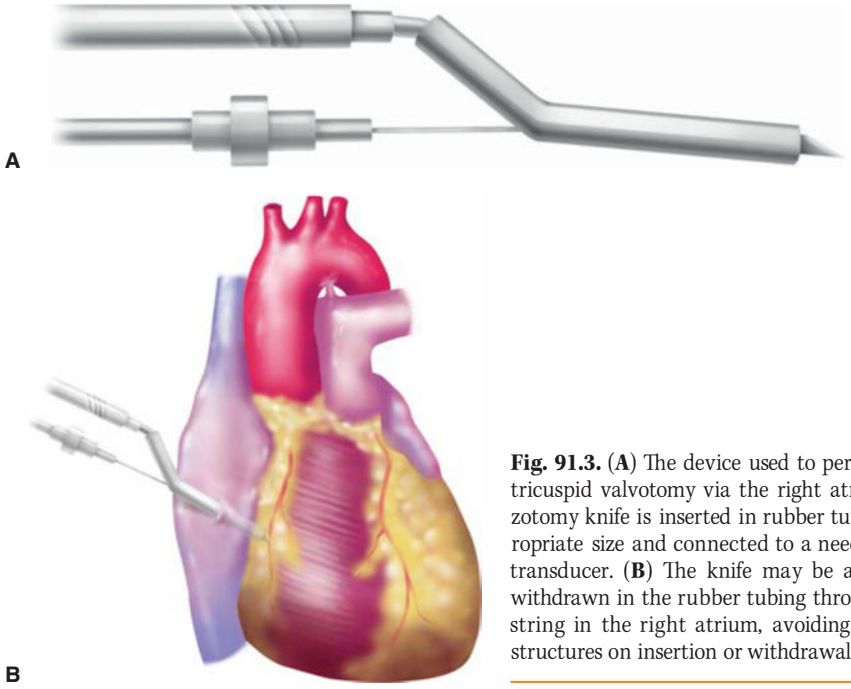


Fig. 91.3. (**A**) The device used to perform closed tricuspid valvotomy via the right atrium. A rhizotomy knife is inserted in rubber tubing of appropriate size and connected to a needle pressure transducer. (**B**) The knife may be advanced or withdrawn in the rubber tubing through a purse string in the right atrium, avoiding damage to structures on insertion or withdrawal.

have broad-based communications with the RV cavity and the coronary circulation.

The closed tricuspid valvotomy is performed through a median sternotomy. The pericardium is opened, and a purse-string suture is placed around the right atrial appendage. An instrument is made using a rhizotomy knife with a small, curved blade passed through a segment of red rubber tubing (Fig. 91.3A and 91.3B). A pressure-measuring needle is placed into the tubing and connected to a transducer monitor. The tubing is then introduced into the right atrium via the right atrial appendage and passed into the right ventricle while the pressure is monitored. The knife blade is then exposed in the right ventricle and the TV is cut anteriorly. Care is taken to avoid incising the area of the conduction system. When the RV pressure has fallen to one-half systemic pressure or less, the knife is retracted into the tubing, which is then removed from the right atrium. The purse-string suture is tied to achieve hemostasis. We have found in patients without a pulmonary valvotomy that decompression of the right ventricle with a tricuspid valvotomy results in regression of the narrow tortuous types of sinusoids and has not resulted in myocardial ischemia if the native coronary circulation is intact.

Definitive Surgical Treatment and Techniques

Cardiac catheterization is repeated when the patient is 3 to 6 months of age, depending on the anatomy and echocardiographic findings. Further procedures will be based on the anatomic findings at catheterization. These patients are again divided into those with mild, moderate, or severe RV hypoplasia (Table 91.2).

Definitive Procedures for Patients with Mild Right Ventricular Hypoplasia

Some patients with mild hypoplasia treated initially by pulmonary valvotomy may not require subsequent surgery unless the obstruction to the outflow tract has recurred. Relief of obstruction with pulmonary regurgitation and an open atrial communication can produce a large right-to-left shunt resulting in hypoplasia of the TV annulus while the right ventricle develops well due to the regurgitation. This can lead to a discrepancy in the size of the TV and the RV cavity. In a patient with mild RV hypoplasia, definitive repair consists of primary or patch closure of the ASD with a fenestration or an adjustable snare, enlargement of the RV cavity and RV outflow tract by myocardial resection, and pericardial patch augmentation of the RV outflow tract. A monocusp transannular patch with an autologous pericardial valve or a homograft or tissue valve is inserted in the pulmonary outflow tract, depending on the size of the child.

Enlargement of the Right Ventricular Cavity and Right Ventricular Outflow Tract

The RV cavity is enlarged by sharp resection of trabecular myocardium. The procedure is performed using cardiopulmonary bypass

Table 91.2 Approaches for Definitive Repair of Patients with Pulmonary Atresia and Intact Septum

Classification of RV and TV hypoplasia	Definitive repair treatment options
Mild	Closure of ASD (adjustable snare), enlargement of RV and RVOT, and transannular patch with monocusp valve Ligation of previous shunt
	Closure of ASD (adjustable snare), enlargement of RV and RVOT, and pulmonary homograft Ligation of previous shunt
Moderate	Closure of ASD (adjustable snare), bidirectional Glenn shunt, enlargement of RV and RVOT, and transannular patch with oversized stented bioprosthetic valve Ligation of previous shunt
	Closure of ASD (adjustable snare), bidirectional Glenn shunt, enlargement of RV, and pulmonary homograft or Contegra graft Ligation of previous shunt
Severe[a]	Bidirectional Glenn shunt Partial or complete ligation of previous shunt
	Fontan procedure (lateral tunnel or extra cardiac) with adjustable or fixed ASD fenestration

[a]Patients with RV-dependent coronary circulations may be considered for aorta to RV conduit.
ASD, atrial septal defect; RV, right ventricular; RVOT, right ventricular outflow tract; TV, tricuspid valve.

with bicaval cannulation and both antegrade and retrograde blood cardioplegia. The right atrium is opened obliquely and the TV is inspected. The annulus is measured and compared with normal values, and its competence is tested with cold saline. An incision is made longitudinally from the main pulmonary artery, through the annulus. If the outflow tract is hypoplastic, the incision is extended to the RV cavity. Obstructive hypertrophied muscle in the outflow tract is resected. Care is taken to work between the papillary muscles of the TV, which must be preserved. A glutaraldehyde-treated pericardial outflow patch is then placed on the transannular incision. If the RV cavity is adequate and the TV is competent, a pulmonary valve may not be required. In infants, we generally prefer a pericardial monocusp valve. In older children, we have used a pulmonary homograft or porcine valve within the RV outflow tract.

Adjustable Atrial Septal Defect

If the ASD is large, it is closed with a pericardial patch, leaving an open defect adjacent to the right superior pulmonary vein for the adjustable snare. If the defect is small with firm edges, it may be closed with the purse string of the "adjustable atrial septal defect." The adjustable ASD is performed by placing a no. 1 prolene suture as a purse string around the tissue edges of the existing septal defect. A pericardial pledget is used to avoid tearing of the tissue edges. The no. 1 prolene is secured to the edges of the defect with 5-0 prolene interrupted sutures. Both ends of the no. 1 prolene are then brought out through the interatrial groove. An 8-F polyethylene tube is measured so that the end will lie under the linea alba closure. The prolene sutures are passed through the lumen of this tube to create a snare to control the size of the atrial communication. The end of the tubing is sutured to the atrial wall with a single prolene suture. The size of the ASD is adjusted by tightening or loosening the snare. Retracting the prolene sutures in the snare results in closure of the ASD, and pushing on the prolene opens the communication. The prolene is then fixed to the tube with several medium hemoclips. The end of the snare is left under the linea alba in the subxiphoid region, where it can be retrieved under local anesthesia postoperatively for subsequent adjustment. The same technique can be used to create an adjustable defect in a patch repair using pericardium. The defect in the suture line is left on the right side adjacent to the right superior pulmonary vein, where it is encircled by the no. 1 prolene snare as described. The ASD is left open until the patient is weaned from bypass. The ASD is then slowly closed using the snare while the right atrial pressure and the arterial oxygen saturations are monitored. A right atrial pressure of about 10 to 12 mmHg with an oxygen saturation of ≥88% on 100% fraction of inspired oxygen (FiO_2) is the goal. This adjustment of the ASD snare may be facilitated by using transesophageal echo visualization as well (Fig. 91.4A and 91.4B).

Definitive Procedures for Patients with Moderate Right Ventricular Hypoplasia

In a patient with moderate RV hypoplasia, definitive repair is dictated by the previous growth and development of the right ventricle and the TV. If the right ventricle and TV diameter are one-half to two-thirds of normal size, repair consists of partial closure of the ASD with an adjustable snare, enlargement of the RV cavity by myocardial resection, and insertion of a valved connection between the right ventricle and the pulmonary artery. A monocusp transannular patch from native pericardium may be used in younger patients. If the right ventricle and TV diameter is one-third to one-half of normal, repair consists of partial closure of the ASD with an adjustable snare, enlargement of the RV cavity, creation of a bidirectional cavopulmonary Glenn shunt, and insertion of a valved connection between the right ventricle and pulmonary artery. The Glenn shunt reduces the volume load on the small right ventricle and provides an obligatory source of pulmonary blood flow

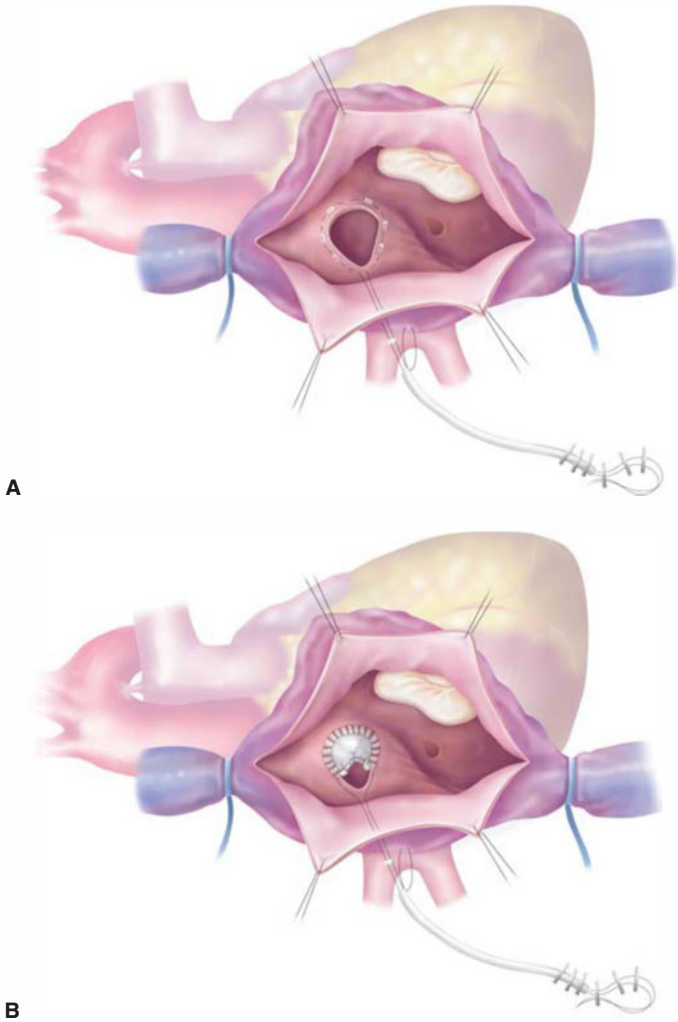


Fig. 91.4. An adjustable atrial septal defect with and without patch closure of the native atrial septal defect. (**A**) A purse string is sewn around the border of the atrial septal defect. Pledgets may be used to reinforce the purse string. The snare is constructed using an 8-F polyethylene tubing placed over the no. 1 polypropylene (prolene) and measured to reach the linea alba. (**B**) If a large secundum atrial septal defect is present, it is closed with a pericardial patch and a defect is left in the lateral wall. The no. 1 prolene is brought through the interatrial septum and placed as a horizontal mattress stitch through the edge of the patch. The no. 1 prolene is then anchored to the patch with a 5-0 prolene suture.

from approximately one-third of the systemic venous return. This has been termed the "one and one-half ventricle repair" or "partial biventricular repair." The ASD is adjustable to create a gradient between the right atrium and left atrium to encourage forward flow through the RV, which will enhance the development of the TV and the right ventricle. Either a two-ventricle repair (with takedown of the Glenn shunt) or a completion of the Fontan reconstruction will follow based on the subsequent growth and development of both the right ventricle and TV.

Tissue Valve Insertion

A transannular incision is made vertically across the pulmonary outflow tract and extended onto the left pulmonary artery and down into the right ventricle. Any residual membrane in the region of the annulus is resected, as well as obstructive muscle in the outflow tract. The distance between the RV outflow tract and the pulmonary artery bifurcation is assessed. If it is short, a stented bioprosthetic porcine valve can be placed under a pericardial or Gore-Tex patch within the RV outflow tract (Fig. 91.5A and 91.5B). A running 3-0 prolene suture is used to insert the valve, which is also sutured to the patch anteriorly. The pericardium is treated with glutaraldehyde for 3 minutes and rinsed with saline. If the distance between the RV outflow tract and the pulmonary artery bifurcation is adequate, an appropriately sized pulmonary homograft may be used. A running suture of 4-0 prolene is used distally just below the pulmonary artery bifurcation. Proximally, the homograft is sutured to the RV outflow tract just below the pulmonary valve annulus with a running suture of 3-0 prolene (Fig. 91.6A and 91.6B). A hood of pericardium or Gore-Tex may be used to complete the reconstruction.

Transannular Pericardial Patch with Monocusp Valve

The technique of a transannular pericardial patch with a monocusp valve is not only used for neonates and infants but can also be used for older children. It is best used for patients with mild or moderate RV hypoplasia with normal pulmonary artery size because the valve will remain competent for a shorter period of time than a tissue valve. It has the advantage, however, of not causing obstruction even when the valve has become incompetent. Insertion of a transannular patch with a monocusp valve is performed through median sternotomy using cardiopulmonary bypass and bicaval cannulation. After harvesting, the pericardium is treated with glutaraldehyde for 5 minutes and then rinsed in saline. Both the transannular patch and the monocusp valve leaflet are outlined on the harvested pericardium using a sterile marking pen. Sizing of the monocusp valve is made using a metal dilator approximately 20% larger than the "normal" diameter for the pulmonary annulus. The width of the monocusp leaflet at its base should be approximately one-half of the circumference of the dilator. It should also correspond with the width and shape of the inferior end of the transannular patch. The superior edge of the monocusp valve leaflet should be attached to the edges of the incised pulmonary artery 5 to 10 mm distal to the area of the true valve annulus. The monocusp valve is attached to the edges of the pulmonary artery and the right ventricle using the same suture that attaches the edges of the transannular

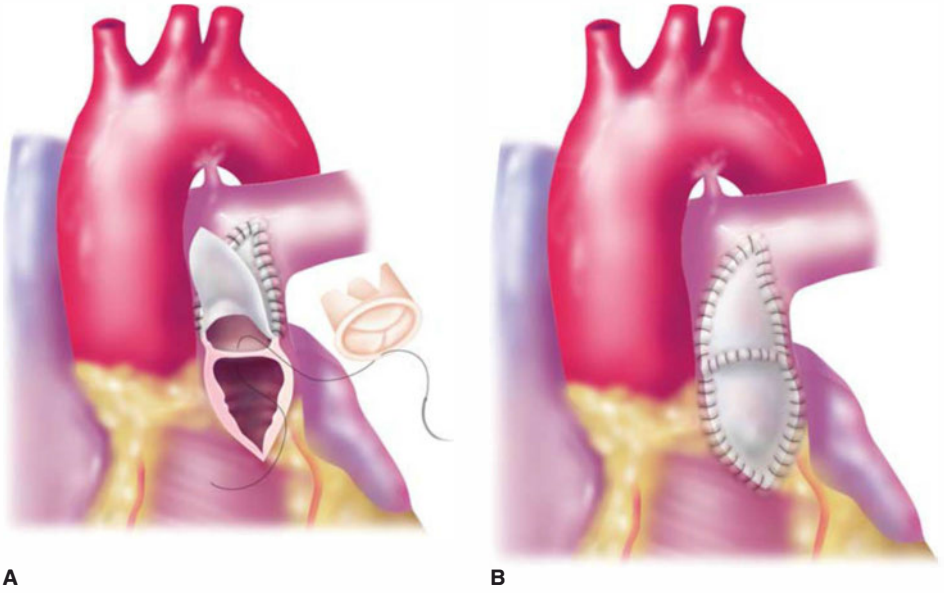


Fig. 91.5. (**A**) Insertion of a porcine valve under a transannular patch for enlargement of the right ventricular outflow tract. A large valve is chosen and is sutured to the annulus posteriorly and to the patch anteriorly with a running suture technique. (**B**) The patch suture line is then completed inferiorly.

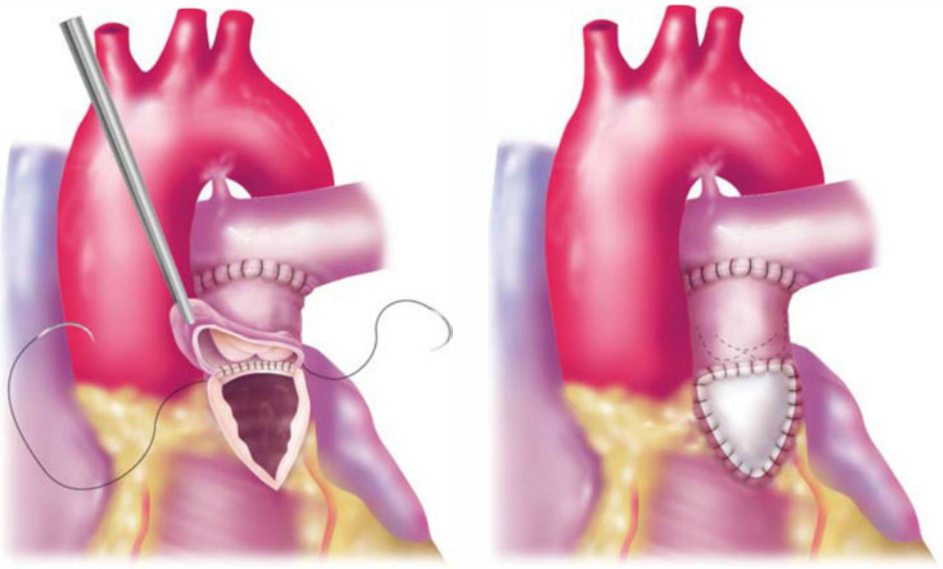


Fig. 91.6. (**A**) A pulmonary homograft is used to enlarge the right ventricular outflow tract. The size of the homograft is carefully selected, and the graft is fashioned on both ends. The homograft is inserted as shown using a running polypropylene (prolene) suture for the proximal and distal anastomoses. (**B**) A pericardial or Gore-Tex hood may be used to augment the proximal anastomosis.

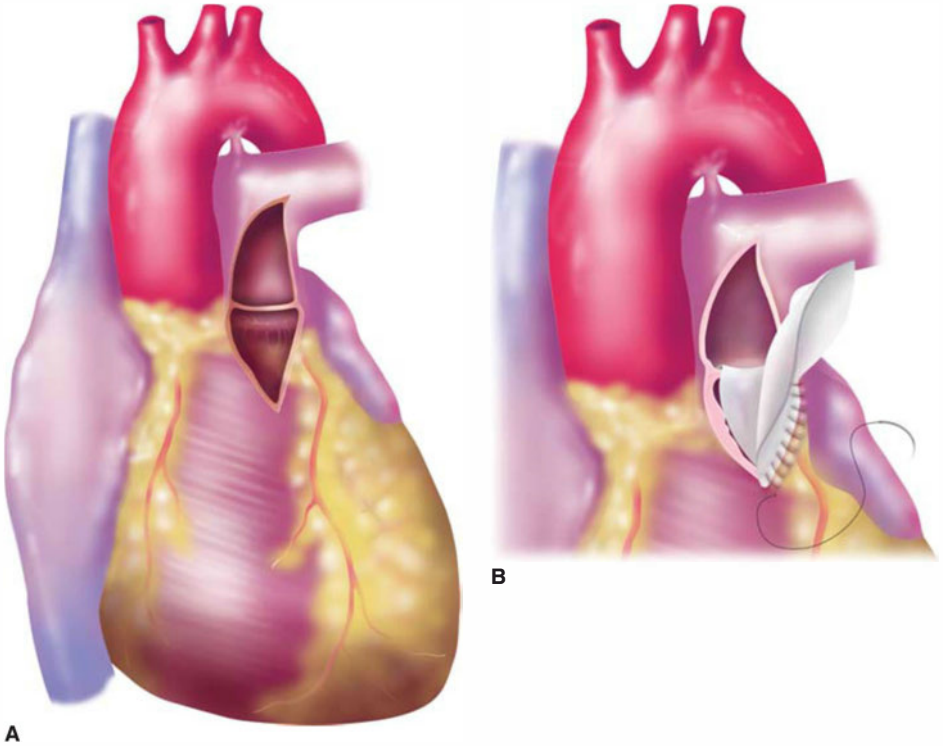


Fig. 91.7. (**A**) A Gore-Tex or pericardial monocusp valve with transannular patch is inserted to augment the right ventricular outflow tract. (**B**) The monocusp valve is sutured to the edges of the myocardium and corresponds with the width and shape of the transannular patch.

patch (Fig. 91.7A and 91.7B). A strip of treated pericardium may also be used to reinforce the suture line in the myocardial edges and promote hemostasis.

Definitive Procedures for Patients with Severe Right Ventricular Hypoplasia

In a patient with severe RV hypoplasia (one-third of normal size or less) and a *Z*-score of −4 to −6, a biventricular repair is usually not possible. Most of these patients will undergo insertion of a central shunt in the neonatal period with or without decompression of the right ventricle depending on whether or not there is an RVDCC. Tortuous sinusoidal connections without coronary stenoses do not usually denote an RVDCC. Decompression of the right ventricle at the time of the bidirectional Glenn shunt will usually result in closure of these sinusoids as opposed to the broad-based fistulous connections. In some cases, the large fistulous connections can be identified on the epicardial surface of the heart and can be suture ligated at the time of the bidirectional Glenn shunt, allowing RV decompression at that time. RV decompression can be performed via the right atrium using the closed technique described previously to incise the anterior leaflet of the TV.

If there is an RVDCC, a bidirectional Glenn shunt is performed at 3 to 4 months of age without RV decompression and without cardiopulmonary bypass. Any additional source of pulmonary blood flow such as a previously placed central shunt is reduced to give an estimated combined Q_p:Q_s ratio of 1:1 from the Glenn shunt and the systemic-to-pulmonary artery shunt. At 2 to 3 years of age, the Fontan procedure is completed with a fenestration or an adjustable ASD. During this final procedure, the atrial septum is excised and the coronary sinus is unroofed. If there are signs of myocardial ischemia, either preoperatively or intraoperatively, the RVDCC may be improved by creating an aorta-to-right ventricle shunt at the time of the Glenn shunt or the Fontan procedure. Early bidirectional Glenn shunting or augmentation of RV flow with an aorta-to-right ventricle connection and an early Fontan procedure results in improved oxygenation of the coronary flow from the RV in these patients with RVDCC.

However, these patients may continue to have problems with ventricular dysfunction and myocardial ischemia, and may have acute myocardial infarction early in life, causing ventricular dysfunction or early death. Thus, in some patients with significant ventricular dysfunction in association with RVDCC, early shunting for stabilization followed by orthotopic cardiac transplantation should be considered.

Bidirectional Cavopulmonary Glenn Shunt

A median sternotomy incision is used for the Glenn shunt. Care should be taken in dissecting out the superior vena cava (SVC) and the right pulmonary artery to stay in the perivascular plane and avoid disrupting lymphatic tissue, which could result in a chylothorax. Injury to the phrenic nerve as it courses along the pericardium on the lateral aspect of the SVC must also be avoided. The pulmonary artery pressure is measured on both sides. The azygos vein is ligated and divided.

A left SVC may be present and is usually found anterior to the left pulmonary artery. Care must be taken not to mistake a left upper-lobe pulmonary vein for a systemic vein. The SVC is then clamped just above the right atrium, and the proximal pressure is monitored. If the pressure does not rise above a mean of 30 mmHg, then a left SVC should be suspected. If there is a left SVC with a venous communication to the right SVC, the pressure will usually not rise above 30 mmHg and bypass or a temporary shunt is not necessary.

If the proximal pressure in the clamped SVC is ≥30 mmHg, a temporary shunt should be used. The use of bypass is avoided if at all possible, particularly if there are ventricular sinusoids. The SVC is dissected from surrounding tissue starting at its right atrial junction and extending to the confluence of the innominate vein. The right pulmonary artery is identified and dissected circumferentially from surrounding tissue both medially and laterally to the site of primary branching. A purse-string suture of 5-0 prolene is placed in the SVC at the junction of the innominate vein. A second purse string is placed in the right atrial appendage. The patient is heparinized, and a temporary bypass shunt is created using two modified aortic cannulas and a Y-connector with a chapeau attachment. The SVC is cannulated with one modified aortic cannula at the junction of the innominate vein. The distal end of this cannula should be beveled so that the opening is as large as possible relative to the venous cannulation site and should face superiorly toward the right internal jugular vein. The second cannula is placed in the right atrial appendage. The circuit is connected after air is completely evacuated from the cannulas and the clamps are removed. A needle can be placed in the chapeau attachment to facilitate the removal of air. Flow should be visualized in

the shunt after removal of the clamps. The cannulas are then supported by towels in a medial position that allows exposure of the SVC and right pulmonary artery.

The SVC is then clamped at its junction with the right atrium and at its junction with the innominate vein. The pressure may be measured in the proximal SVC with the first clamp in place to ensure proper functioning of the shunt. Methylene blue is used to mark the anterior aspect of the SVC and the superior aspect of the PA to ensure proper alignment of the subsequent cavopulmonary anastomosis. The azygos vein is ligated and divided. The SVC is divided at the atrial junction, with care being taken to leave an adequate cuff of tissue with the clamp to allow subsequent closure of this SVC stump. Attention to the rhythm at this point helps to avoid clamp injury to the sinoatrial node. The open end of the SVC is enlarged by an incision in the posterior wall of the vessel. This enlargement ensures a widely patent anastomosis. The right pulmonary artery is clamped with a C-clamp, and a V-shaped incision is made on its superior aspect with the apex pointing anteriorly. The anastomosis is performed with running or interrupted 6-0 or 7-0 prolene (Fig. 91.8A and 91.8B). The cannula in the SVC is removed, followed by removal of the right atrial cannula. If the SVC is narrowed at the site of the purse string, a vascular C-clamp is applied, the purse string is removed, and the incision in the SVC is closed with a running 7-0 prolene suture. Any previously placed systemic-to-pulmonary artery shunt is now reduced in size to give an estimated combined Q_p:Q_s ratio of 1:1 from the systemic-to-pulmonary artery shunt and the Glenn shunt. The change in arterial systolic blood pressure, with the shunt open versus closed, should be no more than 5 to 7 mmHg.

The SVC and pulmonary artery pressures are measured, as well as the arterial oxygen saturations on 100% oxygen. The heparinization is not reversed. If a jugular vein line has been inserted, it is used for measuring the pulmonary artery pressure and for the infusion of pulmonary vasodilators, if necessary. The line is removed within 24 hours to avoid venous thrombosis. A Gore-Tex membrane is left as a pericardial substitute to facilitate reoperation for the Fontan procedure

Lateral Tunnel Fontan with Adjustable Atrial Septal Defect

A Fontan procedure is usually performed as a second-stage operation after the creation of a cavopulmonary Glenn shunt. A median sternotomy is performed, and bicaval venous cannulation is used for total cardiopulmonary bypass. Note that the SVC cannula is placed above the previous Glenn shunt anastomosis. Systemic hypothermia to 28°C is used in addition to cold blood cardioplegia.

A right atriotomy is performed just anterior to the linea terminalis, and the edges are retracted with stay sutures. The coronary sinus is identified and cannulated with a retrograde cardioplegia catheter. A prolene purse string is placed at the opening of the coronary sinus to secure the catheter and achieve more efficient delivery of the cardioplegia. The atrial septum is excised. The SVC orifice is identified from within the right atrium. It is important that this orifice be widely open and not restrictive. The right pulmonary artery is incised adjacent to the opening in the SVC stump. The posterior wall of the anastomosis is achieved by suturing the adjacent pulmonary artery and right atrium together with 5-0 prolene. Anteriorly, the connection may be bridged with a pericardial patch if needed to assure a patulent unrestrictive opening.

After the right atrium-to-pulmonary artery anastomosis has been completed, the lateral tunnel is constructed. A rectangular Gore-Tex patch is cut from 0.8-mm-thick Gore-Tex vascular patch material. The length is carefully measured from the orifice of the inferior vena cava (IVC) to the SVC orifice. The width is left about two-thirds of the length to be trimmed after completion of the posterior suture line.

A running 4-0 prolene suture with an RB1 needle is used for the posterior suture line, which is begun posteriorly at the IVC orifice. The suture line is carried superiorly to the site of the adjustable ASD orifice, where it ends. This site is chosen because there is a natural recess close to the right superior pulmonary vein at the superior and lateral end of the fossa ovalis. A second 4-0 prolene suture line is begun at the superior end of the ASD and carried superiorly around the SVC orifice. The ASD is sized according to the age of the patient and made large in diameter so that it may be reduced in size if necessary after the patient is taken off bypass. As a rule of thumb, the defect size is 4 mm for 2-year-olds and 6 mm for 4-year-olds and above. The patch is trimmed appropriately as the suture line advances. Before the anterior suture line is completed, the snare control is placed for the adjustable ASD. A no. 1 prolene suture is brought through a pericardial pledget, through the interatrial septum at the lower border of the ASD, and through the edge of the Gore-Tex patch. It is then brought back through the upper edge of the Gore-Tex patch and out through the interatrial septum and through the pericardial pledget. An 8-F polyethylene tube is cut to the appropriate length to reach the linea alba, and the prolene sutures are brought through this snare. The no. 1 prolene is sutured to the edge of the Gore-Tex patch with a 5-0 prolene suture, the polyethylene tubing is sutured to the lateral wall with 2-0 chromic catgut, and the prolene is fixed to

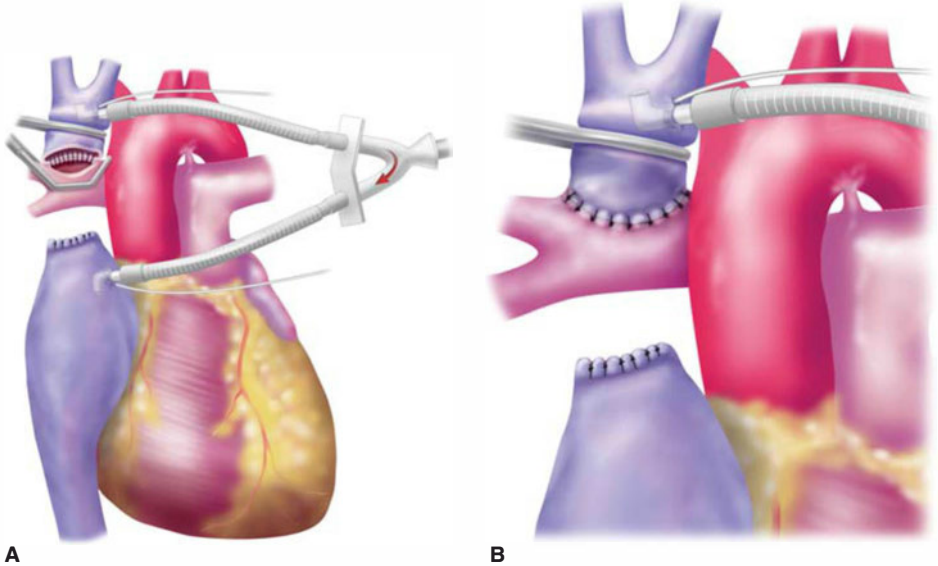


Fig. 91.8. (**A**) A bidirectional Glenn shunt is performed using a superior vena cava-to-right atrium shunt. This technique avoids the use of cardiopulmonary bypass. (**B**) The cannulas are placed in the superior vena cava and the right atrium (as shown), allowing continuous flow of venous return to the right atrium during creation of the cavopulmonary anastomosis.

the tubing with a medium hemoclip. These three points of fixation prevent inadvertent closure of the ASD by tension on the prolene. The patch is now trimmed to create a wide-open connection and to reach just anterior to the linea terminalis. The anterior part of the suture line is completed using full-thickness sutures to avoid a suture line leak. The right atrial incision is then closed with 4-0 prolene (Fig. 91.9A and 91.9B).

The coronary sinus cannula and the purse-string suture are removed, and the right atrium is closed in a two-layer fashion with a running prolene suture. Transthoracic lines are generally placed in the left and right atria. If an internal jugular line is not inserted, then the right atrial line can be inserted to measure the pressure in the pulmonary system via the Fontan tunnel or the Glenn shunt. Air is evacuated from all structures. The patient is warmed to normothermia and subsequently weaned from cardiopulmonary bypass. The adjustable ASD snare is adjusted to achieve arterial saturations of 80% to 85% while an attempt is made to maintain pressure in the lateral tunnel Fontan at or below 15 mmHg. Adjustment of the ASD can be facilitated by visualization intraoperatively with transesophageal echocardiography.

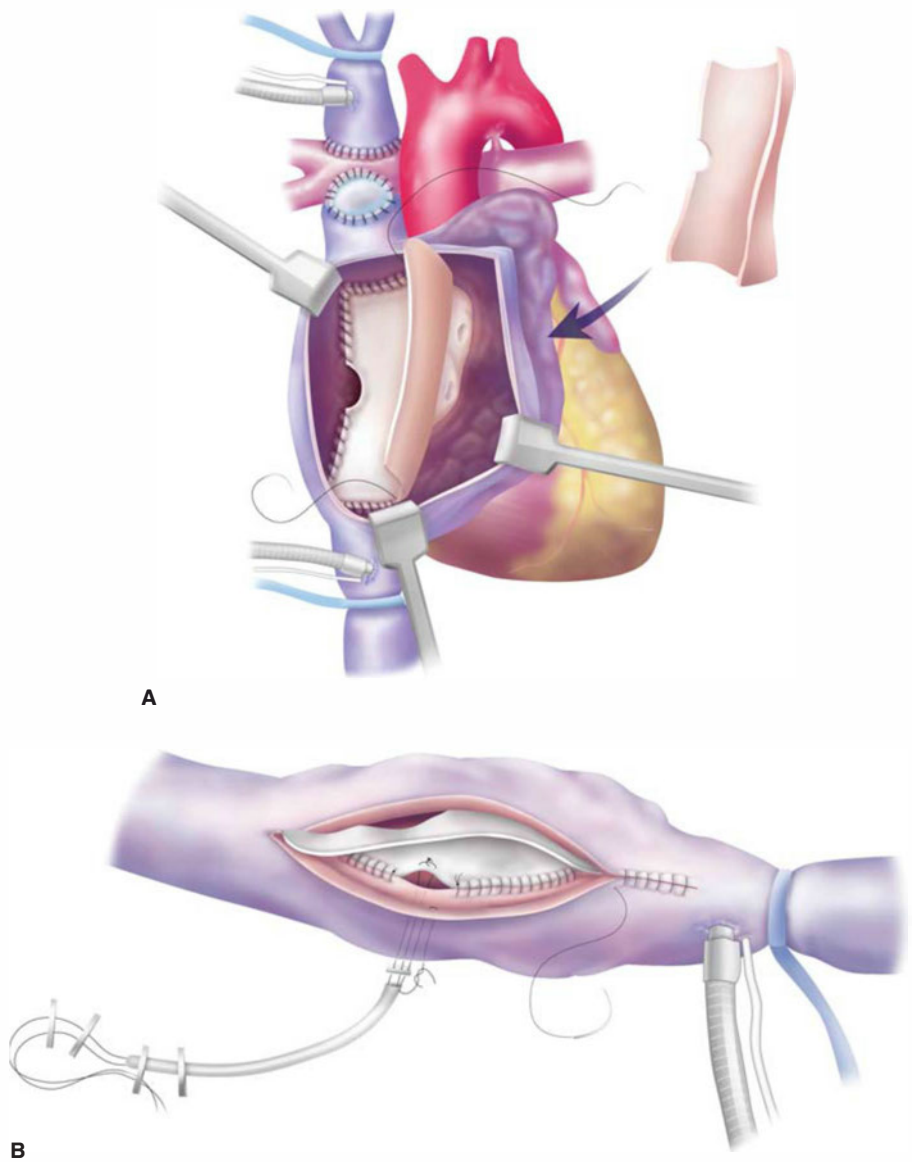


Fig. 91.9. (**A**) A lateral tunnel Fontan procedure with an adjustable atrial septal defect. A tunnel of uniform caliber is created by suturing a piece of polytetrafluoroethylene (Gore-Tex) vascular patch to the orifice of the inferior vena cava and the superior vena cava and to the sinus venosus portion of the lateral wall. (**B**) A defect is left in the lateral tunnel. The adjustable atrial septal defect is created by passing the no. 1 polypropylene (prolene) through the lateral portion of the interatrial septum and through the edge of the Gore-Tex tunnel. The no. 1 prolene is then secured to the edge of the Gore-Tex with a 5-0 prolene suture. The suture is brought back out the interatrial septum and through a pericardial pledget. The snare is constructed with 8-F polyethylene tubing and anchored to the heart through the pledget and through the atrial wall. Medium hemoclips are used at the end of the snare to fix the atrial septal defect.

For Fontan operations in these patients, some have preferred the use of the fenestrated lateral tunnel Fontan with a fixed 4-mm fenestration by a punch technique rather than use of an adjustable defect. While this does not allow postoperative adjustment of the shunt, it does simplify the operative procedure and has been shown to be effective. In most of these cases, the fixed defect will ultimately close spontaneously.

Extracardiac Fontan with Adjustable Fenestration

The extracardiac Fontan is performed through a median sternotomy using cardiopulmonary bypass and bicaval cannulation. The procedure can be completed in most patients without the need for cardioplegic arrest of the heart. A clamp is placed on the IVC near its junction to the right atrium. The IVC is then divided between the snared venous cannula and the clamp. The atrium is repaired and the clamp is removed. The open end of the IVC is anastomosed end-to-end to a Gore-Tex conduit (16 to 20 mm in diameter) using a running Gore-Tex suture. The proximal anastomosis is performed end-to-side between the Gore-Tex conduit and the inferior aspect of the right pulmonary artery. The clamps are released and flow is established between the IVC and the pulmonary arteries. A direct anastomosis can be made between the conduit and the atrial wall to allow shunting through the fenestration (Fig. 91.10A and 91.10B).

To insert an adjustable ASD, a partial occluding vascular C-clamp is placed on the Gore-Tex graft. A direct anastomosis is performed between the extracardiac conduit and the right atrium. A snare is inserted to control the opening and closing of this "atrial septal defect." An alternate method uses a conduit for the defect. With this technique, a short segment of 8.0-mm Gore-Tex graft is anastomosed end-to-side to the middle of the larger conduit. A similar technique is used to create an opening in the right atrium, and the other end of the 8.0-mm graft is anastomosed to this site. A snare is inserted around the smaller conduit. A distinct drawback to the extracardiac Fontan is the need for anticoagulation with warfarin postoperatively for 6 months to 1 year, with subsequent conversion to aspirin therapy.

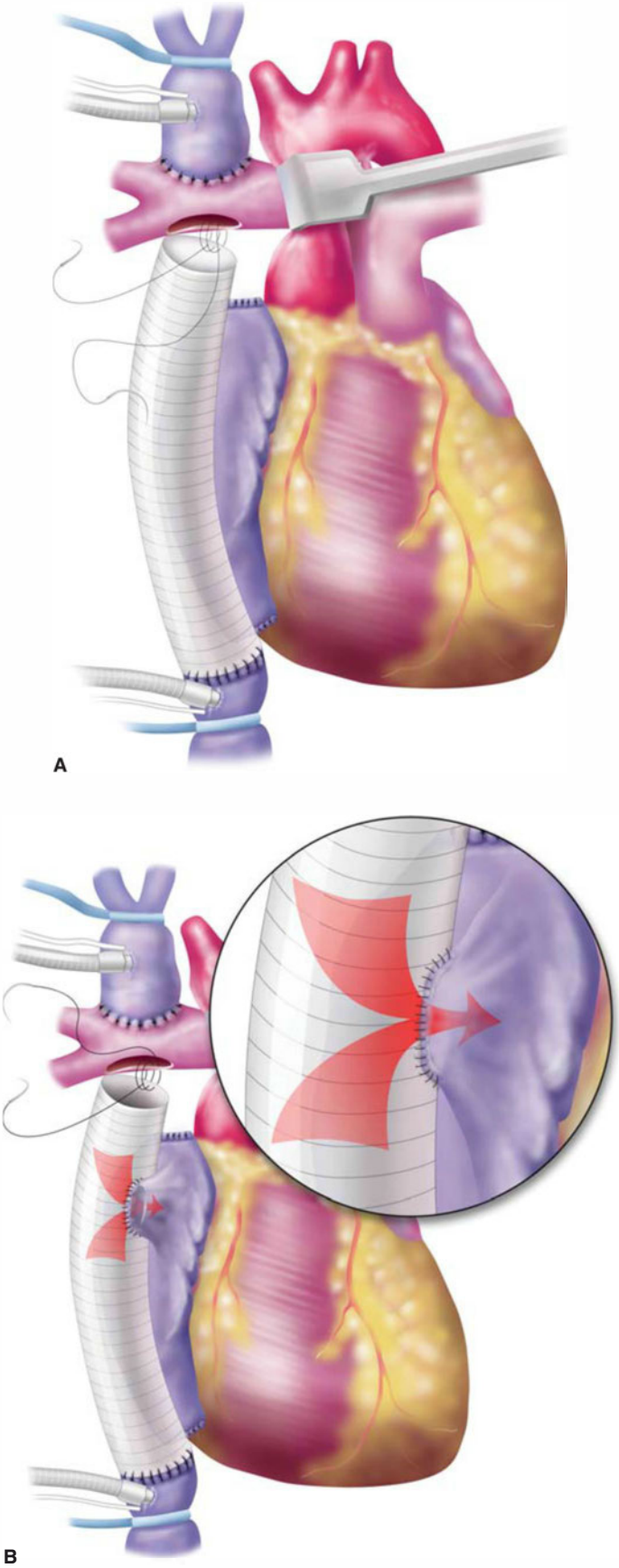


Fig. 91.10. (**A**) An extracardiac Fontan conduit is anastomosed to the inferior vena cava distally and the inferior aspect of the proximal right pulmonary artery proximally. (**B**) A fixed fenestration may be made between the Fontan conduit and the right atrium to allow shunting to the right atrium. An adjustable snare (not shown) can also be used with this type of fenestration.

Aorta-to-Right Ventricle Shunt

The aorta-to-right ventricle shunt is performed through a median sternotomy using cardiopulmonary bypass and bicaval cannulation. Cardioplegic arrest of the heart may or may not be necessary. The shunt is created using a 5.0-mm ringed Gore-Tex graft. A partial occluding clamp is placed on the anterior wall of the ascending aorta. An aortotomy is created, and an end-to-side anastomosis is performed between the graft and the ascending aorta using a running polypropylene suture. A ventriculotomy is made in the infundibular portion of the right ventricle, and the distal end of the graft is anastomosed to this site using a running polypropylene suture.

Postoperative Care and Surgical Complications after the Fontan Procedure

Early management after the Fontan procedure is focused on optimizing cardiac output and reducing the systemic venous pressure. The adjustable ASD or fenestration is useful because it allows as much as one-third of the systemic venous return to traverse the ASD to the left atrium, thus lowering the systemic venous pressure while increasing the cardiac output. A Fontan pressure of ≤15 mmHg is ideally achieved by adjustment of the fenestration.

The pulmonary vascular resistance may be reduced significantly with the use of inhaled nitric oxide. This drug has an advantage over other pulmonary vasodilators because its effect is confined to the pulmonary vascular bed, and the systemic vascular resistance is not reduced. Methemoglobin levels should be monitored carefully with the use of nitric oxide.

Inotropes are routinely used, starting with 5 μg/kg per minute of dopamine and dobutamine. If the systemic vascular resistance is low, dopamine or epinephrine in higher doses is infused via the left atrial line. If these medications are not effective, milrinone or isoproterenol (Isuprel) may be added.

If the hemodynamics are optimal with minimal support, early extubation is attempted. In more critical cases, the patients are sedated and paralyzed for 12 hours before being weaned from the ventilator. If the systemic venous pressure has been high early in the postoperative course, the patient is diuresed before extubation. The patient must be actively monitored for the development of pleural and pericardial effusions, which must be drained immediately. If the venous pressure or the left atrial pressure is high,

echocardiography is performed to assess ventricular and valvular function and to exclude the presence of any obstruction within the systemic venous pathway or pulmonary arteries. Rarely, recatheterization may be necessary for postoperative assessment.

The ASD may require adjustment early postoperatively. In most cases, it is left partially open. In some patients, as the left ventricular function improves, the right-to-left shunt increases, resulting in a decrease in the arterial oxygen saturation. The snare can be exposed under local anesthesia behind the linea alba in the subxiphoid region and is further tightened using medium hemoclips. Betadine is used to irrigate the wound and is injected into the polyethylene snare. The ASD can also be opened further by pushing on the prolene sutures, but this is not as reliable as the ability to close the ASD. The use of an adjustable ASD at complete repair permits right-to-left shunting in those cases in which early ventricular compliance limits forward flow into the pulmonary vascular bed and helps to stabilize these patients in the critical early postoperative period.

At 6 to 12 weeks, an echocardiogram and arterial oxygen saturation measurement are used to assess the size of the defect. If it is small, it is left to close spontaneously. If it is still large, the defect is closed in the catheterization laboratory with a balloon, and if the venous pressures are acceptable, the snare is exposed again behind the linea alba and the defect is closed completely. This allows delayed closure of the ASD without surgical or catheter device intervention.

SUMMARY

In neonates with pulmonary atresia and intact septum, we have found that the surgical classification of RV and tricuspid hypoplasia into mild (more than two-thirds of normal with a tricuspid *Z*-score of 0 to −2), moderate (one-third to two-thirds of normal with a tricuspid *Z*-score of −2 to −4), and severe (less than one-third of normal with a tricuspid *Z*-score of −4 to −6) has been useful in selecting a surgical approach.

A similar classification is used in older children, and patients are stratified into those who will benefit from an attempt to achieve a biventricular repair and those who are best suited to a single ventricle strategy and a Fontan procedure. With this approach, the surgical mortality and morbidity can be reduced.

RESULTS OF SURGICAL TREATMENT

Historically, the surgical treatment of PA/IVS was associated with very high morbidity and mortality. Previous extensive reviews of the clinical and pathologic aspects of this defect have allowed for a greater understanding of the disease and the development of surgical therapy. With increasing surgical experience, a more standardized approach has evolved and outcomes from many institutions have steadily improved. The role of catheter-based interventions in these patients, including balloon valvotomy and stenting of the RVOT and the ductus arteriosus, is evolving and remains to be clearly defined.

In the CHSS multi-institutional study, 408 neonates with PA/IVS were studied from 33 institutions. Overall survival was 77% at 1 month, 70% at 6 months, 60% at 5 years, and 58% at 15 years. Prevalence of end states at 15 years were as follows: successful biventricle repair, 33%; Fontan repair, 20%; one-and-a-half ventricle repair, 5%; heart transplant, 2%; death before reaching definitive repair, 38%; and alive without definitive repair, 2%. They concluded that 85% of all neonates with PA/IVS are likely to reach a definitive surgical repair, with a double-ventricle repair achieved in an estimated 50% and a Fontan repair achieved in 35% in the current era.

FUTURE CONSIDERATIONS

Transcatheter therapy in patients with PA/IVS is becoming a more common approach to initial interventions in neonates. These techniques include radiofrequency ablation valvotomy, PDA stenting, RVOT stenting, and device closure of ASD. As experience increases, these techniques are gaining wider acceptance as alternatives to or complements to surgery in treatment options. A major area of controversy regarding pulmonary atresia and intact ventricular septum is the use of radiofrequency ablation and perforation of the RV outflow tract with balloon dilation in preference to surgical intervention in these neonates. Although the early experience is relatively good with radiofrequency ablation and perforation of the RV outflow tract with balloon dilation (followed by ductal stenting if necessary to maintain pulmonary blood flow), the disadvantage of this technique is the fact that in patients with severely hypoplastic RV outflow tract there is not complete relief of the gradient. Residual obstruction may limit RV growth, and a more radical relief of RV outflow tract obstruction and even division of hypertrophied muscle bundles in the cavity of the right ventricle may permit better growth. The technique can be very effective, however, in patients who have a plate-like pulmonary valve atresia with a reasonable-sized pulmonary annulus and relatively well-developed right ventricle. In these cases, balloon dilation while maintaining the patient on prostaglandin and then gradually letting the ductus arteriosus close can permit total interventional repair with closure of the ASD with an occluder device at a later time. Hybrid therapies, including a Blalock–Taussig shunt and RV outflow reconstruction with later occlusion of the Blalock–Taussig shunt and atrial septum in the catheterization lab, have also been used with good success in selected patients. A comparison of these relatively noninvasive approaches with surgical approaches in all patients with pulmonary atresia and intact septum has not been performed, and therefore the selection criteria for these various interventions are controversial. Various hybrid approaches may emerge combining both surgical and catheter-based interventions. In some studies, however, catheter-based interventions in neonates have rarely avoided surgical interventions.

In utero perforation and dilatation of the PV in mid-gestational fetuses with PA/IVS has been reported in a limited experience. The overall effect of this type of fetal intervention on subsequent growth and development of the right ventricle and its clinical impact on outcomes in patients with PA/IVS also remains to be defined.

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EDITOR'S COMMENTS

Infants with critical pulmonary stenosis and an adequate-sized pulmonary valve annulus are now treated with balloon valvuloplasty rather than surgical intervention. However, patients with severely dysplastic pulmonary valves or significant RV outflow tract obstruction with dynamic infundibular obstruction may still come to surgical intervention and require valvectomy or outflow patching. Transventricular dilation of a stenotic pulmonary valve is rarely necessary but may be applied to certain patients with pulmonary valve atresia.

It is interesting that surgical or transcatheter relief of critical pulmonary stenosis in infants has been associated with a significant incidence of pulmonary insufficiency. As these children have been followed over time, a small percentage have developed right ventricular dilatation sufficient to require valve insertion in the pulmonary outflow tract. It is interesting that the incidence of RV dilation meeting criteria for valve replacement appears to be lower in these patients than in patients who have undergone tetralogy of Fallot repair and who have similar degrees of pulmonary insufficiency. This may suggest that the infundibular incision and resection of muscle in tetralogy of Fallot patients may aggravate the RV dilation and dysfunction in this subset.

The classification scheme described in this chapter relating the relative size of the right ventricle to the surgical approach taken has several advantages. A more classic technique used by the Congenital Heart Surgeons Society has related tricuspid valve diameter (or *Z*-value) to the optimal technique of repair. Because the tricuspid valve size depends on RV size, these two approaches would seem to be similar. It should also be noted, however, that the tricuspid valve is often abnormal in pulmonary atresia, and therefore, tricuspid valve diameter alone may not accurately reflect tricuspid valve function. Thus, some patients with a moderate-sized right ventricle may be limited in long-term repair by abnormal tricuspid valve inflow and require a Glenn shunt and outflow reconstruction or the Fontan operation. In addition, the compliance of abnormal right ventricles with significant ventricular hypertrophy and small size may limit the utility of complete two-ventricle repair in those with moderate hypoplasia. The use of an adjustable atrial septal defect at complete repair as pioneered by Dr. Laks has permitted right-to-left shunting in those cases in which early ventricular compliance limits forward flow into the pulmonary vascular bed and may help to stabilize these patients in the critical early postoperative period.

Decompression of the right ventricle by tricuspid valve incision remains somewhat controversial. Although in the majority of patients tricuspid valve incision/avulsion is well tolerated, patients who have had valvotomies with associated pulmonary insufficiency may not tolerate decompression of the right ventricle well, because the shunt flow may be directed retrograde across the outflow tract to the right ventricle and then into the right atrium, resulting in a steal of blood away from the pulmonary vascular bed. Whereas decompression of the ventricle may result in resolution of some sinusoidal connections to the coronary arteries, in many cases it is not necessary to decompress the ventricle, and late results have not yet shown a significant effect of decompression on arrhythmias or ventricular dysfunction. As noted in this chapter, decompression of a ventricle that has RV-dependent coronary circulation to greater than one coronary system is contraindicated, and these patients have a significant risk of morbidity and mortality from coronary ischemia regardless of the operative approach taken. Thus, in some cases, early bidirectional Glenn shunting or augmentation of RV flow with an aorta-to-right ventricle connection and an early Fontan procedure may result in improved oxygenation of the

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septal defects, atrial septal defects, and atrioventricular septal defects. Rarely, the coronary sinus may also be unroofed in association with this entity. It is said that arrhythmias, including sinus node dysfunction, atrioventricular block, supraventricular tachycardia, and bundle–branch block, are more common under the circumstance of an absent right SVC. During cardiopulmonary bypass, the isolated left SVC must be cannulated directly for the management of venous return if the atrium must be opened.

Left Superior Caval Vein Draining Directly into the Left Atrium

A left SVC may drain directly into the left atrium. Drainage of the vena cava to the left atrium (either the left or the right SVC) was found in <0.2% of the patients in the CHP Cardiology Patient Database. Forty percent of these were patients with atrial isomerism, and 15% were also associated with an atrioventricular septal defect. When the left SVC connects directly to the left atrium, it is usually in association with abnormalities of lateralization with either right or left atrial isomerism. It is slightly more common for the SVC to drain directly into the left atrium in cases of right isomerism. In addition, the left SVC may drain directly into the left atrium without other serious cardiac malformations, causing right-to-left shunting. When this becomes hemodynamically significant, correction of the shunt may require interatrial baffling of the orifice of the left SVC to the systemic atrium or translocation of the left SVC to the right atrium or right SVC. It should be recalled that, in left isomerism, a solitary left SVC may drain directly to the systemic or right-sided atrium or through the coronary sinus.

Left superior caval drainage into the left-sided atrium must be surgically addressed when the systemic and pulmonary venous circulations are to be separated. This is most commonly encountered during the palliation or repair of various forms of the functionally univentricular heart. Under those circumstances, the left SVC may be directly connected to the left pulmonary artery (left bidirectional Glenn anastomosis) as part of a staged or total cavopulmonary connection. Interatrial tunneling of the venous return from the atrial orifice of the left SVC to the right (systemic) atrium may compromise or complicate the intracardiac repair of many defects, especially atrioventricular septal defects. Removal of the left SVC from the left atrium and direct attachment to the right-sided atrium or right SVC may be more efficacious. Cryopreserved homograft tissue may also be used as a patch or conduit to help achieve this extracardiac correction. Another option, in the presence of normal pulmonary hemodynamics, is to attach the left SVC directly to the left pulmonary artery, even when a biventricular repair is contemplated. This will correct the right-to-left shunt and still provide decompression of the left superior caval system.

Presence of a Levoatrial Cardinal Vein

The levoatrial cardinal vein is occasionally confused with the persistence of the left SVC. However, it differs from a left SVC in several ways. The levoatrial cardinal vein is thought to result from the persistence of anatomic channels that connect the capillary plexus of the embryonic foregut to the cardinal veins. It is usually found with a normally present left innominate vein. It ascends from the left atrium or a confluence of pulmonary veins dorsal to the left pulmonary artery, passing between the left pulmonary artery and the left bronchus (Fig. 76.3). When this vein receives pulmonary venous return, the passage of the levoatrial cardinal vein between these two large and potentially fixed structures causes obstruction to the anomalously draining pulmonary venous return.

The levoatrial cardinal vein is most usually recognized in association with left atrioventricular valve stenosis or atresia and with a restrictive or absent interatrial opening. It may provide the only exit for pulmonary venous blood that arrives through normally connected pulmonary veins into the left-sided atrium in patients with left ventricular inflow obstruction. It provides an exit into the systemic venous circulation. An analysis of the CHP Cardiology Patient Database found that 0.04% of the patients had a levoatrial cardinal vein. Fifty percent of the patients had either right or left isomerism, and a small proportion of the patients had other associated anomalies, including double-outlet right ventricle, tetralogy of Fallot, ventricular septal defect, and atrioventricular septal defect.

The levoatrial cardinal vein has little surgical significance. It does not provide a useful venous pathway beyond the early neonatal period, and even then it is too small or obstructive. However, it must be recognized and closed during corrective surgery that demands a separation of the systemic and pulmonary venous circulations. For example, persistence of this vein following a bidirectional Glenn anastomosis would provide an unacceptable collateral route of decompression for the superior venous circulation.

Right Superior Caval Vein to the Left (Pulmonary) Atrium

A right SVC that connects directly to the left atrium has been reported as an isolated anomaly and may cause a right-to-left shunt of approximately 30%. In this situation, the right SVC passes medially and dorsally to the aortic root, connecting to the cephalic portion of the left atrium. It may receive one or more pulmonary veins (Fig. 76.4).

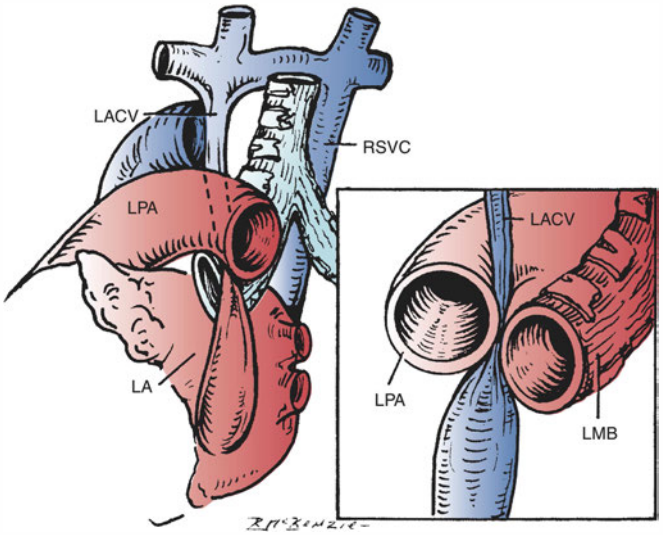


Fig. 76.3. Levoatrial cardinal vein. LA, left atrium; LACV, levoatrial cardinal vein; LMB, left main bronchus; LPA, left pulmonary artery; RSVC, right superior vena cava.

EDITOR'S COMMENTS (*continued*)

coronary flow from the right ventricle. However, these patients continue to have problems with ventricular dysfunction and may have acute myocardial infarction early in life, causing ventricular dysfunction or early death. Thus, in patients with significant ventricular dysfunction in association with RV-dependent coronary circulation, early shunting for stabilization followed by orthotopic cardiac transplantation has been advocated by some centers, and we have performed this procedure in several infants.

For the majority of patients with RV-dependent coronary circulation placement of an aortopulmonary shunt initially followed by bidirectional Glenn shunt and early Fontan procedure seems to be acceptable and the mortality rates do not appear to be higher than the mortality rates with primary cardiac transplantation. Most of the mortality in these patients occurs in the first 3 to 4 months after birth from myocardial ischemia and acute myocardial infarction. Patients who survive to the bidirectional Glenn procedure appear to have relatively good longer term outcomes with the Fontan procedure, and the late mortality rate is relatively low despite the abnormal coronary circulation. The effectiveness of augmenting flow into the right ventricle with aortopulmonary shunts in these patients is not established and often the right ventricle is very small and placement of shunt into the ventricular body difficult. In most cases, we have avoided aortic to right ventricular shunts.

A major area of controversy regarding pulmonary atresia and intact ventricular septum is the use of radiofrequency ablation and perforation of the right ventricular outflow tract with balloon dilation in preference to surgical intervention in these neonates. Although the experience of the Toronto Hospital for Sick Children is relatively good with radiofrequency ablation and perforation of the right ventricular outflow tract with balloon dilation (followed by ductal stenting if necessary to maintain pulmonary blood flow), the disadvantage of this technique is the fact that in patients with severely hypoplastic right ventricular outflow tract, there is no complete relief of the gradient. Residual obstruction may limit right ventricular growth, and a more radical relief of RV outflow tract obstruction and even division of hypertrophied muscle bundles in the cavity of the right ventricle may permit better growth. The technique can be very effective, however, in patients who have a plate-like pulmonary valve atresia with a reasonable-sized pulmonary annulus and relatively well-developed right ventricle. In these cases, balloon dilation while maintaining the patient on prostaglandin and then gradually letting the ductus arteriosus close can permit total interventional repair with closure of the atrial septal defect with an occluder device at a later time. Hybrid therapies, including a Blalock–Taussig shunt and RV outflow reconstruction with later occlusion of the Blalock–Taussig shunt and atrial septum in the catheterization lab, have also been used with good success in selected patients. A comparison of these relatively noninvasive approaches with surgical approaches in all patients with pulmonary atresia and intact septum has not been performed, and therefore the selection criteria for these various interventions are controversial.

The increasingly aggressive use of RV outflow tract ablation and stenting in patients who have moderate tricuspid diameter and RV hypoplasia has dramatically decreased the number of patients who come to surgical intervention for pulmonary atresia with intact ventricular septum. In our center, it is extremely uncommon to be referred to as a patient for RV outflow tract augmentation or for aortopulmonary shunting unless the patient has such a hypoplastic tricuspid valve and right ventricle that eventual Fontan operation is the ultimate goal. To date, there have not been good studies comparing the interventional catheterization approaches for this lesion with conventional surgical therapies. Nevertheless, the majority of patients seem to come to an eventual two ventricle repair after the catheter-based approaches with the occasional need for augmentation of pulmonary blood flow with a shunt or with stenting of the ductus arteriosus.

When outflow tract patches are required, we have elected to use cardiopulmonary bypass in most cases, because it makes the operation simpler. Incision of the RV outflow tract in pulmonary atresia must be done carefully, because even a slight deviation of the incision can cut into the base of the aortic valve leaflet or compromise the left coronary artery. Thus, the use of cardioplegia and cardiopulmonary bypass has resulted in more accurate placement of the incision. In addition, we have elected to place the shunts through a median sternotomy incision rather than a thoracotomy in the majority of patients so that the ductus arteriosus can be ligated at the time of shunt placement. If ductal patency remains, pulmonary overcirculation can occur; therefore, closure of the ductus in the operating room with maintenance of distal saturations is a good sign that the shunt is adequate in size. In addition, competitive flow from the ductus, which may compromise shunt patency in the early postoperative period, is eliminated and retrograde flow into the RV is minimized.

A particularly difficult group of patients are those with pulmonary atresia and Ebstein malformation of the tricuspid valve with severe tricuspid insufficiency. These patients may have significant compromise of the left ventricle by the dilated right ventricle, and the inefficient flow of blood in and out of the right ventricle compromises the overall systemic blood flow. The creation of an aortopulmonary shunt may stabilize pulmonary blood flow in these patients, but the effect of the dilated right ventricle on systemic output remains problematic. Patch closure of the tricuspid valve is not suitable for these patients, because there will be no outflow of the coronary sinus return, and often sinusoidal connections and Thebesian vessels into the ventricle cannot then be decompressed. In these rare instances, the best approach would appear to be early cardiac transplantation. Sano and his colleagues in Okayama, Japan, have suggested either radical plication or complete excision of the right ventricular free wall (with closure of the tricuspid valve orifice) in patients with Ebstein malformation and pulmonary atresia to eliminate the effect of the right ventricle on left ventricular function. These very radical approaches seem to have been successful in a small number of cases and should be considered in patients with pulmonary atresia with

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EDITOR'S COMMENTS (*continued*)

severe Ebstein malformation because patch closure of the tricuspid valve will, as mentioned, often not relieve the dilation of the right ventricle and the secondary effects on left ventricular function. Patch closure of the TV with a small punch hole in the patch may also be considered.

When valve insertion is required, homograft valves can often be used. If the homograft valve is too long from the pulmonary bifurcation to the normal annulus level, it is possible in most circumstances to use a pulmonary homograft and cut the valve just at the commissural attachments, sewing the distal end of the homograft to the pulmonary bifurcation and sewing the proximal homograft in the RV outflow tract to the infundibular septum. In this situation, the pulmonary valve is positioned more inferiorly in the right ventricle but maintains a normal anatomic outflow tract.

When monocusp outflow tract reconstruction is used, we have elected to use either a PTFE patch with a pericardial monocusp fashioned in the operating room or homograft tissue with a contained monocusp, PTFE pericardial membrane, or native pericardium. For Fontan operations, we have preferred the use of the fenestrated lateral tunnel or extracardiac conduit Fontan with a fixed 4-mm fenestration rather than the adjustable ASD used by the authors. In most cases, the fenestration will close spontaneously. The approach to the Fontan operation and hemi-Fontan versus bidirectional Glenn shunt will be commented upon in a later chapter.

TLS

92 Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collaterals

V. Mohan Reddy and Frank L. Hanley

Pulmonary atresia with ventricular septal defect (VSD) and major aortopulmonary collaterals is a complex lesion in which great morphologic variability exists regarding the sources of pulmonary blood flow. The anatomy of the true central pulmonary arteries is also highly variable ranging from normal in size to complete absence. Major aortopulmonary collateral arteries (MAPCAs), probably derived embryologically from the splanchnic vascular plexus, are extremely variable in their size, number, course, origin, arborization, and histopathologic makeup. A given segment of the lung may be supplied solely from the true pulmonary arteries, solely from the aortopulmonary collaterals, or from both, sometimes with connections between the two sources occurring at central or peripheral points and at single or multiple sites. In contrast, the intracardiac morphology of this lesion is relatively straightforward, often with a single anteriorly malaligned VSD, well-developed right and left ventricles, and normal atrioventricular and ventriculoarterial connections. Very uncommonly MAPCAs are present in a similar manner in other lesions like single ventricles, double outlet right ventricle, and transposition complexes.

The ultimate goal of surgical therapy in this lesion is to reconstruct completely separated, in-series pulmonary and systemic circulations. The traditional surgical management strategy for achieving this goal is to embark on a staged reconstruction to centralize the multifocal pulmonary blood supply, recruiting as many lung segments as possible, and then close the VSD and provide egress from the right ventricle to the "unifocalized" pulmonary arterial system. In the past, this always has required multiple operations.

The most important physiologic factor signifying a favorable outcome for these patients after complete repair is the postrepair peak right ventricular pressure. This should be as low as possible. The peak right ventricular pressure depends greatly on the number of lung segments that are unifocalized and on the status of the pulmonary microvasculature in those segments. Another important factor is that the reconstruction must achieve unobstructed delivery of blood from the right ventricle to the pulmonary microvasculature. A number of impediments to achieving this ideal outcome exist. Lung segments can be lost for several reasons. The natural history of these MAPCAs often follows a course of progressive stenosis and occlusion, sometimes making the segment of lung supplied by that collateral inaccessible at the time of unifocalization. Even if accessible, a long-standing severe stenosis of the collateral can lead to distal arterial hypoplasia and underdevelopment of preacinar and acinar vessels and alveoli. In addition, iatrogenic occlusion can occur when these collaterals are unifocalized in stages using nonviable conduits, sometimes resulting in loss of these segments. Finally, MAPCAs without obstruction can rapidly lead to pulmonary vascular obstructive disease in their supplied segments. Similarly, staged unifocalization necessitating the use of modified Blalock–Taussig or central shunts may result in pulmonary vascular obstructive disease.

THERAPEUTIC GOALS AND PATIENT SELECTION

It seems logical that the longer the microvasculature of a given lung segment is left to the hemodynamic vagaries of a MAPCA, the more likely it is that it will either develop pulmonary vascular obstructive disease or involute. Only the "perfectly stenosed" MAPCA may allow normal distal development. Furthermore, stenoses in MAPCAs are well known to progress with time, suggesting that even a "perfectly stenosed" vessel is not likely to remain that way. Extending this logic, it seems clear that the sooner these hemodynamic vagaries can be removed, the greater is the likelihood that the largest number of healthy lung segments can be incorporated into the unifocalized pulmonary circuit. The pulmonary microvasculature taken in aggregate is healthiest at birth and declines thereafter. From these arguments, it seems intuitive that one-stage complete unifocalization and repair early in life gives the greatest chance of achieving a healthy and complete pulmonary vascular bed.

We have prospectively applied a complex surgical management protocol that reflects these goals and principles. The first priority is to completely unifocalize the pulmonary arterial and MAPCA complex via a median sternotomy incision. This removes the MAPCAs from the abnormal physiologic milieu as early as possible. Whenever possible, we also perform intracardiac repair at the first operation. If simultaneous intracardiac repair is not considered advisable because of the concern about unacceptable peak right ventricular pressures, a polytetrafluoroethylene (PTFE) shunt is constructed between the ascending aorta and the fully reconstructed single-compartment pulmonary arterial system. Based on experience with more than 550 patients, this approach of one-stage complete bilateral unifocalization was performed in about 85% of all patients. In 56%, intracardiac repair was also performed. In 29%, a central shunt was created; intracardiac repair was achieved within 2 years in the great majority of patients in whom shunts were created. The remaining 15% of patients typically fall into two general categories. In one group, centrally confluent true pulmonary arteries were present in association with a relatively complete arborization pattern to most or all lung segments. The source of pulmonary blood flow is via MAPCAs that share dual-supply vascular distribution with the true pulmonary arteries. In this anatomic variant, we construct a neonatal

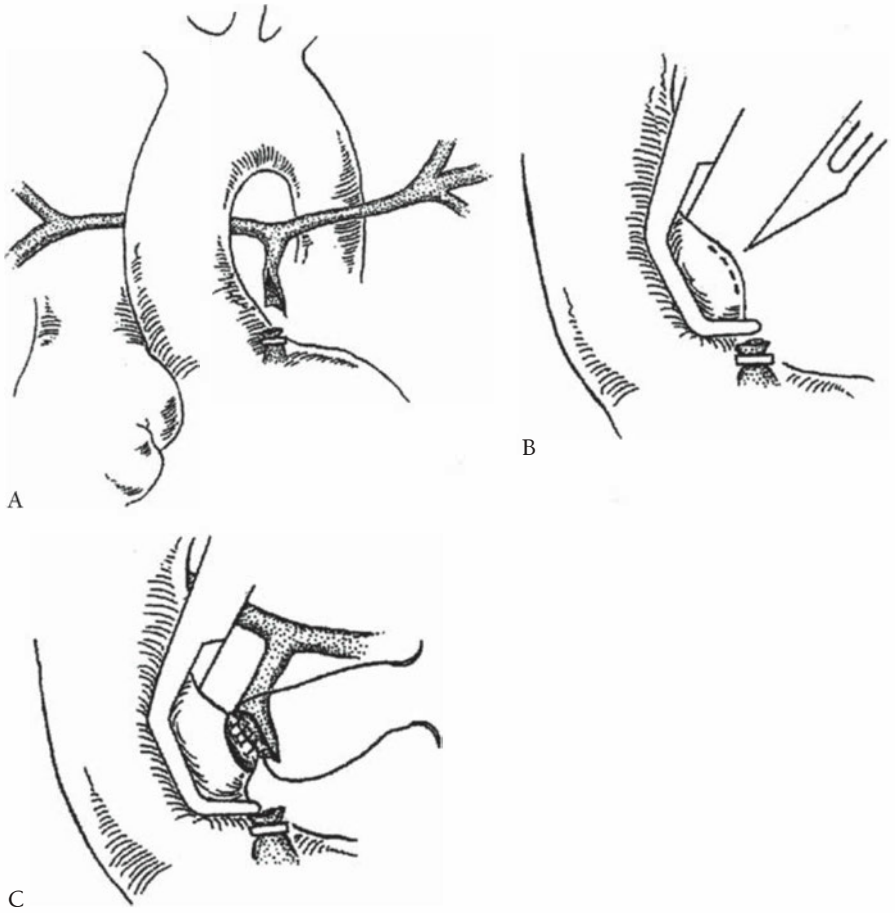


Fig. 92.1. Technique of aortopulmonary window. (**A**) The main pulmonary artery is divided as proximally as possible and spatulated with a longitudinal incision. (**B**) It is important to carefully apply the clamp such that the sequestered portion of the aorta is somewhat posterior to the direct lateral aspect of the aortic circumference. (**C**) Aortotomy is made, and the anastomotic site is enlarged by excising an aortic wall button. Anastomosis is performed with continuous 7-0 monofilament absorbable suture.

aortopulmonary window by removing the blind end of the main pulmonary artery from the infundibulum and anastomose it to the ascending aorta, and we concurrently ligate important MAPCAs (Fig. 92.1). If the patients are selected properly, the central source of pulmonary blood flow will distribute to all lung segments. Patients are then evaluated for intracardiac repair at 3 to 6 months of age. In the second group, true pulmonary arteries may or may not be present. The important factor is that the majority of MAPCAs have multiple stenoses at the segmental and subsegmental branches. In our opinion, these patients are best managed by sequential staged thoracotomies at intervals of 3 to 6 months.

Regardless of which approach is taken, creative surgical techniques are used to achieve native tissue-to-tissue continuity. Allograft patch material is often necessary, but we use it noncircumferentially so that the growth potential of the native tissue is preserved. We limit the use of circumferential nonviable conduits to the central mediastinum.

Ideal age of repair of this lesion based on our extensive experience seems to be between 3 and 6 months in most patients. Our current approach involves the following. If the patient is well balanced physiologically, we prefer to perform this procedure when the patient is between 3 and 6 months of age. However, if the patient is severely cyanotic or has significant pulmonary overcirculation, repair is feasible at an even earlier age. The advantages of early one-stage repair are numerous. Early normalization of cardiovascular physiology and correction of cyanosis is achieved. Protection against pulmonary hypertension, related to high flow either through collaterals or through systemic shunts, is accomplished. The number of operations is reduced. The use of nonviable material in the periphery of the lung is completely eliminated in the great majority of cases. With this approach, the number of patients who can be completely repaired is substantially enhanced.

TECHNIQUE: ONE-STAGE COMPLETE UNIFOCALIZATION AND INTRACARDIAC REPAIR

Surgical access to the mediastinum is via a generous midline incision and a median sternotomy (Fig. 92.2). A subtotal thymectomy is performed and the pericardium is harvested and preserved in glutaraldehyde solution. The primary dissection is in the central mediastinum around the trachebronchila tree, subcarinal space,

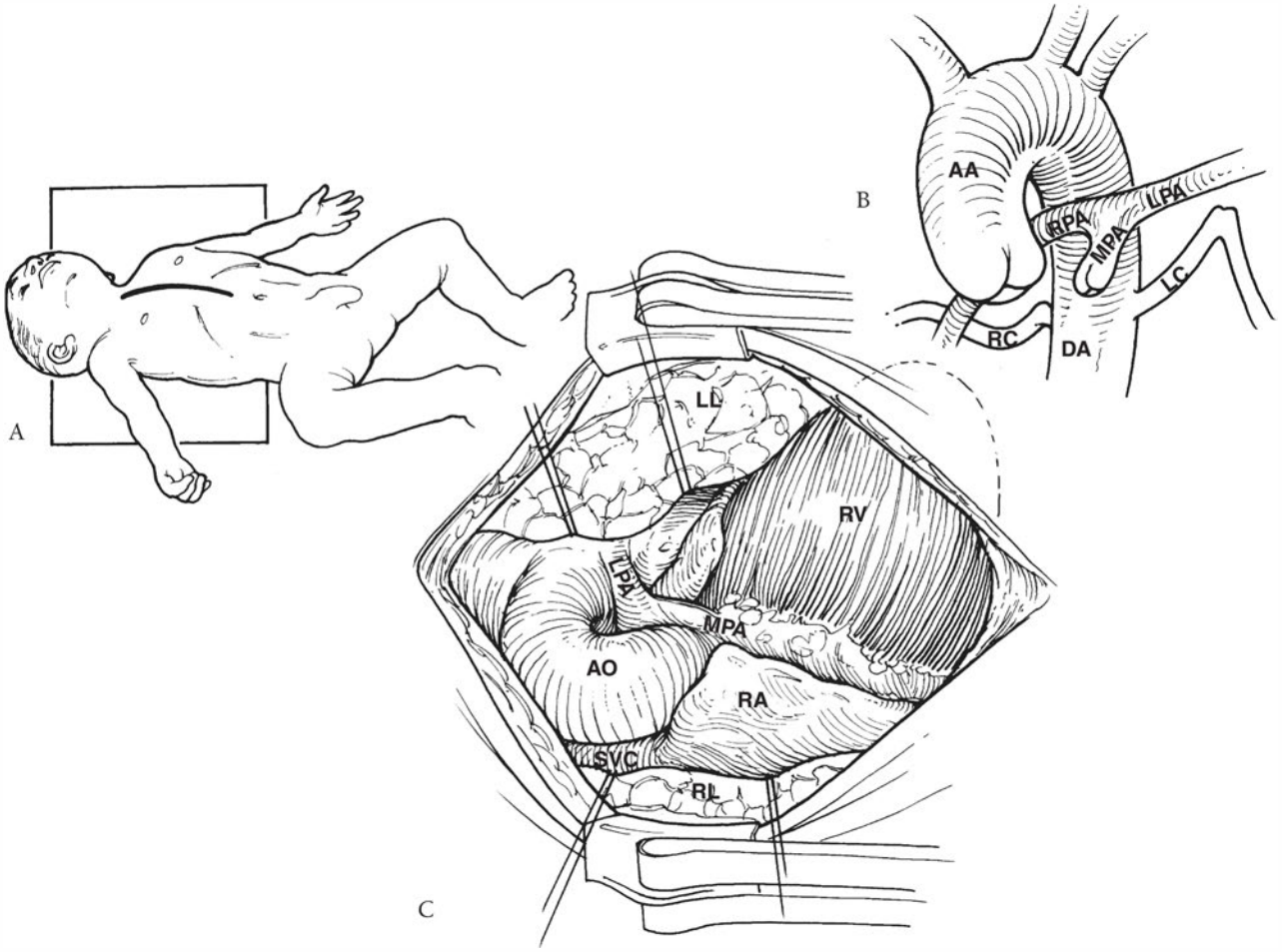


Fig. 92.2. (**A**) An extended midline incision and sternotomy are performed to facilitate generous sternal retraction. (**B**) The pulmonary blood supply. The pulmonary arteries are small: the right pulmonary artery (RPA) supplies only the right lower lobe, and the left pulmonary artery (LPA) supplies only the upper lobe. The collaterals supply the remainder of the lung segments. (**C**) View from the surgeon's side after harvest of the pericardial patch. The pericardial edges are suspended with silk stay sutures and held with hemostats to facilitate hilar dissection. AA, ascending aorta; AO, aorta; DA, descending aorta; LC, left collateral; LL, left lung; MPA, main pulmonary artery; RA, right atrium; RC, right collateral; RL, right lung; RV, right ventricle; SVC, superior vena cava.

and the space between the superior vena cava and the aorta. The native pulmonary arteries, if present, are dissected out (Fig. 92.3A). Most collaterals from the upper descending aorta can be identified and dissected in the subcarinal space (between the tracheobronchial angle and the roof of the left atrium) by an approach between the right superior vena cava and the aorta (Fig. 92.3B). The floor of the pericardial reflection in the transverse sinus is opened, and the posterior mediastinal soft tissues are dissected using cautery. The descending aorta is then exposed in the posterior mediastinum, and all the collaterals from it are identified, dissected, mobilized, and controlled. The subcarinal approach is an important maneuver not only to gain access to collaterals, which typically arise from this location, but also to provide the most direct avenue for collateral rerouting for direct tissue-to-tissue anastomosis, which would otherwise be impossible. In addition, in some cases collaterals arising from the aortic arch or the neck vessels are exposed and dissected in the superior mediastinum along the trachea or through the pleural cavities. Avenues for collateral rerouting are also developed by opening the pleura on both sides posterior to the phrenic nerves in the hilar regions (Fig. 92.4). If the collaterals need only to be ligated and not unifocalized, then the mediastinal dissection can be minimized by the pleural approach. In such cases, the right pleura is widely opened anterior to the phrenic nerve, the right lung is lifted out of the pleural cavity, and the right-sided collaterals are identified and dissected. Similarly, the left pleura is opened and the left-sided collaterals are identified and dissected. The pleural approach is also useful to quickly gain control of all the collaterals in unstable patients before or immediately after cardiopulmonary bypass is instituted.

In stable patients, as many collaterals as possible are permanently ligated at their origin, mobilized, and unifocalized without cardiopulmonary bypass (Fig. 92.3C and 92.3D). When the patient's oxygenation reaches a compromising level, cardiopulmonary bypass is instituted, and the remainder of the collaterals is unifocalized at mild-to-moderate hypothermia with the heart beating. A calcium-supplemented blood prime is used in the cardiopulmonary bypass pump circuit to maintain normal cardiac function. During

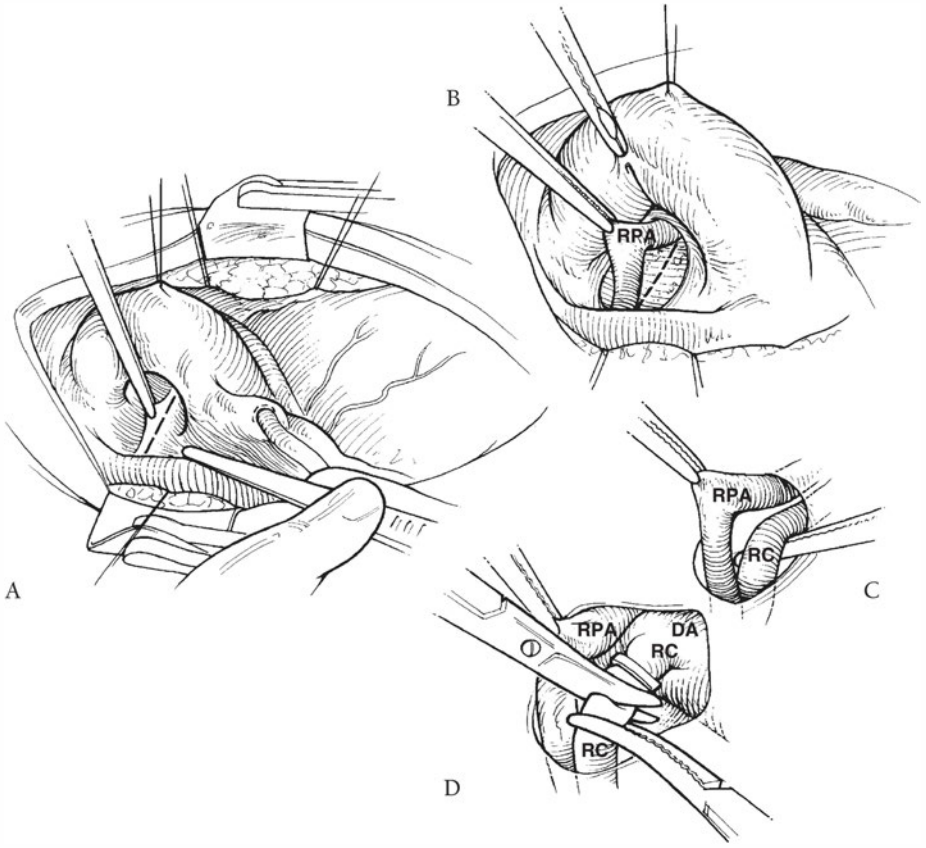


Fig. 92.3. (**A**) The pericardium is incised over the right pulmonary artery. (**B**) The right pulmonary artery (RPA) is mobilized extensively all the way into the hilum. Transverse sinus dissection is performed in the area shown by the dashed line. (**C**) The right collateral (RC) is identified and mobilized. (**D**) The aortic end is occluded with a large hemoclip, and the collateral is transected. As much length of the collateral as possible is taken to facilitate tissue-to-tissue reconstruction of the neopulmonary arteries. DA, descending aorta.

the unifocalization process, the emphasis is on avoiding synthetic or allograft conduits in the periphery and on achieving unifocalization by native tissue-to-tissue anastomosis. One or more of the following techniques of unifocalization are generally used in these patients:

1. Side-to-side anastomosis of the collateral to the central pulmonary arteries, thereby augmenting the hypoplastic central pulmonary arteries
2. Side-to-side anastomosis of collateral to collateral or of collateral to peripheral native pulmonary artery
3. End-to-side anastomosis of collateral to collateral or of collateral to native pulmonary artery
4. End-to-end or end-to-side anastomosis of collateral to central conduit
5. Allograft patch augmentation of stenotic distal segments of the collaterals

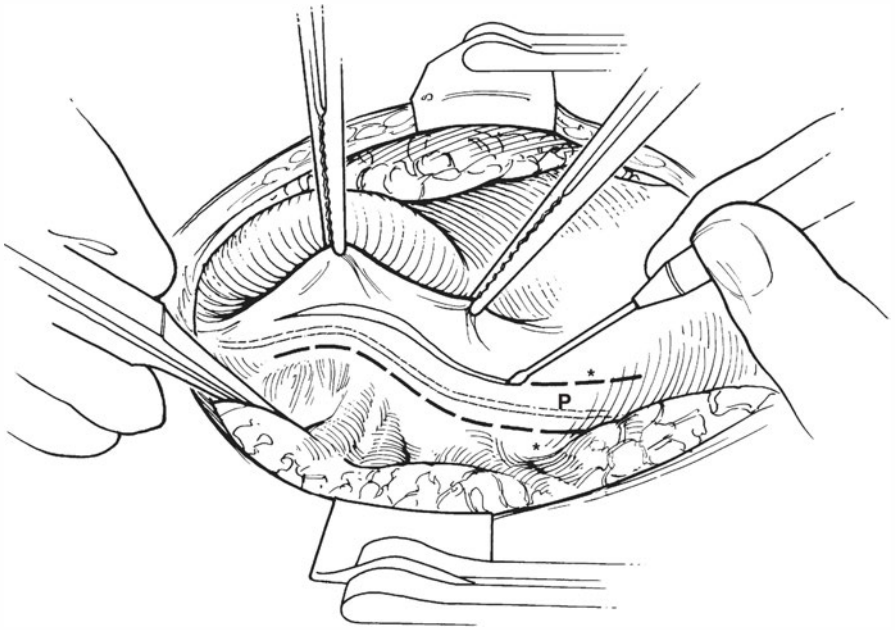


Fig. 92.4. The pericardium and parietal pleura on both sides are incised longitudinally above and below the pericardium (the phrenic nerve is left on a strip of tissue with careful attention not to injure the nerve during the entire procedure). This facilitates hilar dissection for isolating, rerouting, and unifocalizing the collaterals. P, phrenic nerve. * indicates the extent of incision.

6. Allograft patch augmentation of the reconstructed neocentral pulmonary arteries

These anastomoses are achieved directly by bringing collaterals through the transverse sinus or below the lung hilum or occasionally above the hilum, using as much of the collateral length as possible. Collateral length is given the highest priority to achieve tissue-to-tissue anastomosis. For example, if a discrete stenosis is present in the midportion of a collateral, the entire collateral would still be used. The stenosis is managed by side-to-side reconstruction at the necessary level or, if that is not possible, by patching. Some times even collaterals that had a dual supply to a lung segment along with true pulmonary artery supply are unifocalized to build up the size of the reconstructed pulmonary arteries. However, particularly difficult aspects of unifocalization are occasionally completed after cardioplegia is induced and aortic cross-clamping at moderate hypothermia. The important concepts necessary to achieve this type of unifocalization are flexibility regarding reconstruction, aggressive mobilization, maximizing the length of the MAPCAs, and creative rerouting (Figs. 92.5–92.8).

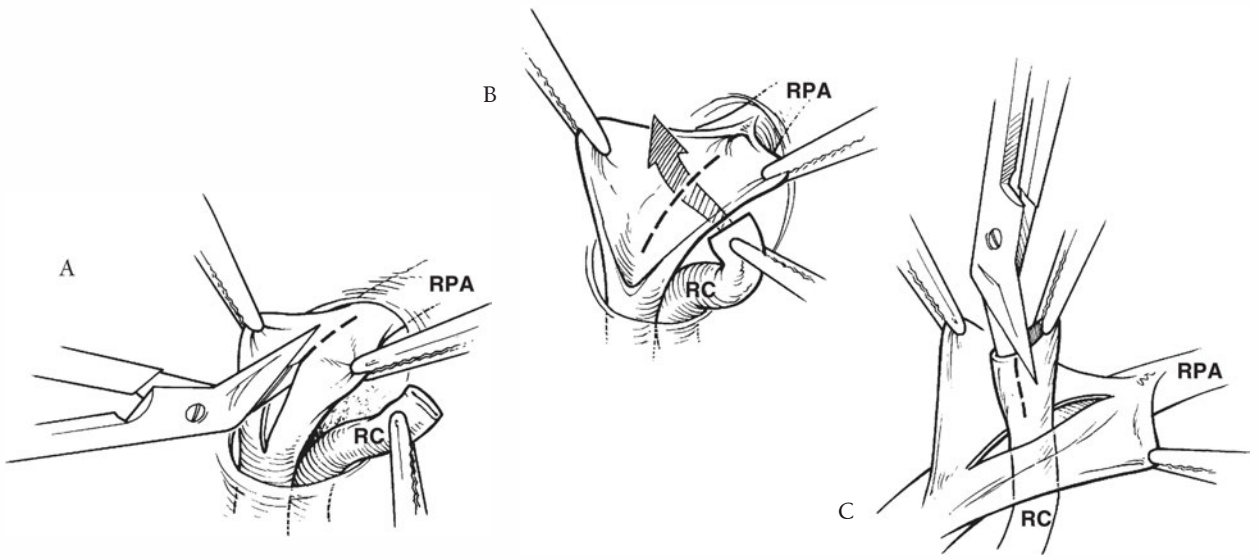


Fig. 92.5. (**A**) The right pulmonary artery (RPA) is opened. This arteriotomy is later extended to augment the RPA. (**B**) An arteriotomy is then made in the posterior wall of the RPA. (**C**) The end of the collateral vessel is splayed open. LPA, left pulmonary artery; MPA, main pulmonary artery; RC, right collateral.

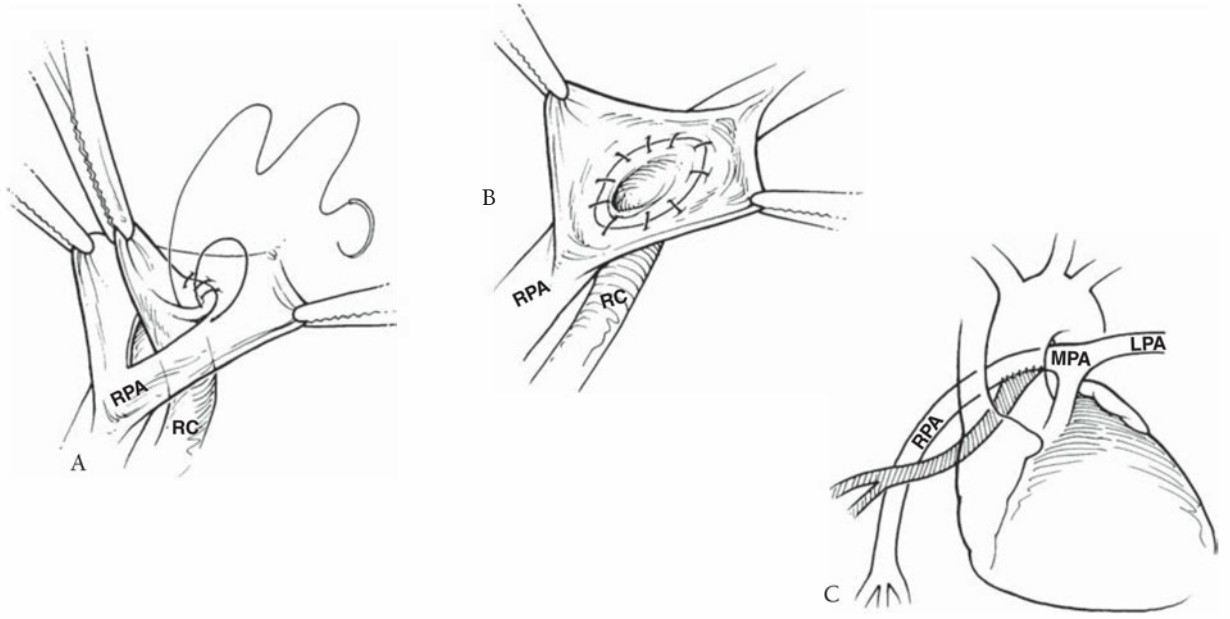


Fig. 92.6. (**A**) The collateral is anastomosed to the right pulmonary artery (RPA) posteriorly using 7-0 absorbable monofilament suture. The site of the anastomosis varies depending on the position and course of the collateral. (**B**) The unifocalized collateral through the RPA. The opening should be unobstructed, wide, and without any kinks. (**C**) The appearance of the right-sided pulmonary arteries after unifocalization. LPA, left pulmonary artery; MPA, main pulmonary artery; RC, right collateral.

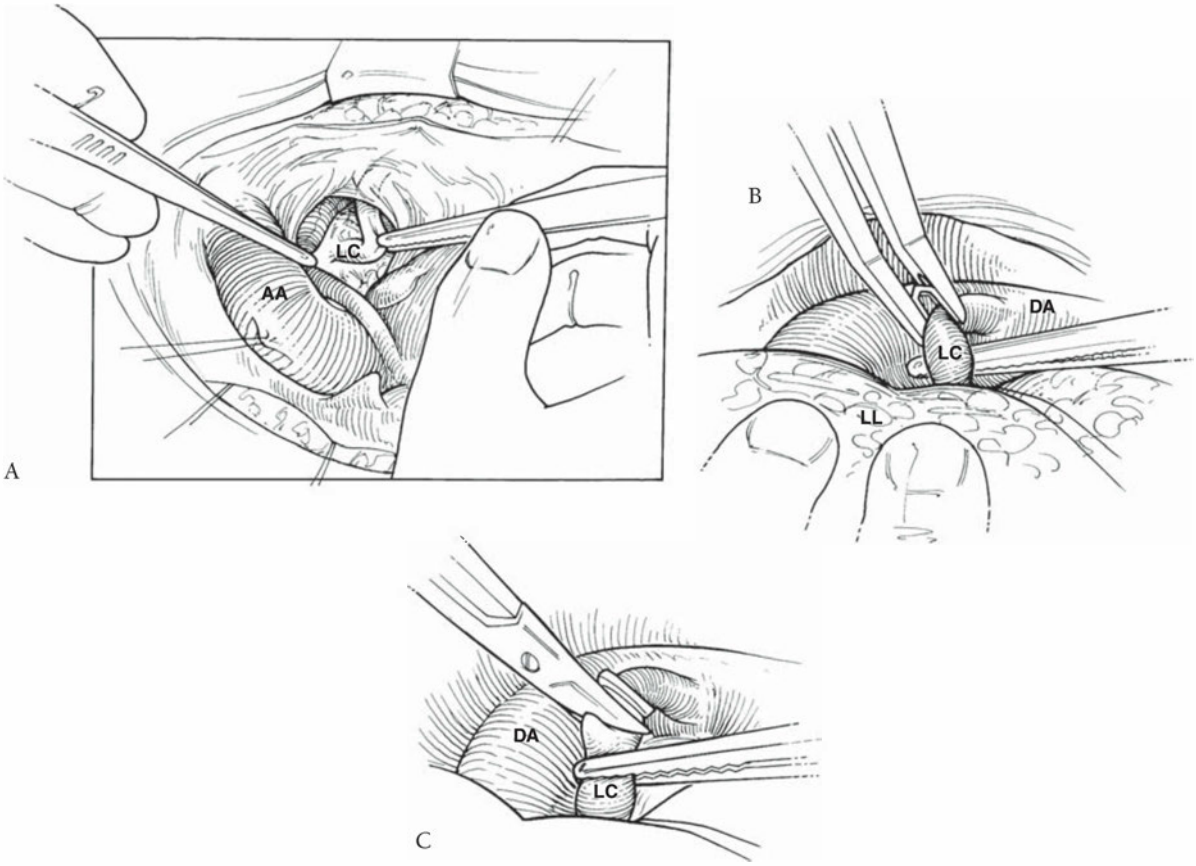


Fig. 92.7. (**A**) The aorta is retracted to the right side. The left pulmonary artery (LPA) is shown being held by the forceps on the left, and the left collateral is shown being held by the forceps on the right. The left collateral (LC) is dissected and mobilized as much as possible in the mediastinum through the transverse sinus. (**B**) The left lung (LL) is retracted out of the pleural cavity and the descending aorta is exposed. The LC is dissected at its origin, mobilized, and occluded with hemoclips close to the origin from the aorta. (**C**) The collateral is further mobilized and transected. AA, ascending aorta; MPA, main pulmonary artery.

After unifocalization is completed, the new single-compartment pulmonary arterial system is assessed for suitability for VSD closure (see section "Criteria for Closure of the Ventricular Septal Defect"). If VSD closure is indicated, a longitudinal ventriculotomy is made in the right ventricular infundibulum and the hypertrophied muscle bundles are resected (Figs. 92.9–92.12). The VSD is closed with a glutaraldehyde-fixed autologous pericardial patch or a polyester (Dacron) patch using continuous nonabsorbable monofilament running suture and sometimes reinforced with interrupted pledgeted mattress sutures. The right atrium is opened to inspect the atrial septum. An atrial septal defect or patent foramen ovale, if present, is partially closed to leave a small unidirectional interatrial communication as a "pop-off" valve for venous blood in case of postoperative right ventricular dysfunction. In some cases with intact atrial septum, a small one-way interatrial communication is created. At this stage, rewarming is started.

An allograft valved conduit is tailored and used to connect the right ventricle to the reconstructed neopulmonary arterial system (Fig. 92.10). The distal conduit is anastomosed to the reconstructed pulmonary arteries. If needed, a distal tongue of tissue is shaped to augment the reconstructed central branch pulmonary arteries.

In a total of >2000 unifocalized collaterals, only a small number have been reconstructed with circumferential nonviable conduit (expanded PTFE; Fig. 92.13). In patients with absent or stringlike true pulmonary arteries, sometimes a second, nonvalved allograft conduit may be necessary to reconstruct the central left and right pulmonary arteries. In such patients, in whom growth potential is an issue, the hilar regions are reconstructed only with native tissue using the techniques described, and this second conduit serves as the main left and right pulmonary arteries only with the conduit limited to the pericardial cavity (Fig. 92.14). In patients with adequate collateral length, the collaterals alone are used to reconstruct the central main, right, and left pulmonary arteries, without the need for a second conduit (Fig. 92.13). The proximal right ventricle-to-conduit anastomosis is performed with a running nonabsorbable monofilament suture. A pressure-monitoring catheter is placed through the right atrium into the right ventricle or pulmonary arteries across the tricuspid valve. The right ventriculotomy is then closed with a pericardial or an allograft patch shaped like a hood, extending from the proximal conduit onto the right ventricle.

After separation from cardiopulmonary bypass, aortic, right ventricular or pulmonary arterial, and atrial pressures are measured continuously. A transesophageal echocardiogram is performed to ensure that there are no significant residual defects.

Bilateral pleural and mediastinal tube drains are placed and the sternum is closed. If bleeding or ventilation is an issue, we electively leave the sternum open and close the chest wound with a silicone rubber

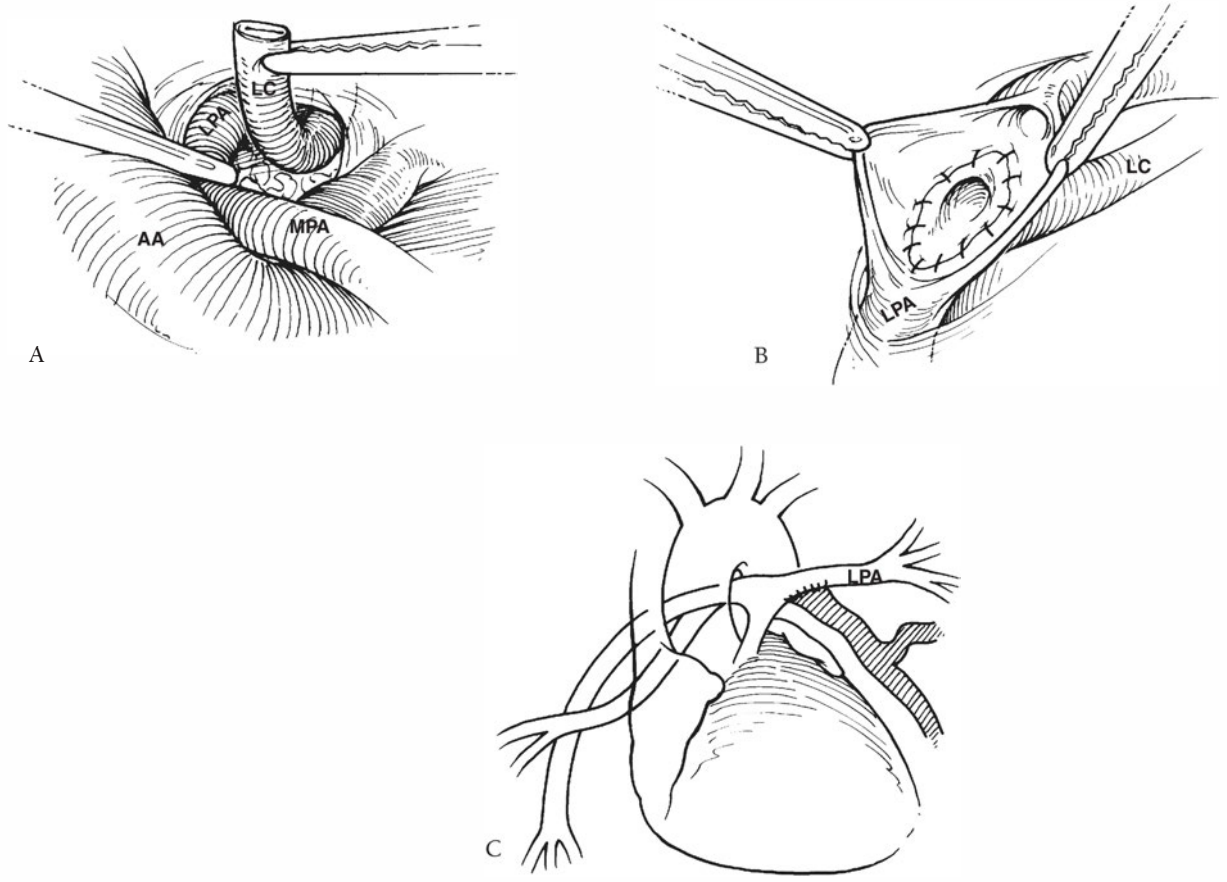


Fig. 92.8. (**A**) The transected left collateral (LC) is routed into the central mediastinum through the transverse sinus. (**B**) The collateral is then unifocalized to the left pulmonary artery (LPA) using the technique described for the right side. (**C**) The appearance of the pulmonary arteries after the unifocalization is completed. AA, ascending aorta; MPA, main pulmonary artery.

(Silastic) patch. Secondary sternal closure is performed on the second or third postoperative day.

CRITERIA FOR CLOSURE OF THE VENTRICULAR SEPTAL DEFECT

Once the complete unifocalization has been performed, while the patient is still on cardiopulmonary bypass, the total resistance of the pulmonary vascular bed is assessed in some patients by an intraoperative pulmonary flow study. The pulmonary vascular bed is cannulated and perfused using a calibrated pump head from the cardiopulmonary bypass machine, with gradually increasing flow leading up to the equivalent of at least 3 L/m^2 cardiac output. At the same time, a pressure catheter is placed in the pulmonary artery system and the left atrium is vigorously vented. If the mean pulmonary artery pressure is $\leq$25 mmHg, the decision is made to close the VSD. If the mean pulmonary pressure is $>$25 mmHg, a central shunt is created.

POSTOPERATIVE MANAGEMENT

The significant postoperative events are phrenic nerve palsy, severe episodic

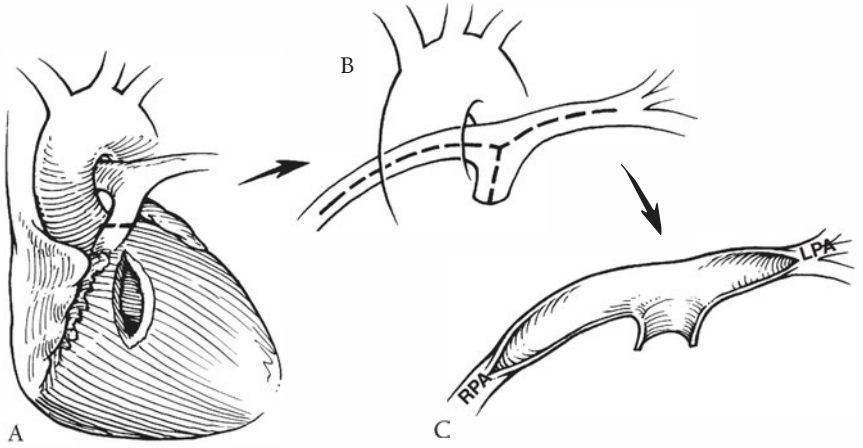


Fig. 92.9. (**A**) The hypoplastic main pulmonary artery is disconnected from the heart and (**B** and **C**) opened into both branch pulmonary arteries all the way into the hilum. LPA, left pulmonary artery; RPA, right pulmonary artery.

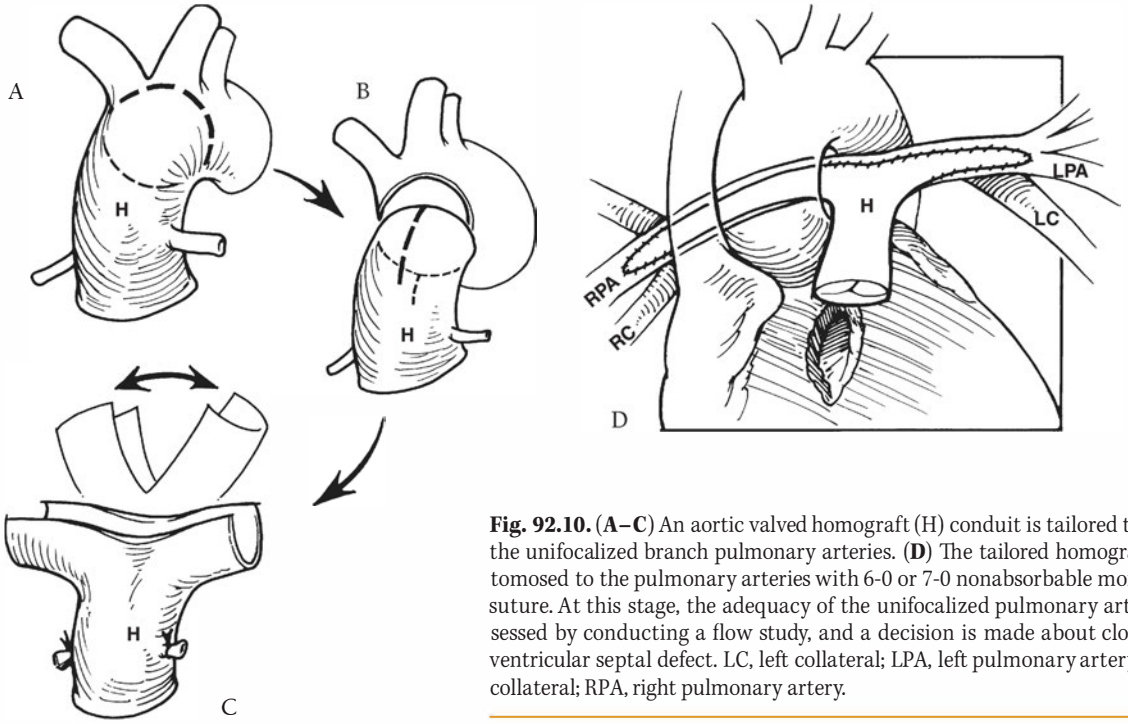


Fig. 92.10. (**A–C**) An aortic valved homograft (H) conduit is tailored to augment the unifocalized branch pulmonary arteries. (**D**) The tailored homograft is anastomosed to the pulmonary arteries with 6-0 or 7-0 nonabsorbable monofilament suture. At this stage, the adequacy of the unifocalized pulmonary arteries is assessed by conducting a flow study, and a decision is made about closure of the ventricular septal defect. LC, left collateral; LPA, left pulmonary artery; RC, right collateral; RPA, right pulmonary artery.

bronchospasm, pulmonary parenchymal reperfusion injury, and pulmonary hemorrhage. Rarely, splanchnic end-organ injury has occurred.

With proper attention to the phrenic nerve, we have minimized this complication. Severe bronchospasm is probably caused by the extensive dissection and disruption of lymphatics and blood vessels around the tracheobronchial tree. It is also possible that the autonomic nerve balance is affected because of the dissection. Another important observation is the development of reperfusion injury of the lung. This is generally limited to the segments that are severely underperfused before unifocalization. The splanchnic end-organ injury is manifested in the form of acute hepatic insufficiency and rarely bowel necrosis. Aggressive monitoring of serum potassium, serum glucose, and hepatic enzyme profiles is strongly recommended. Hyperkalemia and hypoglycemia should be promptly detected and aggressively treated in such cases. The cause of this phenomenon is not completely known, although in the latter half of our series this complication has been extremely rare. We believe that maintaining perfusion pressure >40 mmHg on cardiopulmonary bypass has been an important factor in the elimination of this complication.

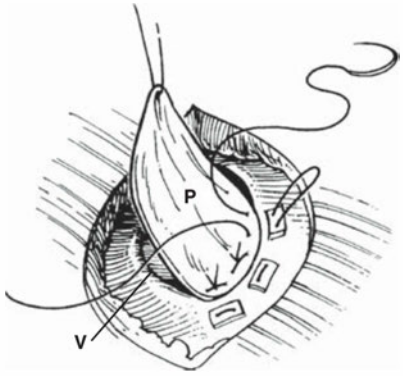


Fig. 92.11. The ventricular septal defect is closed with a glutaraldehyde-fixed pericardial patch (or a polyethylene terephthalate [Dacron] patch; P) and interrupted pledgeted 5-0 or 4-0 braided polyester sutures. V, ventricular septal defect.

FOLLOW-UP MANAGEMENT

All completely repaired patients are followed clinically, and a cardiac catheterization is performed at about 1 year postoperatively or earlier if necessary. In addition, during the first year the patients are followed every 3 months by echocardiography and nuclear pulmonary flow scan to promptly detect and manage alterations in pulmonary blood flow distribution or pulmonary hypertension. In patients where the VSD is left open after one-stage complete unifocalization, a follow-up cardiac catheterization is performed electively at 3 to 6 months postoperatively and assessed for possible VSD closure. Some patients require one or more balloon angioplasty interventions or surgical pulmonary artery rehabilitation before VSD closure. If the ratio of pulmonary to systemic blood flow (Q_p/Q_s) at catheterization is >2:1, the VSD is closed. If the Q_p/Q_s ratio is <2:1, in all patients with a second-stage closure of the VSD, an intraoperative pulmonary flow study is performed to assess the feasibility of VSD closure.

FOLLOW-UP RESULTS

To date approximately 700 operations have been performed in over 500 patients. Early mortality in the last 10 years of our experience is about 2%. However, the late mortality continues to be around 5%. The question always posed is whether the early low RV pressures are maintained during follow-up and whether the collaterals continue to grow. Our experience suggests that the unifocalized collaterals continue to grow and the good early hemodynamics continue to be maintained in the majority of patients. In a recent review of 80 patients who underwent conduit replacement over the last 10-year period, we found that the peak RV-to-peak LV pressure ratio was 0.36 compared with the early postoperative ratio of 0.38. It is true that some of these patients have undergone surgical or catheter intervention to relieve unifocalized pulmonary artery stenosis. But the fact

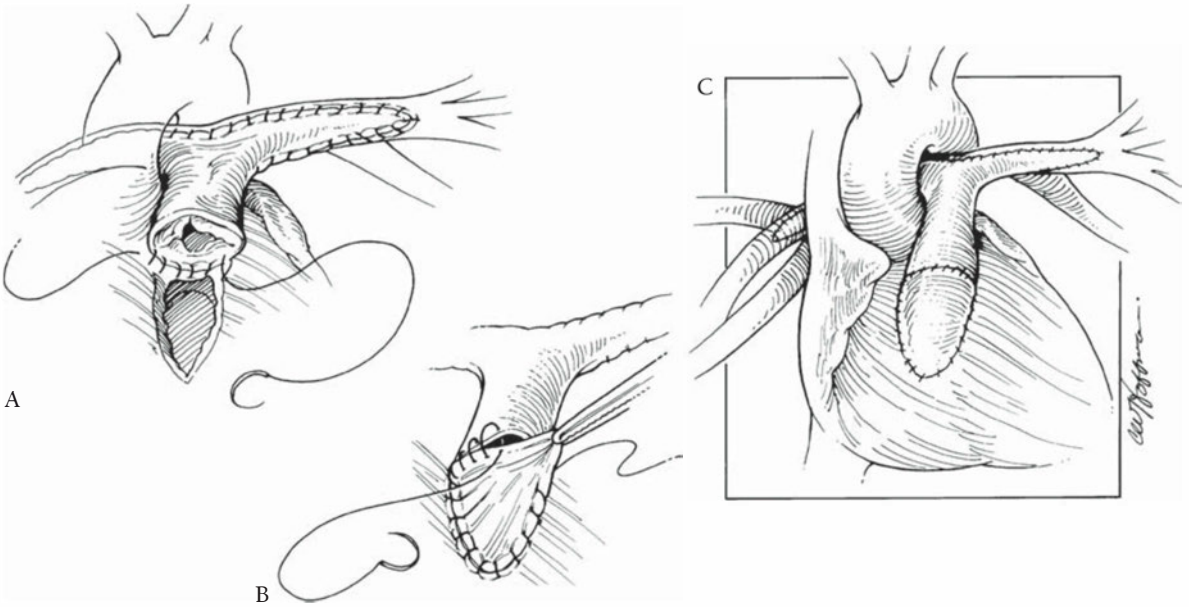


Fig. 92.12. (**A**) The homograft conduit is anastomosed to the right ventricle with 4-0 or 3-0 nonabsorbable monofilament suture. (**B**) The ventriculotomy is closed with a hood of homograft or glutaraldehyde-fixed pericardial patch and 4-0 or 3-0 nonabsorbable monofilament suture. (**C**) The final appearance after complete repair.

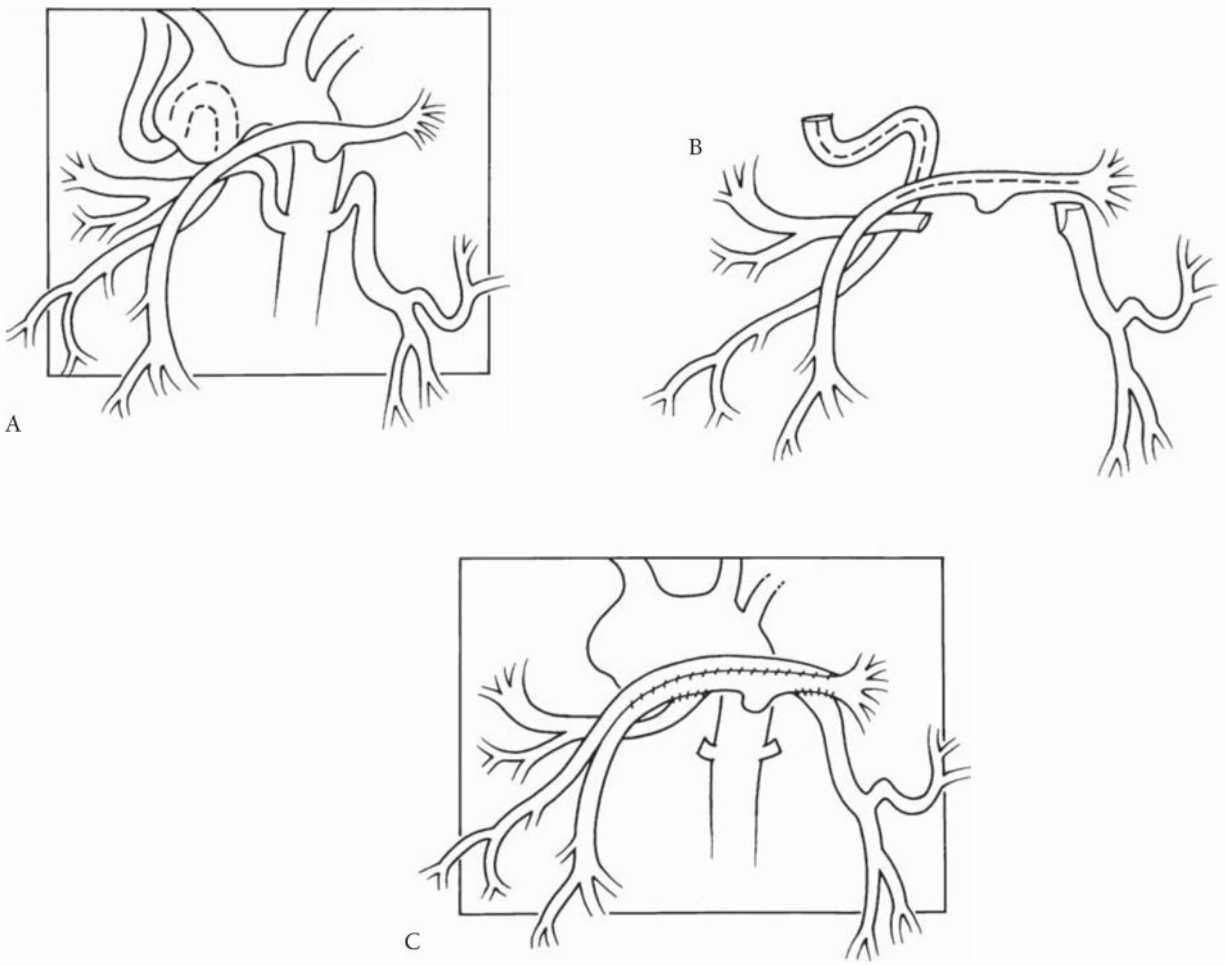


Fig. 92.13. A variation in the technique of reconstruction. (**A**) Figure of MAPCAS drawn from an actual angiogram of a patient. (**B**) Shows the tailored MAPCAS which are to be flayed open along the dashed lines. (**C**) Completed reconstruction with only native tissue. Here, the central neopulmonary arteries are created using only the collaterals and without any homograft patches. In cases in which there are no true pulmonary arteries and the collaterals are of adequate length and size, we reconstruct the central pulmonary arteries with similar techniques using the collateral tissue only.

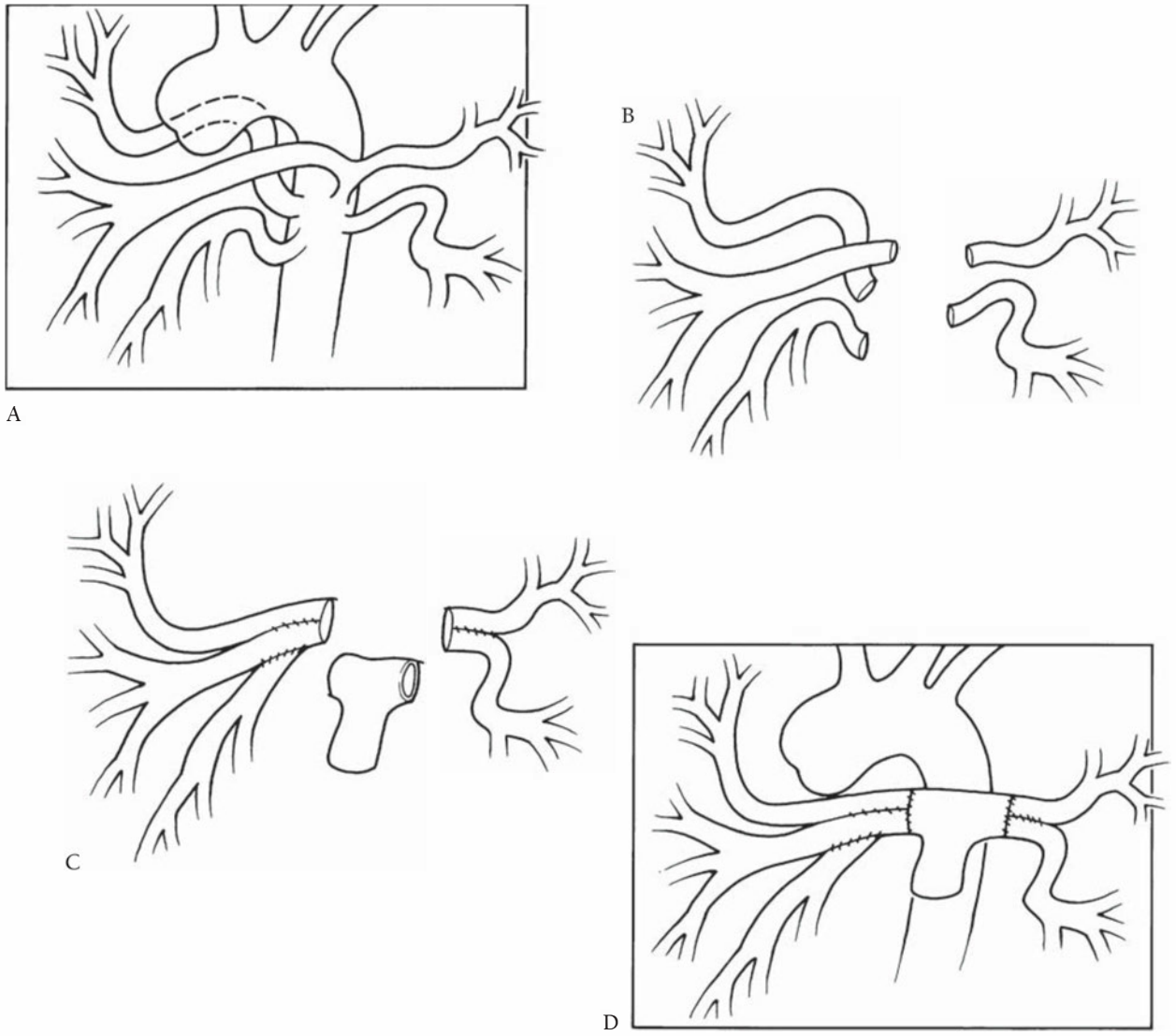


Fig. 92.14. Another technical variation. In this instance, (**A**) there are no true pulmonary arteries. After the (**B** and **C**) collaterals are unifocalized, part of the (**D**) central pulmonary arteries is reconstructed entirely with pulmonary homograft (or sometimes with a second homograft conduit).

that RV pressures remain is an indication that the unifocalized pulmonary arteries continue to grow and the distal pulmonary vasculature is healthy.

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EDITOR'S COMMENTS

Surgical repair of pulmonary atresia with ventricular septal defect and aortopulmonary collateral sources of blood flow is a technical challenge. The aortopulmonary collaterals may supply only a relatively small portion of the total parenchymal volume of the lung or may supply the majority of the pulmonary vascular bed. In addition, some collaterals may be unobstructed, leading to rapid development of pulmonary vascular disease, and others may develop peripheral or proximal stenoses. In the most severe cases, no central pulmonary arteries are present; however, in the majority of cases, there is a very small central pulmonary confluence that may not supply much of the distal parenchyma. Multiple surgical approaches have been applied to this complex congenital heart defect, including staged unifocalizations of the collateral vessels to the lungs by bilateral thoracotomies with pericardial tube reconstruction and unifocalization of the vessels with bilateral shunting and then a repeat operation for central unifocalization and connection to the right ventricular outflow tract, or in cases with diminutive central pulmonary arteries with distal reasonable arborization, connection of the central pulmonary arteries directly to the aorta posteriorly as advocated by Roger Mee.

All of these approaches suffer from the problems of development of distal stenoses and the inability to centrally unifocalize all of the collateral segments at a single setting. In addition, patients who have had bilateral thoracotomies for unifocalization who ultimately fail unifocalization procedures represent a very difficult subset of patients for lung transplantation and cardiac repair at a later time. The severe cyanosis in these patients and the development of significant chest wall collateral vessels have resulted in severe hemorrhage at the time of lung transplant and cardiac repair in our experience and have limited the applicability of lung transplantation in these patients. Therefore, the approach recommended by the authors has appeal because direct intervention to unifocalize all major aortopulmonary collaterals is performed at an early age through a median sternotomy incision, which will, it is hoped, limit the development of aortopulmonary collaterals from the chest wall to the pulmonary parenchyma. Thus, if failure of unifocalization occurs, it may be possible in these patients to undertake lung transplantation with cardiac repair at a later time without the problem of severe hemorrhage from the pleural spaces.

The extensive nature of operations to provide complete unifocalization in infancy is well described in this chapter. The problems with organ system dysfunction from long bypass times for these complex operations and the difficulties in preventing steal from the collateral vessels with initiation of bypass are well described. Nevertheless, in most cases, central unifocalization can be performed with reconstruction to the right ventricular outflow tract or a central aortopulmonary shunt, which has the advantage of providing access to the distal pulmonary vasculature for catheter interventional procedures to deal with distal stenoses.

Despite the outstanding results with surgical treatment of pulmonary atresia, VSD, and MAPCAs presented by Dr. Hanley's group, these continue to be very difficult for patients. Even when excellent pulmonary flow can be achieved with VSD closure and a relatively low ventricular pressure in the operating room, development of distal stenosis and distortions of the pulmonary vascular bed can result in gradual and progressive elevation of pulmonary resistance with right ventricular failure. The long-term outlook of patients who have VSD closure and unifocalization remains to be completely elucidated. Nevertheless, the approaches described by Dr. Hanley and associates have resulted in a VSD closure rate that is higher than has been traditionally seen with other staged surgical approaches, suggesting that early intervention with extensive patching of any areas of stenosis may result in better overall long-term outcomes. Nevertheless, these patients require a great deal of attention and observation for the development of distal stenosis that will require catheter or surgical intervention.

The series of one-stage unifocalization and cardiac repair of Drs. Hanley and Reddy has convinced the community of pediatric cardiothoracic surgeons that this approach is favored to staged reconstruction in virtually all cases. The pioneering work by Dr. Hanley and associates has shown that despite the technical complexity of these operations even in very small infants, unifocalization of multiple aortopulmonary collaterals can be performed with technically excellent results and a long-term outlook that is clearly superior to either staged unifocalization techniques or the overall natural history of progressive cyanosis and ventricular failure seen in patients with this condition.

In patients with inadequate collateral vessels to the lungs, severe progressive cyanosis, and a distal pulmonary vascular bed that is not amenable to unifocalization and reconstruction, we use bilateral lung transplantation and cardiac repair. This procedure is best performed in patients who have not had extensive previous pleural surgery because of the severe adhesions and hemorrhage at reoperation. Hemorrhage has been the primary cause of operative mortality with transplantation in this setting. Thus, avoidance of thoracotomy incisions if at all possible at reconstruction in these patients is preferable if later lung transplantation or heart–lung transplantation is contemplated.

TLS

93 Contemporary Surgical Therapy for Tetralogy of Fallot

Charles D. Fraser, Jr. and Muhammad S. Khan

SECTION I

An Overview

The development of effective treatment strategies for variants of tetralogy of Fallot (TOF) is inextricably linked to modern advances in the surgical treatment of congenital heart disease. In many ways, this lesion should be viewed as the benchmark against which other treatment strategies for congenital cardiac defects are compared. In fact, no other condition offers the scope and breadth of treatment history, particularly if one considers those congenital abnormalities involving biventricular circulation and abnormalities of connection of the right ventricle to the pulmonary arteries. In this regard, it is incumbent upon the current era of cardiac surgeons to pay particularly close attention to the evolution of strategies for surgical treatment of TOF and, very specifically, contemporary outcomes as well as long-term follow-up series. Quite clearly, these data should be used to further refine treatment strategies in the present and future eras.

Prior to the advent of surgical therapy, the natural history of untreated TOF offered a dismal prognosis for patients. In the early part of the 20th century, therapeutic options were meager and included avoidance of stressful situations and, in rare circumstances, phlebotomy for those patients with advanced chronic polycythemia.

Without doubt, the field of palliative congenital heart surgery was ushered in with the advent of the successful application of a surgically created systemic to pulmonary artery shunt (Fig. 93.1). The Blalock shunt was enormously successful in augmenting pulmonary blood flow, bringing significant symptomatic, and in some cases durable relief to the chronic sequelae of cyanosis (Fig. 93.2). This was an enormous conceptual advance in surgery and arguably should be heralded as transformative in its impact on surgical thinking about developing other strategies to treat congenital heart disease. The much heralded and lingering debate over credit for the development of this operation probably should be put to rest, in which it appears that the historical record is quite clear. During his tenure on the faculty at Vanderbilt University School of Medicine in Nashville, TN, Dr. Alfred Blalock developed a renowned surgical laboratory, primarily involved in the investigation of shock. In that context, Blalock was introduced to a young man who would become part of one of the most productive surgical investigational relationships of that century when he hired Mr. Vivien Thomas to work in his lab. Much has been written about this storied relationship and the reader is referred to the bibliography for further references of the details of the relationship. However, it is important to emphasize that Mr. Thomas was a disadvantaged African-American man, denied access to formal medical education because of economic hardships during the post-Depression era in the southern United States. Through this providential relationship and the recognition of mutual strengths and aspirations, Blalock and Thomas effectively collaborated on a number of landmark investigations. Of special note is work that was published by Blalock in 1933 on the prospect of the creation of a model of pulmonary arterial hypertension in the canine model. The surgical technique involved the division of the left subclavian artery and a direct end-to-end anastomosis into the divided left pulmonary artery. While unsuccessful in creating hypertension in this animal model because of the extremely low pulmonary vascular resistance in the canine, without doubt the technical nuances of the delicate systemic to pulmonary artery anastomosis were worked out in the research laboratory at Vanderbilt University. After being recruited to become the Chairman of Surgery at the Johns Hopkins Hospital, Dr. Blalock collaborated with Dr. Helen Taussig in applying the previously developed surgical technique to the palliation of children with disorders limiting pulmonary blood flow. The much heralded first clinical application and the series thereafter established the Johns Hopkins Hospital as the early epicenter of surgical treatment of congenital heart disease. The shared credit for this successful series of operations rests with Blalock, Taussig, as well as Thomas and the surgical and medical practitioners of the current era should respectfully acknowledge the contributions of all three of these remarkable individuals and their impact on the treatment of children with heart disease.

Emboldened by the early successes with surgical palliation, Drs. John W. Kirklin at the Mayo clinic and C. Walton Lillehei at the University of Minnesota began to develop successful operations to directly correct the intracardiac pathology in TOF. The first series of surgically corrected patients were reported by these two pioneers in the mid-to-late 1950s. While surgical techniques have been improved and perioperative management has afforded patients a very favorable progress in the current era, it must be emphasized that the cohort of these earliest successful surgeries, now being followed for almost 60 years, represents an unusual, perhaps unique, window of observation into the natural history of surgically repaired congenital heart disease. In the context of TOF, this observation cannot be overemphasized and will be the focus of subsequent commentary in this chapter as it relates to timing and approach for surgery.

Anatomy and Physiology

By way of definition, this chapter will be limited to discussion of classic TOF. This lesion, originally described by Étienne-Louis Arthur Fallot in a postmortem series in late 1888, represents the prototypical cyanotic congenital heart disease. As is well articulated by leading cardiac morphologists, including Drs. Stella and Richard Van Praagh at Boston Children's Hospital and Dr. Robert H. Anderson at the Royal Brompton Hospital and the

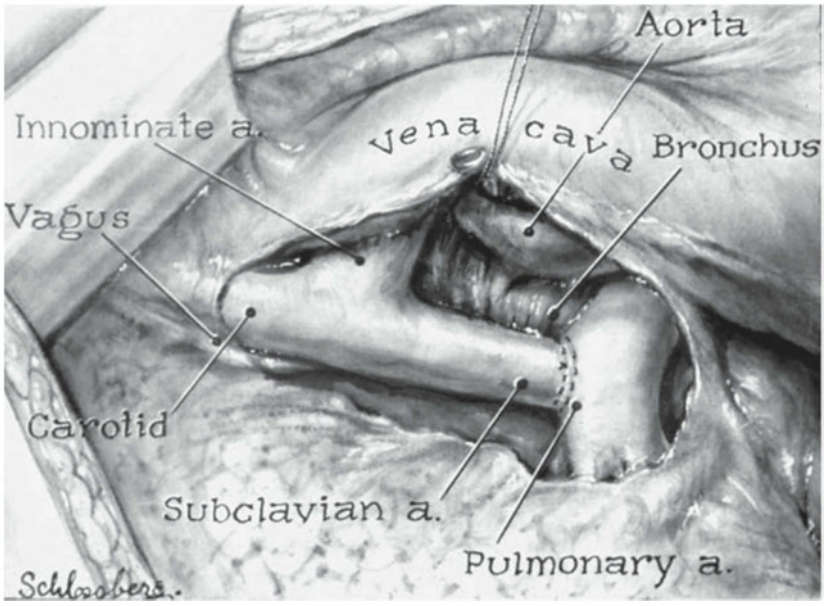


Fig. 93.1. The original illustration of the first BT shunt by Leon Schlossberg. (Image used with permission from The American College of Surgeons.)

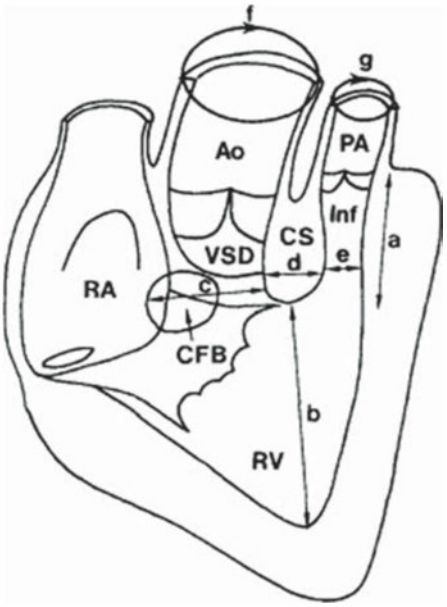


Fig. 93.3. A diagram showing the morphometric measurements of the tetralogy of Fallot heart. RA, right atrium; CFB, central fibrous body; RV, right ventricle; VSD, ventricular septal defect; Ao, aorta; CS, coronal septum; PA, pulmonary artery; Inf, infundibulum. The letters **a** through **g** indicate the dimensions recorded as described in the original article. (Diagrams used with permission from Becker AE, et al. Tetralogy of Fallot: a morphometric and geometry study. Am Card 1975;35:402-412.)

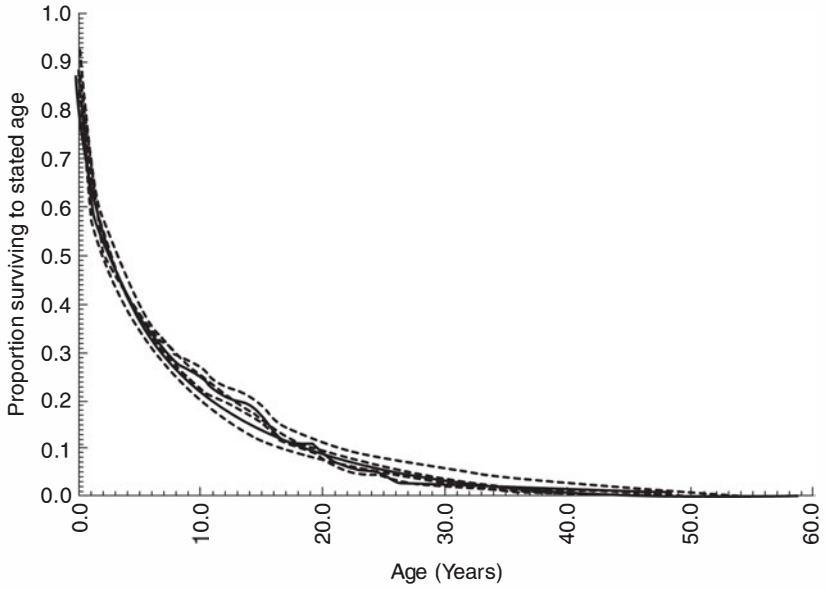


Fig. 93.2. The natural survival for unrepaired tetralogy of Fallot patients. (Reprinted with permission from Bertranou EG, Blackstone EH, Hazelrig JB, et al. Life expectancy without surgery in tetralogy of fallot. Am J Cardiol 1978;42(3):458-466.)

National Heart Trust of the United Kingdom, the central morphologic feature of TOF relates to anterior malalignment of the infundibular septum relative to the conotruncus resulting in a large subaortic ventricular septal defect ("malalignment VSD") and varying degrees of right ventricular outflow tract obstruction (Fig. 93.3). The additional classic elements of TOF include secondary right ventricular hypertrophy and some degree of aortic override or malalignment relative to the interventricular septum. Again, for the purposes of this chapter, comments will be limited to conditions involving balanced ventricles with atrioventricular concordance, malalignment VSD, typically of perimembranous location and pressure nonrestrictive designation, and the presence of prograde pulmonary blood flow into identifiable native branch pulmonary arteries and the absence of major aortopulmonary collateral arteries and significant arborization abnormalities of the lungs which should be discussed in a separate chapter. Mention will be made, however, of patients with TOF with complete atrioventricular septal defect (AVSD) and double outlet right ventricle with subaortic VSD and RVOT obstruction (tetralogy-type double-outlet right ventricle). It is important to note that the degree of aortic override in this chapter will not be used to distinguish between TOF and double-outlet right ventricle, and all patients with the presence of aortomitral fibrous continuity irrespective of the degree of aortic override are to be considered in the spectrum of TOF (Fig. 93.4A, and 93.4B).

Associated Conditions

Conditions frequently found associated with TOF include a persistent left superior vena cava, most commonly to an intact coronary sinus. In some patients, in addition to the outlet VSD there may be additional muscular VSDs. A patent foramen ovale is typically present. The ductal configuration is typically associated with normal ductal insertion, although it is not infrequent to observe ductal entrapment of the proximal left pulmonary artery which may ultimately lead to severe proximal left pulmonary artery stenosis or ductal coarctation.

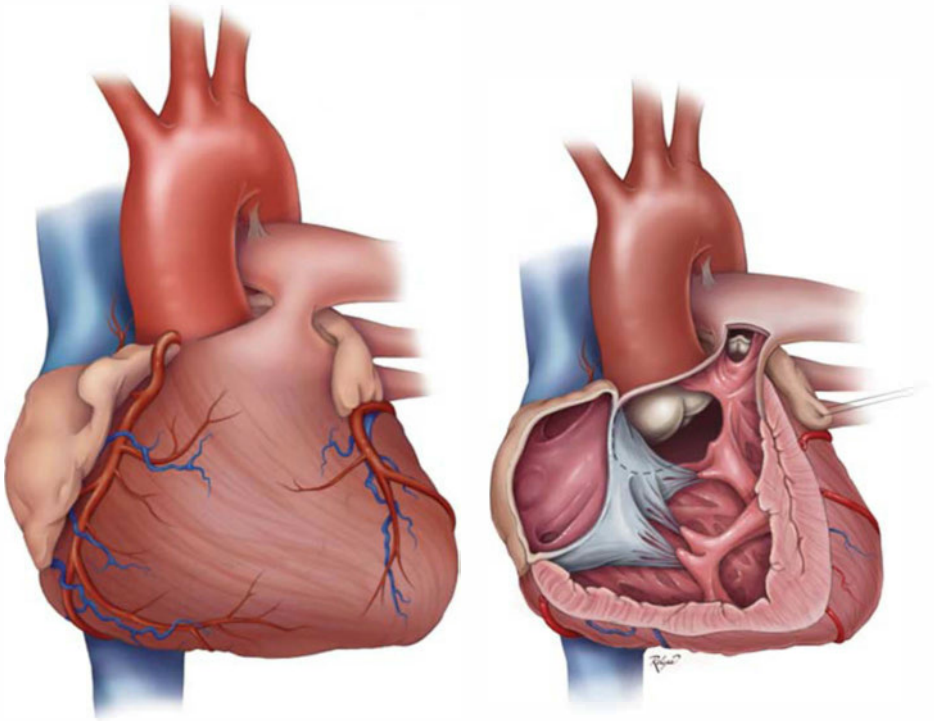


Fig. 93.4. The external (left) and internal (right) anatomy of the heart of a patient with tetralogy of Fallot. (Used with permission from Charles D. Fraser Jr., MD, Congenital Heart Surgery, Texas Children's Hospital, Houston, TX)

A significant anomaly that may definitely affect the surgical approach is found in the presence of anomalous origin of the coronary arteries. The most frequent anomaly includes anomalous origin of the anterior descending coronary artery from the right coronary artery coursing across the subpulmonary infundibulum. Less frequently, a dominant left main coronary ostium gives rise to a large right coronary artery that originates from the left main trunk which travels around the pulmonary infundibulum and across the anterior surface of the RVOT. An associated right aortic arch is found in 25% of patients.

Embryology

For understanding the embryologic development of TOF, we refer the reader to the normal development of the heart. The truncus arteriosus is divided by the aorticopulmonary "spiral" septum that grows down from the midline at 7 to 8 weeks of fetal life to fuse with the ventricular septum. This essentially creates the two great vessels and also isolates the right and left ventricles. In the case of TOF, it is thought that the septum is shifted toward the right leading to three of the four tetralogy defects, that is overriding aorta, pulmonary stenosis, and a malalignment VSD. The fourth defect, that is right ventricular hypertrophy, develops to overcome resistance to blood flow caused by the combination of the earlier mentioned three defects as well as right ventricular pressure load.

TOF is seen in association with a number of chromosomal abnormalities and syndromes. Approximately 15% of patients with TOF have chromosome 22q11.2 deletion syndrome (the presentation of which includes DiGeorge syndrome), whereas 7% of Down's syndrome (trisomy 21) patients have TOF. Other syndromic presentations include Pentalogy of Cantrell also known as thoraco abdominal syndrome, Alagille syndrome, and CHARGE (Coloboma of the eye/central nervous system anomalies, Heart defects, Atresia of choanae, Retardation of growth/development, Genital/urinary defects, Ear anomalies) syndrome. Certain genetic associations have also been found to exist in tetralogy patients that include mutation in *NKX2.5*, *JAGGED1*, *ZFPM2/FOG*, *VEGF*, and *CHD7* genes.

Pathophysiology

The symptomatology of TOF presents a clinical spectrum related to the severity of RVOT obstruction at the infundibular, valvar, supravalvar, or branch pulmonary artery level. Newborns with severe infundibular and valvar obstruction may present with profound cyanosis and ductal-dependent pulmonary blood flow. However, the majority of patients remain relatively asymptomatic in the early period of life. Progressive hypertrophy of the right ventricle and infundibulum participate in a gradual progression toward a degree of right-to-left shunting at the level of the VSD and propensity for desaturation. Frank "tetralogy spells" representing an elevated catecholamine state and reactive infundibular obstruction may be severe but typically are not present until later in infancy. Nonetheless, the natural history in most patients portends a progressive propensity for cyanosis and ultimately, secondary morbid sequelae from that cause. On the other end of the spectrum, however, are the individuals who have anatomy morphologically consistent with TOF with anterior malalignment VSD, but in whom there is little to no obstruction of the pathway of pulmonary blood flow. These patients may present with normal saturations, and in some cases pulmonary overcirculation. These patients with "pink tetralogy" may actually present with signs and symptoms of congestive heart failure relative to a large left-to-right shunt (Fig. 93.5).

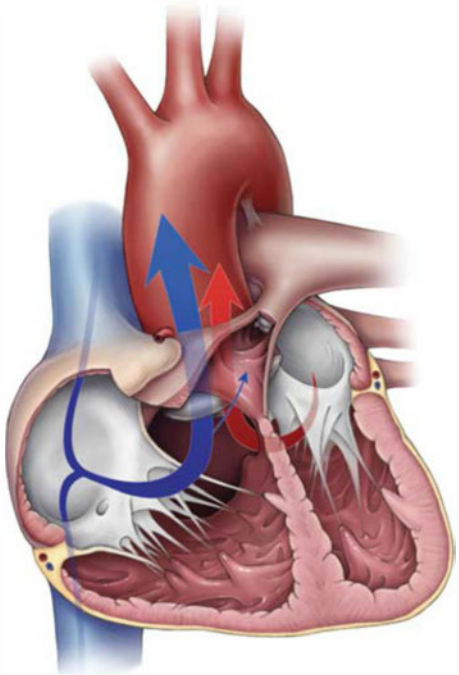


Fig. 93.5. A diagram documenting the directionality of pulmonary blood flow (*blue arrows*) during a tetralogy spell (*red arrow* indicates aortic blood flow). (Used with permission from Charles D. Fraser Jr., MD, Congenital Heart Surgery, Texas Children's Hospital, Houston, TX.)

SECTION II

Diagnostic Evaluation

Physical Examination

The level and degree of obstruction in the RVOT governs the symptomatology as well as the findings on physical examination. Those patients presenting with severe obstruction may have evidence of cyanosis, although this may, at times, be very difficult to discern, particularly in dark-skinned individuals. Additionally, many patients with cyanosis present with a dusky or even pale appearance, and the deep plethora of cyanosis is atypical, particularly in the current era with early intervention. Furthermore, the late sequelae of chronic cyanosis, including finger clubbing and profound nailbed cyanosis are also very uncommon in developed countries with the capability of early diagnosis and treatment of the patients. A point of emphasis merits attention here. In our observation, it is often very difficult for parents and even pediatricians to discern cyanosis on physical exam. Thus, in a conservative management strategy, it is unwise to rely too much on parents' ability to detect desaturation.

Auscultation of patients with severe infundibular obstruction in patients with well-developed branch pulmonary arteries is often consistent with high-velocity flow through those structures, including high-pitched systolic murmur and most often best auscultated at the left sternal border. Paradoxically, patients who present with profound hypercyanosis or frank spells often have diminution or even absence of the murmur as the pulmonary blood flow becomes severely restricted. On the other hand, patients with limited infundibular obstruction will often have very faint murmurs and may, in fact, present with signs of pulmonary overcirculation, including pulmonary crackles and a gallop rhythm.

Chest radiography classically reveals the *coeur en sabot* or boot-shaped heart configuration consistent with a limited pulmonary trunk shadow and a hypertrophied right ventricle and upturned apex. The arch sidedness may be right or left on chest radiography. In case of decreased pulmonary blood flow, the lung fields will show decreased vascular markings (oligemia).

In the current era, the primary diagnostic modality for patients presenting with TOF is transthoracic echocardiography. Important diagnostic features include the presence of prograde pulmonary artery flow, branch pulmonary artery size and distribution, precise delineation of VSD, and delineation of systemic and pulmonary venous connection. In those centers performing primary neonatal tetralogy repair for all patients, irrespective of symptomatology, the presence of an anomalous anterior descending coronary artery may be of significance and at some centers an extensive effort is made to discern this association, although at our hospital this has not been the standard approach. This issue will be expanded upon further later in this chapter.

The majority of patients with TOF do not require diagnostic cardiac catheterization. At our institution, catheterization now has limited application in TOF except in those cases where delineation of the pulmonary artery architecture has not been achieved through other imaging modalities, and it is perceived that this will have some relevance to the surgical timing and approach (see below). The theoretical application of interventional methods in the palliation of symptomatic TOF will be discussed in subsequent sections.

Computed Tomography Scan/Magnetic Resonance Imaging

These imaging modalities are of limited necessity in the primary diagnosis of TOF, although in some circumstances they may be used to ascertain the pulmonary artery architecture and origins and to screen for the presence of significant aortopulmonary collateral arteries.

Timing and Preoperative Care

The subjects of optimal timing and specific surgical technique remain sources of significant dialog and controversy in patients with symptomatic and asymptomatic TOF. It is this author's opinion that an individualized strategy designed to optimize both early cardiac and neurodevelopmental function as well as long-term preservation of cardiac function and anatomy should be central features in any center's perioperative management strategy. It is widely known that the late outcomes of patients with previously repaired TOF are predicated on important factors including tricuspid valve function, rhythm disturbance, right ventricular function, endocardial scarring, pulmonary artery distortion, pulmonary valve insufficiency, and chronic right ventricular pressure–volume overload as well as left ventricular dysfunction. Also of note, there is increasing evidence of late propensity of chronic aortic root dilation, progressive aortic valve insufficiency, and frank aneurysmal degeneration of the ascending aorta. As such, it appears logical that all current treatment strategies should take into account the prospect of affecting these late pathologic sequelae, and every effort should be expended to mitigate these circumstances. This is particularly true of right ventricular function where the timing and mode of therapy as well as the associated surgical "side-effects" must be taken into consideration.

In the current era of pediatric cardiac surgery, the substantial improvements in perioperative mortality rates has allowed for a refined focus on long-term neurodevelopmental outcome, and considerable ongoing effort is being expended in assessing the potential to optimize childhood neurologic potential. In this context, numerous investigators including our own institution have focused efforts on the risk profile for children born with congenital heart disease and whether the timing and mode of cardiac surgery can positively or negatively impact neurodevelopmental potential. Beyond the scope of this chapter, it is important to briefly note that current data would suggest that gestational age, perinatal stress, perinatal cyanosis, and application of cardiopulmonary bypass (including the adjunct of hypothermic circulatory arrest) have potential deleterious consequences on long-term neurodevelopmental outcomes. It is unclear at present whether further strategic refinement of timing and mode of repair can ultimately influence long-term neurodevelopmental potential in these patients, although it is highly intuitive that this is the case. In fact, the early arguments for promoting patients for aggressive primary neonatal repair of TOF were predicated on concerns about early perinatal exposure to cyanosis and the potential deleterious effects of that prospect. In contrast, however, recent evidence has also suggested that prudence is warranted in unnecessarily exposing newborns to the potential negative impact of perinatal stress. This is most markedly evident in the premature brain where numerous studies have demonstrated the vulnerability of the premature oligodendrocytes to stress. While definitive data are lacking and clearly enormous work needs to be done in carefully assessing the variables potentially relevant in brain injury and development, it is reasonable to conclude that surgeons in the current era must carefully consider the risk/benefit ratio of surgical approach, not only in terms of acute mortality but also the effects of neurodevelopmental potential.

Preoperative Preparation

Ductal-dependent newborns with severe RVOT obstruction and near pulmonary atresia being put forward for surgery will be maintained on a prostaglandin (PGE1) infusion until the time of operation. Typically, we favor a small bore percutaneous central catheter to provide durable intravenous access prior to surgery and the PGE_1 is then discontinued at the initiation of cardiopulmonary bypass.

Infants and older children who have demonstrated a propensity toward cyanotic spells are often started on oral beta blockers by the referring cardiologist. In general, we have believed that patients who have been deemed to require beta-blocker therapy should be put forward for operation as soon as possible as hypercyanotic spells are unpredictable in many patients.

For patients in whom beta-blocker therapy has been initiated, there is a propensity toward perioperative sinus tachycardia, and our approach has been to continue the oral beta-blocker therapy up to the morning of surgery. These patients are also typically started on an infusion of short-acting beta blockers (Esmolol) prior to weaning from cardiopulmonary bypass. Frequently, during the preparatory stages there is a tendency for these patients to develop some degree of hyperthermia. This is, in general, not a favorable situation, and a mild degree of ambient hypothermia may be helpful in reducing the propensity for tachycardia and a catecholamine surge. In this setting, the anesthesiologist should work very closely with the surgeon to derive a management plan that is designed to limit the potential for hypercyanosis prior to the surgical correction. This may include oral or intravenous hydration, and as mentioned previously, intravenous beta-blocker therapy. Shunt-dependent newborns or infants referred for surgery should be admitted to hospital the night before operation and intravenous access should provide an opportunity for ongoing hydration as hypovolemia prior to operation may induce a cascade of problems, including acute shunt occlusion or hypercyanosis.

Newborn vs. Non-Neonatal Infant Repair

Much has been written about the appropriate timing for referring patients for surgery with TOF. In fact, it is believed in many institutions that the standard of care is a strict policy of referral for all patients with this diagnosis for primary newborn repair, irrespective of symptomatology and anatomy. While several notable investigators and institutions have opined that this approach now represents standard of care, this in fact is a highly debatable conclusion. A recent survey of data from the Society of Thoracic Surgeons confirms that the majority of TOF repairs are not performed in the newborn period. Nonetheless, there is much logic in considering primary repair in a newborn patient. Theoretically, this offers the potential to avoid or mitigate the propensity for cyanosis and the deleterious effects of that issue on organ system function, including neurologic development. As noted previously, this issue is not resolved. A compelling argument for promoting patients to surgery at the time of primary diagnosis rests in the implied follow-up responsibility should one not refer a patient at that time. Quite clearly, patients with ductal -dependent circulation require operation, and in later sections of this chapter, we will discuss that issue specifically. Nonetheless, most patients do not present with ductal dependent circulation and may be managed expectantly from a symptomatology viewpoint. It must be emphasized, however, that the management team, which should include a close interaction between the attending cardiologist and cardiac surgeon, must be highly vigilant for any evidence of progress toward cyanosis or spelling. This should also include careful education of the parents for diligent follow-up and timely intervention. As such, in those settings where these elements cannot be assured, and there is some risk of improper follow-up and timely referral for operation, it is entirely logical to promote these patients for surgery as a newborn. Whether or not this represents the best timing for surgery with regard to the long-term function of the right heart remains an issue of debate and should be the focus of ongoing longitudinal patient follow-up and refinement of surgical practice.

Our institutional policy at Texas Children's Hospital since 1995 has been to perform non-neonatal infant repair of TOF except in those newborns who have ductal-dependent circulation or have morphology and symptomatology highly indicative of a propensity toward cyanotic spelling. In the current era, there appears to be no justification in delaying complete repair of TOF beyond infancy, and we typically favor performing this somewhere around 6 months of life (Fig. 93.6A–93.6D).

Late Presentation

While increasingly rare, there are patients who present undiagnosed late in childhood or even in adult life. These patients can, in fact, present significant management challenge. In particular, adults of advanced age with untreated TOF typically have developed extreme right ventricular hypertrophy, making intraoperative access challenging and perioperative management more complex.



POSTOPERATIVE MANAGEMENT

Successful postoperative management of patients undergoing surgical repair of TOF is predicated on a thorough understanding of the nuances of acute postoperative right heart dysfunction. Successful complete repair of TOF involves elements of manipulation of right heart structures, which will be reflected on in more detail at a later point but that may, to a greater or lesser degree, exacerbate propensity for right heart dysfunction. In this regard, any management strategy that is additive to that in a negative sense will adversely impact patient prognosis through diminution of cardiac output and systemic venous congestion. In this regard, many surgeons, particularly those who strictly perform neonatal repair, will often leave a small patent foramen ovale or atrial septal defect to mitigate the effects of right heart distention and dysfunction. In our experience with selective, individualized management, this is not necessary, and we have not favored leaving an atrial level communication.

The management team in the intensive care unit should be vigilant about issues which affect right heart function and the management approach should be adjusted accordingly. In general, we favor an indwelling left atrial catheter as right and left heart filling pressures are often incongruous in the early perioperative period and accurate and effective fluid resuscitation must be evaluated in terms of right and left heart filling. There is a consistent tendency in practitioners less familiar with TOF to excessively volume resuscitate these patients which will exacerbate the propensity for right heart failure, hepatic congestion, pleural effusions, and prolonged recovery.

Inotropic management should be directed toward limiting tachycardia and systemic vasoconstriction. A vicious cycle that can be initiated by an uninformed or inexperienced management team is highly repeatable in patients with TOF. This, of course, is all the more prevalent in patients who have residual lesions, severe right heart dysfunction, tricuspid regurgitation, and dysrhythmia. Such patients may develop a central hyperthermia with

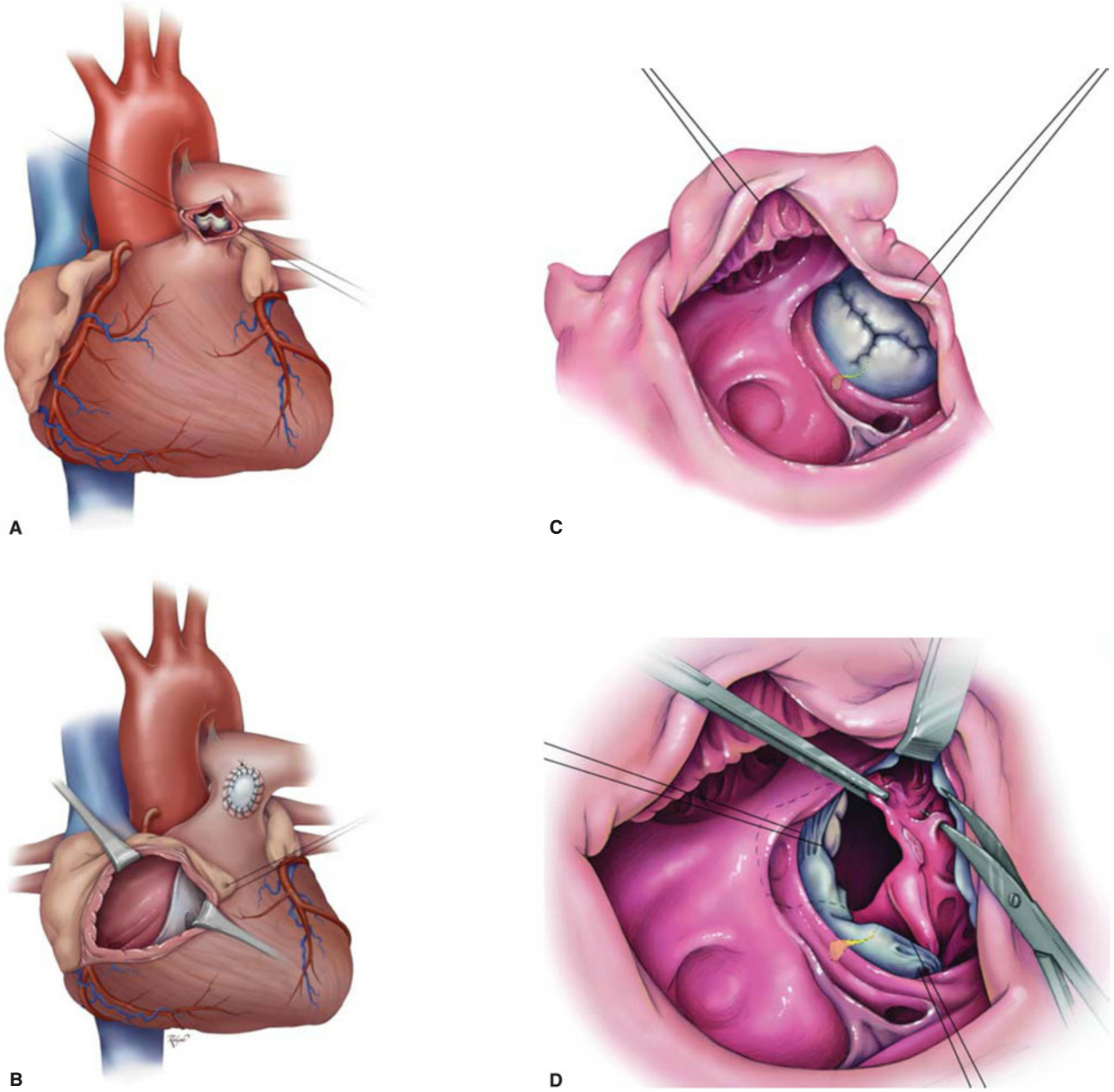


Fig. 93.6. (**A–D**) Illustrations describing the various aspects of transatrial/transpulmonary repair of tetralogy of Fallot. (From Morales DL, Zafar F, Heinle JS, et al. Right ventricular infundibulum sparing (RVIS) tetralogy of Fallot repair: a review of over 300 patients. Ann Surg 2009;4:611-617.)

profound peripheral vasoconstriction. These patients often have a progressive tachycardia and the propensity for junctional ectopic tachycardia (JET) is quite significantly increased (to be discussed further at a later point). In this setting, the tendency is to escalate inotropic support and excessively volume resuscitate the patient. Our view is that every effort should be made to improve peripheral cardiac output and reduce the core hyperthermia. This may be effectively achieved with the addition of intravenous milrinone in addition to other systemic vasodilators. In this setting, we have used low-volume, room temperature peritoneal dialysis as well, usually with a reduction in the heart rate and improved diastolic filling, the cardiac output will improve. The goal is, of course, to avoid this cycle.

Our favored inotropic regimen includes the initiation of intravenous milrinone prior to separation from cardiopulmonary bypass, along with a low-dose esmolol drip (25 to 50 μg/kg/min) to reduce tachycardia. We have tended to avoid intravenous epinephrine as contractility is typically not an issue and we have preferred to avoid the secondary consequences of tachycardia and peripheral vasoconstriction.

Junctional Ectopic Tachycardia

An escalating junctional rhythm (His Bundle tachycardia, JET) can become extremely problematic in children with TOF. This has long been recognized to be the result of mechanical trauma at the time of operation. Quite clearly, the propensity for this rhythm to be induced perioperatively is greatly increased in newborn tetralogy repairs as has been well-documented in

the literature. In our own series of infant tetralogy repair, the incidence of junctional tachycardia is significantly lower and JET, by and large, is an avoidable complication. In those patients in whom the rhythm does become problematic and is not responsive to core cooling, intravenous amiodarone loading has proven quite efficacious.

Complete Atrioventricular Block

In the current era with wide recognition of the location of the conduction system, the incidence of surgically induced atrioventricular block should approach zero. In our own series from Texas Children's Hospital in over 300 patients, no patient has suffered complete heart block and hence, this complication should be avoidable. Right bundle branch block is frequent in TOF patients after surgery and this is typically the result of the location of the suture line for the VSD closure. There are surgeons who favor complete transection of the moderator band; in these instances, there is obligate right bundle branch block, and this has been implied in late functional impairment of the right ventricle. Methods have been described to avoid the right bundle branch at the time of the repair, although this may be quite challenging.

Intraoperative Transesophageal Echocardiography

There appears to be wide variability in the application of intraoperative transesophageal echocardiography (TEE) in the management of patients undergoing surgical repair of TOF. In our own institution, the use of TEE has become standard practice, and we have found it useful for assessing the completeness of the repair, including avoidance of residual VSDs, preservation of the tricuspid valve function, and assessment of the RVOT. There are some surgeons who claim that this is not necessary except in selected cases; however, it is not clear that this is the case. In skilled and experienced hands, the risk profile of performing a transesophageal echo study should be extremely low, and in our view there is no strong reason against performing this study in this group of patients.

SECTION III

Surgical Considerations

The basic surgical elements of complete repair of TOF include correction of the anatomic and functional defects and preservation of overall right ventricular function. Given the wide variety of approaches used by surgeons to accomplish a complete repair, it is quite clear that there is considerable variability in opinion about the optimum surgical approach. Our approach at Texas Children's Hospital has been to utilize a methodology that minimizes trauma and irreparable damage to the right ventricle. However, the long-term superiority of this approach is, as of yet, unproven.

Patients undergoing TOF repair are typically approached through a standard midline sternotomy incision, although some surgeons have favored an inframammary incision for female patients. We have believed that the inframammary approach is an imprudent approach for these children. To our view, it must be acknowledged to parents in the preoperative consultation that all patients undergoing successful TOF repair face a life-long risk of possible surgical reintervention. As such, we have favored a midline sternal incision. Typically, a partial thymectomy is performed to facilitate exposure for cardiopulmonary bypass. We typically harvest a pericardial patch and have favored glutaraldehyde-prepared autologous pericardium for patch material for the surgical repair, although surgeons have reported successful series using a wide variety of materials including polytetrafluoroethylene (PTFE, Gore-Tex®) patch, polyethylene terephthalate (Dacron®) patch, heterologous pericardium and, more recently, decellularized small intestinal submucosa. It appears that there is, at present, no documented superiority of one material over another, and this choice largely appears to be a matter of surgeon preference. We have favored pericardium because of its availability, theoretical resistance to infection, and lack of porosity which eliminates the potential of early perioperative patch shunting as may be seen in patients having Dacron® patch closure. Patients typically undergo standard cardiopulmonary bypass cannulation. We have favored separate vena caval cannulation with angled cannulas of appropriate size. In those patients with a persistent left superior vena cava, we typically will cannulate this directly, although it is also acceptable to place a vent/drain into the superior vena cava via the coronary sinus. We have completely eliminated the use of hypothermic circulatory arrest in our practice of TOF repair. It is still observed that some surgeons favor using DHCA for facilitation of intraoperative exposure, and in those centers performing exclusive newborn TOF repair, the application of hypothermic circulatory arrest is more widely practiced. Myocardial protection in our unit is achieved through multidose, antegrade, crystalloid cardioplegia with intermittent topical hypothermic normal saline. We have favored moderate hypothermia for our cardiopulmonary bypass strategy, typically cooling patients to a nasopharyngeal temperature of 30°C. We have also favored intraoperative physiologic/neurologic assessment including bicortical near-infrared spectroscopy used as an adjunct to optimized cardiopulmonary bypass strategy.

The basic elements of TOF repair include precise closure of the VSD with preservation of the atrioventricular node and the Bundle of His, relief of RVOT obstruction, preservation of pulmonary valve function if possible, relief of pulmonary artery obstructions and closure of the atrial level communication. For this operation, there have been a significant number of variations of surgical technique that have been adopted and promoted. It must be emphasized that at present no surgical technique has been demonstrated to be clearly superior over another, although there is accruing intermediate term data to support the logical conclusion that operations designed to preserve, as much as possible, native right ventricular contractile function and pulmonary valve competence have a lower incidence of need of intermediate-term reoperation. Thereby, these operations would theoretically confer a life-long outcome benefit. The various techniques will be reviewed in detail. It must be acknowledged that it is the author's contention that the transatrial/transpulmonary approach offers a superior methodology, although the pros and cons of this versus other surgical methods will be discussed.

Classic Transventricular Repair

Historically, the majority of patients undergoing complete repair of TOF have undergone a transventricular approach, which is well described in many outstanding textbooks. This operation is approached through a RVOT infundibulotomy. This gives very good access to the VSD, which can be readily patch-closed with either running or interrupted suture technique. This method is applicable to all ages of patients and appears to be the preferred method for those units committed to all neonatal primary tetralogy repairs. The infundibulotomy also provides access to the obstructing RVOT musculature, and where necessary, a transannular pulmonary valvotomy can be extended up onto the hypoplastic main pulmonary artery. Surgeons then have

the option of either primary closure of the RVOT or a patch augmentation of the infundibulotomy and where necessary, the main and branch pulmonary arteries. This is a very straightforward operation, giving the surgeon excellent view of the critical features of the TOF repair, and is the most widely practiced methodology. There are several important points of concern about this approach. First, it is intuitive that relative to the size of the patient and thereby the size of the heart, the infundibulotomy becomes incrementally longer in smaller children. In the author's experience, this has translated into patients presenting late in life after transventricular repair with, essentially, the entire anterior wall of the right ventricle being nonfunctional due to extensive right ventricular incision. Second and by necessity, large coronary arteries traversing the anterior wall of the infundibulum including the first conal branch must be transected. In fact, some surgical textbooks note that this is an important component of the technique. This renders a significant portion of the anterior wall of the right ventricle ischemic (see illustration, Fig. 93.7A, and 93.7B). Next, in the setting of a large infundibulotomy which extends across the pulmonary valve annulus, the patient is exposed to an acute compromise of overall right ventricular forces and, as such, it is widely recognized that in those newborns and small infants undergoing transventricular repair, it is often a necessity to leave an atrial level communication to mitigate the effects of acute right ventricular dysfunction.

It is important to re-emphasize that the transventricular operation represents the "gold standard" complete TOF repair technique against which all other methodologies must be compared. Historically, this is the most widely practiced form of TOF repair, and thereby the outcome information which is progressively accruing concerning the long-term outlook for patients after TOF repair, notably progressive right ventricular dilation/dysfunction and pulmonary valve insufficiency are related to this methodology.

Modified Transventricular Repair

To attempt to ameliorate the effects of pulmonary valve insufficiency on both acute and chronic right ventricular performance after TOF repair, a number of investigators have developed techniques to minimize or eliminate the need of a transannular RVOT incision. Mavroudis and Backer have published an interesting series of patients in whom an infundibulotomy was performed for resection of RVOT muscle and VSD closure, but then the pulmonary annulus was spared and an operation involving enlargement of at least two of the pulmonary valve sinuses as per a technique similar to repair of supravalvar aortic stenosis is accomplished. The authors note that in their series, the incidence of pulmonary transannular incision was much lower than previously reported; however, they have accepted in their patients a much higher right ventricular to systemic postoperative pressure ratio of 0.53 ± 0.13.

A more controversial approach has been recently promoted by Bacha and colleagues originally from Boston Children's Hospital and now more recently from Morgan Stanley Children's Hospital. This involves a longitudinal right ventriculotomy and transventricular closure of the VSD. The pulmonary valve is left intact and an on-table balloon pulmonary valvotomy is performed, essentially fracturing the pulmonary annulus, but with the theoretical benefit of preserving pulmonary valve competence. The early data for this approach are somewhat encouraging, although by definition it will be many years before follow-up data can be used to compare this against other techniques. To this author's viewpoint, a concerning feature of this approach is the very extensive right ventriculotomy which is performed to allow access to the VSD and relief of the RVOT obstruction, and to provide access to the pulmonary valve annulus.

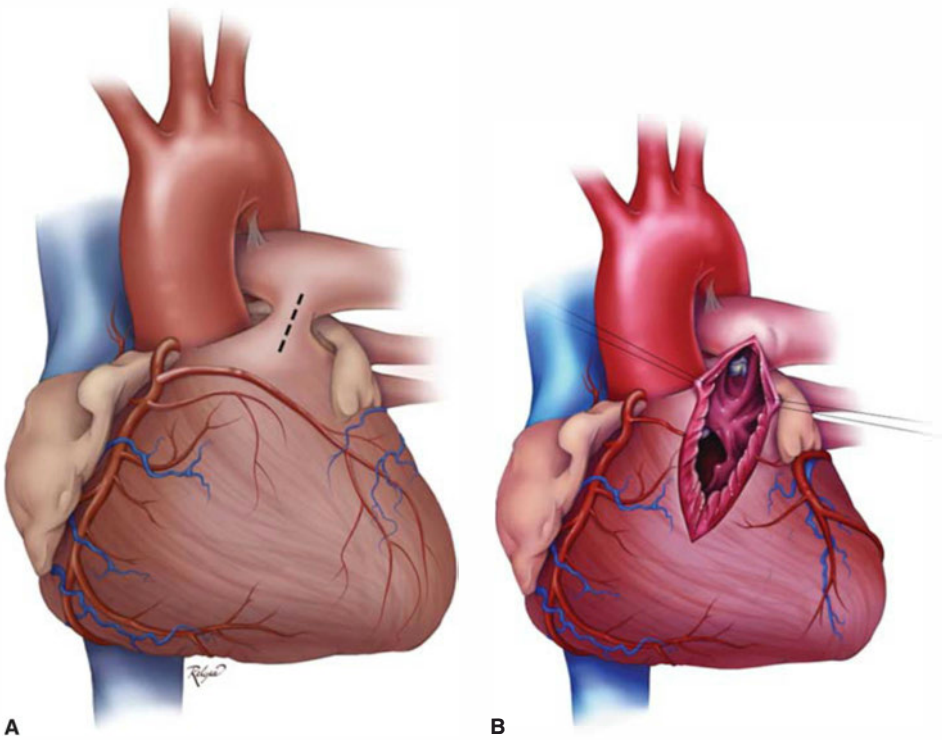


Fig. 93.7. (**A** and **B**) Illustrations showing the accessory anterior descending coronary artery and the transection during transventricular repair. (From Morales DL, Zafar F, Heinle JS, et al. Right ventricular infundibulum sparing (RVIS) tetralogy of Fallot repair: a review of over 300 patients. Ann Surg 2009;4:611-617.)

Transatrial/Transpulmonary Repair

This is the preferred methodology at the author's institution over the last 16 years, and as such will be the most detailed technical description (Fig. 93.6A–93.6D and Table 93.1). The transatrial/transpulmonary approach was originally described by Kawashima and later further championed by Pacifico at the University of Alabama, and then broadly applied by Mee and Karl. The later investigators published the first large series of non-neonatal transatrial/transpulmonary repair of TOF. The central feature of this operation is complete avoidance of a transmural RVOT incision. In fact, in our series of operations from Texas Children's Hospital, a transmural infundibulotomy has been avoided in all but three patients in a series of now approximately 450 cases (Table 93.2).

The transatrial/transpulmonary operation is approached through median sternotomy and full-flow cardiopulmonary

Table 93.1 Experience of TOF Repairs from July 1995 to June 2008 at Texas Children's Hospital

Total TOF repairs	***n* = 304**
Demographics	
Median age	9 mo (2 d–23 y)
Females	132 (43%)
Median weight	8 kg (3–62 kg)
Chromosomal abnormality	38 (12%)
Trisomy 21	19
DiGeorge syndrome	7
Other	12
Prerepair systemic-PA shunt	51 (17)
Operative	
TA/TP repair	301 (99%)
Mini-transannular	222
No ventricular incision	79
Transventricular repair	3 (1%)
Median CPB time	167 min (71–450 min)
Median cross-clamp time	98 min (9–242 min)
DHCA	2 (<1%)
Postoperative	
Arrhythmias	10 (3%)
Heart block	0
Junctional ectopic tachycardia	7 (2%)
NS ventricular tachycardia	2 (<1%)
Fascicular tachycardia	1 (<1%)
Bleeding requiring reoperation	7 (2%)
Renal failure requiring dialysis	3 (1%)
Neurological injury by imaging or clinical assessment	2 (<1%)
Median ICU stay	3 d (1–90 d)
Median hospital stay	7 d (3–256 d)
30 day hospital survival	99.7%
Hospital survival for TA/TP repair	100%
Follow-up	
Survival	
1 year	97%
7 years	96%
Freedom from reoperation	
1 year	98%
7 years	96%
Arrhythmias at 7 y (*n* = 65)	3%
Normal RV function on echo at 7 y	95%

DHCA, deep hypothermic circulatory arrest; NS, nonsustained; PA, pulmonary artery; RV, right ventricle; TA/TP, transatrial/transpulmonary.
Data published by: Morales et al. Right ventricular infundibulum sparing (RVIS) TOF repair: a review of over 300 patients. Ann Surg 2009;250:611-617.

bypass with mild-to-moderate systemic hypothermia. As stated previously, the central feature of this operation which does govern timing of the surgery is strict avoidance of a transmural infundibulotomy. Several critical observers of the non-neonatal approach to TOF have offered the viewpoint that it is illogical to avoid neonatal TOF repairs, given that it is standard practice to perform repair of truncus arteriosus, and in many centers, TOF with pulmonary atresia in the newborn period. The authors acknowledge this point, but it must be strictly recognized that TOF and the long-term effects of right ventricular dysfunction on morbidity and mortality are the issues of focus. From the standpoint of right ventricular function, the long-term prognosis of patients with surgically repaired truncus arteriosus and TOF with pulmonary atresia cannot possibly be as favorable as TOF. Furthermore, the long-term data are incomparable given the different eras of initial surgical repair. Finally, the presenting pathophysiology in these conditions is much different than most patients with TOF, and thereby arguing for comparing approaches, to our view, has inherent flaws.

The operative surgeon approaches the majority of the transatrial/transpulmonary repairs through a standard right atriotomy incision. We typically place a left atrial vent through either the patent foramen ovale or through a surgically created atrial septal defect. The RVOT is then assessed through the tricuspid valve and the obstructing infundibular musculature is resected with great care being exercised to avoid "button-holing" the right ventricular free wall. The surgeon must also carefully assess the papillary muscle and chordal support of the tricuspid valve, and to avoid significant traction on the atrioventricular node and the bundle of His. Using this approach, the surgeon can effectively perform an adequate infundibular resection up to the level of the pulmonary valve annulus. In very small newborns, this access is extremely limited by stearic hindrance from the operating surgeon's hands and instruments. This is one of the several reasons (to be discussed later) that most patients disqualifies newborns from a strict transatrial/transpulmonary repair strategy. In performing the infundibular resection, the operative surgeon is prudent to be conservative. Particularly in the younger patients, there is ample evidence that after relief of the primary obstruction, the secondary infundibular hypertrophy will regress and, over time, the infundibular

Table 93.2 Comparison of the Two Common Techniques of TOF Repair

Transatrial/Transpulmonary	Transventricular
Age usually >3–4 months	Any age
Applicable for all anatomic variations	Applicable for all anatomic variations (conduit for AADCA)
Smaller field of vision	Larger filed of vision
Difficult to teach	Not as difficult to teach
Two procedures for complete repair if shunted	Single procedure for complete repair
Palliative shunt for selective cases	No palliation required
Limited infundibulotomy	Extended infundibulotomy
Risk of injury to tricuspid valve	Risk of injury to tricuspid valve
Longer optimes	Shorter optimes
No circulatory arrest required	DHCA may be required for smaller patients
No ASD left unrepaired	ASD may be necessary
Improved RV function	High risk of acute RV dysfunction
Mortality <1%	Mortality <1%
Arrythmias <5% (JET)	Arrythmias >5% (JET)
Lower midterm reoperation rate	Higher midterm reoperation rate

AADCA, accessory anterior descending coronary artery; ASD, atrial septal defect; JET, junctional ectopic tachycardia; RV, right ventricle.

musculature becomes less problematic. We have adopted the viewpoint that, "You can't put back what you have taken out," and this has been effective in our experience. Typically, in the relaxed heart, the goal of the infundibular resection is to achieve a RVOT dimension of 2-mm larger than the mean expected pulmonary annular size for the patient. After performing the infundibular resection, attention is then turned to the VSD. VSD repair is addressed through the right atrium, and in all patients the VSD can be reached through the tricuspid valve either with or without a tricuspid valve incision. In our experience, the need for tricuspid valve incision is extremely rare, including those patients with severe aortic override, if one is diligent about the visualization. We have favored interrupted polypropylene sutures, buttressed with Teflon-felt pledgets as the most secure methodology to achieve a complete closure and carefully avoid the conduction system (Fig. 93.8A and 93.8B). A hallmark of an effective TOF repair is a complete VSD closure with no peripatch leaks. Following completion of the VSD closure, in those patients who have an adequate pulmonary valve annulus ascertained by probing through the RVOT, the operation is completed. This includes de-airing of the left heart and primary closure of the atrial septal defect. In those patients in whom the pulmonary valve annulus is grossly abnormal, a longitudinal pulmonary arteriotomy is performed, and where necessary a mini-transannular incision is performed. This may involve the division of the anterior commissure of the pulmonary valve, but the transannular incision is not designed to enlarge the infundibulum. In most patients, the transannular incision only extends through the annulus for a distance of 2 to 5 mm. Having done this, the RVOT is probed again, and if adequate the patient is warmed and the remainder of the operation is accomplished with the heart beating. We make every effort to preserve native pulmonary valve function by conservation of available leaflet tissue. In some settings, this involves enlarging a corresponding sinus, and then the pulmonary arteriotomy is closed, typically with an untreated autologous pericardial patch. In the cases where there is proximal branch pulmonary artery stenosis, we have favored direct autologous pericardial patching during rewarming with the heart beating.

The transatrial/transpulmonary approach is applicable to all anatomic variations, including those patients in whom an anomalous anterior descending coronary artery is present. This technical modification has been well described by us and others and is quite advantageous in comparison to a remote right ventriculotomy which is required for those units in whom a transventricular approach is applied. It must also be noted as an advantage of the transatrial/transpulmonary approach that a right ventricle to pulmonary artery conduit is not necessary in these operations. Furthermore, it is also of theoretic benefit (and this has been opined by several observers) that an additional advantage of this technique is avoidance of injury to any of the major right ventricular coronary arteries. This includes complete avoidance of transecting any of the major conal branches, which is mandatory in the transventricular methodology.

The authors recognize that there are several important limitations to strict adherence to the policy of performing a transatrial/transpulmonary approach. If one is careful in assessing the morphology of the patient's lesion as a newborn, and in particular those patients with severe RVOT obstruction with near pulmonary atresia, strict adherence to the transatrial/transpulmonary policy may necessitate an intervening palliative procedure. This, of course, is the subject of intense debate and a point of criticism of this policy. We believe that judicious use of an intervening palliation to allow the patient to achieve a sufficient size (in our series, typically between 4 and 6 kg depending on morphology) allows a unit to aspire to a program of uniform utilization of the transatrial/transpulmonary approach. There will be more commentary about palliative operations in a subsequent section.

The next significant disadvantage of the transatrial/transpulmonary approach is that it is extremely difficult to teach. The reason for this is quite simple. There is only one person at the operative field who can see the operation, and that is the operating surgeon. The first assistant is severely limited in visualization. In our unit, in teaching the congenital heart surgical fellow, we have typically had that person either scrubbed or unscrubbed, standing immediately behind the operating surgeon, to give the trainee the best view of the specific nuances of the repair, before graduating the trainee on to primary surgeon.

A final observation is that the operative surgeon must be very diligent about limiting the potential for traction injury on the heart in achieving adequate exposure. This risk, of course, is exacerbated in the very small patients where visualization may be challenging. The surgeon should be extremely diligent about avoiding excessive operative traction which can cause significant injury to the tricuspid valve, as well as to the conduction system. In our series,

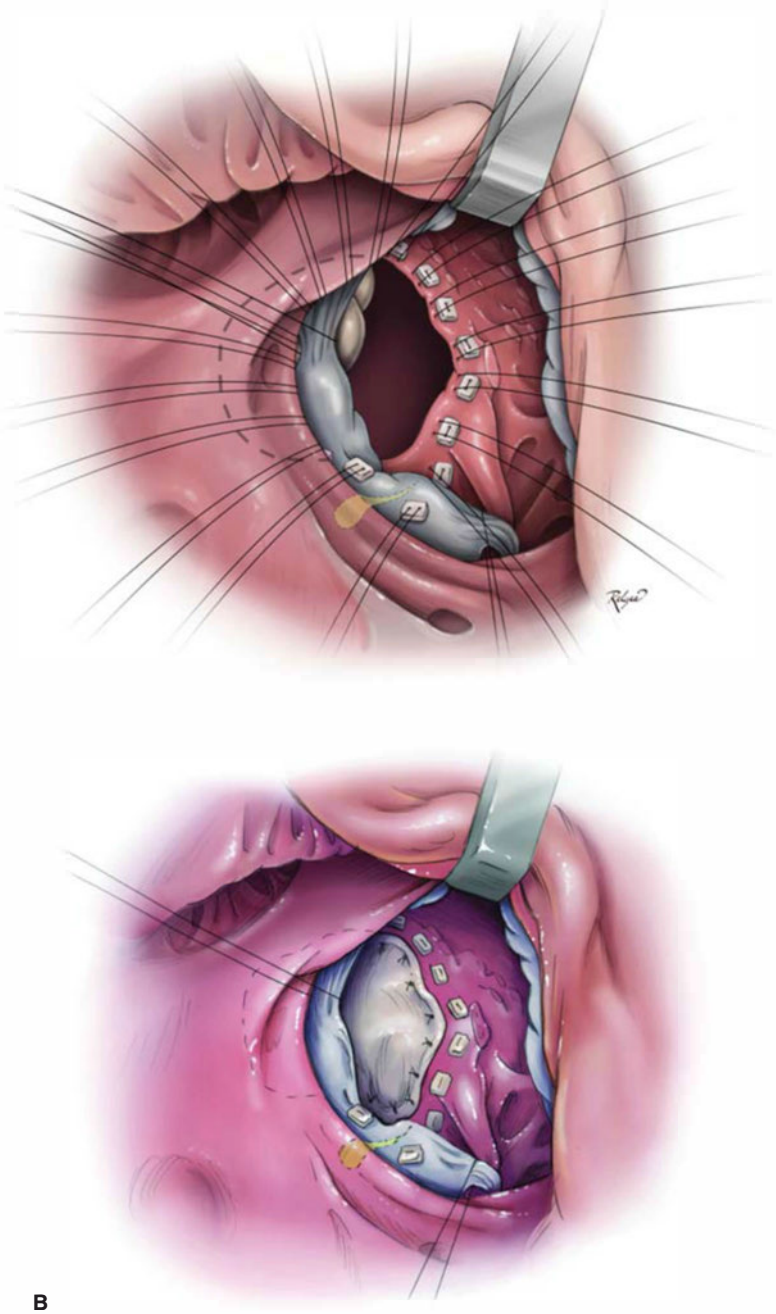


Fig. 93.8. (**A** and **B**) Illustrations describing the ventricular septal defect closure in a patient with tetralogy of Fallot with sutures avoiding the conduction system. (From Morales DL, Zafar F, Heinle JS, et al. Right ventricular infundibulum sparing (RVIS) tetralogy of Fallot repair: a review of over 300 patients. Ann Surg 2009;4:611-617.)

with a very diligent approach to the tenets of the operation, the incidence of complete atrioventricular block has been 0, and the incidence of perioperative JET has been lower than other large reported series (2% vs. 14%).

Finally, there is no question that the transatrial/ transpulmonary approach is technically more demanding than the transventricular approach. Operative times tend to be longer as well as myocardial ischemic times. This does not appear to have translated into any demonstrable disadvantage and the published results have been favorable in comparison to other contemporary series. The unanswered question is whether strict adherence to this approach, and thereby the theoretical improvement of acute and intermediate term right ventricular performance, will confer incremental long-term improvement in outcomes in comparison to other methodologies. Due to limited duration of longitudinal follow-up, definitive data are lacking. However, intermediate data from our own and other contemporary series are encouraging. Even if one considers the additional influence of an intervening palliative operation, for patients enrolled in a strict transatrial/ transpulmonary methodology, the incidence of intermediate-term reoperation is lower than the incidence of reoperation in patients undergoing a strict neonatal approach. This provides inferential evidence confirming that early right ventricular performance is improved. Indeed, Geva and colleagues, as well as other investigators have confirmed the lack of a functional infundibulum in patients after a TOF repair as an important diagnostic feature portending poor long-term right ventricular performance. It is, therefore, logical that any operation that preserves infundibular contribution to cardiac output would translate into a long-term benefit.

Intervening Palliations

Given the current high level of sophistication of perioperative diagnostic imaging, including high-resolution transthoracic echocardiography, an individualized operative strategy can be derived for all patients with TOF. In those limited cases where there is poor delineation of the branch pulmonary architecture, other forms of imaging may be necessary and, depending on the institutional experience and expertise, this may include magnetic resonance imaging, contrast-enhanced computed tomography, or cardiac catheterization.

In patients in whom there is clear ductal dependence of pulmonary blood flow as a newborn, a management algorithm should be derived to optimize long-term right ventricular performance along with acute outcomes. In newborn patients with limited aortic override and limited muscular infundibular obstruction, a transatrial approach can be achieved even in small babies. Typically, those cases are handled as per previous descriptions with either limited or no transannular pulmonary incision depending on morphology. On the other hand, in patients with extreme aortic override and severe infundibular obstruction, the ability to perform a transatrial/transpulmonary approach is compromised in tiny babies. We have, in those patients, continued to advise the use of judicious shunting as an intervening staging palliative operation. In the units that promote patients for an intervening shunt, a strict and careful assessment of outcomes is mandatory. This includes

adherence to diligent follow-up and prompt referral for a complete repair. The palliative operation or intervention is not designed as a durable solution, and as previously mentioned, we typically favor promoting patients for a transatrial/transpulmonary operation somewhere around 6 months. In our unit, we have favored approaching shunt construction through the side of the morphologic right lung. We believe that this is best approached through a right thoracotomy incision, as this offers the operative surgeon the opportunity to place the shunt more peripherally on the subclavian artery, and thereby avoid overshunting and this has worked out quite well for our institution. Of course, other surgeons have recommended sternotomy, and this is certainly acceptable, again with the tenet that this program and strategy is predicated on diligent follow-up and prompt re-referral for definitive repair in infancy.

In some units, including our own, there is growing enthusiasm about the possibility of interventional procedures to stage patients for complete repair. Methods that have been employed include stenting of the ductus arteriosus and stenting of the RVOT. While these are very interesting technical modifications, at present, they cannot be construed as anything other than experimental and these patients should be followed very carefully for deviation from accepted morbidity and mortality data. In the authors' opinion, there are several concerning features of these methodologies. Stenting the RVOT has the inherent challenge of needing to traverse the tricuspid valve in a very small baby with delicate valvar mechanism. In our experience, there have been patients in whom the tricuspid valve mechanism has been distorted or injured during other interventions as newborns including dilation of the pulmonary valve in patients with critical pulmonary stenosis. It is, therefore, entirely logical that this same approach applied to babies with TOF does place the tricuspid valve mechanism at risk, which of course is one of the features of long-term outcome for TOF patients which has to be considered. Ductal stenting in patients with duct-dependent pulmonary blood flow requires discontinuation of PGE_1 and progressive ductal constriction. This places the child at risk of a hypercyanotic event and can be quite a precarious undertaking. Again, the emphasis in performing such a procedure on a child should be that it is largely experimental and very accurate perioperative and longitudinal follow-up data must be collected if this approach is to be employed.

Finally, any unit that is employing a staged management approach to children with TOF has an obligate follow-up responsibility to ensure that these patients undergo timely reintervention. Interstage attrition is unacceptable in this condition, and if there is an issue in that regard, this management strategy is unjustifiable.

Tetralogy of Fallot with Anatomic Pulmonary Atresia

As mentioned in the Introduction, comments concerning this condition will be limited to those patients in whom there are no major arborization defects/major aortopulmonary collateral artery dependent bronchopulmonary segments.

In patients with TOF with pulmonary atresia, our goal has been to accomplish a repair which, as much as possible, achieves the anatomic and functional result that would be aspired to in patients undergoing a transatrial/transpulmonary approach for TOF with pulmonary stenosis. This can be achieved in many cases. We have favored early palliative shunting and infant/non-neonatal referral for an infundibulum sparing operation where the RVOT resection and VSD closure are approached as we would standardly perform for children with TOF with pulmonic stenosis. Typically, a very limited transannular pulmonary infundibular incision can then be augmented with a small pericardial patch avoiding the need of a right ventricle to pulmonary artery conduit. This has allowed many patients to be effectively treated without the obligate need of conduit reoperation in early childhood.

An alternate approach favored in most centers, and in certain settings in our own unit, is a primary newborn repair for this condition. This is quite achievable with good results, but with the premise that any extensive right ventricular incision places the patient at ultimate long-term risk of impaired right ventricular performance as is seen in the longitudinal follow-up of patients after TOF with pulmonary stenosis repair.

Tetralogy of Fallot with Complete Atrioventricular Septal Defect

While beyond the scope of this chapter, some discussion of TOF with complete AVSD should be included here. As a general principle, these repairs are more challenging in patients with TOF with pulmonary stenosis repair for several very obvious reasons. As mentioned previously, the elements affecting perioperative morbidity and mortality in TOF repair are related to overall performance of right ventricular forces. As such, patients with TOF with AVSD are, by definition, at increased risk. First, partitioning of the atrioventricular valve tissue with the goal of achieving two ideal atrioventricular valves is always a challenge in these patients and in patients with a very significant malalignment component where there may be severe aortic override, and the surgeon may be in a position of having to compromise in the partitioning of the atrioventricular valve tissue. Of course, in a patient with a large VSD patch and a marginal right atrioventricular valve, there is considerable potential for compromised right heart performance. In general, we have favored, as with other tetralogy patients, a program of non-neonatal repair to optimize the intracardiac repair and thereby, right ventricular performance. This has included a two-patch technique with avoidance of division of the superior bridging component of the common atrioventricular valve. It is beyond the scope of this chapter to focus on the specific elements of AVSD repair, but noteworthy that any element of distortion of the right atrioventricular valve places patients with tetralogy with AVSD at increased risk.

CONCLUSIONS

In the modern era, the acute postoperative morbidity and mortality after TOF repair should be minimal with many series reporting operative mortality rates of $<1\%$. This, therefore, is the outcome expectation from which all management strategies should be derived. With the acceptance of the necessity of accomplishing complete repair with very low perioperative morbidity or mortality, the focus should be further refined on optimizing long-term right ventricular performance. As such, the operative surgeon is obliged to take into consideration evolving methodologies and accruing data, which allow us to further improve our operative techniques in optimizing these patients' long-term prognosis. In counseling parents and ultimately patients with TOF, the operative surgeon is part of an important management team emphasizing our current optimistic prognosis outlook for these patients; but furthermore, the need for diligent long-term follow-up, and where necessary, timely re-referral for intervention as needed including pulmonary valve replacement which is discussed in other chapters in this text.

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EDITOR'S COMMENTS

The optimal approach to tetralogy of Fallot remains somewhat controversial. Although the number of operative deaths has decreased with tetralogy of Fallot repair and approaches zero in many series, the optimal age at repair is unknown. In centers where neonatal repair is commonplace, operative intervention for complete repair of tetralogy of Fallot can be undertaken at any age. We have elected to perform tetralogy repair at the age of presentation when patients have significant cyanosis. If this approach is used, there is a small subset of patients who present early in neonatal life with significant cyanosis and ductal dependence of pulmonary blood flow. These patients often have the most severe forms of tetralogy of Fallot with significant outflow tract obstruction, small pulmonary arteries, and significant risk for left branch pulmonary artery stenosis after repair. Nevertheless, direct primary repair in these patients may have better results than staged reconstruction with palliative shunting because involution of ductal tissue in these patients often results in pulmonary artery discontinuity when a shunt is placed, and then the advantages of forward flow in the pulmonary arteries may be lost.

Anomalous origin of the left coronary artery has not generally been a contraindication to complete repair in our experience. Short transannular incisions are now the rule and often are able to avoid anomalous coronaries in the right ventricular outflow tract (RVOT). Sometimes a flap of left atrial appendage can be used to cover the coronary artery on the RVOT to maintain an autologous tissue connection to the pulmonary arteries, or in unusual cases the pulmonary artery can be divided and brought down to the ventriculotomy incision directly, bridging the anomalous coronary vessel. It, therefore, is not generally necessary to use a valved conduit in reconstruction in these patients. When neonatal repair is undertaken, we have generally left the foramen ovale open or partially closed the secundum atrial septal defect to allow a pop-off of pressure in the right atrium, which may result from elevation of pulmonary resistance and decreased RV compliance in the newborn. In older patients, we agree with Dr. Fraser that closure of the atrial septal defect should be performed to prevent right-to-left shunting and cyanosis in the early postoperative period due to right ventricular compliance decrease.

The primary controversy in repair of tetralogy of Fallot is the age of the patient at primary repair and the use of transatrial/transpulmonary approaches to complete reconstruction with avoidance of transannular patching. Patients with extensive transannular patches may develop progressive late right ventricular dilation because of progressive pulmonary insufficiency, and these patients have a significant late morbidity and mortality from ventricular arrhythmias and right and ultimately left ventricular dysfunction. Pulmonary valve replacement may be necessary. It is unclear, however, whether pulmonary regurgitation is a result of extensive right ventricular muscle resection or extensive patching as a result of tubular hypoplasia of the RVOT. The exact cause of right ventricular dysfunction and pulmonary insufficiency is not clear. It seems logical, however, to limit the extent of right ventriculotomy and transannular patching to the minimal amount possible to prevent late right ventricular dysfunction. In many cases, even with transannular patching, pulmonary regurgitation is only modest, and right ventricular dilation does not occur. Although it is important to relieve RVOT obstruction if at all possible even at the cost of more extensive transannular patching, the extensive studies of late ventricular function performed by Dr Andrew Reddington and his associates suggests that the characteristics of the right ventricle are more important than the pulmonary insufficiency alone in late function of the right ventricle. These studies suggest that residual stenosis is well tolerated and can limit the magnitude of pulmonary insufficiency with resultant ventricular dilation and late RV failure. Thus, the trend is to leave more residual RV outflow tract obstruction than has typically been the case in the past with the anticipation that there will be a gradual decrease in the ventricular systolic pressure over time and that mild degrees of obstruction (20 to 30 mm) are well tolerated over a lifetime. Thus, minimal transannular patches are becoming more standard and attempts to preserve pulmonary valve function are becoming increasingly desirable.

Nevertheless, the relative advantages of leaving residual obstruction that may in fact progress over time and require reoperation and the problems of placing monocusps or other valve-bearing or valve-creating procedures in the RV outflow tract with the potential need for reoperation make continued follow-up of these patients mandatory. If postoperative residual pulmonary hypertension is anticipated, we have also used monocusp valves in the RVOT to aid in the postoperative hemodynamic stability of these patients. Primary repair in the neonatal or infant period without previous palliative procedures such as systemic-to-pulmonary shunts has been possible in the majority of cases.

Transatrial and transventricular approaches to the ventricular septal defect are both suitable; however, transatrial repair allows the greatest exposure to avoid the conduction tissue. Closure of the ventricular septal defect before muscle resection allows adequate endocardium for anchoring the patch and avoids having to sew the patch for the ventricular septal defect to raw divided muscle, which may not hold sutures well. We have elected to use pulmonary homograft material or PTFE for RVOT reconstruction when necessary. Although nonfixed homologous pericardium can be used. If a small residual ventricular septal defect is present, a jet directed toward the outflow patch can result in aneurysmal dilation and require reoperation. Therefore, if outflow tract reconstruction is necessary, we have generally preferred to use either glutaraldehyde-fixed autologous pericardium or homograft or Gore-Tex material, which has less tendency to stretch.

Patients with particularly difficult situations are those with tetralogy of Fallot and absent pulmonary valve. In these patients, the ductus arteriosus is typically absent and the to-and-fro blood flow in the main pulmonary arteries causes severe central pulmonary artery dilation and bronchial compression. Ventricular septal defect closure and extensive posterior and anterior internal plication of the pulmonary arteries to a normal

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EDITOR'S COMMENTS (*continued*)

diameter with the placement of a valved homograft from the right ventricle to the pulmonary arteries may result in the most optimal repair of this defect. The use of a valved conduit decreases the pulsatility in the central pulmonary arteries and therefore decreases the tendency for bronchial compression. Extensive posterior internal plication of the pulmonary arteries results in a more normal anatomic diameter of the pulmonary arteries and adds substance to the pulmonary artery, which can support the underlying bronchial wall. In spite of these technical modifications, some patients will require long-term ventilatory support because of distal arborization abnormalities and distal tracheomalacia with significant developmental abnormalities of the lung. Nevertheless, we believe that obtaining the most normal hemodynamic and physiologic conditions of the RVOT at the initial operation will maximize early recovery and the ability to wean the patient from ventilatory support. It is not uncommon in these patients to require reoperation for additional pulmonary artery plication in the first 2 to 3 months after initial repair because pulmonary artery size can continue to enlarge even with adequate initial plication and can compromise lung function late after the procedure. There has been increasing interest in the use of a Lecompte maneuver to bring the dilated central pulmonary arteries anterior to the aorta in tetralogy of Fallot with absent pulmonary valve syndrome to avoid bronchial compression. It is not clear how this technique affects the malacia that has been assumed to be present in the bronchi because this would not be addressed by this technique. Nevertheless, the results with this approach seem to be good and can avoid the need for placing a competent valve in the right ventricular outflow tract in patients with respiratory compromise. Presumably also progressive dilation of the pulmonary arteries will not result in increased airway symptoms as has been seen in the more traditional plication approaches. The exact physiology of tetralogy of Fallot with absent pulmonary valve syndrome and dilated pulmonary arteries and the effect of the pulmonary pulsatility on the airways have not been extensively studied. Prone positioning in these patients seems to result in some improvement, even when the VSD is open, suggesting that rather than specific airway compression by the dilated pulmonary arteries, distribution of blood flow in the lungs may be a more important factor.

Repair of tetralogy of Fallot now is associated with mortality approaching zero in many large series from several pediatric cardiovascular surgical centers. The optimal age at repair is unknown; however, we have generally elected to repair these patients early in infancy in the majority of cases. Because symptoms generally occur early in neonates with the most complex forms of tetralogy of Fallot, operative intervention, therefore, must be initiated early in these patients. Asymptomatic neonates with tetralogy of Fallot in our center generally are followed until they are 2 to 4 months of age, at which time elective complete repair can be undertaken with very low morbidity and mortality.

As noted by the authors of a chapter, tetralogy of Fallot has become benchmark against which other treatment strategies for congenital heart disease must be compared. It was one of the first conditions successfully treated by open intracardiac repair and as such one of the defects for which we have the longest follow up. As these patients have aged, the issues related to preservation of ventricular performance and the need for reoperation have become evident. Perhaps, there is no area in congenital heart surgery currently where there is a greater amount of debate and surgical modification. Because of the well-recognized issue of late development of right ventricular dilatation and its affect on exercise performance and late left ventricular function, there has been an increasing interest in trying to preserve the pulmonary valve as much as possible at the initial operation. In spite of the logic that would suggest that preservation of valve function will improve overall right heart function with time, this hypothesis has not been completely studied. It is unclear whether extensive infundibular resection with a transatrial and transpulmonary approach has similar effects on infundibular function as limited infundibular incision and patch augmentation or primary closure. Preservation of epicardial conal branches when intramural coronaries are disrupted by extensive infundibular resection may not in fact preserve overall ventricular performance in the long term. Newer approaches to try to balloon dilate the pulmonary valve through a ventriculotomy or to augment the pulmonary valve to maintain valve function are creative, but there are no data currently that these approaches can result in any better long-term outcome and in the majority of valve-preserving operations, valve function is preserved over a relatively short-term period. Patients who have extensive malalignment of the ventricular septum with tubular hypoplasia of the right ventricular outflow tract and main pulmonary artery will still require more extensive patching to relieve obstruction, and the incidence of reoperation for recurrent right ventricular outflow tract obstruction appears to be greater in series of patients who have had more valve-preserving operations. Thus, tetralogy of Fallot is probably the condition in congenital heart surgery now where there is the greatest amount of controversy and opinion but there is relatively little good data comparing one technique versus another.

Most centers have largely avoided aortopulmonary shunting in patients with tetralogy of Fallot. The indications for aortopulmonary shunts appear to be quite limited. Despite the excellent results with the series from the authors with the use of shunts in newborns, similar long-term outcomes have been reported in centers who have not performed primary shunting. In the majority of centers now the mortality rate for tetralogy of Fallot repair even in infancy approaches 0% to 1%.

There is increasing interest as was described in this chapter, in the use of interventional techniques for palliation in tetralogy of Fallot patients. Stenting of the right ventricular outflow tract and the ductus arteriosus has been recommended by some centers and has been increasing utilized in a few. There are

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EDITOR'S COMMENTS (*continued*)

concerns, however, with the use of these techniques. Stenting of the right ventricular outflow tract in tetralogy of Fallot may allow for improved oxygen saturation and to allow a patient to escape the neonatal period to undergo a repair at a later time in infancy but has the disadvantage of creating a rigidity of the RV outflow tract that may make intraoperative repair quite difficult. I have experienced situations where the stent in the outflow tract has become incorporated enough into the myocardium that it is not been able to be completely removed without significant disruption of underlying structures including portions of the tricuspid valve. If the stent extends down into the ventricle across the infundibulum, then the ability to manipulate the cardiac structures to gain exposure of the VSD through the right atrium can be extremely limited due to the rigidity of the stent fixing the infundibular septum superiorly. With the outstanding results of even neonatal repair of tetralogy of Fallot in the current era, it would seem that the use of these techniques should be limited to patients who have significant contraindications to the primary surgical repair.

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94 Cavopulmonary Shunts and the Hemi-Fontan Operation

Marshall L. Jacobs and Robert D. Stewart

HISTORY AND RATIONALE

The earliest palliative operations that were developed to treat various forms of congenital heart disease include the systemic-to-pulmonary artery shunt, first described by Blalock and Taussig in 1944, and the pulmonary artery band, first described in 1952 by Muller and Damman. In the earliest cases, these palliative procedures were not performed as elements of a planned approach that anticipated eventual definitive reconstruction, but rather in an attempt to prolong survival and improve the functional status of markedly compromised patients with either severe cyanosis or profound congestive heart failure. During the same era of innovation by pioneer cardiovascular surgeons, a third approach to palliation was explored by a number of investigators and surgical teams. This approach involved diverting some or all of the systemic venous return directly to the pulmonary arteries, bypassing the heart. The concept of cavopulmonary anastomosis may have had its origin in early studies of the concept of the "dispensable right ventricle." Rodbard and Wagner in Chicago in 1949 had experimentally anastomosed the right atrial appendage to the pulmonary artery and ligated the main pulmonary artery in dogs, demonstrating the feasibility of excluding the right ventricle from the circulation. The first report of experimental cavopulmonary connection was published in 1950 in Italy by Carlo Carlon of Padua who hypothesized that an "advantage would be received if the blood of the superior vena cava (SVC) should reach the capillary region of the right lung by way of a convenient anastomosis between the great venous trunk and the arterial system of the right lung." Carlon's canine preparation was an end-to-end anastomosis between the proximal end of the divided azygous vein and the right pulmonary artery with pre-atrial ligation of the superior vena cava. In 1951, in the first English language report of these experiments, Carlon and associates wrote, "We are not aware that anyone else has foreseen and studied the problem of oxygenation of the pulmonary blood under venous pressure and without cardiac output." Carlon's first clinical experience was not reported until 1964. In the meantime, Glenn and Patino at Yale published their first report of experimental cavopulmonary shunts in 1954, and in 1955 reported a large study of 59 operated dogs, with six long-term survivors. Glenn's experimental preparations explored the feasibility and physiology of both superior cavopulmonary connections and inferior cavopulmonary connections, noting the occurrence of chylothorax in some instances of the former strategy and ascites in the latter. The first clinical report by Glenn was published in 1958. It is following that report, and a subsequent series of publications by the same authors, that the superior cavopulmonary anastomosis has become widely known as the "Glenn shunt." Interestingly, however, a fundamentally similar operation was at the same time being developed and evaluated in Budapest by Frances Robiscek and associates, in Russia by Galankin and Darbinian and by Meshalkin, in the United States by Shumacher, and in Italy by Carlon and others, as stated above. In retrospect, it appears likely that the first attempts at clinical cavopulmonary shunts were by Harris B. Shumacher in 1954. His two young patients, one with truncus arteriosus and one with transposition, both had markedly elevated pulmonary vascular resistance and both died within hours of operation.

The "Glenn shunt" (unidirectional superior cavopulmonary anastomosis) found a unique and important place in the management of cyanotic heart disease in general, and functionally univentricular hearts in particular. The resulting physiology was fundamentally different from that achieved by creation of systemic-to-pulmonary artery shunts. The cavopulmonary anastomosis was capable of increasing pulmonary blood flow and thus systemic arterial oxygen saturation without increasing the volume load on the systemic ventricle. It also proved the feasibility of transpulmonary flow without complete dependence on a subpulmonary ventricle. During the 1960s and early 1970s, the classic "Glenn shunt" was used extensively, with good results, to palliate patients with tricuspid atresia, and to a lesser extent other forms of functionally single ventricle. Unfortunately, it became apparent after a number of years that cyanosis would eventually recur in a high proportion of patients. This was in part attributable to maldistribution of blood flow in the involved lung (favoring flow to the lower lobe), and in part to the development of arteriovenous shunting within the lung that was perfused exclusively with superior vena caval blood. The "unidirectional" Glenn shunt was eventually modified in a variety of ways by Dogliotti, Haller, and Azzolina to allow the flow of superior vena caval blood into both pulmonary arteries. They utilized either an end-to-side or side-to-side anastomosis of the superior vena cava to the right pulmonary artery with maintenance of continuity between right and left pulmonary artery branches. This technique, which preserves the confluence and the integrity of the central pulmonary arteries, eventually supplanted the classic Glenn anastomosis as preferred palliation for functional single ventricles. Currently, without particularly good reason, the bidirectional superior cavopulmonary anastomosis is generally and widely referred to as the "bidirectional Glenn shunt." It is most widely used as a preliminary or interim procedure (following neonatal palliation) in patients with functionally univentricular hearts for whom a staged approach to reconstruction culminating in completion of the Fontan circulation is anticipated. Occasionally, construction of a superior cavopulmonary

connection, together with maintenance of a physiologically restrictive source of antegrade pulmonary blood flow from the functionally univentricular heart or from a small patent arterial duct or shunt, is considered as a final or definitive palliative arrangement, as an alternative to the Fontan procedure. Other applications of the bidirectional superior cavopulmonary connection are less common. They include those circumstances where mild-to-moderate degrees of hypoplasia of the right ventricle and its associated valves permit the use of the right ventricle as a subpulmonary ventricle that handles venous return from the inferior vena cava, while superior caval return is diverted directly to the lungs. This arrangement has come to be known colloquially as "one-and-a-half ventricle repair." Occasionally, in the setting of complex malformations with atrioventricular discordance, the superior cavopulmonary connection has been used in conjunction with a simplified atrial baffle procedure that deals only with inferior caval return. Finally, superior cavopulmonary connection has, in rare instances, been used to achieve right ventricular volume unloading in the setting of ischemic right ventricular failure. It has been suggested that it may have an analogous role in patients with reduced right ventricular function who are candidates for left ventricular mechanical support.

ROLE OF THE SUPERIOR CAVOPULMONARY CONNECTION IN THE MANAGEMENT OF FUNCTIONALLY UNIVENTRICULAR HEARTS

By the 1980s, refinement of the technical aspects of modified Fontan operations and development of guidelines for patient selection led to a decrement in the overall mortality associated with these procedures. Nonetheless, a troublesome degree of morbidity and early mortality occurred following modified Fontan operations, even among those who met all criteria for "favorable candidate" status. Too often, even among carefully selected patients with adequate preoperative ventricular function and pulmonary vascular anatomy and resistance, the postoperative course was characterized by elevation of central venous pressure, tachycardia, with sequestration of fluid in the pleural and peritoneal spaces, and hypotension which was only transiently responsive to volume administration at the expense of further increases in venous pressure. Little improvement was seen with administration of inotropic agents and vasodilators, or with external compressive devices to promote venous return while minimizing volume administration. Echocardiography frequently showed a striking degree of ventricular wall thickness relative to cavity volume, vastly out of proportion to the preoperative degree of myocardial hypertrophy. This appearance of a thickened ventricle with relatively small cavity volume was associated with tachycardia and poor perfusion, regardless of the contractile state of the myocardium. It became evident that the conversion from the unoperated or palliated state to the post-Fontan circulation was associated with rapid removal of a chronic volume load on the ventricle. The change in loading conditions took place abruptly, while regression of myocardial mass proceeds slowly. As a consequence, a maladaptive response to removal of the ventricular volume load occurs, characterized by markedly increased ventricular myocardial mass-to-volume ratio. The persistence of increased muscle mass in the setting of acutely diminished ventricular volume results in increased ventricular wall thickness and decreased cavity dimensions. This change in the geometry of the ventricle can result in significant alterations in both systolic and diastolic functions. Perhaps most importantly, the diminished compliance of the acutely thickened ventricle results in poor filling, at the expense of elevated pressure in the cavopulmonary pathway and diminished cardiac output. This realization shed light on earlier observations that older age (longer duration of volume loading) and increased ventricular hypertrophy (increased muscle mass) were risk factors for mortality in association with the Fontan operation. More importantly, it leads to the hypothesis that dividing Fontan's operation into two procedures could accomplish early reduction of the volume work of the single ventricle, at the same time minimizing the impact of changes in ventricular geometry on outcome and survival. Following construction of a superior cavopulmonary connection and elimination of all other sources of pulmonary blood flow (the first stage), a gradual adaptive process characterized by normalization of the mass-to-volume relationship of the functionally single ventricle would make it possible to perform the completion Fontan procedure (the second stage) with the expectation of minimal change in ventricular geometry and favorable conditions with regard to the ventricle's diastolic properties (compliance).

At the end of the decade of the 1980s, and in the early 1990s, several groups explored the potential benefits of a two-staged approach to Fontan operation. For some, the protocol of performance of an early bidirectional superior caval pulmonary anastomosis followed later by a completion Fontan procedure was a pathway selected specifically for high-risk Fontan candidates. Others, notably Norwood and Jacobs in Philadelphia, pursued a two-staged approach to Fontan operation for virtually all patients. They postulated that some of the morbidity and mortality associated with Fontan operation would be eliminated by staging. Performance of a superior cavopulmonary connection in the first year of life achieved construction of part of the Fontan pathway. More importantly, the duration of the palliated state was minimized, and the volume load on the single ventricle was reduced as early in life as is practical. Thus in 1989, Norwood introduced the hemi-Fontan procedure, as the first step in a two-stage process of achieving total cavopulmonary connection. Obligating superior vena caval return to pass through the lungs before returning to the functional single ventricle, the hemi-Fontan operation is the physiologic equivalent of the bidirectional Glenn anastomosis. Because it is planned as an intermediate step before an anticipated completion Fontan procedure, it differs technically from a bidirectional Glenn anastomosis in ways that simplify the eventual completion Fontan.

INDICATIONS AND PREOPERATIVE ASSESSMENT

The most common indication for a cavopulmonary shunt is anatomy that is not amenable to eventual biventricular repair as a means of achieving systemic and pulmonary circulations in series. Superior cavopulmonary connections are generally considered as a preparatory stage for any unoperated or palliated patient older than about 3 months of age in which a univentricular repair is anticipated, particularly for those patients with risk factors for a Fontan-type procedure. In these situations, a cavopulmonary connection may be combined with a procedure to address factors that may increase the risk of Fontan operation, such as systemic ventricular outflow tract obstruction, atrioventricular valve (AV) insufficiency, excessive ventricular volume load associated with systemic-to-pulmonary artery shunts, impediments to pulmonary venous return, or pulmonary artery distortion. It may also be a rational

staging procedure for selected patients for whom the ultimate choice between univentricular and biventricular repair is not clear. Finally, some patients with functionally univentricular hearts may be destined for heart transplantation because of either poor myocardial function or severe organic systemic AV valve abnormalities. In these rare instances, reduction of the volume burden on the ventricle by means of superior cavopulmonary connection may stabilize the physiology while awaiting transplantation and may even result in improved ventricular function and/or lessened AV valve regurgitation.

The cavopulmonary connection has several advantages as an interim procedure in preparation for an eventual completion Fontan procedure. First, it can be performed with a lower mortality and morbidity than a Fontan procedure or a total cavopulmonary connection, particularly in infants from 3 to 12 months of age. Second, in contrast to a systemic-to-pulmonary artery shunt, a cavopulmonary shunt increases effective pulmonary blood flow by directing fully desaturated blood into the pulmonary circulation. Third, the risk of developing pulmonary vascular disease is reduced by lowering pulmonary arteriolar pressure. Fourth, the risk of pulmonary artery distortion is less than with a systemic-to-pulmonary artery shunt. Finally, and perhaps most important, the cavopulmonary shunt, when combined with removing systemic-to-pulmonary artery shunts or antegrade pulmonary blood flow, reduces ventricular work by reducing the volume load on the single ventricle. As noted above, this often improves ventricular function and reduces systemic AV valve regurgitation, which would compromise candidacy for a Fontan procedure.

Careful assessment by echocardiography is undertaken as part of the preoperative planning prior to surgery to create cavopulmonary connections. Comprehensive evaluation of the anatomy includes assessment of the segmental relations and connections, the morphology and function of the ventricle(s), presence and degree of AV regurgitation, presence and degree of systemic ventricular outflow tract obstruction (including aortic arch obstruction), and the nature of systemic and pulmonary venous connections. In particular, the existence of bilateral superior caval veins and/or the possibility of interrupted inferior vena cava with azygos continuation must be considered. All potential sources of pulmonary blood flow (antegrade from the heart, patent ductus, surgically created shunt(s), aortopulmonary collaterals) should be evaluated and the architecture of the intrapericardial pulmonary arteries must be assessed. Cardiac catheterization is indicated when these details cannot be reliably assessed by means of echocardiography or cardiac magnetic resonance imaging, or when a physiologic assessment is critical to surgical decision making or risk stratification. Determination of pulmonary vascular resistance may be important in the uncommon circumstance presented by an operated or palliated older patient. Measurement of pulmonary venous wedge pressure is generally informative if direct measurement of pulmonary artery pressure and pulmonary blood flow is not easily accomplished. Occasionally, imaging studies will suggest the presence of abnormalities of venous anatomy that may best be managed by catheter-directed intervention prior to surgery. An example would be venovenous collateral connections from systemic veins to pulmonary veins, or the presence of a small secondary (usually left-sided) superior vena cava or cardinal vein, which is considered too small to be effectively incorporated into bilateral superior cavopulmonary connections.

Significant pulmonary artery distortion or stenosis should be identified during preoperative evaluation so that it can be addressed as part of the cavopulmonary shunt operation. Some centers rely on determination of pulmonary artery size prior to performance of cavopulmonary connection. We do not generally exclude patients from superior cavopulmonary connection on the basis of hypoplasia of the intrapericardial pulmonary artery branches. Superior cavopulmonary anastomosis has been accomplished successfully in patients with Nakata indices as low as 70 mm^2/m^2.

OPERATIVE TECHNIQUE

The Unidirectional Superior Cavopulmonary Anastomosis ("Classic Glenn Shunt")

The "unidirectional" or "classic Glenn" cavopulmonary shunt is largely of historical interest and is rarely performed in the current era. Typically, the procedure was undertaken without cardiopulmonary bypass support. The surgical approach was most often a posterolateral thoracotomy incision through the fourth intercostal space. Once this exposure has been accomplished, the pleura and pericardium are incised posterior to the phrenic nerve, which is left attached to the widely mobilized anterior pleuropericardium to avoid excessive traction directly on the nerve (Fig. 94.1). The superior vena cava is dissected free from the level of the

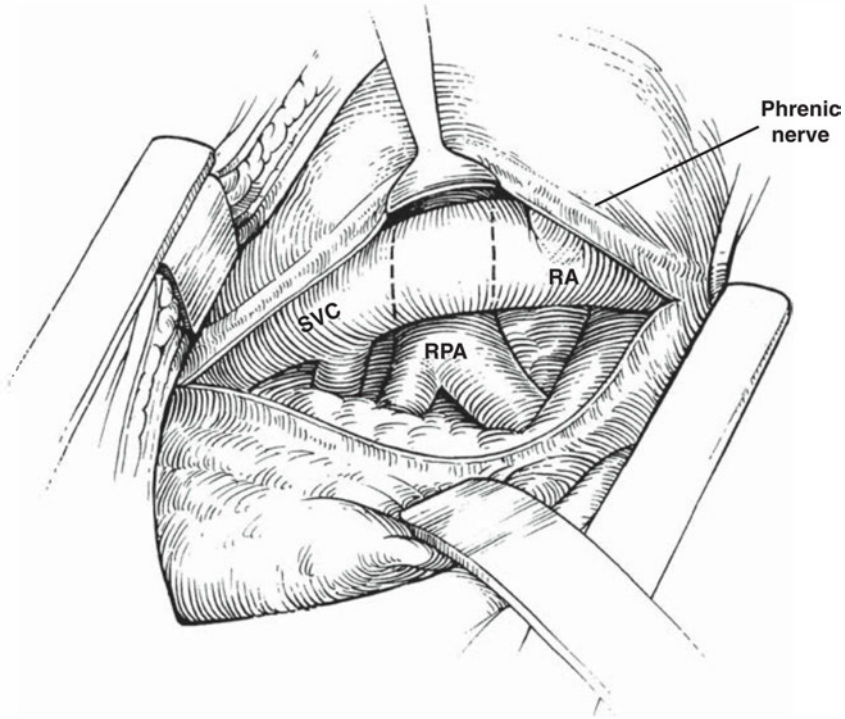


Fig. 94.1. Exposure for a unidirectional or "classic Glenn" cavopulmonary shunt. RA, right atrium; RPA, right pulmonary artery; SVC, superior vena cava.

innominate vein to the right atrium. The azygos vein is ligated and divided to provide better mobility of the superior vena cava and to prevent collateral venous connection with the inferior vena cava. The right pulmonary artery is dissected from its central origin to beyond the first hilar branches. After intravenous administration of heparin, the proximal right pulmonary artery is occluded with a vascular clamp. Oxygen saturation at this time is dependent entirely on flow to the contralateral lung. If oxygenation and hemodynamics are acceptable in the face of this test occlusion of the right pulmonary artery, the distal branches of the right pulmonary artery are occluded with silastic vessel loops or small atraumatic clamps. The right pulmonary artery is then divided at a point medial to the superior vena cava, the proximal end is oversewn, and the clamp is removed. The orifice of the right pulmonary artery may be enlarged slightly by extending the opening inferiorly. A partially occluding (side-biting) vascular clamp is placed on the superior vena cava from above the divided azygos vein to just above the right atrium, isolating a long segment of vena cava (Fig. 94.2). Careful placement of the clamp avoids complete obstruction of superior vena caval drainage to the right atrium. An incision is made in the lateral aspect of the superior vena cava, beginning where the inferior margin of the right pulmonary artery crosses the vena cava and extending superiorly through the orifice of the divided azygos vein. End-to-side anastomosis of the right pulmonary artery to the superior vena cava is then accomplished using fine monofilament suture. Care is taken to avoid "purse stringing," including the placement of a few interrupted sutures. Distal vessel loops are released, followed by removal of the side-biting clamp from the superior vena cava (Fig. 94.3). Following a period of manual ventilation to fully inflate the right lung, the superior vena cava is temporarily occluded above its junction with the right atrium. It is customary to make a direct measurement of pressure in the superior vena cava at this point, before proceeding with definitive ligation of the superior vena cava above its junction with the right atrium. An alternative surgical method of creating a unidirectional superior cavopulmonary anastomosis involves transection of the superior vena cava above its junction with the right atrium and direct end-to-end anastomosis of the cava to the divided right pulmonary artery. This approach may be facilitated either by the use of a decompressing shunt from the innominate vein to the right atrium or by a brief period of cardiopulmonary bypass support.

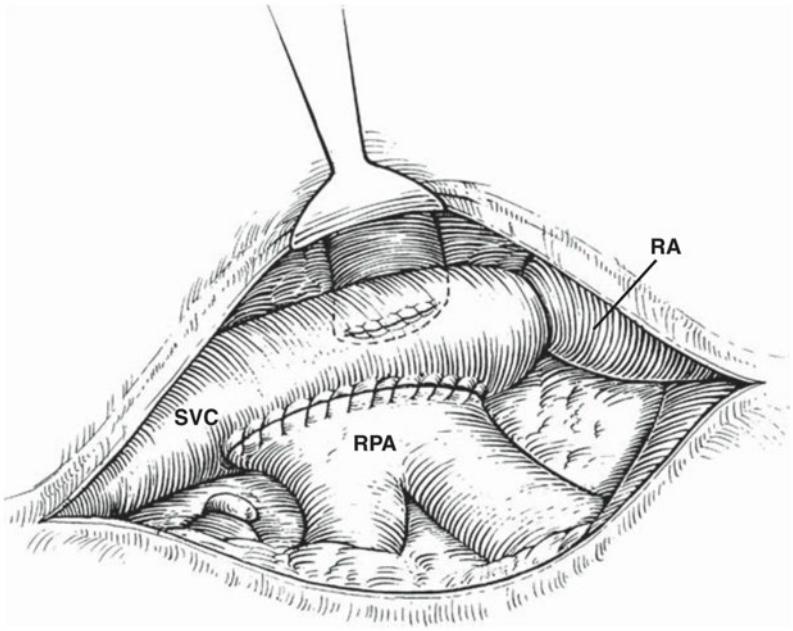


Fig. 94.3. A completed "classic Glenn" cavopulmonary shunt with ligation of the lower superior vena cava (SVC). RA, right atrium; RPA, right pulmonary artery.

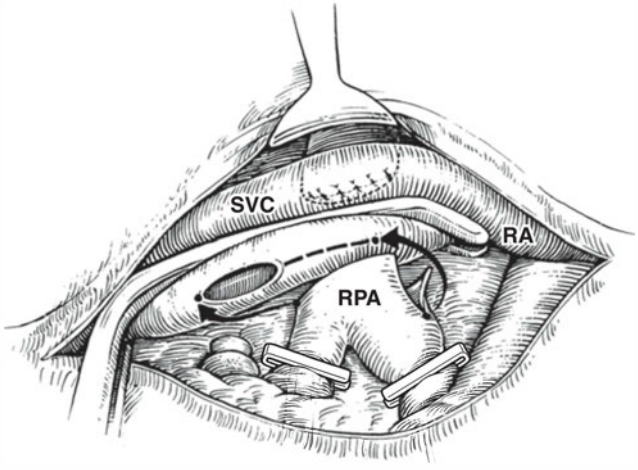


Fig. 94.2. Preparation for end-to-side anastomosis of right pulmonary artery to superior vena cava (SVC) in performance of the unidirectional or "classic Glenn" cavopulmonary shunt. RA, right atrium; RPA, right pulmonary artery.

The unidirectional Glenn anastomosis involves iatrogenic disruption of continuity between the right and left pulmonary arteries, which is viewed as disadvantageous in patients for whom eventual Fontan circulation is anticipated. A rare circumstance where the unidirectional cavopulmonary anastomosis might be performed today would be in a patient with an isolated or very hypoplastic left pulmonary artery that is being "resuscitated" with a systemic-to-pulmonary artery shunt. Superior vena caval flow may be entirely diverted to the right pulmonary artery in order to achieve reduction of the volume load on the functionally single ventricle. In general, awareness of technical issues related to the unidirectional Glenn shunt is primarily of importance today because of the likelihood of encountering older patients who underwent such palliative procedures in the past and the need to reestablish pulmonary artery continuity in the context of a Fontan procedure, a Fontan conversion, or even heart transplantation.

The Bidirectional Superior Cavopulmonary Anastomosis ("Bidirectional Glenn")

The bidirectional cavopulmonary anastomosis is most frequently constructed as part of the staged palliation of functionally univentricular heart disease in a patient for whom eventual completion of the Fontan circulation by means of an extracardiac conduit type of completion Fontan procedure is anticipated. Median sternotomy is by far the most common surgical approach. Cardiopulmonary bypass support is used in the majority of instances. Cardiopulmonary bypass can be avoided selectively, in cases where no intracardiac procedures are required, as long as blood flow to the contralateral lung is sufficient to maintain adequate systemic arterial saturation during clamping of the ipsilateral branch pulmonary artery, and adequate cerebral blood flow is maintained with the avoidance of severe cerebral venous hypertension. In general, this involves the use of a temporary shunt that diverts blood from the superior vena cava to the right atrium. This is accomplished under conditions of systemic heparinization, by the placement of a cannula in the innominate vein and another in the right atrium, and connecting them. The use of cardiopulmonary bypass support is required if total occlusion of the ipsilateral proximal pulmonary artery results in an unacceptable degree of systemic arterial desaturation, if an ipsilateral aortopulmonary shunt is present and is the principle source of pulmonary blood flow, if superior vena cava pressure is too high following caval occlusion despite the presence of the cava to right atrium shunt, or if cerebral oxygen saturation falls to an unsafe level. Cardiopulmonary bypass is required, of course, if concomitant intracardiac procedures are anticipated. Many, but not all surgeons, prefer to routinely use cardiopulmonary bypass support for cavopulmonary shunt operations.

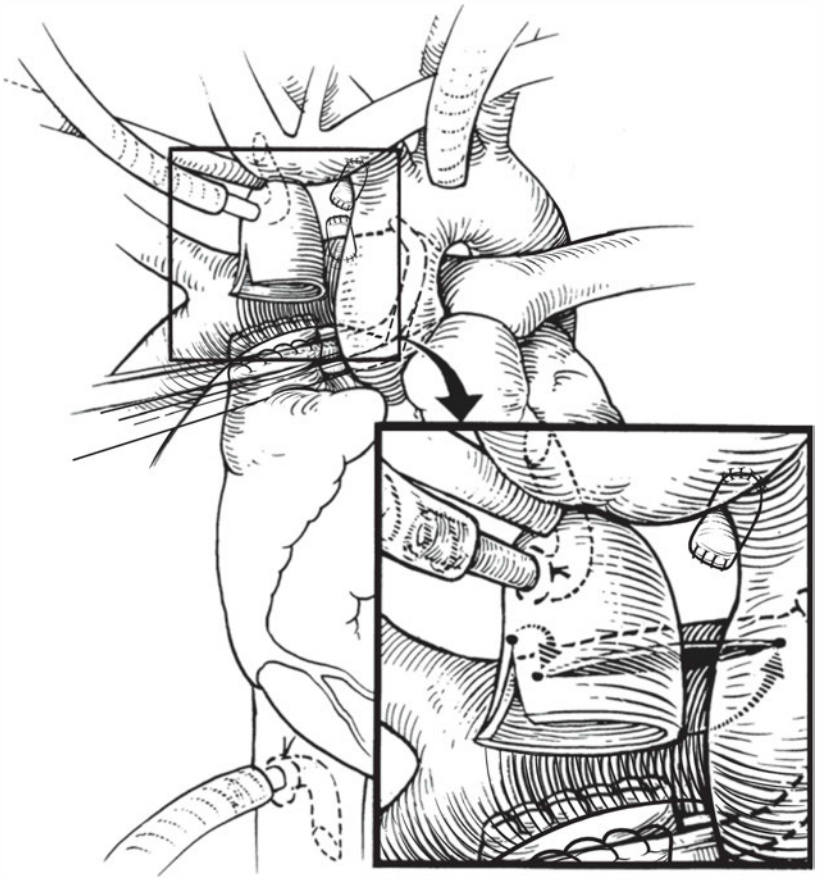


Fig. 94.4. Exposure and cannulation for a bidirectional cavopulmonary anastomosis. In this instance, the orifice of the vena cava is enlarged slightly by cutting back on the lateral side (inset).

Cardiopulmonary bypass support for the construction of a superior cavopulmonary anastomosis generally involves placement of one cannula in the superior vena cava near its junction with the innominate vein or in the innominate vein directly. A second drainage cannula is positioned in the right atrium, or in the inferior vena cava, if intracardiac procedures are anticipated (Fig. 94.4). Once satisfactory parameters are present with either a cava-to-right atrium shunt or a conventional cardiopulmonary bypass, it is customary to ligate and divide the azygos vein. If an ipsilateral aortopulmonary shunt is present, it is occluded and divided. The superior vena cava is occluded with a tourniquet or clamp at or just below the site of cannula placement. A vascular clamp is placed just above the junction of the superior vena cava with the right atrial junction, avoiding injury to the sinoatrial node and its blood supply. The superior cava is transected at the level of the inferior border of the right pulmonary artery and the cardiac end of the vena cava is oversewn. The upper end of the divided superior vena cava is prepared for anastomosis to the pulmonary artery. The orifice of the vena cava may be enlarged slightly by cutting back on either the lateral or the medial side. A vascular clamp is placed across the origin of the right pulmonary artery for proximal control. The distal branches may be controlled with silastic vessel loops or atraumatic vascular clamps. A longitudinal pulmonary arteriotomy is created along the superior aspect of the right pulmonary artery. If a previously placed systemic-to-pulmonary artery shunt has been occluded and divided, the location of the shunt determines whether it is preferable for the arteriotomy to extend through the shunt site or be separate from it. The placement of traction sutures in the pulmonary arteriotomy and in the divided superior vena cava may be helpful in orienting the anastomosis. Many surgeons use a combination of running and interrupted monofilament sutures to accomplish the cava to pulmonary artery anastomosis (Fig. 94.5). What is most important is avoidance of purse stringing and avoidance of torsion or twisting of the vessels. If for any reason it appears that the anastomosis may be narrowed, incorporation of a small patch of pericardium may be advisable. When performed under cardiopulmonary bypass, the bidirectional cavopulmonary anastomosis can generally be accomplished with a beating heart under conditions of normothermia or mild hypothermia. Moderate or greater degrees of hypothermia and cardioplegic arrest are generally utilized when concomitant intracardiac procedures are performed. The decision regarding elimination or maintenance of additional sources of pulmonary blood flow is a matter or surgeon's preference and

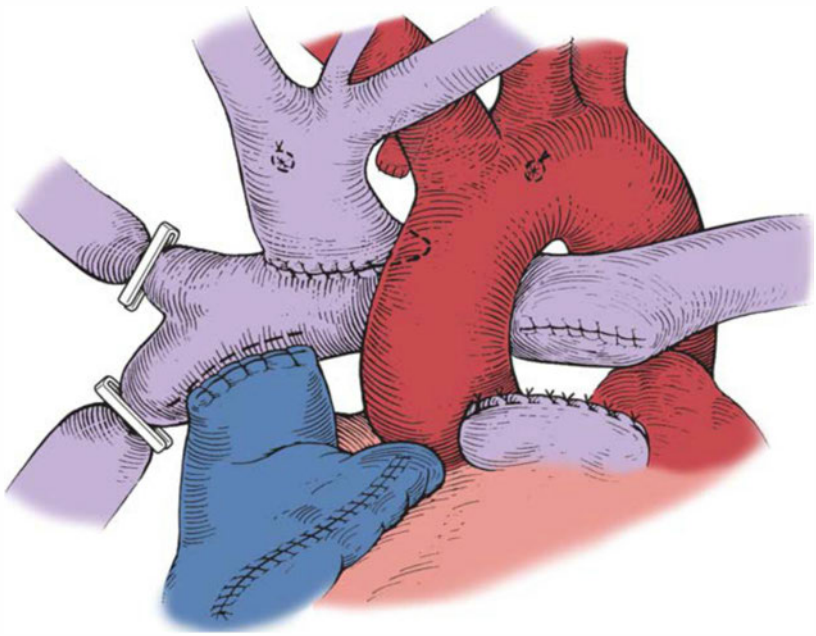


Fig. 94.5. A completed bidirectional cavopulmonary anastomosis with elimination of other sources of pulmonary blood flow. The right modified Blalock–Taussig shunt has been divided and the pulmonary artery end of the shunt has been excised. The main pulmonary trunk has been divided and oversewn. The proximal suture line incorporates the pulmonary valve leaflets, in order to preclude the possibility of creating a blind cul-de-sac of main pulmonary artery, where thrombus might develop.

may be determined by specific patient-related issues. As a general policy, we eliminate all other sources of pulmonary blood flow when performing superior cavopulmonary anastomoses in patients who will eventually go on to completion of the Fontan circulation. Elimination of antegrade pulmonary blood flow from the heart via the pulmonary valve should be accomplished not by simple ligation but by means of division of the pulmonary trunk and oversewing the cardiac end at the level of the valve leaflets, in order to avoid leaving a blind-ending pouch of proximal pulmonary artery, which is known to be a risk factor for thrombus formation secondary to stasis. At the completion of any superior cavopulmonary anastomosis procedure, ventilation should be sufficient to eliminate atelectasis. Hyperventilation and hypocarbia should be avoided, however. Optimal systemic oxygenation is often accomplished with a mild degree of hypercarbia, as the slight elevation of $PaCO_2$ maximizes cerebral blood flow. Maximizing the fraction of total cardiac output that perfuses the upper body and returns via the superior vena cava (thus passing through the lungs before returning to the heart) seems, in most cases, to be of greater importance than the mild degree of pulmonary vasodilation that may be achieved as a result of hyperventilation.

The Hemi-Fontan Procedure

As previously noted, Norwood introduced the hemi-Fontan procedure in 1989 as the first step in a two-stage process of achieving total cavopulmonary connection. Though the hemi-Fontan operation is the physiologic equivalent of the bidirectional Glenn anastomosis, it differs technically from a bidirectional Glenn anastomosis in ways that simplify the eventual completion Fontan. While any type of completion Fontan procedure can be accomplished in a patient who has undergone a hemi-Fontan operation, it is optimal preparation for a Fontan connection of the lateral atrial tunnel type. In the hemi-Fontan operation, the natural connection of the right atrium with the superior vena cava is maintained, but the caval orifice is first enlarged (to be as large as the inferior vena caval right atrial connection), and then closed with a patch that is easily removed at the final stage. Other sources of pulmonary blood flow including systemic-to-pulmonary shunts and antegrade flow to the pulmonary arteries are eliminated. Architecture of the central pulmonary arteries is optimized by augmentation of the central confluence with homograft vascular patch material. Other potential hemodynamic burdens such as systemic ventricular outflow tract obstruction or AV valve regurgitation are addressed as part of the hemi-Fontan procedure. Proponents of the bidirectional Glenn anastomosis argue that the hemi-Fontan is a technically more complex procedure, which always requires cardiopulmonary bypass and is sometimes performed using hypothermic circulatory arrest. They also argue that the hemi-Fontan procedure involves potential injury to the sinus node or its blood supply, Bove and associates later modified Norwood's original operative technique, using an incision that does not cross the cavoatrial junction, with the theoretical goal of preserving the integrity of the sinoatrial node and its blood supply, with the hope that this would enhance the likelihood of preserving sinus rhythm.

The operative technique described here is essentially the methodology introduced by Norwood. After the placement of monitoring devices, the infant is positioned supine and prepped for median sternotomy. Surface cooling is begun, including the use of a cooling blanket beneath the patient, the application of ice bags to the head, and cooling of the operating room. After sternotomy, the surgical dissection prior to heparinization is limited to what is needed to achieve access to the aorta and the right atrium for cannula placement. After heparinization, bypass is established with ascending aortic perfusion and drainage via a single cannula in the systemic venous atrium. All systemic-to-pulmonary artery shunts are occluded upon initiation of bypass and flow is adjusted to 150 ml/kg/min. The perfusate is gradually cooled to approximately 16°C. During the cooling phase of cardiopulmonary bypass, the remainder of the dissection is accomplished, with particular attention to freeing the pulmonary artery confluence behind the ascending aorta. When cooling has been completed (generally to nasopharyngeal and esophageal temperatures below 20°C), the aortic cross-clamp is applied and circulation is temporarily discontinued. Blood is drained via the venous cannula into the cardiotomy reservoir, and the cannula is then removed. Cold crystalloid cardioplegia solution (30 ml/kg) is infused into the aortic root. An incision is made in the most superior portion of the right atrium and carried superiorly onto the medial aspect of the right superior vena cava (Fig. 94.6A). Through this incision, the atrial septal defect is inspected and is enlarged if necessary. The confluence of the right and left branch pulmonary arteries is opened anteriorly to a point just medial to the upper lobe branch on each side. If patent, the main pulmonary artery is transected and oversewn proximally at the level of the pulmonary valve. The posterior

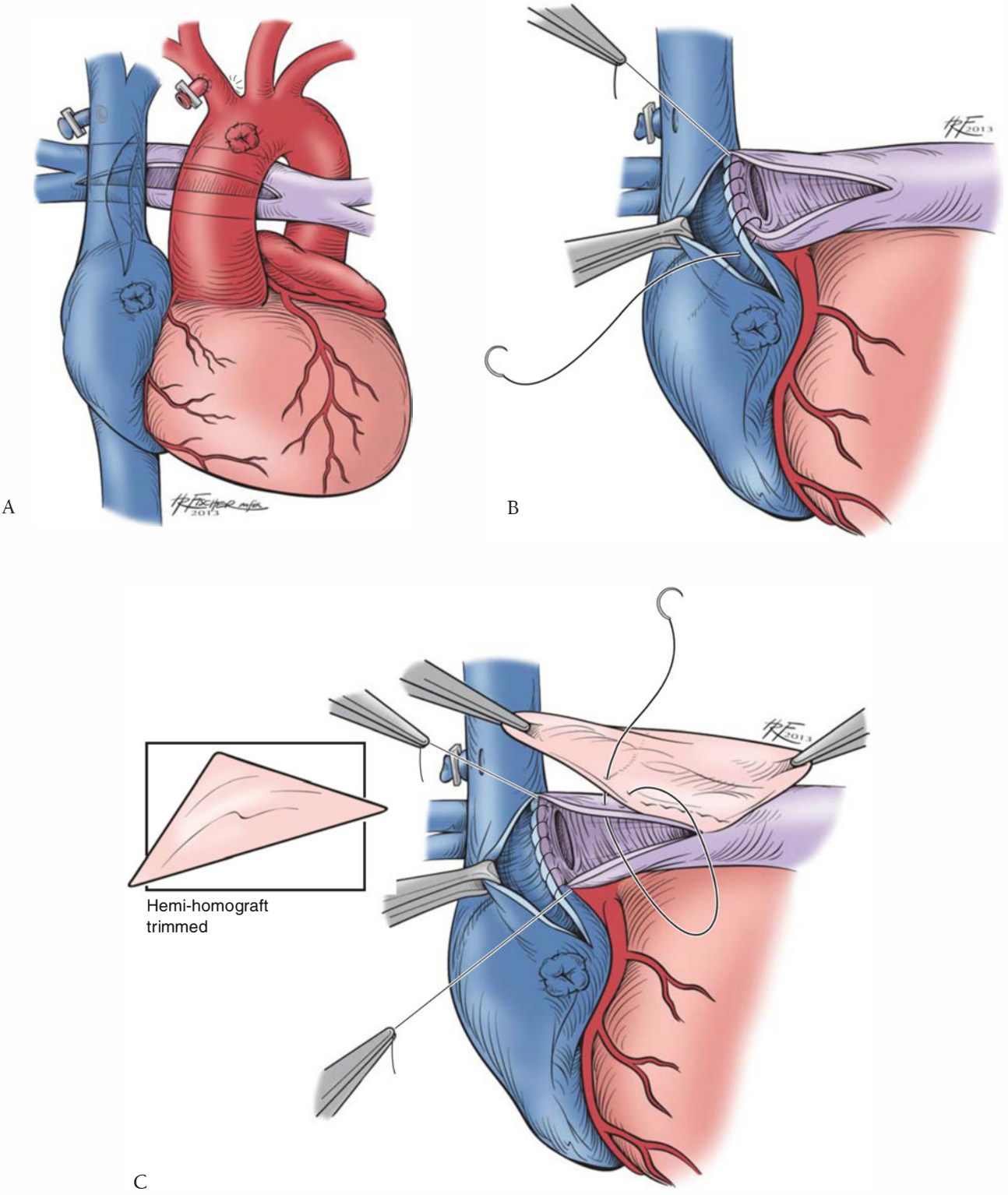


Fig. 94.6. The hemi-Fontan procedure. (**A**) The systemic-to-pulmonary artery shunt is occluded and may be excised. The confluence of the pulmonary arteries is opened anteriorly. (**B**) The right superior vena cava and right pulmonary artery are anastomosed to one another in a side-to-side manner. (**C, D, E, F, G**) A gusset of cryopreserved pulmonary artery homograft tissue is used to augment the confluence of the pulmonary arteries, to create a roof over the large anastomosis of the superior vena cava to the pulmonary arteries, and as a dam to close the surgically enlarged junction of the right atrium with the right superior vena cava. Flow from the superior venacava is into the right and left branch pulmonary arteries.

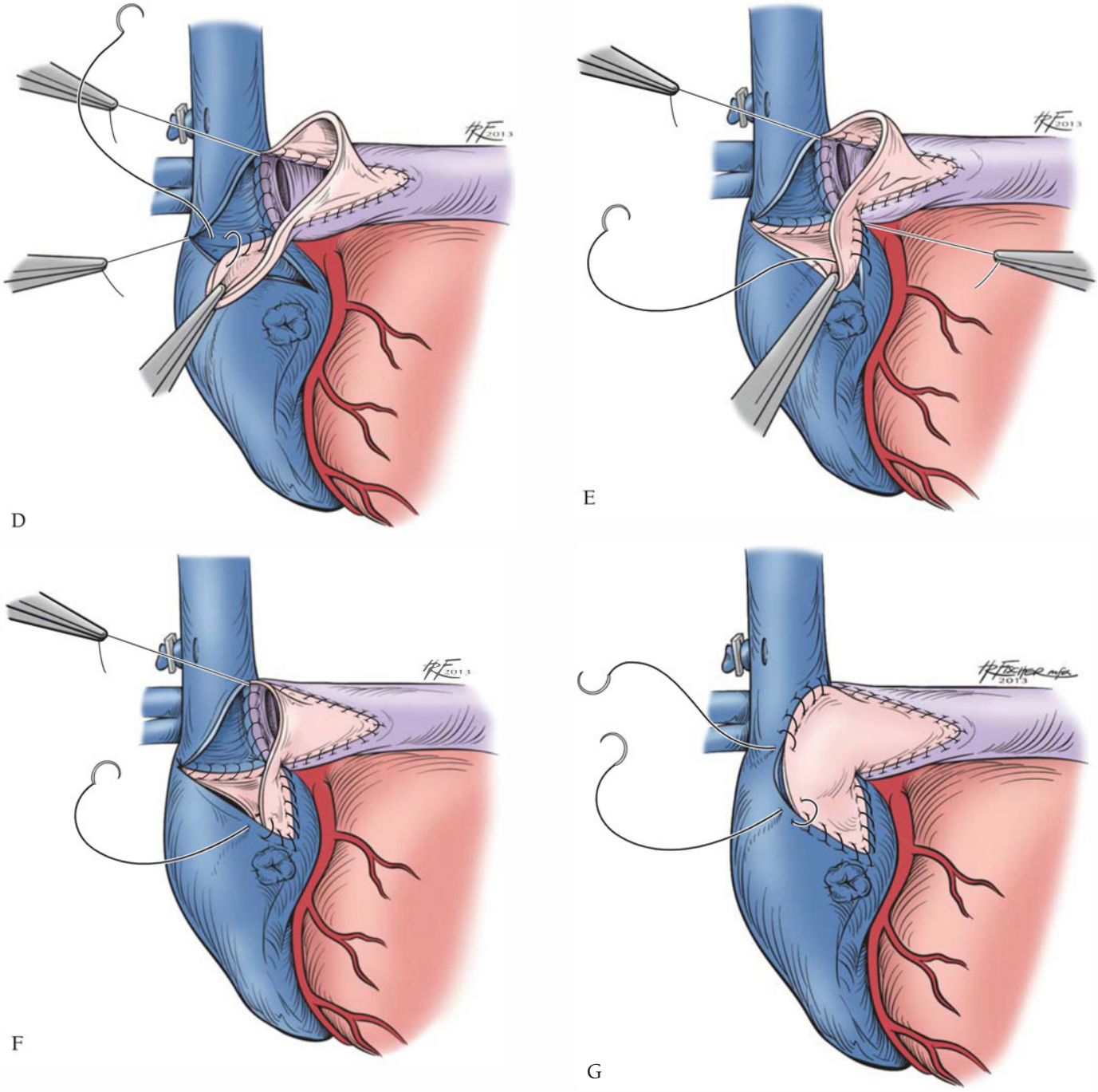


Fig. 94.6. (*Continued*)

lip of the opened right superior vena cava is anastomosed in a side-to-side manner to the right pulmonary artery (Fig. 94.6B). If a left superior vena cava is present, it too is opened along its medial aspect, occluded at its cardiac end, and anastomosed in a side-to-side manner to the ipsilateral branch pulmonary artery. A single patch of cryopreserved pulmonary artery homograft tissue is used to augment the confluence of the pulmonary arteries anteriorly, to create a roof over each anastomosis of the vena cava(e) to the branch pulmonary arteries, and to occlude the inflow of the right superior vena cava into the right atrium (Fig. 94.6C,D). The technique of enlarging and then patching the junction of the right superior vena cava with the right atrium effectively enlarges this connection to a caliber equivalent to that of the junction of the inferior vena cava with the right atrium, but temporarily occludes this connection that will eventually be reopened at the time of the completion Fontan procedure. While some surgeons have stressed the importance of ligation and division of the azygos vein (which may enhance mobilization of the superior vena cava for simple bidirectional Glenn anastomosis), we have not appreciated any important disadvantage in those instances where we have performed hemi-Fontan procedures and left the azygos vein intact. When the cavopulmonary amalgamation and any concomitant procedures have been completed, bypass is resumed and the cross-clamp is removed. In general, the hemi-Fontan procedure itself can be accomplished during a period of about 30 minutes, either

under conditions of hypothermic circulatory arrest or on continuous cardiopulmonary bypass support. During rewarming, the lungs are ventilated to eliminate atelectasis and to minimize impedance to transpulmonary flow. Conventional ultrafiltration may be accomplished while rewarming. After rewarming, separation from bypass is generally accomplished with minimal inotropic support. Modified ultrafiltration may then be accomplished. Following decannulation, catheters inserted through the cannulation purse string in the right atrium are used for infusions and to monitor cardiac filling pressure (percutaneous central venous lines are not used). Sternal closure is accomplished routinely.

Postoperative management is the same for the hemi-Fontan procedure as for the bidirectional Glenn anastomosis. This includes judicious administration of fluids guided by cardiac filling pressures and early extubation that can usually be accomplished a few hours after completion of surgery. Diuretics are generally administered beginning on the first postoperative day. Hospitalization usually lasts a week or less. Facial swelling and irritability (which we suspect is attributable to "headache" from transiently elevated venous pressure) often persist for a few days and are more common in younger infants. Pleural effusion and chylothorax are extremely rare. Oxygen saturation in room air at discharge from the hospital is generally in the range of 80% to 84%.

While the construction of a large side-to-side amalgamation of the superior vena cava and the confluence of the pulmonary arteries are facilitated by the absence of a drainage cannula from the superior vena cava, the same surgical result can be achieved using continuous cardiopulmonary bypass with venous drainage accomplished by placing one cannula in the inferior vena cava and one cannula high in the superior vena cava near its junction with the innominate vein if the surgeon wishes to use continuous bypass rather than hypothermic circulatory arrest.

Technical modifications introduced by Bove and others include the use of an incision that does not cross the cavoatrial junction and the use of a separate prosthetic patch (rather than a portion of the homograft as a dam) to occlude the junction of the superior vena cava with the right atrium. It remains to be determined whether these maneuvers actually result in the intended consequence of lowering the incidence of sinus node dysfunction and subsequent arrhythmias.

Other surgeons prefer a modification of the simple superior cavopulmonary anastomosis in which both ends of the divided superior vena cava are anastomosed to the right pulmonary artery (superiorly and inferiorly), and the cavoatrial junction is occluded with a prosthetic patch. Such modifications of the "bidirectional Glenn" operation are often erroneously referred to as hemi-Fontan procedures.

SPECIAL SITUATIONS

Bilateral Superior Caval Veins

Bilateral superior caval veins are present in many patients with functionally univentricular hearts. In most instances of situs solitus (normal arrangement of the atria), the left-sided superior vena cava drains to the coronary sinus. In cases of visceroatrial heterotaxy with atrial isomerism, it is common for the left-sided superior caval vein to drain directly to the left-sided atrium or left side of the common atrium. In the vast majority of cases with bilateral superior caval veins, it is advisable to include both veins in the cavopulmonary amalgamation, either as described above for the hemi-Fontan operation or as simple bilateral end-to-side cavoatrial anastomoses, if the so-called "bidirectional Glenn procedure is preferred. Alternatively, if the less dominant of the two superior caval veins is tiny, it may simply be ligated, particularly if there is a crossing vein (innominate vein) that allows it to drain into the dominant caval vein. Some studies have suggested that bilateral superior caval veins are a risk factor for adverse outcomes following superior cavopulmonary anastomosis, with one report from Toronto suggesting that bilateral superior vena cava-to-pulmonary artery anastomosis is associated with an increased risk of thrombus formation and unfavorable growth in the central pulmonary arteries. Van Arsdell and associates suggested the construction of a centrally located "V-shaped" amalgamation of the right and left caval veins and the pulmonary arteries. A recent risk factor analysis of mortality and morbidity associated with the bidirectional Glenn operation by Kogon and associates in Atlanta did not reveal any association between bilateral bidirectional cavopulmonary anastomoses and adverse outcomes.

Interrupted Inferior Vena Cava and Azygos Continuation

Approximately 80% of patients with heterotaxy syndrome of the left isomerism variety (polysplenia syndrome) have suprarenal interruption of the inferior vena cava. In most such cases, the infrarenal vena cava drains via the azygos vein (or a hemi-azygos vein) to a superior caval vein (most often left-sided). The hepatic veins enter the atria directly, either though a single confluence or in two or more groups. This systemic venous arrangement often occurs in the setting of functionally univentricular heart—frequently with common AV valve and with double-outlet right ventricle (DORV). In 1984, Kawashima and associates described a new type of total cavopulmonary shunt operation, which consisted of end-to-side anastomosis between the superior vena cava with azygos or hemiazygos continuation and the confluence of the pulmonary arteries. The operation was initially proposed as a definitive form of palliation. Experience at several centers, however, soon revealed that these patients appeared to be at risk for recurrent cyanosis due to the development of significant and progressive right-to-left shunting via pulmonary arteriovenous malformations. In many cases, the problem was ameliorated by subsequent direction of the hepatic vein effluent to the pulmonary circulation, with the observation of gradual improvement in arterial saturation and evidence of regression of the pulmonary arteriovenous malformations. Consequently, patients who undergo Kawashima's total cavopulmonary shunt operation should have planned completion of the Fontan circulation. This can be accomplished by diverting the hepatic vein flow to the pulmonary arteries using either lateral atrial tunnel or extra-cardiac conduit technique, or by creating a connection between the hepatic veins and the azygos vein. In most instances, these procedures should be completed not more than a year after the Kawashima operation, and sometimes within a few months.

One and a Half Ventricle Repair

For patients with a marginal subpulmonary ventricle, conventional therapeutic pathways include the establishment of functionally univentricular circulation or pursuit of a high-risk biventricular repair. Another alternative is the establishment of biventricular physiology with partial bypass of the subpulmonary ventricle by means of a superior cavopulmonary connection. This "one and a half ventricle repair" reduces the volume work of the subpulmonary ventricle, which then must handle only the inferior caval return. Theoretical advantages include avoidance of the degree of pressure elevation in the sub-diaphragmatic venous circulation that would accompany complete Fontan circulation, together with the preservation of pulsatile pulmonary blood flow and some contribution to total cardiac

output by the marginal subpulmonary right ventricle. A theoretical disadvantage is the possibility of transmission of elevated pressure to the superior vena cava, sometimes with systolic flow reversal. That complication can sometimes be addressed by gentle banding of the proximal right pulmonary artery. The one and a half ventricle repair was originally proposed for cases of mild-to-moderate right ventricular hypoplasia in the setting of pulmonary atresia with intact ventricular septum. It has subsequently been extended to other anomalies including unbalanced atrioventricular canal defect, straddling AV valves, and as an adjunct to repair of Ebstein's anomaly. Another unique circumstance where concomitant superior cavopulmonary connection has proved useful in the setting of biventricular physiology is in association with the double switch operation for congenitally corrected transposition of the great arteries. The use of the cavopulmonary anastomosis allows for a simplified approach to the atrial baffle repair portion of the double switch operation. Hanley and associates refer to the modified atrial procedure that baffles inferior vena caval return to the tricuspid valve in conjunction with a bidirectional Glenn operation as "a hemi-Mustard procedure." Theoretical advantages include potential reduction of baffle- and sinus node-related complications. It also makes the atrial baffle repair much less challenging in cases with atrial *situs solitus* and dextrocardia.

RESULTS AND COMPLICATIONS

The physiology that results from superior cavopulmonary connection should be the same regardless of whether one chooses the technique of bidirectional Glenn anastomosis or hemi-Fontan operation, as long as the cavopulmonary connection is nonrestrictive and all areas of pulmonary artery distortion or obstruction are addressed. Computational fluid dynamic studies have failed to show a significant or consistent advantage of one type of connection over the other, with respect to energy conservation and distribution of flow. The difference between the two procedures probably becomes most important once the Fontan circulation has been completed. In studies by Migliavacca and associates, the hemi-Fontan and bidirectional Glenn procedures performed equally well, but important differences in energy losses and flow distribution were found after the completion Fontan procedures. They observed superior hydraulic performance of the lateral tunnel Fontan operation *after* the hemi-Fontan procedure relative to any other type of Fontan connection, including all of those with simple bidirectional cavopulmonary anastomosis. They suggested that this observation may be explained by more optimal caval offset achieved in the surgical reconstruction that includes the hemi-Fontan connection. It is important to emphasize that these inferences are derived using three-dimensional models of typical hemi-Fontan and bidirectional Glenn operations based on anatomic data derived from magnetic resonance scans, angiocardiograms, and echocardiograms. Other investigators have used a variety of other strategies to model the cavopulmonary shunt and Fontan circulation and have drawn different conclusions.

When originally proposed as a strategy for high-risk Fontan patients or as part of a plan to accomplish the Fontan circulation in two stages, the superior cavopulmonary connection was typically created in the second half of the first year of life. In 1996, Jacobs and Norwood reported on 400 hemi-Fontan procedures with mean age at operation of 8.5 months (range: 2 to 24 months). Operative mortality was 8% for the entire cohort. For the last 200 patients, operative mortality was 4.0%. Younger age at operation was not an independent risk factor for operative mortality. Urgent operation in the presence of a hemodynamic burden requiring concomitant procedures was associated with increased mortality. Numerous studies since that time have demonstrated the safety and efficacy of early superior cavopulmonary connection. The age at which elective bidirectional Glenn shunt or hemi-Fontan procedures are undertaken has declined at many centers from 6 to 8 months down to 3 to 4 months, particularly for patients with hypoplastic left heart syndrome and related anomalies. For these patients, the period between initial palliation and second stage surgery is perceived as being a time of significant risk. At any rate, in patients with appropriately low pulmonary vascular resistance, it has been possible to accomplish superior cavopulmonary connections as early as 8 to 10 weeks. Recent series have reported operative mortality rates for hemi-Fontan and bidirectional Glenn operations in the range of 1% to 4%. While younger age has not generally been identified as a risk factor for mortality, younger patients do appear to be at greater risk for perioperative morbidity, including transient periods of lower systemic oxygen saturation.

Most patients undergoing superior cavopulmonary connections can be liberated from ventilatory support and have tracheal extubation within several hours after surgery. While it is not our specific goal, management at some centers includes extubation within the operating room. Arterial oxygen saturation may be as low as the 70% range early after surgery, but generally stabilizes in the low-to-mid 80% range within the first few days. This is consistent with the fact that superior cavopulmonary connection in infants generally results in a pulmonary-to-systemic blood flow ratio (Q_p:Q_s) of approximately 0.6:1. In older patients, a smaller fraction of total cardiac output goes to the upper body, and Q_p:Q_s after superior cavopulmonary connection is proportionately lower. Profound or persistent hypoxia following surgery should be investigated to rule out the presence of an undiagnosed left superior vena cava, other venous anomalies including connections resulting in diversion of superior vena caval blood away from the lungs, or a baffle leak between the superior vena cava and the right atrium following a hemi-Fontan procedure.

Early postoperative complications are rare. A mild-to-moderate degree of systemic hypertension is common early after surgery. While this may be treated pharmacologically, it is generally self-limited. Transient sinus node dysfunction is observed commonly following both the bidirectional Glenn anastomosis and the hemi-Fontan procedure. Nearly all patients recover sinus rhythm or a normally conducted atrial rhythm prior to hospital discharge. A mild degree of facial swelling and upper body plethora are not uncommon, and generally resolve spontaneously. Superior vena cava syndrome with significant upper body edema suggests the possibility of high superior vena cava pressure that may be the result of a restrictive anastomosis, high transpulmonary gradient, poor ventricular function, or AV valve regurgitation. Pleural effusions are uncommon after superior cavopulmonary anastomosis. Pleural fluid collections that cannot be accounted for by postoperative bleeding, that are large, and that persist in the face of diuretic administration should be aspirated to determine whether they are chylous in nature. The potential for phrenic nerve injury should always be kept in mind during the course of surgical dissection. While phrenic paresis is a rare complication of superior cavopulmonary connection operations, it can have a dramatic effect on overall physiology and contribute importantly to morbidity following the eventual completion Fontan procedure.

The time interval between superior cavopulmonary connection and completion of the Fontan circulation is important. While the degree of arterial desaturation associated with superior cavopulmonary connection is generally well tolerated, it appears that longer intervals between this procedure and the completion of the Fontan circulation are associated with the development of systemic-to-pulmonary arterial collaterals, in particular from vessels such as the internal thoracic arteries and other branches of the brachiocephalic vessels. In addition, the complete absence of hepatic vein effluent from the pulmonary circulation appears to be associated with time-related risk of development of pulmonary arteriovenous malformations, which eventually account for clinically important hypoxemia. While this process may be accelerated in the setting of heterotaxy, it nonetheless appears to be a general phenomenon and has been observed in patients with a variety of underlying cardiac diagnoses, including hypoplastic left heart syndrome and others where atriovisceral arrangements are entirely normal. The phenomenon is undoubtedly related at least in part to the role of an as-yet-unidentified hepatic factor but is likely to be multifactorial.

ADDITIONAL SOURCES OF PULMONARY BLOOD FLOW

The issue of whether or not all additional sources of pulmonary blood flow should be eliminated at the time of creation of superior cavopulmonary connection(s) remains unresolved. With respect to the strategy of creating the Fontan circulation in two stages, the idea of early reduction of the volume load of the functionally single ventricle, followed by a period of adaptation with normalization of mass-to-volume ratio and thus ventricular compliance, is consistent with a strategy of elimination of other sources of pulmonary blood flow when the superior cavopulmonary connection is created. The improved results of Fontan procedures achieved using a staged approach have been attributed, at least in part, to this concept. On the other hand, proponents of preserving additional sources of pulmonary blood flow (patent ductus, restrictive right ventricular outflow tract, or tight pulmonary artery band) argue that the additional flow can promote pulmonary artery growth and that comparisons of patients with and without additional sources of pulmonary blood flow after a bidirectional cavopulmonary anastomosis have shown no negative effect on the outcome of the eventual Fontan procedure. Importantly, these comparisons are based on populations of patients who have tolerated the additional pulmonary blood flow and been considered acceptable for completion of the Fontan connection. Some patients do not tolerate additional sources of pulmonary blood flow in the setting of superior cavopulmonary connection and show signs of superior vena cava syndrome (occasionally with chylothorax) or ventricular dysfunction. In such instances, the superior cavopulmonary connection must be investigated to rule out anatomic problems. Then, the additional source(s) of pulmonary blood flow should be eliminated by either catheter intervention or surgical means. Finally, some believe that superior cavopulmonary connection together with accessory source(s) of pulmonary blood flow should be considered definitive palliation and an alternative to completion of the Fontan circulation.

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EDITOR'S COMMENTS

The use of early cavopulmonary shunts and the hemi-Fontan operation in staged reconstruction for single-ventricle physiology has resulted in marked improvement in late outcomes after the Fontan procedure. The primary reason for this salient benefit is the ability to address pulmonary artery distortions and minimize excessive pulmonary blood flow early to prevent problems with pulmonary vascular resistance. Thus, in many cases, staged reconstruction with an initial cavopulmonary shunt is the preferable approach to a one-stage complete Fontan procedure. In very favorable patients with optimal ventricular anatomy and protected pulmonary vascular beds, a one-stage Fontan procedure can be contemplated; however, the staged reconstruction has particular benefit in patients with hypoplastic left heart syndrome in whom a high incidence of left pulmonary artery hypoplasia is noted. In addition, decreasing volume loading of the functional right single ventricle early may improve late ventricular function and minimize the significance of tricuspid valve regurgitation in these patients. While Dr. Jacobs has clearly described the use of the hemi-Fontan operation and interim cavopulmonary shunts as a way to decrease the late morbidity of the Fontan operation, the strategy he has described is based on the concept of the earliest completion of the Fontan circulation that is possible. The primary need to complete the Fontan operation is the consideration that creating a normal systemic oxygen saturation will decrease the development of aortopulmonary collateral arteries, which increase the volume load on the single ventricle. For many years, it was assumed that the creation of a cavopulmonary shunt early in life with elimination of systemic-to-pulmonary shunts would decrease the volume load on the single ventricle and improve ventricular performance at the Fontan operation. It is now known that most of these patients, even after creation of cavopulmonary connections, will develop significant aortopulmonary collateral flow due to continued cyanosis. Thus, it is not completely true that the volume load on the single ventricle is eliminated by the creation of cavopulmonary shunts. Early completion of the Fontan may mitigate the development of more aortopulmonary collateral flow; however, this has not been directly examined. It is now becoming common in our center for patients to undergo MRI calculation of aortopulmonary collateral flow with elimination of additional sources of pulmonary blood flow aggressively in the catheterization laboratory prior to the completion Fontan operation at 1½ to 2 years of age. This approach seems to decrease the development of pleural effusions early postoperatively; however, the effect on long-term function of the Fontan circulation and the redevelopment of additional sources of pulmonary blood flow have not yet been completely determined.

Although the unilateral cavopulmonary anastomosis as advocated by Glenn can be accomplished technically without the use of cardiopulmonary bypass, there is little use for this technique at present. The bilateral cavopulmonary connection has supplanted the standard Glenn anastomosis in virtually all circumstances. The use of a unilateral cavopulmonary connection has been associated with significant development of late arteriovenous malformations of the lung. An additional disadvantage of unilateral cavopulmonary connections is the fact that the connection of the superior vena cava to the right pulmonary artery directly generally supplies the larger right pulmonary vascular bed with a smaller amount of blood flow from the upper body, leaving the smaller left pulmonary vascular bed supplied by the larger flow from the inferior vena cava or right ventricle. Thus, there is a relative flow distribution abnormality with the use of these unidirectional Glenn shunts. Even with bilateral cavopulmonary connections and certainly with the Kawashima operation (in which the majority of inferior vena caval flow is directed to the lungs without contributions from hepatic venous return), development of arteriovenous malformations of the lung is commonplace and results in progressive cyanosis. Early conversion of these patients to the completed Fontan procedure with fenestration is advocated to maintain some hepatic flow to the lung to diminish or resolve the development of arteriovenous malformations.

Although the bidirectional cavopulmonary anastomosis can be accomplished simply with the use of either decompressing shunts from the superior vena cava to the right atrium or with the use of cardiopulmonary bypass (as is preferred in our institution), we continue to use the hemi-Fontan procedure for some patients with single-ventricle physiology. This operation uses a homograft patch to widely enlarge the pulmonary bifurcation and create a dam between the superior vena cava and the right atrium without actual disconnection of the superior vena cava from the atrium at operation. Although this operation is extensive and is often done under a brief period of circulatory arrest, the reconstruction of the pulmonary bifurcation eliminates causes of maldistribution of pulmonary blood flow and permits a very simple completion Fontan procedure, which is generally done when the patient is 1½ to 2 years of age. More complex types of hemi-Fontan reconstruction with a Gore-Tex dam in the superior vena caval orifice in the right atrium followed by division of the superior vena cava with anastomosis to the right pulmonary artery superiorly and inferiorly result in additional multiple suture lines and a longer operative procedure. In addition, the superior vena cava at the right atrial junction is often augmented with additional pericardium to create a larger opening into the pulmonary artery. It is unclear whether this technique offers any distinct advantage over the hemi-Fontan procedure. The only relative advantage is the lack of an incision crossing the superior vena cava-to-right atrial junction, which can interfere with the blood supply to the sinus node and potentially increase the risk of late development of atrial arrhythmias. While the hemi-Fontan operation often will divide the sinus node artery, it is not clear that the late incidence of atrial arrhythmias is higher with the use of this technique and, in fact, the bidirectional Glenn shunt has also been associated with junctional rhythm early postoperatively, possibly due to disruption of nerve pathways along the superior vena cava and right atrium. The contribution of sinus node injury to late development of atrial arrhythmias has not been clearly determined.

Whereas the azygos vein is usually ligated or divided with creation of bidirectional Glenn shunts or the hemi-Fontan operation, it has been left open in many patients in our center and has not resulted in significant decompression of the superior vena cava in most cases. It is not clear that decompression through the azygos vein will occur in the majority of cases after the cavopulmonary anastomosis if

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EDITOR'S COMMENTS (*continued*)

left open. It appears that if the pulmonary vascular resistance remains low, leaving the azygous vein open is not a disadvantage and does not decompress the superior venous system. The azygous vein, however, can become an important steal of blood from the upper body and away from the pulmonary vascular bed if patients have elevated pulmonary vascular resistance or AV valve regurgitation and, therefore, in the majority of cases disruption of the azygos vein is performed at the time of cavopulmonary anastomosis to eliminate a potential source of decompression and potential desaturation. Most decompression of superior vena cava-to-pulmonary anastomoses occurs when there is a left-sided vein of Marshall, which can decompress to a low-pressure chamber such as the right atrium through the coronary sinus.

Although the Nakata index has been useful for defining the extent of the pulmonary vascular bed for completion Fontan operation, we have not used this index in our center. In most cases, patients with an adequate oxygen saturation with an aortopulmonary shunt and a pulmonary venous wedge pressure that is low can undergo a cavopulmonary anastomosis with satisfactory results, even with a low Nakata index number. It is not uncommon for patients with even a single pulmonary arterial supply to have satisfactory results with a cavopulmonary anastomosis in the presence of discontinuous pulmonary arteries if pulmonary vascular resistance is low in the remaining vascular bed.

In our operative techniques, no monitoring lines are placed in the superior vena cava or jugular veins. Such monitoring lines can result in thrombosis and have catastrophic consequences if pulmonary embolization occurs after a cavopulmonary anastomosis. We have used transthoracic lines positioned through the hemi-Fontan reconstruction into the pulmonary artery directly or into the pulmonary venous atrium by placing these catheters on either side of the homograft baffle at the superior vena cava-to-right atrium anastomosis. These lines are generally left in place for $<$24 hours. In all cases, operations are performed through a median sternotomy incision because there is little benefit to the thoracotomy incision for the construction of the cavopulmonary anastomosis. The majority of these patients require a later completion Fontan procedure and, therefore, the ease of creation of the anastomosis by sternotomy is not a disadvantage. In addition, we use right-sided systemic-to-pulmonary artery shunts at initial palliation and take down the shunt and place the cavopulmonary anastomosis at the shunt insertion site at reoperation through the same sternotomy incision. This eliminates the need for left-sided aortopulmonary shunts and the difficulty in takedown of these shunts with its potential risk to the left phrenic nerve.

A significant addition to the creation of cavopulmonary shunts or hemi-Fontan procedures is the use of modified ultrafiltration postoperatively. In our experience, the use of modified ultrafiltration markedly decreases the risk of pleural effusions after these operations and is associated with a mortality rate that approaches zero.

Currently, almost all patients undergoing the Fontan procedure have had intermediate staging with a cavopulmonary shunt. As the results with surgery for hypoplastic left heart syndrome have improved, the cavopulmonary shunts have been done at a younger age to decrease the risk of interstage mortality between the first-stage reconstruction and the second stage. It is unclear which age is the minimum at which one can complete a cavopulmonary shunt with low morbidity. Many centers routinely perform the cavopulmonary shunt at 3 to 4 months of age rather than 6 months of age, which was our practice in the past. In rare cases and with documented low pulmonary resistance, even earlier cavopulmonary shunt can be contemplated in some cases. Although the mortality does not appear to be higher with early cavopulmonary shunt, the morbidity does appear to be slightly greater, with a longer hospital stay and possibly more venous congestion early after the procedure. Nevertheless, early conversion to a cavopulmonary connection may decrease the morbidity of shunt thrombosis from aortopulmonary shunts and improve oxygen saturations in patients who have shunt stenosis and limited pulmonary blood flow.

At the Children's Hospital of Philadelphia, we continue to use the hemi-Fontan procedure in patients in which extensive pulmonary arterial reconstruction is necessary at the time of the second-stage reconstruction for hypoplastic left heart syndrome or other single ventricle abnormalities. With the increased use of the right ventricular to pulmonary artery shunt in the first-stage reconstruction for hypoplastic left heart syndrome, we have seen distortion at the pulmonary bifurcation to the left of the neoaorta in many patients. This can be very readily addressed with the hemi-Fontan operation where patch augmentation of the pulmonary bifurcation is a routine part of the operative procedure. The hemi-Fontan operation is designed to make the completion Fontan with a lateral tunnel procedure extremely straightforward. Thus, the use of a hemi-Fontan operation at second-stage reconstruction generally predisposes the patient to a lateral tunnel completion Fontan operation. If a bidirectional Glenn shunt is used as a second-stage reconstruction, then extracardiac Fontan completion operation is generally preferred. Some centers have divided the superior vena cava and anastomosed both ends to the right pulmonary artery with a dam in the atrial portion of the connection. This seems to be a suboptimal approach in our opinion since the superior vena cava orifice is not adequately sized to carry the entire vena caval flow with the use of a lateral tunnel connection. In some centers at the completion Fontan operation, the cavoatrial junction is enlarged. This seems to be an unnecessary step to us if the hemi-Fontan is used as a second-stage reconstruction. In addition, the enlargement of the cavoatrial junction has the same disadvantage of disruption of sinoatrial node blood supply, which can increase the risk of junctional rhythm early after the Fontan completion procedure.

An area of continued controversy regarding cavoatrial shunts is the potential decrease of right-sided AV valve regurgitation with the elimination of

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volume load on the single ventricle. There are conflicting reports in the literature regarding the effect of a bidirectional Glenn shunt on tricuspid regurgitation in patients with single ventricle physiology. In most cases, it does not appear that volume unloading of the ventricle actually significantly changes the amount of AV valve regurgitation. In only a small percentage of patients can significant reduction of AV valve regurgitation be documented.

The increasing use of the extracardiac conduit Fontan procedure has limited the use of hemi-Fontan operations in most centers. The long-term comparison of lateral tunnel Fontan operations done with a previous hemi-Fontan operation and extracardiac Fontan operations has not been performed. It appears that in both cases, the incidence of arrhythmias is now relatively lower postoperatively at long-term follow-up than in previous reports, and the use of extra cardiac Fontan connections has not completely eliminated the incidence of late arrhythmias.

TLS

95 Tricuspid Atresia/Single Ventricle and the Fontan Operation

Stephanie Fuller and Pradeep Kaushik

INTRODUCTION

Atresia of any heart valve, atrioventricular (AV) or semilunar, results in single-ventricle physiology. A variety of anatomic defects should be considered as a single ventricle from the standpoint of surgical interventions (Table 95.1). Patients born with a single functional ventricle have a dismal long-term prognosis without eventual surgical intervention. But the results of staged surgical palliation, culminating in the Fontan procedure as the final palliative surgical procedure for these patients, are generally quite good.

CLASSIFICATION OF SINGLE-VENTRICLE ANATOMY

The classic defect for which the Fontan operation was first performed is tricuspid atresia. In this defect, there is no identifiable tricuspid valve tissue or valve remnant. Instead, the floor of the right atrium is completely muscular, and the floor is separated from the ventricular mass by the fibrofatty tissue of the AV sulcus. Generally, in association with the atretic right AV valve, there is a total absence of the inlet and varying portions of the trabecular portion of the right ventricle.

There is almost always an associated ventricular septal defect (VSD), which is frequently restrictive. The great vessels may be transposed with the aorta arising from the infundibular chamber or normally related with the aorta arising from left ventricle (LV). The VSD is frequently restrictive, which results in restriction of pulmonary blood flow when the great vessels are normally related but results in subaortic stenosis when there is transposition of the great arteries (TGA). When transposition is present, even if the bulboventricular foramen (BVF) is initially large and unrestrictive, with time it can be expected to become smaller, resulting in subaortic stenosis later in life. If there is a coarctation present, then it is very likely that the BVF will be restrictive in the newborn period.

The natural history in tricuspid atresia is quite variable, but it is predicted by the underlying pathophysiology as in all single-ventricle hearts. Most patients with tricuspid atresia and normally related great arteries have some degree of obstruction to pulmonary blood flow. This generally progresses as the VSD becomes more obstructive or infundibular obstruction increases. As a result, there is increasing cyanosis. Without surgical intervention, over 90% of patients will die by 1 year of age from complications of hypoxia. Early palliation is, therefore, advocated for every neonate with tricuspid atresia. In the neonatal and early infantile period, when pulmonary vascular resistance (PVR) is high and pulmonary blood flow is inadequate, a systemic-to-pulmonary artery (PA, modified Blalock–Taussig) shunt is performed.

In contrast, those patients with tricuspid atresia and ventriculoarterial discordance (associated TGA) generally have a worse prognosis. Unobstructed pulmonary blood flow results in congestive heart failure and death within the first year of life. Subaortic obstruction complicates the scenario, further shortening the survival time. In the extreme case, there is ductal dependency of the systemic circulation in the newborn. In the case of excessive pulmonary blood flow, it is imperative to limit the pulmonary blood flow, usually with a PA, to protect the patient from developing pulmonary vascular disease and ventricular dysfunction caused by chronic volume overload.

Some children with single ventricles who present in infancy or later develop systemic ventricular outflow tract obstruction (subaortic stenosis). The resultant myocardial hypertrophy can adversely affect the suitability for a Fontan procedure. Patients at risk are those in whom the aorta arises above a small outlet chamber, such as in tricuspid atresia with double-inlet LV, rudimentary right ventricle, and TGA, particularly if the VSD is small or if there is coexisting aortic arch obstruction. A modified Damus–Stansel–Kaye procedure should be performed to establish unobstructed systemic arterial outflow.

PLANNING FOR SINGLE-VENTRICLE PALLIATION

Because the ultimate success of the Fontan operation depends on a suitably low PVR and adequate PA architecture, it is critical to commence the preparation for a Fontan procedure in the newborn period by appropriately regulating pulmonary blood flow and allowing for adequate growth of the PAs. The ultimate goal of palliative interventions leading up to Fontan operation are to improve clinical symptoms with simultaneously providing optimal PA architecture with low PVR, preserve systolic and diastolic ventricular function, avoid AV valvar regurgitation, and relieve systemic ventricular outflow obstruction. Thus, surgical management of patient with tricuspid atresia is based on the concept that systemic venous blood can be made to pass through healthy mature lungs without the assistance of a ventricular pump.

Physiologically, after the first few months of life, PVR generally falls, and then the goals of subsequent surgical interventions should be to reduce the volume load on the single ventricle. Once systemic arterial-to-PA shunts are eliminated, pulmonary blood flow is reestablished by creating direct connections between the venous system and the PAs (by means of a bidirectional cavopulmonary artery shunt or hemi-Fontan). Any residual anatomic problems such as PA distortion, obstruction to systemic blood flow, AV valvar regurgitation, or a restrictive atrial septal defect should be dealt with at the same procedure to simplify the "definitive" management (Fontan operation). Staging of the Fontan procedure is performed because of the high incidence of pleural effusions and ventricular failure that occurred when patients were taken

Table 95.1 Structural Defects That Are Managed by Staged Palliation

Tricuspid atresia with normally related great arteries	Hypoplastic left heart syndrome
Tricuspid atresia with transposition of the great arteries	Double-outlet right ventricle with mitral atresia
Double-inlet left ventricle with normally related great arteries	Unbalanced atrioventricular canal with hypoplastic left ventricle
Double-inlet left ventricle with transposition of the great arteries	Heterotaxy syndromes
Unbalanced atrioventricular canal with hypoplastic right ventricle	
Pulmonary atresia with intact ventricular septum	

directly from a neonatal single-ventricle palliation to the Fontan.

STAGE II OPERATION

Indications for Stage II Palliation

Infants who have had neonatal palliation become candidates for a second-stage operation by 3 to 6 months of age. Younger age at the time of second-stage surgery is associated with better exercise performance than when surgery is delayed until a later age. Adequate ventricular function remains a prime determinant for a successful Fontan circulation. Indications for second stage include cyanosis secondary to inadequate pulmonary blood flow after initial palliation, congestive heart failure from ventricular volume overload caused by either severe AV valve regurgitation or an elevated Q_p:Q_s. Prior to cavopulmonary anastomosis, an echocardiogram should be performed. If there is concern regarding the anatomy, patency, or tortuosity of the PAs, then cardiac catheterization is an indication. Cardiac catheterization is also recommended if there is a clinical concern that can be treated interventionally after initial palliation such as aortic arch obstruction, or cyanosis resulting from either decompressing veins or stent stenosis.

The two options for a stage II palliation are the bidirectional Glenn (BDG) and the hemi-Fontan. While they are similar physiologically, each procedure has some unique advantages and different surgical techniques. Typically, our choice at Children's Hospital of Philadelphia is to perform a BDG on most patients unless a pulmonary arterioplasty is required in which case a hemi-Fontan is the natural choice. Ultimately, the choice of stage II operation will dictate the type of Fontan performed. The BDG is converted to an extracardiac fenestrated Fontan, whereas the hemi-Fontan is easily converted to a lateral tunnel fenestrated Fontan.

Bidirectional Glenn

Glenn performed the first cavopulmonary connection. He used a unidirectional (classic) superior cavopulmonary anastomosis to the right PA. Eventually, this operation was modified to allow continuation of flow to both PAs. This has now become the standard of care for infants with single-ventricle physiology and route to end-stage completion.

The Glenn is performed on normothermic cardiopulmonary bypass, typically with a beating heart. Bicaval cannulation is performed and, once on cardiopulmonary bypass, the shunt is ligated. The azygos vein is ligated and divided. It is important to then maintain proper orientation of the superior vena cava (SVC) to avoid twisting and obstruction of the vessel. The SVC is divided from the right atrium and the cardiac end is oversewn. The distal end of the SVC is anastomosed to the ipsilateral PA in an end-to-side manner (Fig. 95.1). Typically, no prosthetic material is used when performing a Glenn. In cases of pulmonary distortion or narrowing, however, a separate patch of homograft may be necessary to augment the PA. However, if augmentation is necessary, we typically prefer to perform a hemi-Fontan in the cases of PA narrowing.

Hemi-Fontan

The hemi-Fontan procedure (anastomosis of the SVC with the PAs, augmentation of the PAs to eliminate any areas of distortion or narrowing, and elimination of systemic-to-pulmonary shunts or any other sources of pulmonary blood flow) is generally performed at the same time of the Glenn, ideally by 6 months of age. The main differences

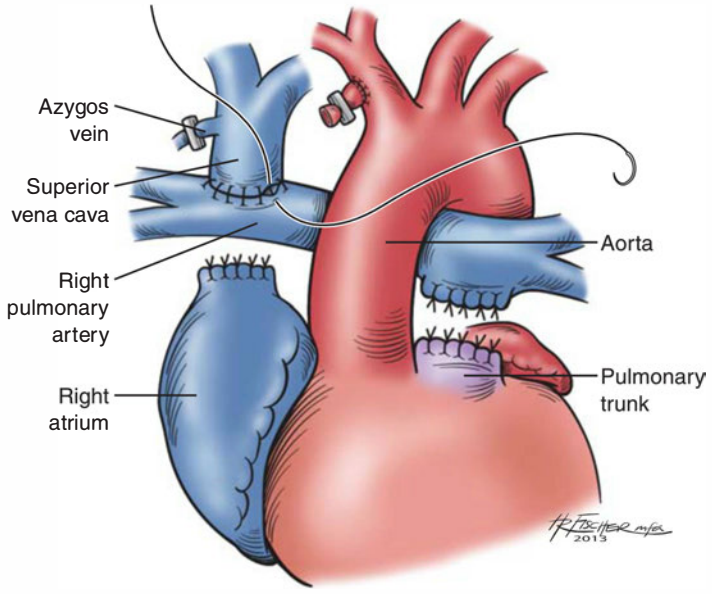


Fig. 95.1. Bidirectional superior cavopulmonary anastomosis (bidirectional Glenn). The superior vena cava (SVC) is transected at the level of the right pulmonary artery (RPA), the cardiac end is oversewn, and the cephalad portion of the SVC is anastomosed end-to-side into the proximal right pulmonary artery. Ao, aorta; IVC, inferior vena cava; PA, pulmonary artery.

between the hemi-Fontan and the Glenn are (1) mandatory use of homograft material for PA augmentation, (2) use of deep hypothermic circulatory arrest, and (3) no division of the SVC to right atrial junction. Alternatively, a "dam" is created using a homograft patch in the orifice of the SVC to obstruct draining directly into the atrium and force blood into the PAs.

The hemi-Fontan procedure is accomplished using hypothermic cardiopulmonary bypass combined with surface cooling before use of deep hypothermic circulatory arrest allowing for adequate visualization of the complex baffle. The PAs are freed from behind the aorta, and a longitudinal incision is made in the anterior aspect of the PAs extending from the level of the origin of the right upper lobe branch to the level of the origin of the left upper-lobe branch (Fig. 95.2A). An incision is made in the most superior portion of the right atrium and is extended onto the medial aspect of the SVC.

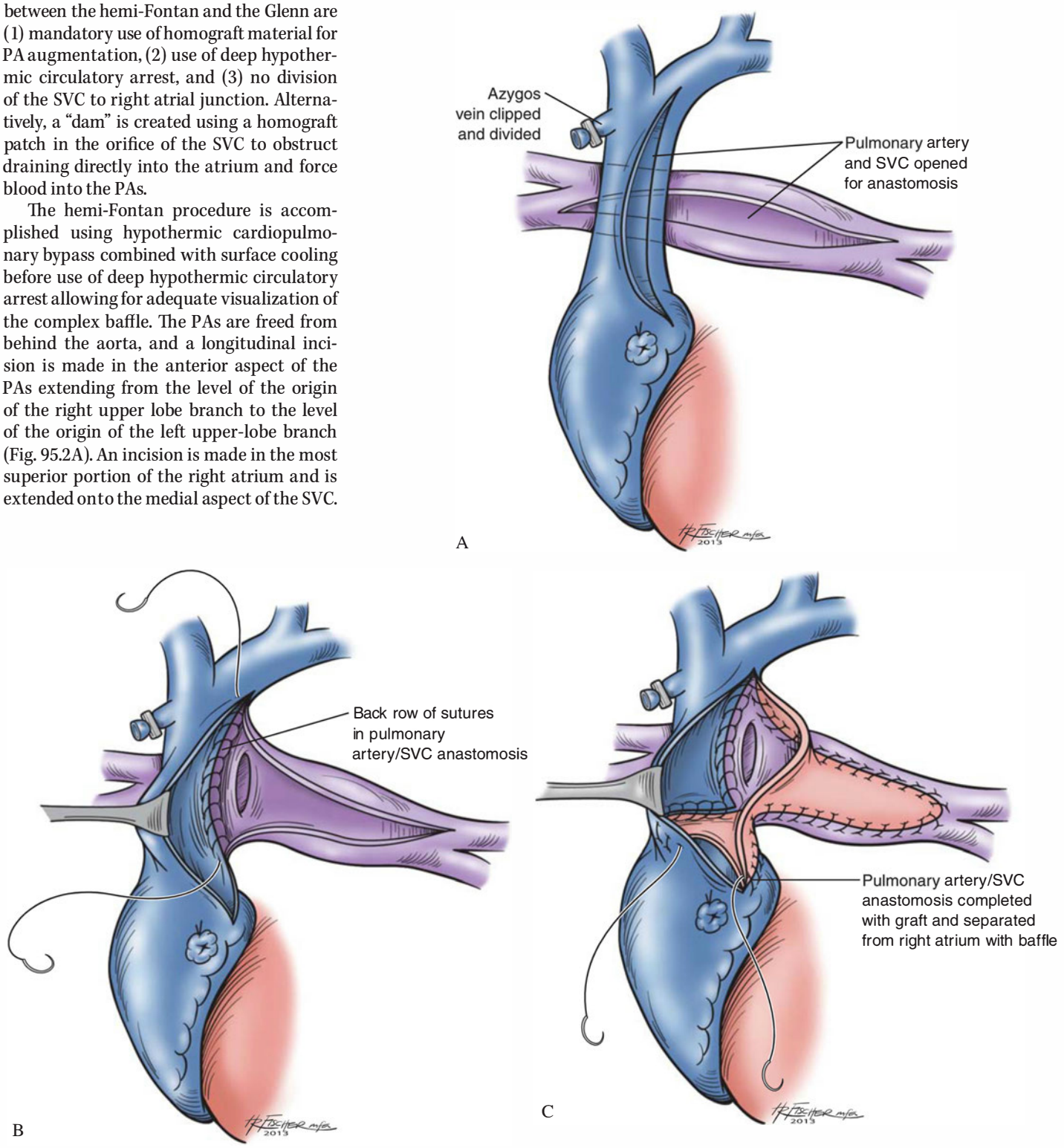


Fig. 95.2. Steps of hemi-Fontan procedure. (**A**) An incision is made in the pulmonary artery (a.) from hilum to hilum to allow the widest possible opening of the pulmonary bifurcation. An incision is also made in the medial aspect of the superior vena cava (SVC) across the cavoatrial junction onto the right atrial appendage, as delineated in this figure. The azygos vein (v.) is ligated and divided. (**B**) With the pulmonary artery widely opened and the incision in the SVC—right atrial junction, suturing is initiated connecting the posterior aspect of the SVC and right (R.) atrium to the right pulmonary artery by using a running 6-0 polypropylene suture. In this manner, a wide connection between the SVC and pulmonary artery can be ensured. (**C**) After the posterior suture line is created, a large triangular patch of pulmonary homograft is used to augment the pulmonary artery bifurcation. As the suture line along the inferior aspect of the pulmonary bifurcation is created, the suture line is carried into the cavoatrial junction, using the pulmonary homograft to create a dam between the SVC and right atrium. The suture line is brought completely across the cavoatrial junction to the anterior incision across the cavoatrial junction. Next, the homograft is folded down to create the dam at the level of the atrium and secured with a 5-0 polypropylene suture. Care must be taken with this suture line to incorporate the doubled flap of pulmonary homograft completely along the right lateral aspect of the suture line to prevent any baffle leaks entering the right atrium after the repair.

At the most rightward extent of the pulmonary arteriotomy, the right PA is anastomosed to the posterior lip of the opened SVC (Fig. 95.2B). A gusset of cryopreserved PA homograft is used to augment the confluence of the PAs and to create a roof over the patulous anastomosis of the PAs to the SVC. A portion of the same homograft gusset is rotated posteriorly and used as a dam to close the junction of the right atrium with the SVC, obligating superior vena caval return to flow exclusively through the branch PAs (Fig. 95.2C). When bilateral superior vena cavae are present, each is anastomosed to the ipsilateral branch PA using cryopreserved homograft to augment the anastomosis in a manner similar to that described above.

Concurrent Procedures

In some cases, additional surgical revision is required at the time of the second-stage operation. One of the most common lesions requiring revision is recoarctation of the aorta. In many centers, this is diagnosed by echocardiography and the diagnosis is confirmed by cardiac catheterization. We favor balloon dilation of the coarctation once detected as long as the patient is approximately 8 weeks from surgical reconstruction. If distal aortic arch recoarctation is neither suitable nor responsive to balloon dilation, patch augmentation at the time of the cavopulmonary anastomosis may be required.

Another indication for more extensive operation at the second stage is AV regurgitation. Often, unloading the volume stressed ventricle and performing the cavopulmonary anastomosis ameliorate the severity of regurgitation. However, in cases of severe regurgitation, inspection and possible intervention upon the AV valve is recommended. Commissuroplasty, annuloplasty, choral shortening, and leaflet extension are common valvuloplasty techniques to minimize regurgitation.

Postoperative Management

Conversion from an aortopulmonary shunt to a stage II operation decreases the work of the single ventricle by decreasing the volume load on the ventricle. Postoperative management includes judicious administration of fluids guided by cardiac filling pressures and early extubation, which usually can be accomplished in the operating room or within 3 to 5 hours after completion of surgery. Positive pressure ventilations can cause increased airway pressures and adversely affect both PVR and ventricular filling. Spontaneous breathing likely leads to an increase in PCO_2 that promotes cerebral blood flow therefore increasing SVC return to the lungs or pulmonary blood flow.

Typically, hemodynamics are robust. In fact, transient hypertension has been observed after the cavopulmonary shunt. One of the goals of postoperative management is to minimize the transpulmonary gradient to allow passive blood flow through the lungs guaranteeing return of oxygenated blood to the common atrium. An elevated transpulmonary gradient may cause cyanosis and could result from elevated PVR, downstream pulmonary venous obstruction, or from impaired lung expansion due to hemo- or pneumothorax. Elevation of PVR due to cardiopulmonary bypass may be decreased by the administration of nitric oxide as a pulmonary vasodilator.

Facial swelling and irritability (which is attributable to "headache" or "hangover" from transiently elevated venous pressure) often persist for a few days and are more common in younger infants. Also common, patients may develop demarcation of the upper body secondary to venous congestion in the upper chest, bilateral arms, and head. Persistent facial swelling, particularly of the fontanelle and eyelids, or the development of a chylothorax should prompt investigation for SVC narrowing. The gradient can usually be estimated by echo and cardiac catheterization pursued if necessary.

Diuretics are generally administered beginning on the first postoperative day. Hospitalization usually lasts a week or less. The late development of cyanosis can be the result of the development of atriovenous malformations or collateral venous drainage. Both may be treated by transcatheter coil embolization. Although it is a more extensive operation, the hemi-Fontan allows for the technically simpler later lateral tunnel Fontan procedure. It also has the potential advantage of allowing nonsurgical completion of the Fontan circulation in the cardiac catheterization laboratory.

FONTAN OPERATION

The last stage of reconstructive surgery for children with single-ventricle physiology is the Fontan completion. The modified Fontan circuit allows systemic return from both the upper and lower extremities to return to the PAs passively and directly by bypassing the heart. Therefore, there is complete separation of the systemic and pulmonary circulations. Improved patient selection, multiple technical modifications, and better postoperative management have reduced the mortality of the operation to <2% in many centers with acceptable postoperative morbidity and decreased length of stay.

History of the Fontan

Fontan and Baudet reported the first "physiologic" correction of tricuspid atresia in 1971. Shortly thereafter, in 1973, Kreutzer and colleagues reported the first Fontan procedure with a direct connection of the right atrium to the PA. In 1983, Norwood and colleagues reported the first successful Fontan in an HLHS patient.

The early Fontan operation, a direct atriopulmonary anastomosis was modified to an intra-atrial lateral tunnel because of the high incidence of tachyarrhythmia in the atriopulmonary circuits. The most recent modification is the insertion of an extracardiac conduit from the IVC to the PA. This technique eliminated the need for suture lines in the right atrium, thereby minimizing the incidence of sinus node dysfunction and both atrial flutter and fibrillation after the Fontan.

In those patients who went directly to a Fontan without the intermediate superior cavopulmonary anastomosis, outcomes were not as good. The acute decrease in single-ventricle end-systolic and end-diastolic volume without a change in ventricular mass or wall thickness results in impaired diastolic function and low cardiac output. Many of these patients suffered from prolonged pleural effusions ultimately resulting in hypoalbuminemia, volume overload due to replacement, and Fontan failure. Over the last two decades, the most significant modification was the formation of a fenestration in either pathway. The fenestration allows for a communication between the systemic venous and pulmonary venous pathways, creating a right-to-left shunt. At the cost of desaturation, approximately the same as that with a cavopulmonary anastomosis pre-Fontan, the fenestration reduces mortality and morbidity by decreasing the incidence of pleural effusion and increasing the ventricular preload by shunting.

Indications for Surgery

Traditionally, candidates for the Fontan completion include those with normal ventricular function, minimal AV valve

regurgitation, normal PAs, and low PVR. Yet many less than "ideal" candidates are rendered suitable for the Fontan operation given proper perioperative management. Risk factors for poor outcome after the modified Fontan operation include ventricular hypertrophy, elevated PVR or PA pressure, PA distortion, AV valve regurgitation, and ventricular dysfunction. Severe ventricular dysfunction and fixed elevated PVR (>4 Wood Units) remain the most significant contraindications to surgery. Historically, patients underwent preoperative echocardiogram and cardiac catheterization to assess anatomic and hemodynamic suitability. Any lesions that can be intervened upon by catheterization such as recoarctation of the aorta, PA coarctation opposite the Fontan site or cavopulmonary narrowing is addressed at the time of catheterization.

Lateral Tunnel Fontan

For the lateral tunnel Fontan completion, which patients undergo approximately 18 to 24 months after the hemi-Fontan, minimal dissection is needed because of the preparative nature of the hemi-Fontan stage (Fig. 95.3A). We choose to perform this operation under a brief period of deep hypothermic circulatory arrest with cannulation of the aorta and the common atrium. An incision is made in the lateral wall of the right atrium, anterior and parallel to the sulcus terminalis, extending inferiorly to the level of the Eustachian valve of the IVC and superiorly to within a few millimeters of the junction of the right atrium and the SVC, thus exposing the homograft dam that divides the right atrium and SVC. The portion of the PA homograft that was used as a dam is excised to expose internally the SVC and the branch PAs (Fig. 95.3B). An appropriate length segment is then cut from a portion of a 10-mm polytetrafluoroethylene (PTFE) tube graft that has been opened longitudinally (Fig. 95.4). This is used to create a lateral atrial tunnel or intra-atrial baffle by which inferior caval flow is directed to the reopened atrial inlet of the SVC and thus to the PAs (Fig. 95.5). This suture line is then completed by incorporating the walls of the right atrium into the superior edge of the lateral tunnel to close the atriotomy (Fig. 95.6).

Two important features of this type of lateral atrial tunnel Fontan are (1) ensuring that the opening of the right atrium into the SVC and PAs is as large as the inferior vena caval opening and (2) avoiding direct suturing to the sulcus terminalis, which has been implicated in the development of postoperative tachyarrhythmia's. In most

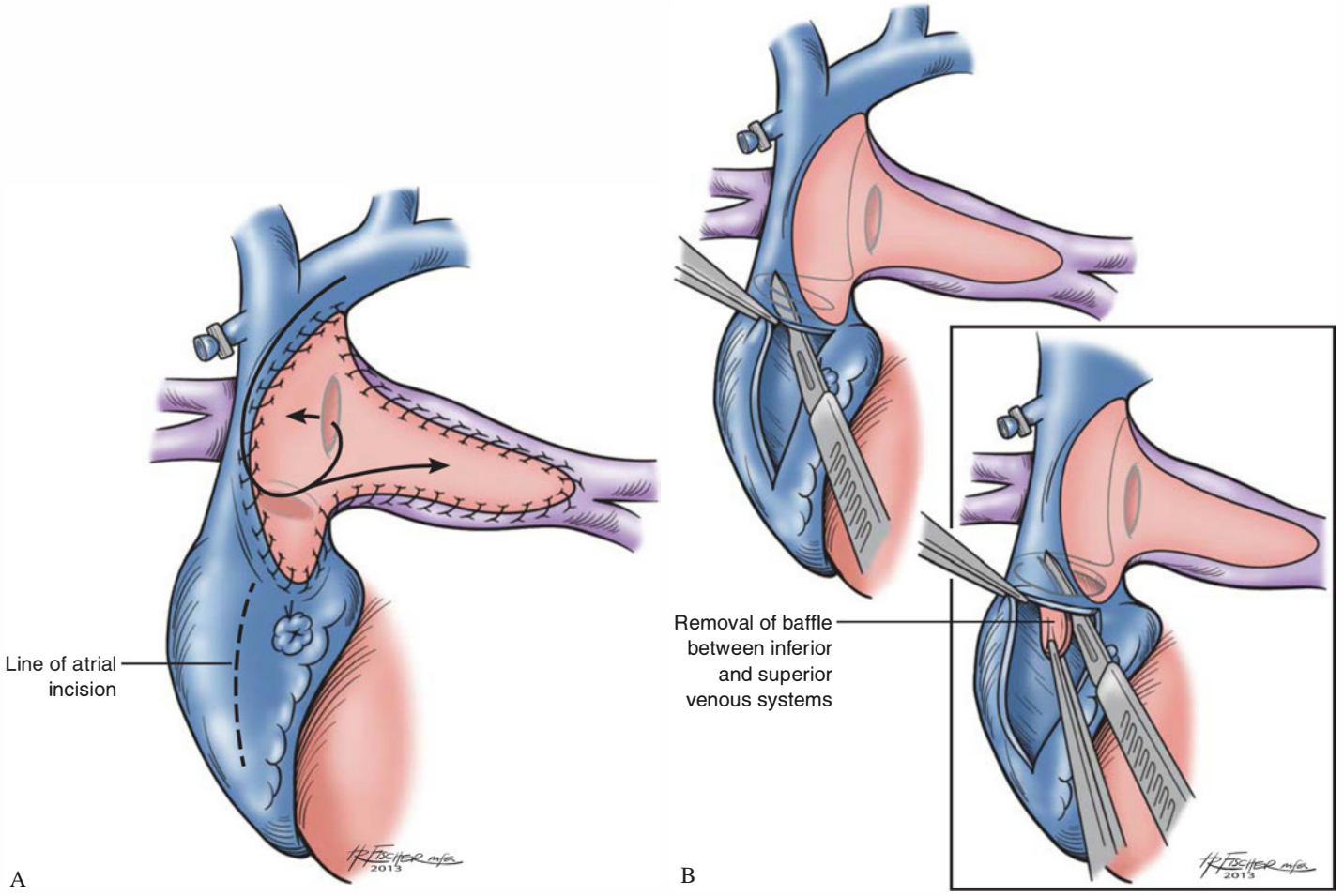


Fig. 95.3. Lateral tunnel completion Fontan. (**A**) Working through the right atrial incision, the homograft dam between the superior vena cava and right atrium is readily identified. (**B**) This dam of tissue is excised under direct vision, creating an opening that is adequate in size for baffling of the inferior vena cava flow into the pulmonary arteries. This opening is created in such a way that a rim of tissue can be used for the suture line of the intracardiac lateral tunnel (*inset*).

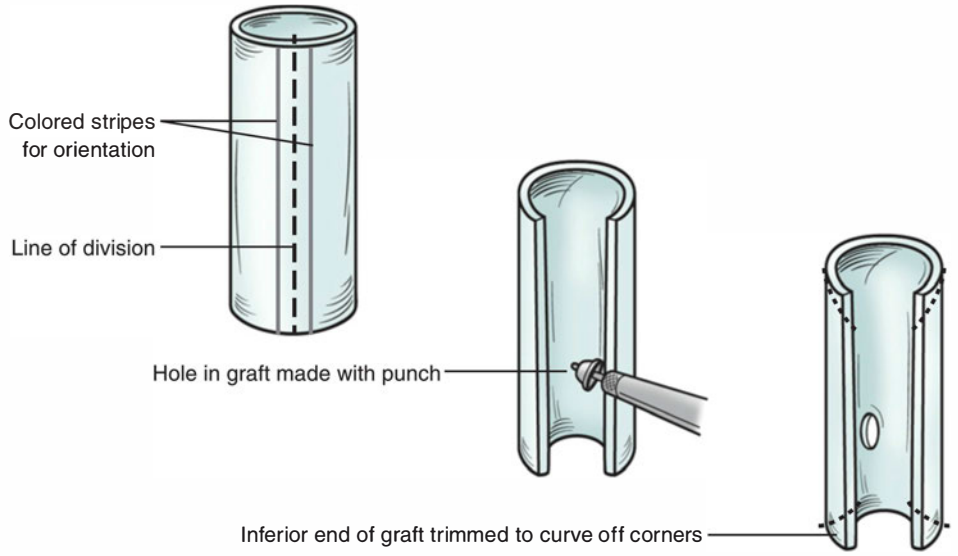


Fig. 95.4. Lateral tunnel completion Fontan. A polytetrafluoroethylene graft 10 mm in diameter is opened longitudinally, so that a larger lateral tunnel can be created than the diameter of the graft used. Before implantation, an incision is made in the left lower portion of the graft, and a 4-mm punch hole is made in the graft, which can be easily performed before implantation and avoids manipulating the graft after it is sewn in place. The inferior rim of the graft is then trimmed to a gentle curve and sewn into the lateral aspect of the right atrium.

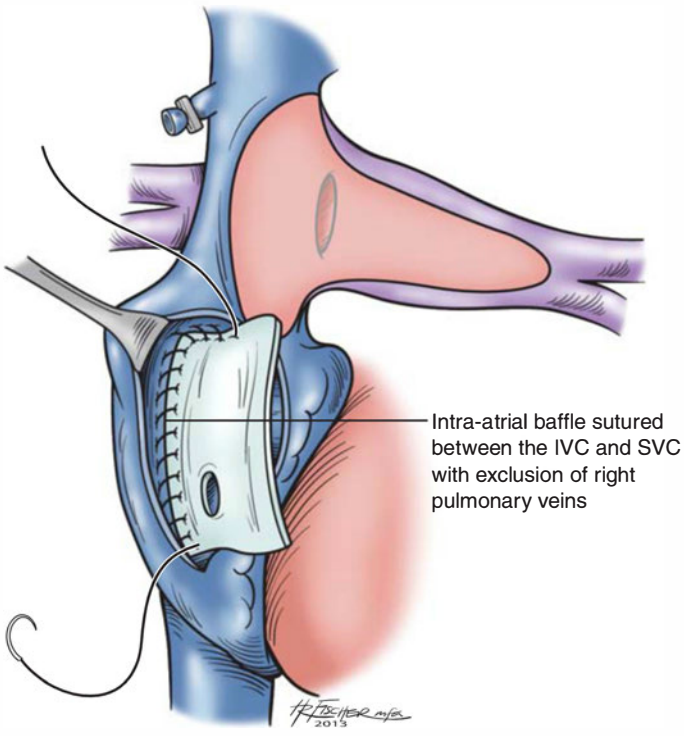


Fig. 95.5. Lateral tunnel completion Fontan. The opened polytetrafluoroethylene graft with the previously created fenestration is secured in the right atrium, excluding the right pulmonary veins behind the baffle. The suture line along the posterior aspect of the right atrium, which is then carried down inferiorly to the eustachian valve, where the graft is sutured along the eustachian valve up to the margin of the incision in the right atrium. Superiorly, the graft is sutured along the wall of the atrial septum up to the level of the excised dam in the cavoatrial junction. Then the graft is brought to the right and sewn to the margins of the previous excision of the homograft dam up to the level of the incision in the right atrium. The graft at this point can be trimmed if it is excessively wide and then the suture line continued, sandwiching the graft-free margin between the two walls of the right atrium to complete the lateral tunnel suture line. After completion of this suture line, the atrium can be filled with the saline solution and the venous cannula reinserted. IVC, inferior vena cava; SVC, superior vena cava.

instances, we choose to fenestrate the lateral tunnel prior to insertion and usually accomplish this by creating one hole in the gusset using a 4-mm punch device (Fig. 95.4). Fenestrations of this size promote decompression of the venous pathway and enhance ventricular filling postoperatively. They usually close spontaneously, avoiding the need for device placement in the venous pathway. As previously mentioned, several studies have shown a significant decrease in pleural effusions and hospital stay compared with nonfenestrated patients.

The completion of Fontan procedure as described here can generally be done with a period of hypothermic circulatory arrest of about 20 minutes duration. Total cavopulmonary connection had mainly laminar flow patterns. In this design, the superior vena caval blood drains directly into the PA, and inferior vena caval blood is baffled through a straight intra-atrial conduit into the PA. Additional theoretical advantages are a reduced risk of thrombosis because of less blood stasis, and exposure of only a limited portion of the right atrium to high

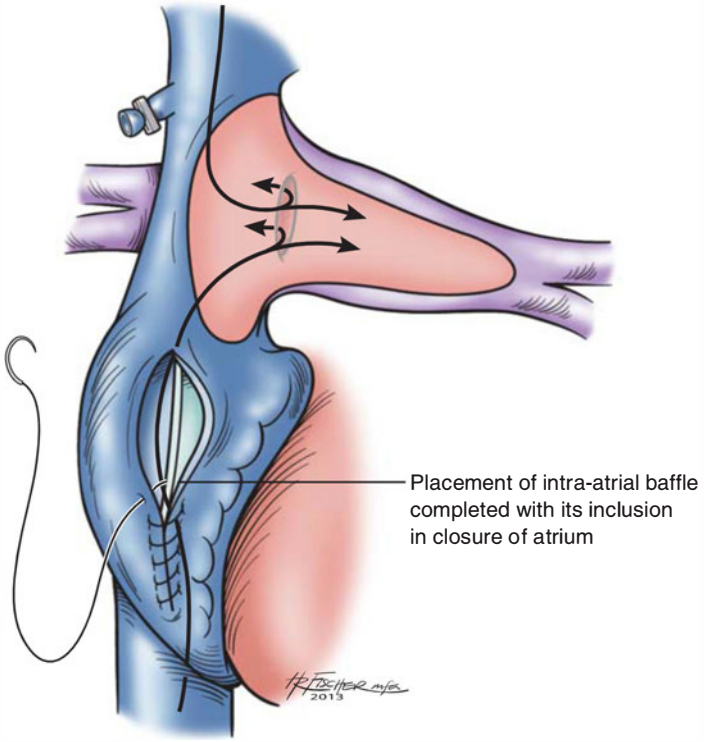


Fig. 95.6. Lateral tunnel completion Fontan. An advantage of incorporating the wall of the lateral tunnel in the suture line is the ability to place lines easily for postoperative monitoring. Transthoracic atrial lines are positioned on either side of the polytetrafluoroethylene baffle, which places one line on the left atrial side of the baffle and the other on the pulmonary arterial side for the measurement of pressures postoperatively. In this manner, no lines must be placed directly in the internal jugular vein, innominate vein, or superior vena cava, reducing the risk of postoperative caval thrombosis.

venous pressures, thus reducing the risk of arrhythmias Also, because the coronary sinus remains in the lower pressure pulmonary venous atrium, there is unobstructed myocardial venous drainage.

Extracardiac Fontan

For those children who have undergone a BDG as their second-stage operation, we favor an extracardiac Fontan completion. Similar to the lateral tunnel Fontan completion, this is performed at approximately 2 years of age. Although performed using bicaval cannulation at some institutions, we prefer to perform the extracardiac Fontan under deep hypothermic circulatory arrest as well. An 18- or 20-mm PTFE conduit is used and a 4-mm punch for fenestration on the medial aspect of the conduit at approximately the lower half of the conduit. In preparing the conduit, the outer layer of PTFE is stripped from around the hole to avoid thrombogenicity at the anastomosis of the fenestration. The IVC must be extensively mobilized at the level of the hepatic veins and divided at its entrance into the right atrium. The atrium is closed using a running suture to approximately the last centimeter of atrium. And end-to-end anastomosis is completed between the transected IVC and the distal conduit (Fig. 95.7). A side-to-side anastomosis is performed between the fenestration in the conduit and the atrium. The underside of the right PA directly across from the Glenn anastomosis is incised and the proximal graft sutured to the inferior aspect of the PA. Typically, the graft is beveled toward the main PA segment. In addition, the underside of the graft is cut with a concave curvature to allow more natural filling of the PAs and slight elevation of the graft over the right pulmonary veins. The completed extracardiac Fontan is shown in Figure 95.8.

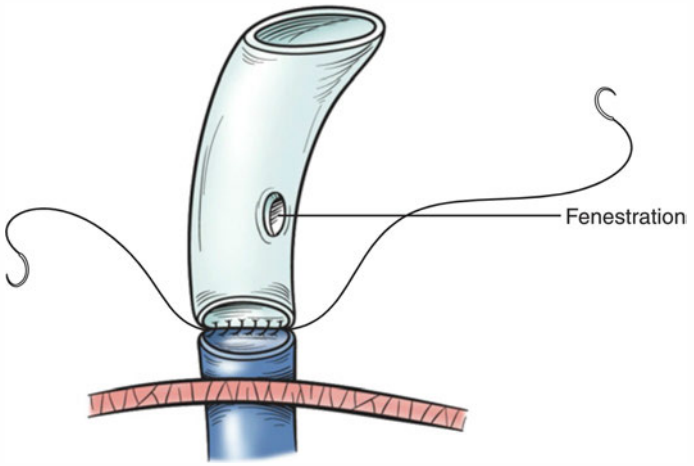


Fig. 95.7. Extracardiac Fontan completion. The cardiac end of the transected inferior vena cava (IVC) is doubly oversewn with running 4-0 or 5-0 nonabsorbable monofilament suture. The extracardiac conduit of polytetrafluoroethylene vascular tube graft is then tailored. The size of the conduit usually ranges from 18 to 22 mm in diameter. The cross-sectional diameter of the conduit is usually slightly oversized with respect to the diameter of the IVC. An end-to-end anastomosis between the conduit and the IVC is performed with 4-0 or 5-0 monofilament suture. The length of the conduit is then tailored to the undersurface of the right pulmonary artery. The superior orifice of the conduit is beveled at an angle of ≤45 degrees relative to the long axis of the conduit. This allows an oblique conduit-to-pulmonary artery anastomosis with a substantially greater anastomotic surface area rather than with direct end-to-side connection. In addition, this allows greater offsetting of the superior and inferior cavopulmonary connections, which has been shown by computational fluid dynamics to minimize energy losses at the cavopulmonary junction. Furthermore, this oblique anastomosis effectively serves as a central pulmonary arterioplasty, and the angle of the bevel can be adjusted to maximize this effect.

Associated Lesions

Patients undergoing a third stage are at risk of the same associated lesions as at the time of the second stage. With an older and larger patient, however, many of these can be managed interventionally during cardiac catheterization. The most common associated lesions are left PA stenosis either from compression by the neoaorta or constriction of the ductus arteriosus, AV valve regurgitation, and neoaortic arch obstruction. Therefore, if necessary, patients should undergo left pulmonary

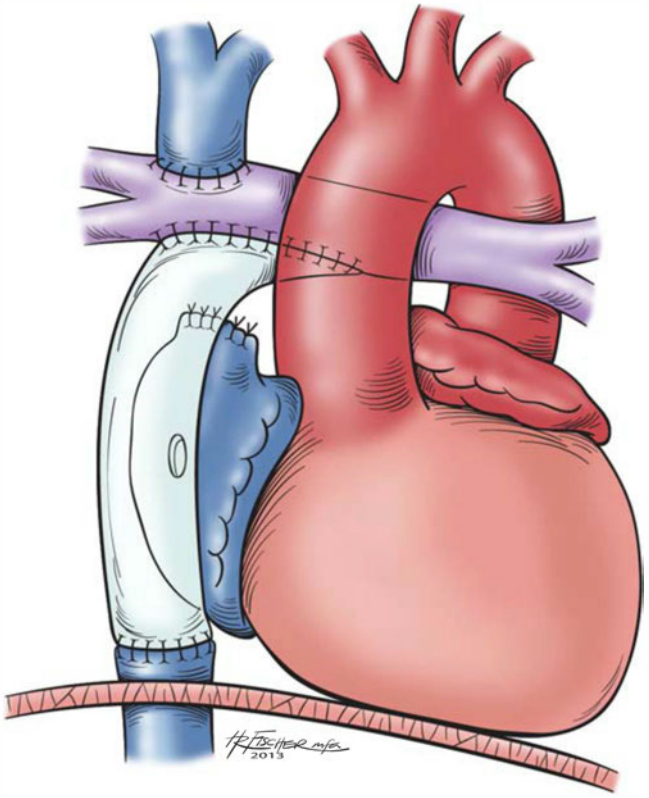


Fig. 95.8. Extracardiac Fontan completion. The completed extracardiac conduit Fontan is shown. A fenestration is placed by performing a 4- to 5-mm side-to-side anastomosis between the conduit and the right atrial free wall.

arterioplasty, valve repair, or arch augmentation at the time of the Fontan.

Postoperative Management

Postoperative care for the Fontan is not so different from that for a stage II operation. Ideally, patients should maintain high cardiac output with low atrial filling pressures. However, the same ventricular changes occur when offloading the ventricle at the Fontan as do at the BDG or hemi-Fontan. Immediate issues are maintaining hemodynamic stability with minimal inotropic support, hemostasis, and early extubation. Low cardiac output may be attributable to hypovolemia or inadequate preload, elevated PVR, anatomic obstruction of the systemic venous pathway, or ventricular failure. Atrial catheters in the systemic atrium and the Fontan pathway help to delineate the etiology.

RESULTS

Early mortality after the Fontan procedure has steadily come down to 0% to 5.5%. This is in spite of liberalizing patient selection criteria and extending the procedure to many forms of complex single ventricle. Multiple factors contributed to this improved early outcome. Certainly, volume unloading of the single ventricle by an early BDG or hemi-Fontan procedure has markedly improved the suitability for a subsequent Fontan operation. The introduction of total cavopulmonary connection with its energy efficient circulation, with the use of lateral tunnel or extracardiac conduit, has clearly been a huge advance in the improved outcome of the Fontan operation.

Pleural effusions constitute the most common early morbidity after the Fontan operation and often dictate the length of stay. The use of a baffle fenestration has been reported to reduce the duration of postoperative chest tube drainage. Replacement of the fluid with intravascular saline, albumin, clotting factors, and immunoglobulins may all be necessary in cases of prolonged drainage. Correctable causes such as baffle obstruction, innominate or SVC vein thrombosis, fenestration occlusion, and PA distortion should all be ruled out. Thoracic duct ligation or coil embolization with lymphangiogram is rarely needed.

After the Fontan operation, patients are at risk for venous thrombosis secondary to transient liver dysfunction and coagulation abnormalities due to the low flow state through the cavopulmonary circulation, presence of atrial arrhythmias and fresh suture lines. This can lead to increased incidence of perioperative stroke. It is important to avoid dehydration and maintain adequate preload. Anticoagulation is, therefore, usually advocated after the immediate postoperative period with aspirin and in some cases warfarin.

Sinus node dysfunction and atrial arrhythmias occur frequently after the Fontan procedure, particularly in patients with heterotaxy syndrome. Atrial pacing may be required in the perioperative period. Many studies support, based upon both univariate and multivariate analysis, that patients undergoing extracardiac Fontan are more likely to maintain sinus rhythm compared with those undergoing lateral tunnel Fontan.

Rogers et al. looked at our single institution experience with the Fontan in 771 consecutive patients. Despite increasing patient complexity, mortality was low at 3.5% throughout the series but only 1% since 1996, the current era. The likelihood of morbidity, mainly persistently draining pleural effusions, was similar to that experienced in other contemporary series. Many other institutions have published their contemporary results, as depicted in Table 95.2, and there clearly appears to be an era effect.

The incidence of late sequelae and complications related to the Fontan procedure and single-ventricle physiology are not yet completely known nor the mechanisms thereof understood. Patients are at risk for the development of protein-losing enteropathy and plastic bronchitis. Despite normal systolic function and even in cases of preserved cardiac output, Fontan patients will have failing physiology as a result of diastolic dysfunction and noncompliance of the single ventricle.

CONCLUSION

The mortality rate of the Fontan operation for tricuspid atresia and other complex forms of single-ventricle physiology has been drastically reduced by advances both technical and perioperative. The second-stage procedure allows early reduction of the volume work of the single ventricle and remodeling of ventricular geometry before completion of Fontan operation. The introduction of the fenestration to maintain ventricular preload has led to a marked decline in postoperative complications, namely, the development and duration of prolonged pleural effusions.

Table 95.2 Summary of Contemporary Reports of Early Postoperative Outcomes

	Cohort years	*N*	Mortality (%)	Takedown (%)
Rogers et al.	1992–2009	771	3.5[a]	1.0
O'Brien et al.	1997–2008	145	5.5	2.8
Hirsch et al.	1992–2007	636	4.0	3.0
Tweddell et al.	1994–2007	256	2.0	0.8
Salvin et al.	2001–2005	226	0.9	0.9
Petrossian et al.	1992–2005	285	1.1	1.4

[a]1% since 1996.

The long-term results of the Fontan procedure are not yet known. While early failure of the Fontan requiring takedown is now rare, late morbidities are pervasive. Additional palliative strategies have been developed. For example, the development of late arrhythmias in lateral tunnel patients can be treated by Fontan conversion to an extracardiac tunnel. And late development of either semilunar or AV valve regurgitation may be treated with replacement. However, for those patients who have no further palliative options, cardiac transplant is recommended. It is important to note that all those patients must undergo thorough evaluation of hepatic function and fibrosis to rule out the need for dual organ transplant secondary to chronic hepatic changes as a result of the Fontan circulation.

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EDITOR'S COMMENTS

The Fontan operation has been a major advance in the treatment of complex congenital heart disease. It is applicable to multiple forms of complex defects, including all forms of single ventricle and situations in which biventricular repair is either not feasible or is associated with long-term complications and late reoperation. Multiple modifications of the Fontan procedure have permitted a marked improvement in late morbidity and mortality, such that the Fontan operation is associated with a mortality of <5% in most centers and long-term results remain quite good despite the lack of a ventricle pumping blood through the pulmonary circulation.

In spite of these very salient features of the Fontan operation, multiple modifications have been recommended to decrease late morbidity. Perhaps, the most significant advance in the Fontan operation has been the advent of a fenestration in the intra-atrial baffle, which has been shown to maintain ventricular preload in situations of elevated pulmonary vascular resistance and improve cardiac output. Initial use of the fenestration in high-risk Fontan procedures at Children's Hospital Boston was associated with a marked improvement in the incidence of pleural effusions and a decreased incidence of takedown of the Fontan operation even in "high-risk" Fontan candidates. Although initial approaches were directed at closure of the fenestration at a later catheterization procedure, it is interesting that in all patients with temporary occlusion of the fenestration, cardiac output dropped, suggesting that there is a potential benefit of maintaining ventricular preload with a fenestration. For these reasons, we use fenestrations in all Fontan operations, and although spontaneous closure occurs in at least 60% of patients with a 4.0-mm fenestration of an intra-atrial baffle, it is unclear whether closure of the fenestration is necessarily beneficial in all patients. This controversy is heightened by the fact that patients who have developed protein-losing enteropathy after the Fontan operation or who have persistent effusions that are not resolving with medical management may have complete resolution of the problems by creation of a fenestration in a previously nonfenestrated atriopulmonary connection. This fact would suggest that the optimal hemodynamics with fenestration of the atrial baffle may be very beneficial in the long term for Fontan patients. Arterial desaturation may be an acceptable price to pay for adequate oxygen delivery with improvement in cardiac output in these patients if late morbidity is in fact improved.

A major issue for long-term function after the Fontan operation is the incidence of atrial flutter and fibrillation, which can compromise ventricular function and cardiac output. Much attention has been directed to the contribution of atrial

(*continued*)

EDITOR'S COMMENTS (*continued*)

suture lines to the development of atrial arrhythmias. The creation of extensive atrial suture lines has resulted in areas of conduction block that may contribute to atrial flutter. For this reason, some centers have advocated the use of extracardiac connections from the IVC to the pulmonary artery to completely eliminate pressure loading of the right atrium and to eliminate suture lines in the right atrium. Long-term follow-up of these extracardiac connections is not available, however, and longitudinal growth may result in constriction of the conduits and require late conduit replacement in some cases. In addition, it is difficult to fenestrate extracardiac Fontan connections with a satisfactory degree of long-term patency. Direct anastomosis to the right atrial wall results in rapid spontaneous closure, and a graft from the extracardiac conduit to the right atrium may also thrombose rapidly. We have modified our technique for creation of a fenestration with an extracardiac conduit by creating a medial fenestration in the extracardiac graft and then suturing the residual opening from the IVC entrance into the right atrium around the graft, approximately 4 to 5 mm from the edges of the fenestration. In this manner, the fenestration is exposed directly to the interior of the right atrium with a small rim of graft material separating it from the suture line. This appears to decrease the risk of stenosis and thrombosis of the fenestration. Nevertheless, in our own experience of over 770 Fontan operations, the use of an extracardiac conduit with fenestration results in a less consistent fenestration and a slight increase in the duration of postoperative pleural effusions. The use of a lateral tunnel with a fixed fenestration of 4 mm has been associated with the lowest incidence of prolonged pleural effusions and the shortest hospital stay in our experience. If a hemi-Fontan procedure done with a homograft baffle from the SVC to the right atrium is used as an initial-stage procedure, extracardiac Fontan connection can be performed with the creation of a fenestration through the "dam" between the SVC and right atrium at the hemi-Fontan connection and results in adequate fenestration patency over the intermediate term in most cases, although a lateral tunnel Fontan approach is certainly preferable.

The long-term benefits from Fontan reconstruction remain to be determined; however, it is apparent that the late results with single-ventricle repairs have been markedly better than most surgeons would have anticipated. Although a certain population of patients will develop protein-losing enteropathy late after the operative procedure, the incidence of this condition at intermediate follow-up is relatively low. Thus, the Fontan operation has become a mainstay of repair for complex congenital heart disease, and continued refinement is being undertaken to optimize late hemodynamic results. Modification of atrial suture lines and division of potential conduction zones for atrial flutter may be added to the Fontan operation in the future to prevent late arrhythmias. In addition, diversion of inferior vena caval flow preferentially to the right lung with superior vena caval flow directed to the left lung may optimize flow distribution in the pulmonary arteries after the operation. The use of lateral tunnel techniques has decreased the incidence of significant stasis in the right atrium, and giant right atrium after the operation, although rare, must be taken down to create the lateral baffle superior to the right pulmonary vein entrances to prevent compression of the pulmonary veins and late stenosis. Failure of Fontan operations requiring takedown is now extremely unusual with attention to staging of the operations and addressing of significant sources of pulmonary artery distortion before a complete Fontan procedure.

There has been great interest in modifying the Fontan operation to provide the most optimal hemodynamic efficiency of flow into the pulmonary vascular bed. Many studies using MRI flow calculations and distributions have suggested that various Fontan modifications can be adapted to individual patients to improve postoperative hemodynamics. In some cases, the use of the Y-shaped graft can optimize flow into each pulmonary artery from the inferior vena cava and in situations of heterotaxy syndrome with interrupted inferior vena cava and azygos continuation, direct connection of the hepatic veins to the azygos vein to ensure distribution of hepatic flow to both lungs has been suggested. It is now possible to model the cardiac anatomy in patients prior to the completion Fontan operation in order to define the best individual anatomy for the patient. In most cases, an extracardiac conduit is the simplest way of reconstructing the Fontan circulation since it allows for any pulmonary venous anatomy and can be directed to either pulmonary artery if necessary. While there has been a great deal of interest in the extracardiac conduit as an improvement over lateral tunnel operations in terms of late development of atrial arrhythmias, long-term follow-up studies with contemporary patients using both surgical techniques have not been reported. Most series that have compared the lateral tunnel with the extracardiac conduit Fontans have a significantly longer follow-up in the lateral tunnel group. It is known that atrial arrhythmias are more frequently followed by the longer patients after the Fontan operation and therefore this may be a confounding factor. Nevertheless, fewer atrial suture lines and the lack of exposure of the atrium to high pressure are likely to improve the incidence of late arrhythmias. It should, however, be noted that even in the extra cardiac Fontan patients, previous atriotomy and atrial septal resections have been performed in most cases and these are highly arrhythmogenic incisions. The late follow-up of the extra cardiac Fontan has continued to show arrhythmias in a significant proportion of patients.

The late complications of the Fontan operation including protein-losing enteropathy and AV valve regurgitation are concerning. Patients who develop protein-losing enteropathy may often have a progressive course to die within 3 to 5 years of diagnosis. Many attempts have been made to improve the hemodynamics in these patients including creation of a fenestration (which is poorly tolerated in a patient who has not been previously cyanotic), infusion of anticoagulants, pacing to improve cardiac output, and recently the use of enteric steroid therapy, which may improve the enteral protein loss. Most of these therapies have been largely palliative but can in some cases significantly improve the patient's quality of life.

TLS

96 Hypoplastic Left Heart Syndrome

Peter J. Gruber and Thomas L. Spray

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) comprises a wide spectrum of anatomic abnormalities with the common features of left ventricular hypoplasia and hypoplasia of the ascending aorta. At one end of the spectrum, there may be some mild left ventricle hypoplasia, mild aortic stenosis, and aortic coarctation. At the other end of the spectrum, however, there is complete absence of the left ventricle, aortic atresia, and aortic arch hypoplasia or even interrupted aortic arch.

HLHS is a uniformly fatal disease if untreated. It represents 5% of all congenital heart disease and is responsible for nearly 25% of cardiac deaths in the first week of life. Of 10,000 live births, approximately 1.8 will be born with HLHS, with a slight male predominance. Of these, 25% will also have a noncardiac anomaly and 5% a chromosomal abnormality (trisomies 13, 18, and 21). Syndromic lesions are rare, with Turner syndrome (monosomy X) the most common. The recurrence risk is 2.2% for one affected sibling and 6% for two affected siblings, suggesting some genetic predisposition but arguing against a simple effect.

Surgery for HLHS is one of the great successes in the management of congenital heart disease. Before the 1980s, HLHS was a uniformly lethal condition. However, over the last 25 years the repair of HLHS has become a standard operation in nearly all institutions. In 1952, Lev first described maldevelopment of the left-sided cardiac structures in combination with a small ascending aorta and transverse arch. By 1958, Noonan and Nadas had further defined the syndrome to describe a variety of cardiac malformations of left heart structures. The first report of any attempt to palliate a patient with mitral atresia was by Redo in 1961, who performed an atrial septectomy using inflow occlusion through a right thoracotomy; the patient died soon after the operation. In 1968, Sinha outlined the management principles still in use today that include creation of an unobstructed atrial communication, unrestricted ductal flow, and control of pulmonary blood flow. Cayler described an anastomosis between the right pulmonary artery (RPA) and ascending aorta with banding of both right and left pulmonary arteries (LPAs). It is of interest that 35 years later, pulmonary artery (PA) banding is being used in certain centers for selected children who present with a medical or anatomic situation unsuitable for stage 1 Norwood reconstruction; this first-stage hybrid procedure involves stenting the ductus arteriosus and atrial septal defect and using bilateral PA bands. Litwin, Mohri, and others performed operations that were variations of the principles of palliation that were unsuccessful but contributed to the development of the knowledge of the disease and its repair. In 1977, Doty described primary reconstruction that included atrial septation and a right atrium (RA)-to-PA Fontan circuit. Again, although no patients survived, this experience established the principle that one-stage reconstruction with a Fontan repair would not be successful due to high neonatal pulmonary vascular resistance. Levitsky, Behrendt, and others described multiple variations of surgical procedures that, although they demonstrated no long-term success, established the principle of staged reconstruction with initial palliation followed by later separation of the systemic and pulmonary circulations. However, it was Norwood who in 1980 first achieved successful palliation in infants. In 1983, he described the first successful staged approach culminating in a Fontan repair. Today, the Norwood procedure remains the primary reconstructive approach.

ANATOMY

Patients with HLHS are currently categorized on the basis of atrioventricular (AV) and semilunar valvular morphology into three primary subsets: (1) aortic atresia with mitral atresia (40%), (2) aortic stenosis with mitral stenosis (30%), and (3) aortic atresia with mitral steno sis (30%) (Fig. 96.1).Aortic stenosis with mitral atresia is rare. HLHS variants include malaligned AV canal, double-outlet right ventricle with mitral atresia, tricuspid atresia with transposed great arteries, and univentricular heart with aortic stenosis. There is frequent leftward and posterior deviation of the septal attachment of the septum primum, but this feature is unlikely to be a causal developmental mechanism because it is commonly seen in other congenital heart disease phenotypes. Usually, the superior vena cava (SVC) and inferior vena cava (IVC) are normally connected to the RA, although in about 15% of patients a left SVC-to-coronary sinus is present. Other structural abnormalities of the heart are rare, with $<5\%$ of patients demonstrating AV valvular dysplasia. Also, rare in nonsyndromic forms ($<5\%$) are abnormalities of pulmonary venous return or an interrupted aortic arch. Abnormalities in brain development are increasingly associated with children with severe congenital heart disease, and these may be a high-risk group for operative repair. The pulmonary vascular tree is also abnormal, with an increase in number of vessels as well as muscularity.

The developmental mechanisms that underlie HLHS are obscure from a molecular standpoint because there are no mutations that have been robustly associated with this condition. Despite the existence of rare family clustering of HLHS, linkage analysis has been largely unproductive. Indeed, it is likely that there exists considerable genetic heterogeneity in this as well as other CHD phenotypes. However, embryologically, there are clues. The severe hypoplasia of left heart structures may be a consequence of limited flow during development secondary to a primary abnormality of either left ventricular inflow or left ventricular outflow. Primary defects of myocardial growth are unlikely to be a mechanism for this disease because the myocardium appears normal. In addition, approximately 5% of patients with aortic atresia demonstrate an unrestrictive ventricular

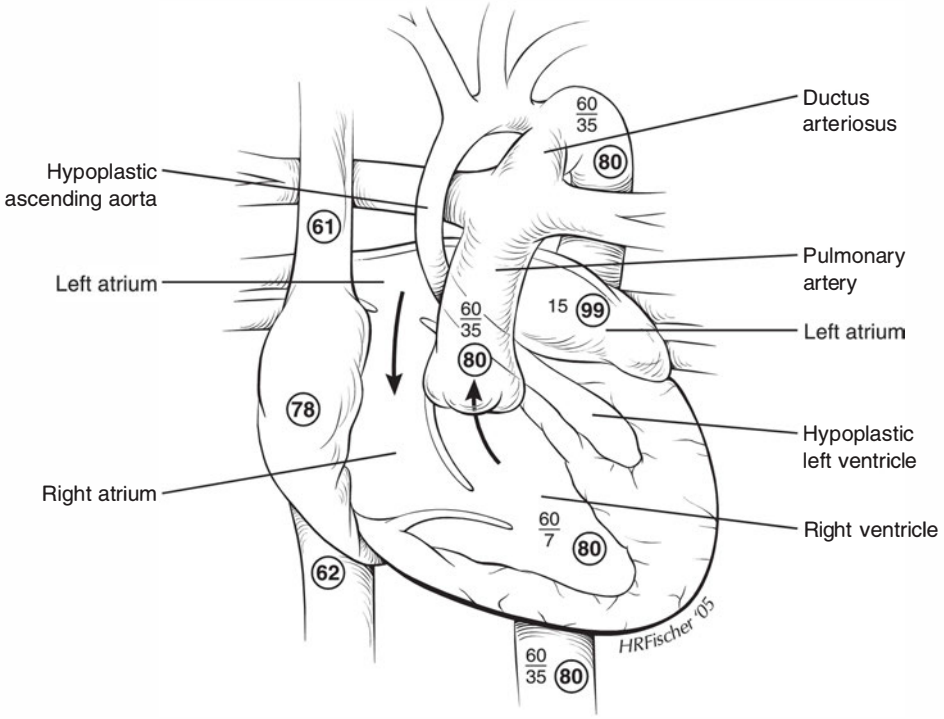


Fig. 96.1. Anatomic features and representative hemodynamic parameters for unrepaired hypoplastic left heart syndrome. Oxygen saturations are enclosed in circles, and blood pressures are indicated by standard nomenclature.

septal defect, and in such cases, there is nearly always normal development of the left ventricle and mitral valve.

PRESENTATION AND INITIAL MANAGEMENT

The normal fetus has a parallel circulation that adequately supports single-ventricle physiology before birth. Three communications (the ductus venosus, foramen ovale, and ductus arteriosus) shunt oxygenated placental blood largely past the hepatic and pulmonary beds to supply the splanchnic circulation. HLHS is well supported in this physiology and is unlikely a cause of fetal demise. More likely, it is a secondary result of early obstructive lesions of either mitral or aortic valvular development. This has been supported in animal models of either mitral or aortic stenosis with resulting left ventricular hypoplasia. However, the primary cause of the obstructive flow lesion leading secondarily to HLHS is unknown. There are no known genetic animal models that fully recapitulate HLHS despite the existence of a large number of mutations that affect valvular development. This argues either for a complex early event that is the result of multiple factors or an early insult influence by subsequent modifiers.

The presentation of infants with congenital heart disease has changed dramatically over the last 15 years. In most large centers, the majority of patients are identified through prenatal echocardiography, although this early identification has not consistently correlated with better outcome. Although some tachypnea and mild cyanosis may be present, it is not until the ductus arteriosus begins to close that the children exhibit impaired systemic perfusion with pallor, lethargy, and diminished femoral pulses. Cardiac examination reveals a dominant right ventricular impulse, a single-second heart sound, and often a nonspecific soft systolic murmur. Electrocardiogram examination reveals right atrial enlargement and right ventricular hypertrophy. Chest X-ray occasionally shows mild cardiomegaly and increased pulmonary vascular markings.

Physical examination of children with HLHS usually appears normal. The examination is determined by the underlying anatomy as well as the duration of the disease. Poor perfusion, weak distal pulses (that may or may not be present, depending on the size of the ductus), acidosis, and a sepsis-like picture may all confound the diagnosis. In the absence of risk factors or laboratory findings consistent with sepsis, one should search for left-sided obstructive lesions. There are no specific laboratory indicators of HLHS, and most patients usually exhibit normal values. With ductal closure and malperfusion, end-organ compromise may be reflected by altered hepatic and renal function tests.

Many mothers of fetuses with HLHS will have had a fetal echocardiogram at 20 weeks with reasonable visualization of cardiac structures. It is neither feasible nor cost effective to screen all pregnancies; therefore a selective approach is taken in which only those mothers at high risk are screened. Frequently, a ventricular size discrepancy is the first hint of impending problems. Certainly, the presence of an intact or restrictive atrial septum with HLHS should prompt term high-risk delivery in an institution in which an urgent postdelivery palliation can be performed safely and rapidly. Emergent operative atrial septectomy is poorly tolerated. The use of prenatal screening improves the prenatal condition of the child but may not improve outcome (at least in cases of transposition of the great arteries or HLHS). After delivery, the infant should undergo two-dimensional and Doppler echocardiography, which defines the anatomy sufficiently for medical and surgical decision-making. It is important to distinguish HLHS from other diseases that may mimic certain features. Chest radiography often demonstrates mild cardiomegaly and excessive pulmonary blood flow. Head ultrasound should be obtained in all patients to rule out intracranial hemorrhage and minimize the risks of heparinization and potential circulatory arrest. Patients with medical necrotizing enterocolitis should ideally have a 7-day course of intravenous antibiotics before repair if they are hemodynamically stable. Preoperative stabilization is critical to the ultimate outcome of patients with HLHS regardless of anatomic subtype, though operation should not be substantially delayed. Nearly all patients with suspected HLHS are transported on prostaglandin E_1 at a dose of 0.01 to 0.025 μg/kg/minute. Two clinically important dose-dependent side effects of prostaglandin E_1 infusion are hypotension and apnea, although these are infrequent. Umbilical arterial and umbilical venous lines are used for central access in most patients. Most patients can ventilate with a natural airway and demonstrate more favorable hemodynamics while extubated. Supplemental oxygen should generally be avoided as it acts as a pulmonary vasodilator, decreasing pulmonary vascular

resistance, increasing the ratio of pulmonary-to-systemic blood flow, and thus decreasing systemic perfusion. Inotropic support is rarely necessary, although it may be required for support in patients who have suffered a perinatal insult. The goal of these maneuvers is to get the patient to the operating room in as stable condition as is possible.

SURGICAL THERAPY

The primary therapy for HLHS is staged reconstructive surgery leading to a modified Fontan–Kreutzer procedure. Over the last 25 years, the Norwood procedure has evolved and is now a standard operation in nearly all institutions for HLHS. There are three primary goals of stage I palliation: (1) establishment of unrestricted interatrial communication to provide complete mixing and avoid pulmonary venous hypertension, (2) establishment of a reliable source of pulmonary blood flow, allowing pulmonary vasculature development and minimizing the volume load on the single ventricle, and (3) providing unobstructed outflow from the ventricle to the systemic circulation.

We offer surgical palliation to nearly all patients with HLHS, including very low birth weight infants and those with nonlethal genetic syndromes. In certain complicating situations, primary transplantation (or other forms of palliation described below) may be considered, such as in severe aortic or AV regurgitation, or dilated cardiomyopathy.

Stage I Reconstructive Surgery

The child is brought to the operating room and ventilated on room air, with care taken to avoid hyperventilation. A full midline sternotomy is performed and a sternal retractor placed. The thymus is removed in its entirety, with care being taken to avoid the phrenic nerves. The pericardium is opened, and an obligatory mediastinal inspection is performed to confirm the echocardiography, especially to identify the abnormalities of the aortic arch and coronary arteries as coronary orientation may alter the surgical approach. The ascending and descending aorta, brachiocephalic vessels, ductus arteriosus, and PAs are extensively mobilized, with care taken to avoid damage to the recurrent laryngeal nerve. No attempt is made to dissect the systemic veins. Purse-string sutures are placed in the proximal main pulmonary artery (MPA) and generously around the right atrial appendage, through which heparin is administered. A previously thawed homograft pulmonary hemipatch is then trimmed in an extended arrowhead shape and set aside (Fig. 96.2A). Two perfusion techniques are commonly used for operative repair: deep hypothermic circulatory arrest (DHCA) and selective antegrade continuous cerebral perfusion. Despite intensive investigation, there is no consensus regarding the superior approach. After the activated clotting time reaches 300 seconds, the patient is cannulated with the arterial cannula at the base of the MPA and a single venous cannula in the RA. Cardiopulmonary bypass is initiated and tapes brought down around the branch PAs. The patient is cooled to 18°C over 15 minutes, during which time any remaining dissection is performed. During this period of cooling, a side-biting clamp is placed on the innominate artery, and a polytetrafluoroethylene (PTFE) graft (usually 4.0 mm for patients $>$3.2 kg and 3.5 mm in smaller infants) is anastomosed in an end-to-side manner. The clamp is removed and flow assessed. If blood does not briskly flow from the open shunt, the anastomosis should be revised. A large hemoclip is placed to temporarily occlude the shunt. On initiation of circulatory arrest, tapes are brought down around the brachiocephalic vessels and a vascular clamp is placed on the descending aorta distal to the ductal insertion site. Cardioplegia is administered antegrade through a side port on the arterial cannula. After draining the patient of blood, all cannulas and PA tapes are removed. The ductus arteriosus is ligated on the PA side and divided on the aortic side. The atrial septum is completely excised working through the atrial purse string (Fig. 96.2B). Visualization can be improved through a right atriotomy, although this is seldom necessary. Next, the MPA is divided close to the branch PAs, and the defect in the distal MPA segment is closed either with an oval homograft patch or primarily in a vertical manner.

At a point beginning immediately adjacent to the divided MPA, the diminutive aorta is incised medially and the incision carried superiorly along the underside of the transverse arch through the ductal insertion site to a point approximately 1-cm distant. It is important that all redundant ductal tissues be excised from the previous insertion site and the coarctation shelf be debrided (or the segment excised and the remaining vessel reanastomosed). The proximal aortic-to-proximal PA connection is now performed using interrupted, fine polypropylene sutures (Fig. 96.2C). Next, the arch is reconstructed using the homograft patch, carrying this suture line down to complete the Damus–Kaye–Stansel proximally (Fig. 96.2D). The distal Blalock–Taussig shunt–PA anastomosis is now performed to the origin of the RPA, although some surgeons prefer to do this with the cross-clamp off during warming (Fig. 96.2E). The arch is infused with cold saline to assess the geometry or residual obstruction, the atrium is infused with cold saline to deair, and the cannulas are replaced. Cardiopulmonary bypass is begun and the patient warmed to 37°C over 22 minutes. It is important at this point to assess prompt and equivalent filling of coronary distributions. Any perfusion defect should be addressed immediately with revision of the aortic-to-PA anastomosis. During warming, obvious bleeding should be controlled. After the patient has been warmed to 37°C, the clip is removed from the shunt and atrial lines are brought through the chest wall and positioned in the RA. The patient is begun on low-dose dopamine and milrinone support and weaned off cardiopulmonary bypass-modified ultrafiltration is routinely performed. Oxygen saturations should be in the mid-80% range, indicative of adequate pulmonary blood flow. Any base deficit is corrected completely with sodium bicarbonate, and continued persistent acidosis is a relative indication of poor cardiac function requiring examination of the repair. After protamine is administered to reverse the heparin, hemostasis is meticulously obtained. If there are no issues with bleeding, the chest is routinely closed. In approximately 20% of cases, either hemodynamic or respiratory instability or continued potential bleeding results in the potential for cardiac compromise with chest closure. In these cases, a PTFE patch is cut to an appropriate size and approximated to the skin edges, leaving the sternum open for 12 to 24 hours. Postoperative management of these patients includes low-dose dopamine and milrinone, although some centers use phenoxybenzamine as an afterload-reducing agent. The use of afterload reduction results in vasodilation with right ventricular support and generally removes the need for any greater inotropic therapy. In some cases, low-dose epinephrine is used if there is significant hypotension. Patients are lightly sedated and administered either low-dose fentanyl as a continuous drip or intermittent morphine. Patients are not routinely paralyzed with pancuronium unless the sternum is left open. An attempt is made to allow the patient to awaken and to wean the ventilator support such that extubation can be performed on the first or second postoperative day. If the chest is

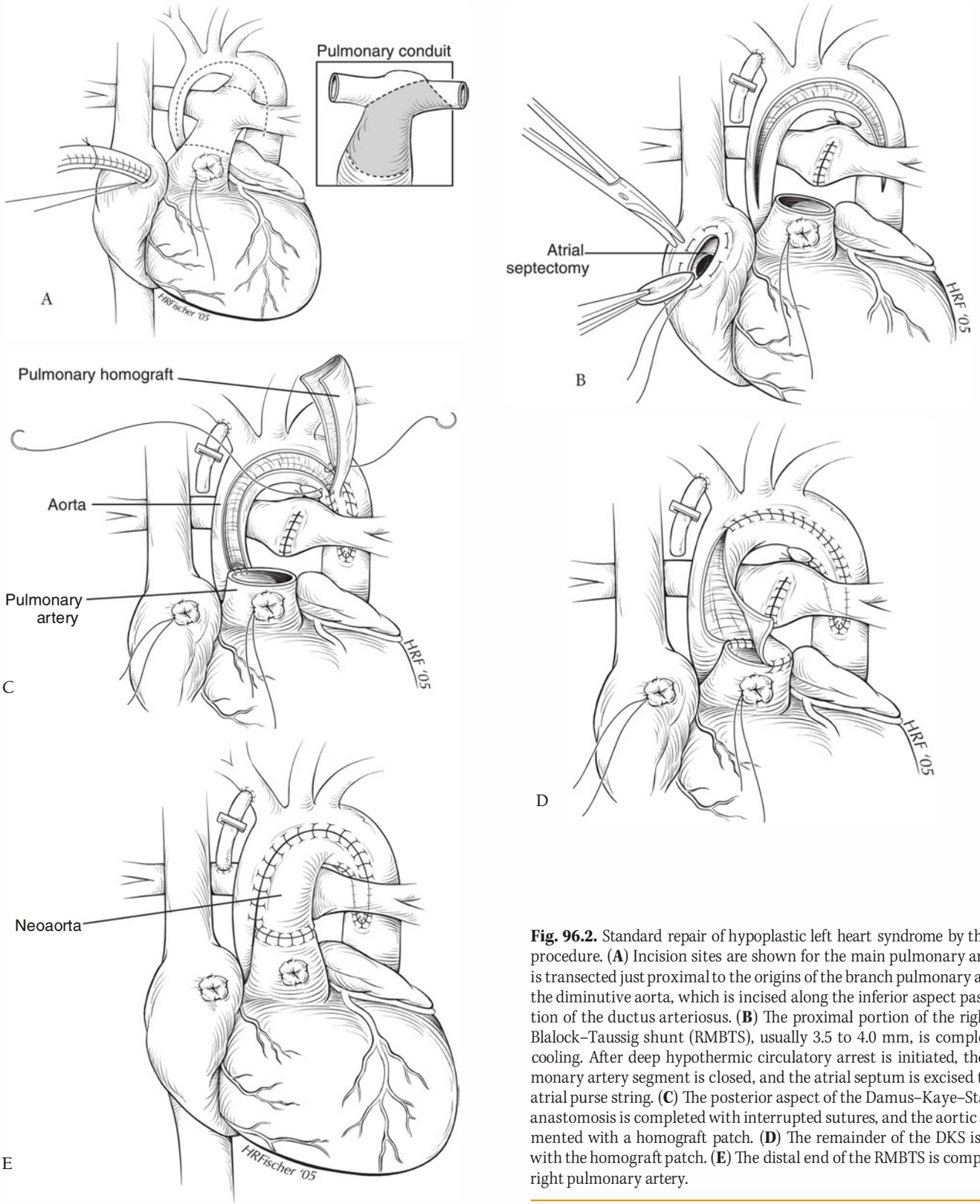


Fig. 96.2. Standard repair of hypoplastic left heart syndrome by the Norwood procedure. (**A**) Incision sites are shown for the main pulmonary artery, which is transected just proximal to the origins of the branch pulmonary arteries, and the diminutive aorta, which is incised along the inferior aspect past the insertion of the ductus arteriosus. (**B**) The proximal portion of the right modified Blalock–Taussig shunt (RMBTS), usually 3.5 to 4.0 mm, is completed during cooling. After deep hypothermic circulatory arrest is initiated, the main pulmonary artery segment is closed, and the atrial septum is excised through the atrial purse string. (**C**) The posterior aspect of the Damus–Kaye–Stansel (DKS) anastomosis is completed with interrupted sutures, and the aortic arch is augmented with a homograft patch. (**D**) The remainder of the DKS is completed with the homograft patch. (**E**) The distal end of the RMBTS is completed to the right pulmonary artery.

left open, the chest is generally closed the day after surgery and the patient allowed to waken, weaning from the ventilator over the next 24 to 48 hours. Rapid deintensification with the removal of nasogastric feedings and removal central lines is preferred. Aspirin is usually administered after the first night when heparin is used at a low dose to decrease the risk of shunt thrombosis. When oral intake is established, low-dose aspirin is administered entirely. Patients are usually extubated between postoperative days 1 and 3. Average duration of hospitalization is between 7 and 21 days, the limiting factor often being establishment of adequate oral caloric intake. In all cases, the principle is continued clinical progress, and any migration from that pathway should prompt investigation into the cause of deviation.

A number of reports including a multi-institutional collaborative effort organized

through the Pediatric Heart Network suggest that a right ventricular-to-PA (RV-PA) shunt as popularized by Sano may improve outcome after stage I reconstruction (Fig. 96.3). However, a number of contradictory studies further comment that before broad adaptation of the RV-PA conduit, other considerations apply. For example, there may be anatomic substrates better suited to one shunt type compared with another. With respect to the RV-PA conduit, there is an increased incidence of shunt reintervention, an earlier return for stage II reconstruction, and no difference in overall mortality. Certainly, consensus exists that staged palliation consists of multiple surgeries that result in the Fontan circulation. Increasingly, an understanding that uniform adaptation of any approach prior to stage III data may be premature.

For high-risk infants, some have advocated a catheter-based hybrid approach: stage I: palliation includes ductal stenting and PA banding; stage II: septectomy, arch augmentation, and cavopulmonary anastomosis; and stage III: catheter-based Fontan completion. Although there have been promising results, these techniques may be limited in certain anatomic subsets such as aortic atresia in which preductal, retrograde coarctation is a significant problem (Fig. 96.4). Palliative catheter-based hybrid approaches may be the most useful in identifiable subsets of patients with HLHS who have very low birth weight or associated cardiac anomalies. These patients are still a high-risk group for stage I reconstruction in most series. Otherwise, catheter-based approaches have no demonstrable advantage over surgical staged palliation.

Stage II Reconstructive Surgery

Two important observations by Norwood and colleagues early in the reconstructive experience prompted the institution of an intermediate stage. The first was that there

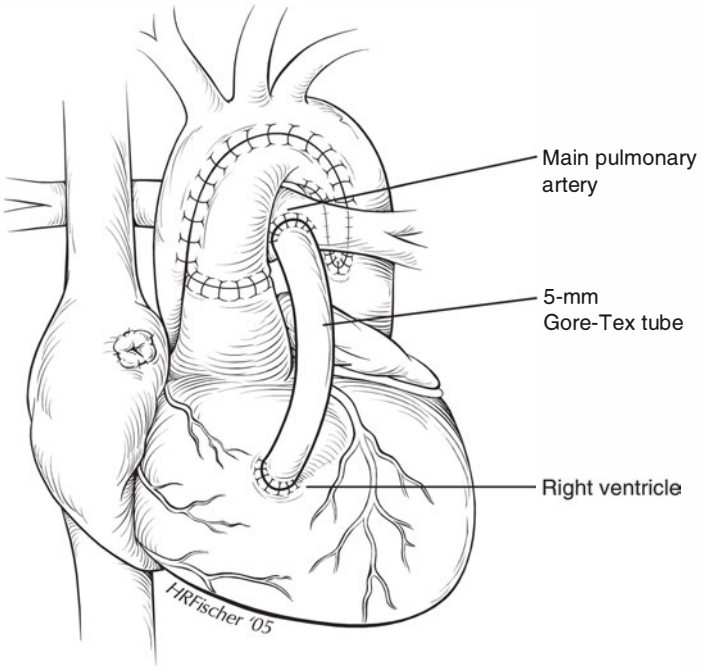


Fig. 96.3. Alternative approach to stage I palliation using the Sano modification. Instead of a right modified Blalock–Taussig shunt, pulmonary blood flow is supplied by a 5.0-mm polytetrafluoroethylene (Gore-Tex) shunt from a right ventriculotomy to the main pulmonary artery.

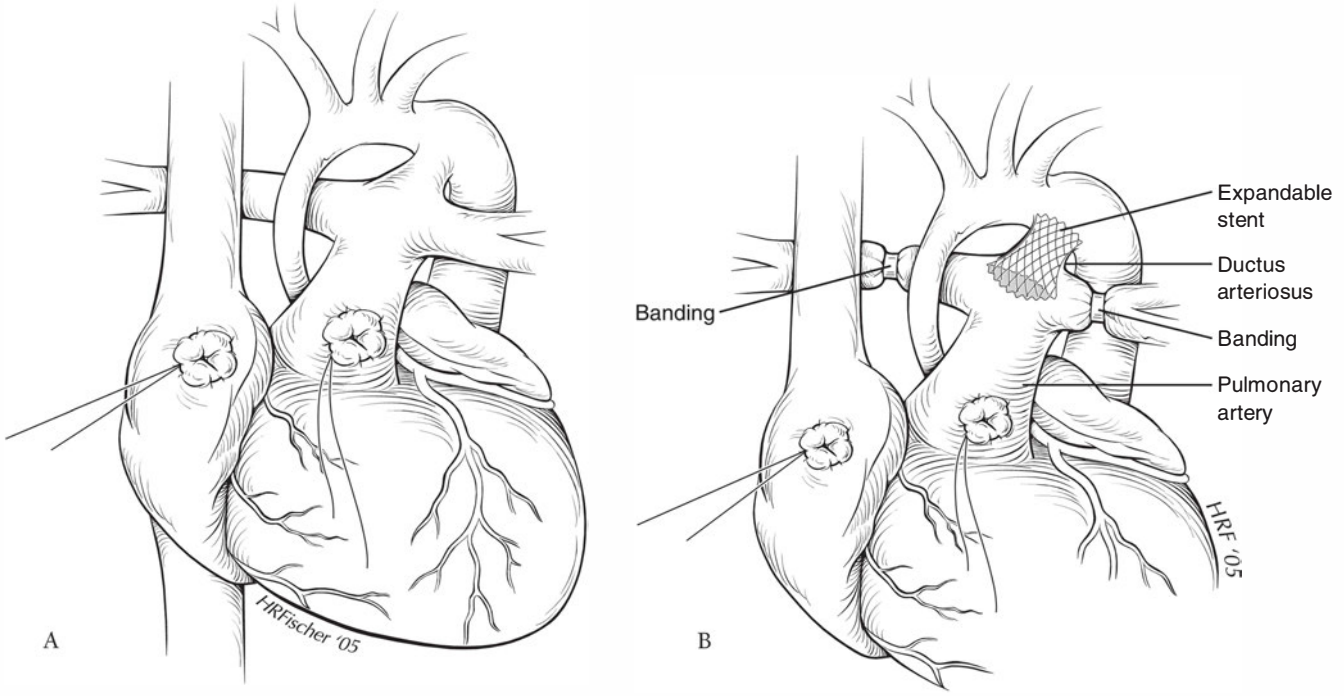


Fig. 96.4. Alternative approach to stage I palliation using ductal stenting and pulmonary artery bands. (**A**) Unrestricted, stable systemic and pulmonary blood flow is created by insertion of an expandable stent in the ductus arteriosus. (**B**) Next, pulmonary blood flow is restricted by placement of bilateral pulmonary artery 3.0-mm bands.

was a time-related interstage mortality. The second was that the chronic volume load of a systemic-to-pulmonary shunt could create diastolic ventricular dysfunction. Thus, an intermediate stage was initiated as either a bidirectional cavopulmonary (Glenn) shunt or a hemi-Fontan procedure. A bidirectional cavopulmonary anastomosis sets the stage for an extracardiac conduit, whereas a hemi-Fontan sets the stage for a lateral tunnel completion Fontan. There is no long-term data that prove the efficacy of one approach over another. In general, at approximately 4 to 6 months of age, stage I survivors are catheterized to evaluate both pressures throughout the heart and the anatomy of PAs. The use of a cavopulmonary anastomosis before approximately 3 months of age is sometimes associated with increased hypoxia and upper-body venous congestion, although bidirectional Glenn shunts have been done in children at even 6 weeks to 2 months of age with good results when there is clearly demonstrable low pulmonary vascular resistance. The technique for a bidirectional cavopulmonary anastomosis is standard (Fig. 96.5).

An alternative operative approach is the hemi-Fontan procedure, which we perform under DHCA. The approach is through a reoperative median sternotomy, during which time care is taken around the dissection of the neoaorta. The patient is cannulated in a standard manner with an arterial cannula in the neoaorta and a single straight venous cannula in the body of the RA. Cardiopulmonary bypass is begun, and the patient is cooled to 18°C. The previous shunt is divided and ligated and the azygos vein ligated. After cardioplegic arrest, the PAs are opened on the anterior aspect and the pulmonary insertion of the shunt excised. If preoperative catheterization revealed PA stenosis, the incisions are carried beyond this point well onto the left PA and onto the right lower lobar branch. Next, the RA is incised superiorly and medially from 12 to 6 o'clock beginning just superior to the cannulation site and ending at the level of the right upper PA (Fig. 96.6). The SVC and the right aspect of the pulmonary arteriotomy are anastomosed with fine monofilament suture. Next, an extended triangular pulmonary homograft patch is used to augment the PAs and create a roof over the anastomosis of the SVC to the RPA, as well as simultaneously create a dam to prevent blood flow between the SVC and RA. The patch-augmented PAs are infused with saline to examine anatomy and deair. The venous cannula is replaced, cardiopulmonary bypass is reinitiated, aortic cross-clamp removed, and warming to 37°C is completed over 22 minutes. Occasionally, additional procedures such as atrial septectomy or arch augmentation may need to be completed in the same setting. One or two right atrial lines are inserted into the body of the RA or into the PA through the suture line. The patient is weaned from cardiopulmonary bypass, and modified ultrafiltration is performed, during which time all suture lines are checked for hemostasis. All cannulas are removed, and protamine is administered. The chest is then closed in a standard manner and the patient returned to the intensive care unit (ICU). In general, these patients can be extubated either in the operating room or soon after returning to the ICU. With reduction in the volume load provided by this procedure, inotropic support is usually brief. Blood–oxygen saturations are generally 80% to 90%, and patients are generally discharged within 5 to 7 days after surgery.

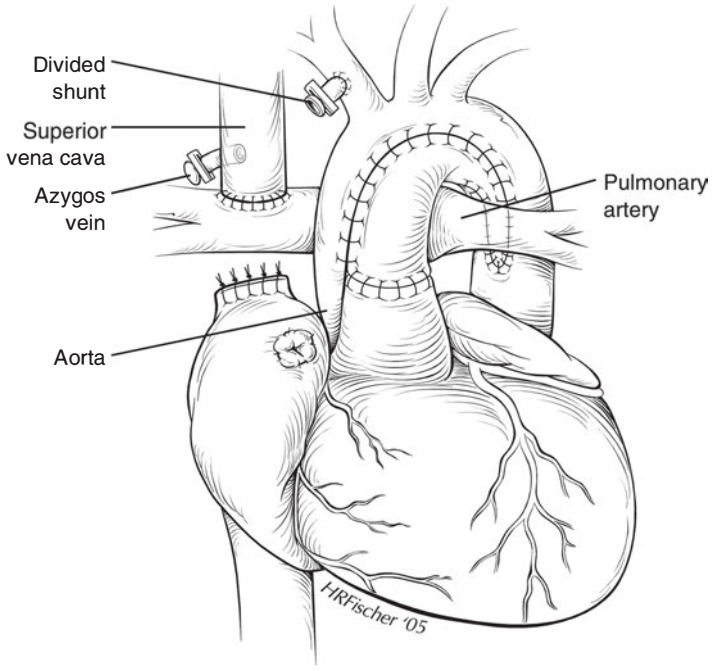


Fig. 96.5. The bidirectional Glenn procedure. The Blalock–Taussig shunt is ligated proximally and removed distally from the right pulmonary artery. The azygos vein is ligated arid the superior vena cava (SVC) is detached from the right atrium, oversewing the atrial portion. Finally, the SVC is anastomosed to the right pulmonary artery.

Stage III Reconstructive Surgery

Between ages 1.5 and 5 years (usually determined by a combination of the patient's weight, growth characteristics, and arterial saturations) the patient is reimaged by either echocardiography or magnetic resonance imaging and, if necessary catheterization. If there are no anatomic issues to be addressed via catheterization (e.g., distal arch coarctation), the patient is referred for Fontan reconstruction via either an extracardiac conduit or lateral tunnel completion Fontan. For the extracardiac conduit, DHCA or continuous bypass with or without aortic cross-clamp is employed, while we use DHCA for the lateral tunnel. The approach is again through a reoperative median sternotomy.

For the extracardiac Fontan procedure, the patient is bicavally cannulated in a standard manner and an arterial cannula placed high in the aortic reconstruction. Cardiopulmonary bypass is begun and tourniquets are applied around the caval cannulas. A vascular clamp is placed at the IVC–RA junction and the IVC divided (Fig. 96.7). The atrial portion is partially closed in two layers with fine monofilament suture. The conduit (18 to 22-mm PTFE) is trimmed to the appropriate length to avoid compression of the right pulmonary vein (shorter than one might expect), and a 4-mm fenestration is punched in the medial aspect near the IVC portion. The IVC-conduit anastomosis is completed with monofilament suture followed by a side-to-side anastomosis of the remaining cardiac portion of the IVC opening with the exterior conduit, leaving a rim of conduit around the fenestration.

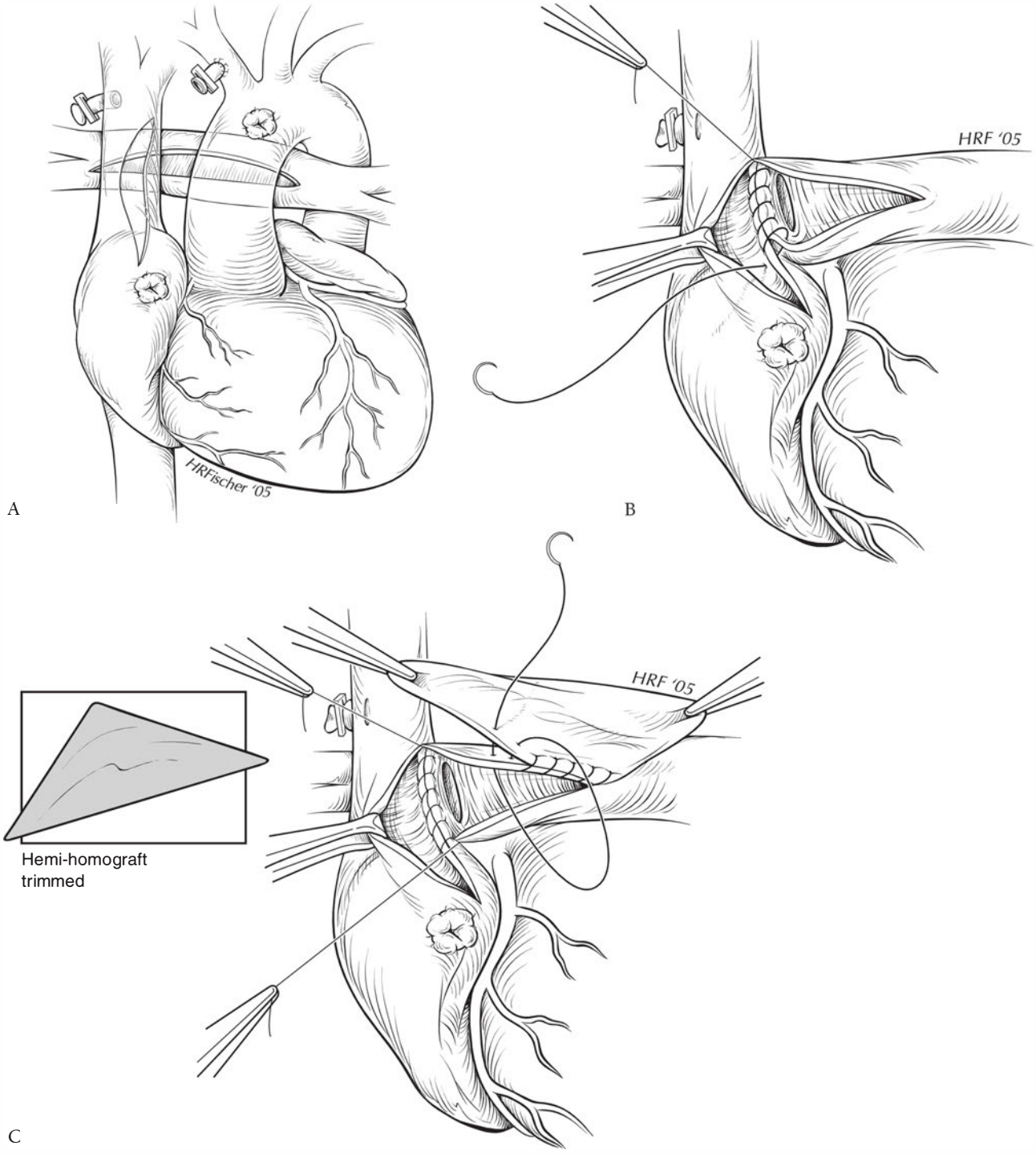


Fig. 96.6. The hemi-Fontan procedure. (**A**) The pulmonary artery is incised widely from the left lower pulmonary artery to the left pulmonary artery. The right atrium is incised vertically in a spiral clockwise manner from the superior portion of the right atrial (RA) appendage into the superior vena cava (SVC) to the superior aspect of the right pulmonary artery. The azygos vein is ligated. (**B**) The posterior aspect of the incision in the SVC is anastomosed to the rightward aspect of the pulmonary arteriotomy. (**C**) An extended triangular homograft patch is trimmed and sewn to augment the pulmonary arteries. (**D**) The homograft patch suture line continues along the SVC–RA junction to create the bottom of the dam. (**E**) The patch is folded on itself to create a triangular-shaped dam. (**F**) The suture line is continued to complete the dam. (**G**) The same homograft patch is used to simultaneously complete the pulmonary artery augmentation and SVC-PA anastomosis.

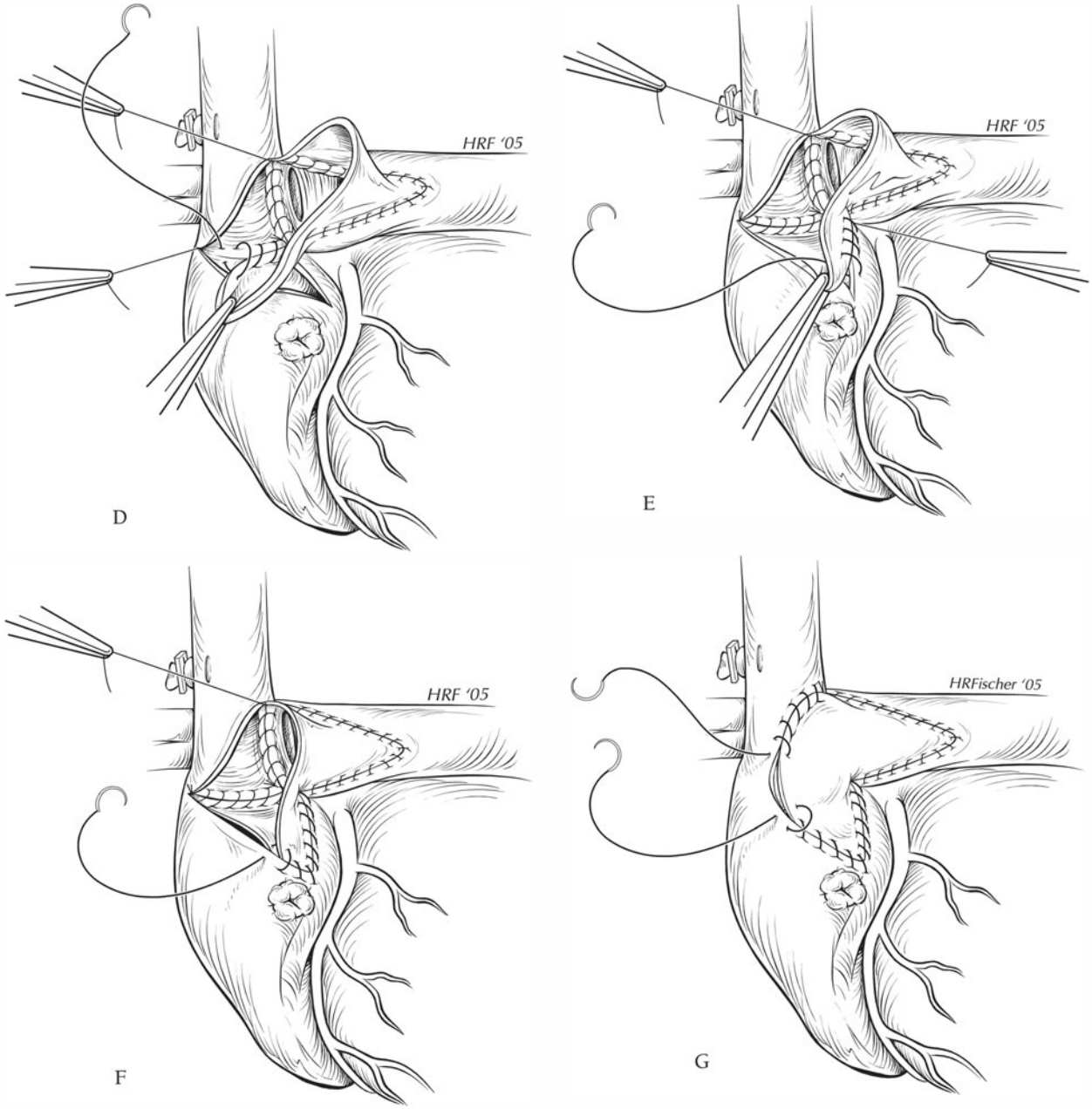


Fig. 96.6. (*Continued*)

Next, the PAs are opened along the inferior margin and inspected directly. If preoperative studies revealed any PA stenosis, and the beveled end of the PTFE conduit will not span the area of stenosis, this is addressed with pulmonary homograft patch augmentation. The conduit is then anastomosed to the inferior aspect of the PAs angled slightly medial to the SVC. Practically, the angled nature of the superior portion of the conduit augments the PAs from LPA to the RPA. The conduit is infused with saline to deair, and the patient is weaned from cardiopulmonary bypass. The SVC cannula is removed, and a period of modified ultrafiltration is begun. All suture lines are checked for hemostasis. At the completion of modified ultrafiltration, the SVC pressure is measured directly with a transthoracic line and subsequently removed. An additional monitoring line is placed in the RA. All cannulas are removed, and protamine is administered. The chest is then closed in a standard fashion and the patient returned to the ICU. In general, these patients are extubated soon after return to the ICU.

An alternative operative approach is the lateral-tunnel Fontan procedure, which we perform under DHCA using a piece of fenestrated PTFE patch to baffle blood from the IVC to the PAs (Fig. 96.8). The patient is cannulated in a standard manner with an arterial cannula in the neoaorta and a single straight cannula in the body of the RA. Cardiopulmonary bypass is begun, and the patient is cooled to 18°C. An aortic cross-clamp is applied and cardioplegic arrest achieved. The patient is drained of blood and the venous cannula removed. A vertically based incision is made in the body of the RA parallel to Waterson groove. The previously constructed PA-RA homograft dam is excised and the Eustachian valve is removed. A 10-mm PTFE tube graft is split longitudinally and trimmed to length, and a 4-mm fenestration is created. Inferiorly, the graft is sewn around the IVC orifice

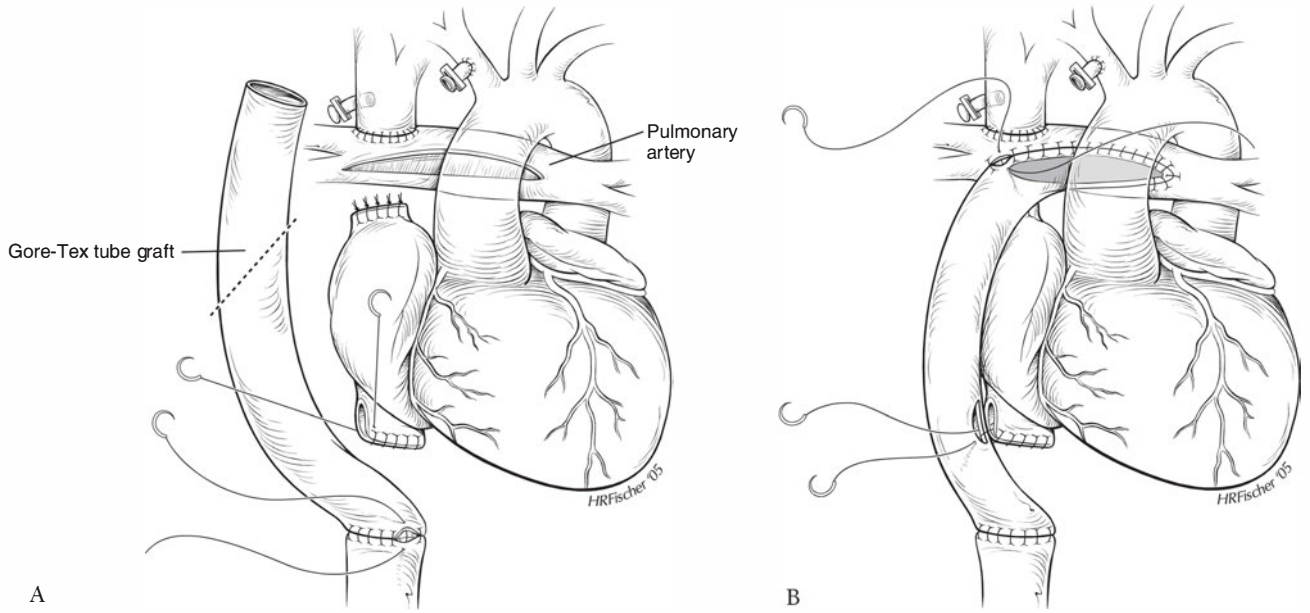


Fig. 96.7. The extracardiac Fontan procedure. (**A**) The inferior vena cava is detached from the right atrium, and the right atrial portion is partially closed. An 18- to 22-mm polytetrafluoroethylene (PTFE; Gore-Tex) conduit is anastomosed to the inferior vena cava (IVC), and a 4.0-mm fenestration is created next to the partially closed IVC. (**B**) A side-to-side anastomosis is fashioned between the fenestration and IVC opening in the right atrium. The distal end of the PTFE conduit is beveled to create a large opening that augments the pulmonary arteries, and the conduit-PA anastomosis is completed.

and the suture line carried superiorly along the line of the interatrial communication. Superiorly, the baffle is sewn around the edge of the newly created opening between the atrium and PAs. Care must be exercised here to assure that the trabecular portion of the atrium contains no leaks. The free, superior edge of the PTFE baffle is then closed in a single sandwich between the two free edges of the RA. Thus, the medial aspect of the lateral tunnel is PTFE, whereas the lateral aspect is native right atrial tissue. The heart is infused with saline to deair and the venous cannula replaced. Cardiopulmonary bypass is reinitiated, and the patient is warmed to 37°C over 22 minutes. Atrial lines are brought into the atrium on either side of the baffle through the suture line. The patient is weaned from bypass and undergoes modified ultrafiltration. Inotropic support is rarely necessary.

RESULTS

Despite continued developments, patients with HLHS continue to present formidable challenges. Since its institution in 1984, results from staged reconstruction have improved significantly. This has been the case across multiple institutions, and many centers report excellent outcomes, often exceeding 90% hospital survival. There is conflicting opinion that variability in outcome may be influenced by anatomy. However, it appears that the HLHS is not a predictor of mortality compared with stage I palliation for other HLHS variants. It is clear that risk factors such as low birth weight, associated cardiac anomalies, longer total support time, and extracorporeal membrane oxygenation (ECMO) or ventricular assist device (VAD) support are predictors of operative mortality. Additional perioperative or operative treatment strategies may improve morbidity and mortality based upon either anatomic of genetic stratification. Home surveillance programs for interstage infants are now common practice in multiple institution and suggest significant benefits toward the reduction of interstage mortality.

Our group compared the outcomes of all neonates who underwent a stage I reconstruction between 2002 and 2004 with use of the RV-PA conduit and modified Blalock–Taussig shunt. In all, 149 infants underwent a stage I reconstruction for HLHS or variants. There was no difference in surgical mortality, time to extubation, or length of hospital stay. However, there was an increased incidence of shunt reinterventions in the patients with the RV-PA conduit. Patients with RV-PA conduit returned earlier for stage II reconstruction but there was no difference in overall mortality. These data have been confirmed in some centers and refuted in others. However, the PHN-based multi-institutional study of single-ventricle reconstruction support the notion that local, team-based factors may outweigh shunt type.

Although promising for certain high-risk populations, catheter-based hybrid approaches are still in an experimental phase. These techniques may be inadvisable in certain anatomic subsets such as aortic atresia in which preductal retrograde coarctation is a significant problem. However, in some groups there is increasing interest in studying this approach compared with Norwood reconstruction.

Results for the final palliation to the Fontan circulation and for the second- and third-stage reconstruction for HLHS continue to improve. Survival rates for the bidirectional Glenn or hemi-Fontan are well above 95% in most centers, and similarly the survival rates for Fontan completion are >95% to 98% in most recent reports. These excellent results confirm the utility of staged reconstructive surgery to the Fontan circulation as the preferred strategy for patients with HLHS, saving transplantation for patients who fail at any stage of the reconstruction sequence. Our experience with the Fontan procedure for HLHS shows a >95% survival with a median hospital stay of 5 to 6 days. Effusions are common though rarely last more than a few days. Prolonged effusions of more than 2 weeks occur in <15% of patients with current strategies.

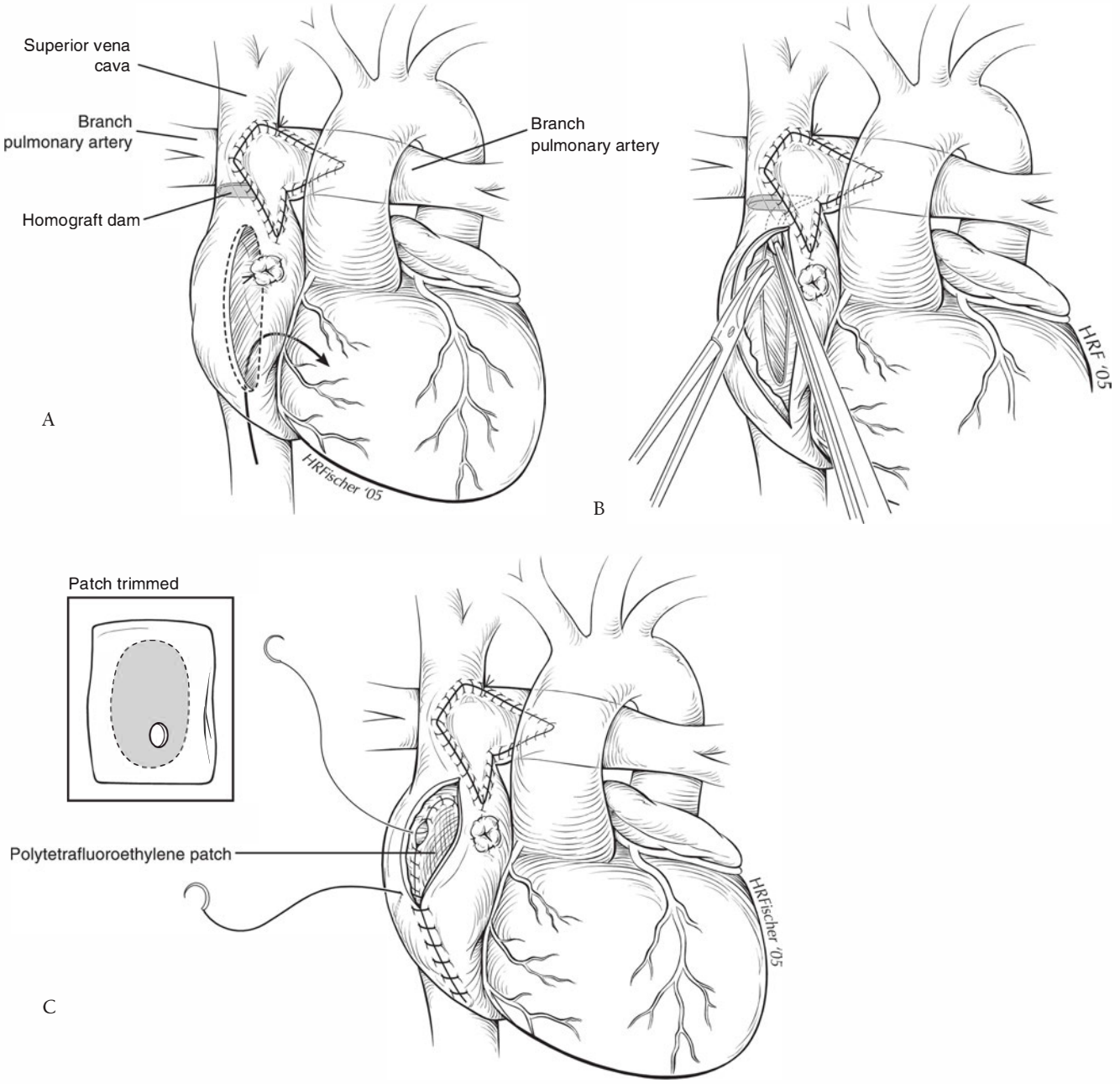


Fig. 96.8. The lateral tunnel Fontan procedure. (**A**) The right atrium is opened in a vertical fashion medial to Waterson's groove. (**B**) The homograft dam is excised, creating an unobstructed opening from right atrium to pulmonary artery. (**C**) A polytetrafluoroethylene (PTFE) baffle is sewn along the posterior rim of the remnant atrial septum, on the medial aspect of the inferior vena cava (IVC), and to the medial aspect of the superior vena cava–pulmonary artery (PA) junction. The free edge of the PTFE dam is sewn in a sandwich to close the right atrium, baffling blood from the IVC to the PA.

This improvement in overall Fontan survival has been reflected in reports from many institutions, suggesting that there has been a marked improvement in cardiac function in survivors of single-ventricle staged reconstruction over the last 15 years. Hospital survival of >90% is common, with guardians reporting their child's overall health as excellent or good. Physical activity is normal in one-third of patients, with one-half reporting only slight physical limitations. School performance varies widely, with about one-third of patients above average, one-third average, and one-third below average.

When comparing outcomes of extracardiac conduit and lateral tunnel Fontan connections, results are conflicting. Some have reported that the lateral tunnel Fontan procedure has a significantly higher incidence of postoperative sinoatrial node dysfunction, supraventricular tachycardia, duration of ICU stay, and ventilator support. However, others have found contrasting data with a lower incidence of sinus node dysfunction in the lateral tunnel versus extracardiac Fontan reconstruction. The potential advantages of extracardiac Fontan reconstruction include the lack of complicated atrial suture lines, although most patients with HLHS will have atrial septectomy as part of their initial procedure. The incidence of late atrial arrhythmias is unknown in this population of patients, and it is not clear whether the extracardiac conduit Fontan will have

significant advantages over the lateral tunnel operation. There have been studies suggesting that the hemodynamic flow characteristics of the extracardiac Fontan may be superior to a lateral tunnel connection. However, the offset created by a hemi-Fontan procedure may be beneficial. These conflicting results have not been correlated with clinical findings and late outcomes.

With the progression of improvement in surgical techniques and pre- and postoperative management, current survival to the Fontan procedure for all patients with HLHS as a group is approximately 75% in centers with the greatest experience. Survival rates of >90% are common in low-risk patients who have normal birth weight and no associated cardiac or noncardiac anomalies. However, the frequent coexistence of low birth weight or tricuspid regurgitation or other noncardiac anomalies continues to create a high-risk group that lowers overall survival in centers who see a large proportion of patients with these comorbidities. Selective application of the variations in the Norwood procedure and hybrid strategies for these subsets of high-risk patients may improve overall survival with greater experience. Developments in preoperative evaluation, operative techniques, and postoperative management based on rigorous controlled trials and rational application or these results will provide continued progress with this challenging disease.

SUGGESTED READINGS

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EDITOR'S COMMENTS

The group at CHOP has really developed most of the changes in technique in hypoplastic heart syndrome. Dr. Norwood initially and now Dr. Spray and Gruber and their colleagues have made this a highly successful operation. The major issue is the delicate of the balance of circulation. There are multiple controversies that have been dealt with well by this chapter. Probably, the most important relates to the standard shunt versus a right ventricular to pulmonary artery conduit as popularized by Sano. The Sano procedure seems to result to a much more stable initial postoperative course. However, as the authors have stated there is usually earlier reintervention and more complications in the mid-term as compared with the standard shunt. Having said that I think in those groups were the volume of these patients is relatively small, the Sano probably allows for easier initial postoperative care.

The second question is the proper kind of Fontan procedure. The authors have preferred the hemi-Fontan followed by a lateral tunnel. The other techniques now available include the extracardiac Fontan and in some cases catheter-based Fontan procedures. The authors make a very good case for the hemi-Fontan followed by the lateral tunnel but certainly they have clearly included all the other possibilities.

The procedure keeps changing and continues to evolve and I think the results will continue to improve.

TLS

97 Anomalies of Pulmonary Venous Return

Carlos M. Mery, James J. Gangemi, and Irving L. Kron

Anomalies of pulmonary venous return form a spectrum of embryologically related congenital heart defects that have in common the failure of the pulmonary veins to unite normally with the left atrium. The lesion may involve anomalous connection of all four pulmonary veins with the systemic venous circulation (total anomalous pulmonary venous return, TAPVR) or abnormal drainage of at least one but not all pulmonary veins into the systemic venous circulation (partial anomalous pulmonary venous return, PAPVR).

TOTAL ANOMALOUS PULMONARY VENOUS RETURN

Historical Aspects

The earliest description of TAPVR was given by Wilson in 1798. In 1942, Brody presented an autopsy series of patients with this anomaly. Muller at UCLA is credited with the initial attempt of surgical treatment of TAPVR in 1951. He described a closed operation consisting of a side-to-side anastomosis between the common pulmonary vein trunk and the left atrial appendage. In 1956, Lewis, Varco, and associates at the University of Minnesota Minneapolis, MN, reported the first successful total correction of TAPVR using hypothermia and inflow occlusion. The same year, Burroughs and Kirklin described their experiences with surgical correction using cardiopulmonary bypass (CPB). The introduction of deep hypothermic circulatory arrest by Barratt-Boyes and colleagues in the early 1970s was a major advance toward accomplishing surgical repair because this technique provides a bloodless operative field. Further advances in early diagnostic modalities, neonatal intensive care, including the availability of extracorporeal membrane oxygenation (ECMO), and pediatric cardiac surgical anesthesia, as well as an awareness of the merits of early surgical intervention, have contributed to the excellent results with TAPVR repair reported recently by many centers.

Embryology

The respiratory system develops as an evagination from the foregut at 26 days. The venous plexus from the early lung buds drains into the anterior cardinal and umbilico vitelline veins, both of which are part of the splanchnic (systemic) venous system. The anterior cardinal veins give rise to the right and left superior vena cavae (SVC), the coronary sinus, and the azygos vein. The umbilico vitelline veins form the inferior vena cava (IVC) and the portal vein. In normal circumstances, the common pulmonary vein develops as an outpouching from the dorsal left atrial wall, eventually fusing with the pulmonary venous plexus at 28 to 30 days. Shortly thereafter, the anterior cardinal and umbilico vitelline venous channels normally undergo involution (Fig. 97.1). Failure of the common pulmonary vein to unite with the pulmonary venous plexus leads to persistence of these embryonic pulmonary venous-to-systemic venous anastomoses, yielding total anomalous pulmonary venous drainage into right atrial tributaries. In some instances, abnormal leftward displacement of the developing atrial septum results in anomalous connection of all four pulmonary venous ostia directly to the right atrium.

Anatomic Defects and Classification

The common underlying anatomic defect in all cases of TAPVR involves anomalous drainage of the entire pulmonary venous circulation into the right atrium, either directly, or via the SVC, IVC, or coronary sinus. The pulmonary venous blood may drain into the systemic venous circulation through a single common channel or by multiple portals of entry. An interatrial communication, usually manifested as a patent foramen ovale (PFO) or secundum atrial septal defect (ASD), is mandatory for shunting of partially oxygenated blood to the left heart.

There are several classification schemes for TAPVR. The most commonly used system is the one described in 1957 by Darling and associates. This four-tier system is based on the site of pulmonary venous drainage into the systemic circulation. In type I, or *supracardiac* TAPVR, all four pulmonary veins form a horizontal common pulmonary venous confluence behind the left atrium that gives rise to a vertical vein that drains into a supracardiac systemic vein (innominate vein, SVC, or azygos vein) (Fig. 97.2). The most common configuration involves an ascending left vertical vein or persistent left SVC that drains into the innominate vein. Type II, or *cardiac* TAPVR, is characterized by total pulmonary venous drainage into a markedly dilated coronary sinus (Fig. 97.3) or, less commonly, directly into the right atrium. Type III, or *infracardiac* TAPVR, involves a more vertical pulmonary venous confluence giving rise to a descending vertical vein that travels through the esophageal hiatus to below the diaphragm, where it most often makes an anomalous connection with the portal vein, one of its branches (Fig. 97.4), or the ductus venosus. In such cases, pulmonary venous blood returns to the right atrium by way of the IVC. Type IV, or *mixed* TAPVR, comprises all mixed defects with connections at different levels.

According to most series, a supracardiac connection is the most common TAPVR variant (45% to 60%), whereas the cardiac and infracardiac types are encountered somewhat less frequently (15% to 30% each) and the mixed type is the rarest (5% to 10%). In general, the infracardiac variants of TAPVR are usually associated with significant obstruction of the anomalous draining vein due to the length of the venous channel and the resistance created in the hepatic portal system. Supracardiac variants are significantly obstructed approximately half of the time while cardiac variants rarely present with significant obstruction. Alternate classification systems have used the embryologic origin of the anomalous connection, the length

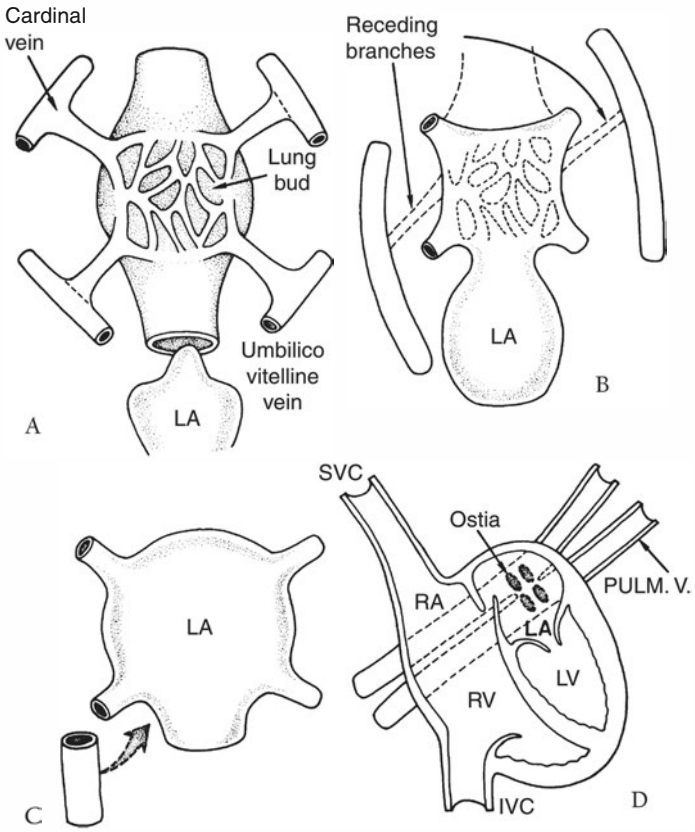


Fig. 97.1. Normal embryonic development of the pulmonary veins. (**A**) Common pulmonary vein evagination from the dorsal left atrial wall growing toward the pulmonary venous plexus surrounding the lung buds. Embryonic pulmonary-to-systemic venous anastomoses still exist. (**B**) The common pulmonary vein fuses with the pulmonary venous plexus as pulmonary-to-systemic communications begin to involute. (**C** and **D**) Complete incorporation of the pulmonary veins into the dorsal left atrial wall with disappearance of embryonic pulmonary-to-systemic venous communications. IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PULM V., pulmonary vein; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

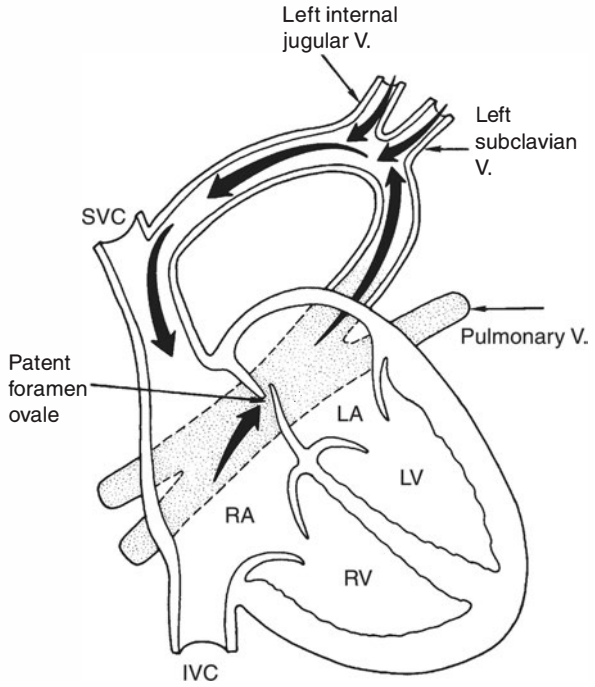


Fig. 97.2. The pathologic anatomy of supracardiac-type total anomalous pulmonary venous return via the ascending left vertical vein. IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava; V., vein.

of the draining veins, and the degree of obstruction between the pulmonary venous and systemic venous pathways. The most complete system, proposed by Herlong and associates as part of the Congenital Heart Surgery Nomenclature and Database Project, describes the anatomic variant, the presence or absence of obstruction, and the type of obstruction (extrinsic or intrinsic compression). Whereas the majority of TAPVR cases are isolated anomalies, the lesion occasionally coexists with other cardiac and extracardiac congenital malformations. TAPVR, especially in autopsy series, has been diagnosed concomitantly with a variety of other acyanotic and cyanotic heart defects, including patent ductus arteriosus, valvular atresia and stenosis, ventricular septal defect, transposition of the great arteries, tetralogy of Fallot, double-outlet right ventricle, and common atrioventricular canal. In addition, there is a well-known association between TAPVR and the heterotaxy syndrome, which includes visceral heterotaxy, isomerism, dextrocardia, and splenic abnormalities (asplenia, polysplenia, hyposplenia). In some series, up to 30% of patients with TAPVR have heterotaxy syndrome.

Pathophysiology

The physiologic consequences of TAPVR depend largely on the presence and magnitude of pulmonary venous obstruction. In those cases with no significant obstruction, TAPVR functions as a large left-to-right shunt. Pulmonary blood flow is greatly increased, and right ventricular overload occurs as a result of both the pulmonary venous and systemic circulations returning to the right atrium. Consequently, pulmonary hypertension, pulmonary edema, right ventricular enlargement, and congestive heart failure supervene. Cyanosis results from the mixing of fully oxygenated pulmonary venous blood with desaturated systemic venous blood. A compensatory right-to-left shunt through a PFO or an ASD is mandatory for survival because this allows shunting of partially oxygenated blood to the left atrium for systemic distribution. Although left atrial and left ventricular volumes are generally small due to shunting, these chambers are rarely hypoplastic.

Obstruction to pulmonary venous blood flow in TAPVR may occur as a result of extrinsic compression, intrinsic luminal narrowing of the vein, a restrictive communication at the atrial level (e.g., a small PFO or ASD), or a combination of

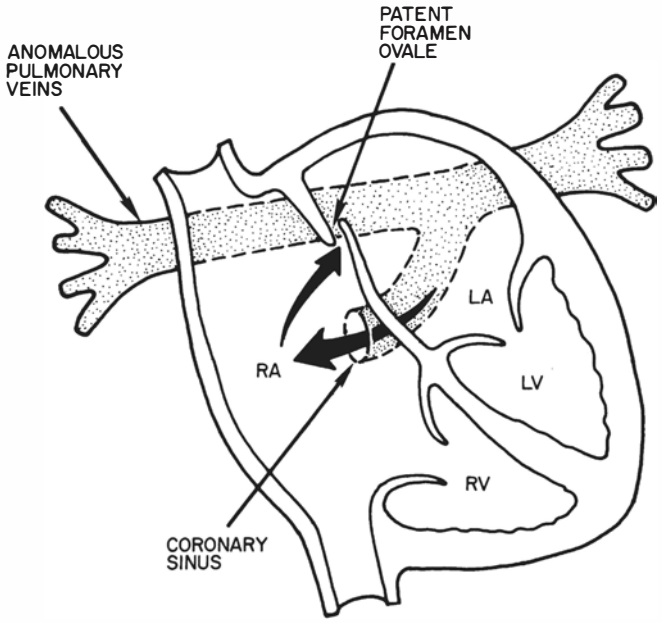


Fig. 97.3. Pathologic anatomy of cardiac-type total anomalous pulmonary venous return via the coronary sinus. (LA left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.)

these. The infracardiac TAPVR is associated with significant obstruction in 80% to 100% of patients. Usually, the descending vertical vein is obstructed at its junction with the portal vein or ductus venosus. In addition, resistance to blood flow through the hepatic sinusoids in the setting of a closed ductus venosus produces a functional obstruction. Supracardiac TAPVR variants may be obstructed due to a "vise-like" compression of the vertical vein between the left pulmonary artery anteriorly and the left mainstem bronchus posteriorly.

In patients with pulmonary venous obstruction, significant pulmonary edema results from the increased hydrostatic pressure in the pulmonary capillary bed proximal to the obstruction. The degree of cyanosis will depend on the balance between systemic and pulmonary blood flow. Patients with significant obstruction will have profound cyanosis due to a significantly decreased amount of oxygenated blood returning to the heart and mixing with the much larger volume of systemic deoxygenated blood. Pulmonary hypertension is usually present and most severe in patients with significant obstruction. In those patients, pulmonary artery pressures approach or exceed systemic pressures.

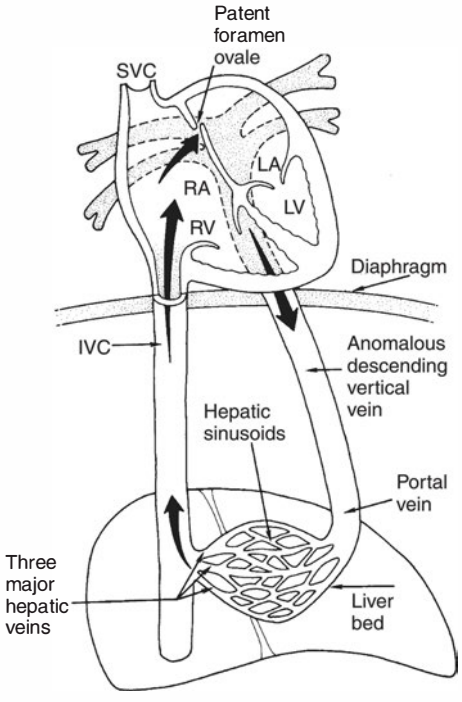


Fig. 97.4. Pathologic anatomy of infracardiac-type total anomalous pulmonary venous return via the descending vertical vein to the portal vein. IVC, inferior vena cava; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

Diagnosis

Signs and Symptoms

The condition of infants with TAPVR is determined largely by the presence and degree of obstruction to pulmonary venous return. Nonobstructed TAPVR often eludes diagnosis at birth and in the early neonatal period. Months later, patients may present with the gradual onset of tachypnea, dyspnea, congestive heart failure, and mild cyanosis. Often, subtle complaints of feeding difficulties and failure to thrive may be the only clues of congenital heart disease in these children. Liver congestion with hepatomegaly and cardiomegaly along with a prominent right ventricular impulse are consistent features.

Cardiac examination may be unimpressive. There may exist either a gallop or a faint diastolic murmur, most often a result of increased flow across the tricuspid valve. Other findings are the hallmarks of increased pulmonary blood flow, such as a systolic ejection murmur over the left second interspace and a prominent fixed S2 component.

In contrast, infants with obstructed TAPVR may present in extremis within hours to days after birth. These infants are profoundly cyanotic and show severe congestive heart failure. Hypotension and metabolic acidosis are frequently present. Findings on cardiac examination are inconsistent, but the right-sided cardiac chambers may be of normal size. The liver may also be of normal span and free of congestion if the ductus venosus remains patent and thus is able to divert pulmonary venous blood directly into the inferior vena cava.

Chest X-Ray

Chest X-ray features of TAPVR are again dependent on the presence or absence of significant pulmonary venous obstruction. Nonobstructed TAPVR is characterized by a normal-sized heart with increased pulmonary vascularity, possibly a distinct pulmonary artery silhouette, and occasional enlargement of the right cardiac silhouette. The classic X-ray images of a "snowman" or "figure-of-eight" in supracardiac connection are rarely appreciated before 6 months of life. In cases of obstructed TAPVR, the cardiac silhouette is usually of normal size but there is marked engorgement of the pulmonary vasculature and diffuse interstitial infiltrates in a reticular pattern indicative of severe pulmonary edema due to venous obstruction (Fig. 97.5).

Echocardiography

Modern two-dimensional echocardiographic technology and techniques have revolutionized the noninvasive diagnosis of TAPVR. In unstable, critically ill neonates, echocardiography prevents any undue delay in diagnosis and surgical therapy. Since the 1980s, echocardiography has become the mainstay for preoperative diagnosis and classification of TAPVR. Two-dimensional echocardiography with Doppler color-flow

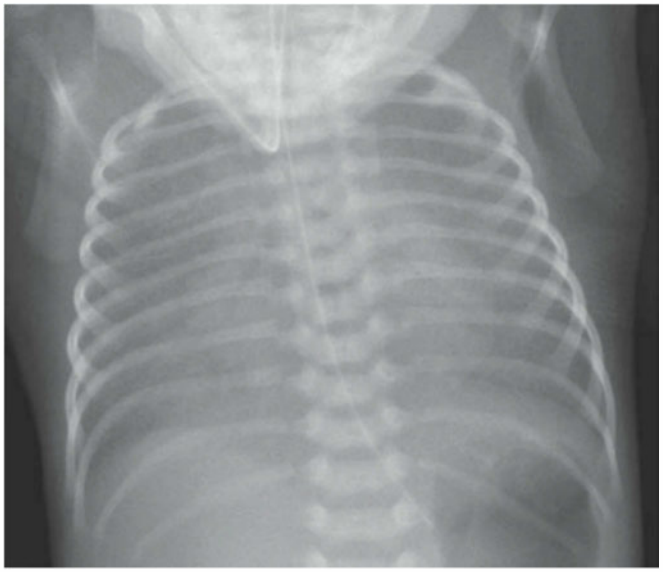


Fig. 97.5. Chest X-ray demonstrating diffuse interstitial pulmonary edema and a normal cardiac silhouette in a patient with obstructed total anomalous pulmonary venous return.

mapping is extremely accurate and reliable in diagnosing TAPVR, delineating the exact drainage pattern of each individual pulmonary vein, detecting the presence and degree of obstruction, and identifying any concomitant cardiac malformations. The absence of pulsatile pulmonary venous blood flow and dilation of the venous confluence or the vertical vein indicate obstruction. The vertical vein is usually well visualized and can be followed to its site of drainage in the systemic circulation.

Computerized Tomography and Magnetic Resonance Imaging

Contrast computerized tomography (CT) and magnetic resonance imaging (MRI) may be useful in patients in which echocardiography fails to delineate the exact anatomy. These modalities can provide important information, particularly in cases of mixed TAPVR or complex anatomy (Figs. 97.6 and 97.7). The downsides of these diagnostic studies (the time needed to obtain them and the administration of contrast) must be weighed against the information to be obtained, especially in patients with obstructed TAPVR that need urgent surgical intervention.

Cardiac Catheterization and Angiocardiography

In the current era, cardiac catheterization is rarely required and is mainly of historical interest. It should be reserved only for cases in which echocardiographic, CT, and MRI findings are inconsistent with the clinical course. The precise site of anomalous pulmonary venous connection is identified by a "step-up" in oxygen saturation in a systemic vein, whereas the exact course of pulmonary venous drainage is delineated during the levophase of selective pulmonary arteriography.

It is common for the blood in all four cardiac chambers to have equal or similar oxygen saturations, which reflects the mixing of pulmonary and systemic venous circulations. TAPVR and transposition of the great arteries are the only two conditions in which the oxygen saturation of blood in the main pulmonary artery is equal to or greater than that in the aorta. Right ventricular and pulmonary artery pressures are frequently elevated in TAPVR but are virtually always so in the presence of obstruction. The adequacy of the PFO in terms of its ability to shunt partially oxygenated blood to the left side of the heart may be estimated by the difference between right atrial pressure and pulmonary capillary wedge or left atrial pressure. The detection of a transatrial pressure gradient suggests a restrictive foramen ovale. Balloon atrial septostomy may be used to temporarily relieve significant obstruction at the atrial level if immediate surgical intervention is not possible.

Preoperative Management and Timing of Surgical Intervention

Infants presenting with obstructed TAPVR represent a surgical emergency. An intensive resuscitation period should be followed by expeditious surgical intervention. Immediate interventions include endotracheal intubation and mechanical ventilatory support. Pulmonary vasoconstriction should be avoided. Although pulmonary vasodilatory measures such as administration of 100% oxygen and hyperventilation to a partial arterial pressure of carbon dioxide ($PaCO_2$) of <30 mmHg have been traditionally used, it is unclear whether they provide any significant benefit. Metabolic acidosis should be treated with sodium bicarbonate or tromethamine infusions. Function of the

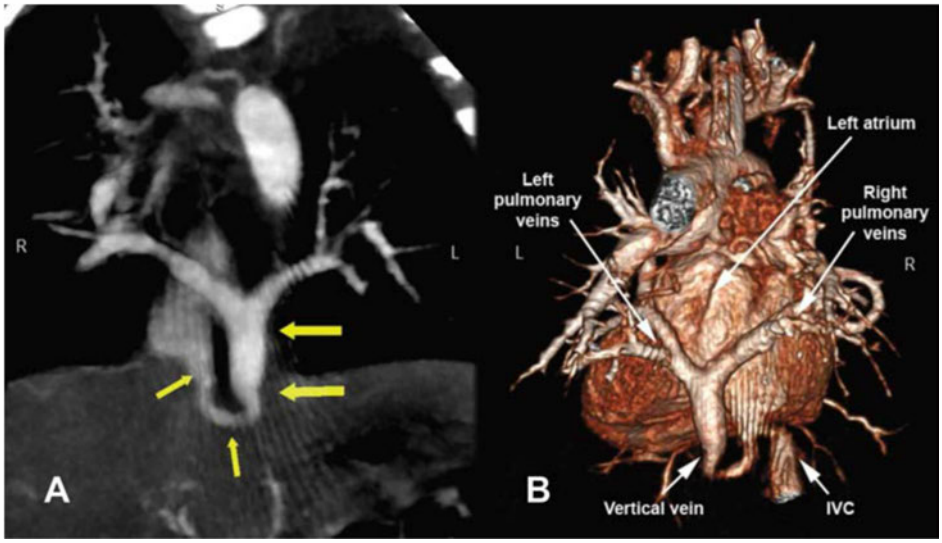


Fig. 97.6. Computerized tomographic angiography on a newborn with infracardiac-type total anomalous pulmonary venous return. The coronal view (**A**) and three-dimensional reconstruction (**B**) show that both left and right pulmonary veins form a confluence on each side of midline and then join to form a descending vertical vein (*arrows*). The vertical vein travels inferiorly to just below the diaphragm where it takes a curvilinear course and drains into the inferior vena cava before it drains into the right atrium. The curvilinear segment of the vertical vein is narrow and with evidence of obstruction.

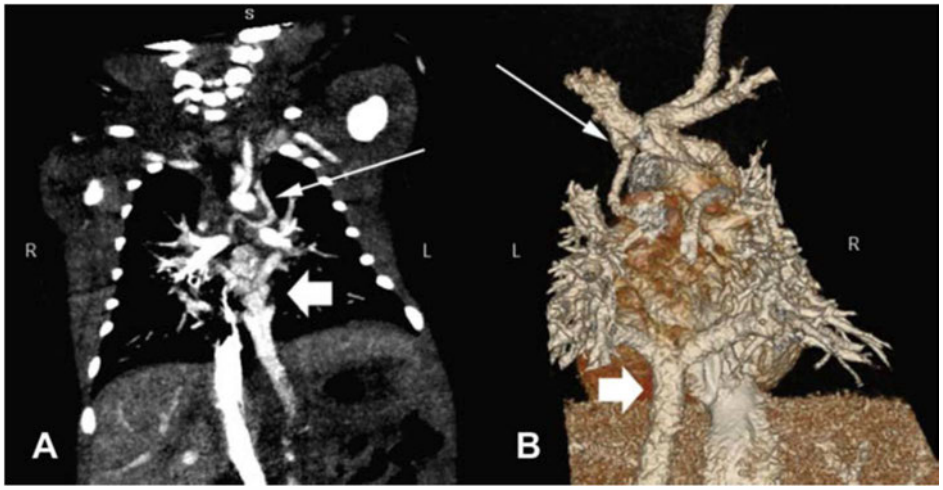


Fig. 97.7. Computerized tomographic angiography on a newborn with mixed-type total anomalous pulmonary venous return. As depicted on the coronal view (**A**) and three-dimensional reconstruction (**B**), all pulmonary veins except for the vein arising from the right middle lobe drain into a descending vertical vein (*thick arrow*) that joins the main portal vein at the porta hepatis. The right middle lobe is drained by a pulmonary vein that courses medially, between the carina and above the left pulmonary artery, to create an ascending vertical vein (*long arrow*) that drains into the left brachiocephalic vein.

failing heart is augmented with the administration of inotropic and diuretic agents. Maintenance of ductal patency with a prostaglandin E_1 infusion may also be of some physiologic benefit.

ECMO has been employed in those infants with severe pulmonary hypertension or cardiac failure refractory to conventional medical measures, although prompt surgical intervention is the mainstay of therapy. A brief period of ECMO may assist in stabilizing these critically ill infants and prevent end-organ dysfunction before operative intervention if immediate surgical intervention is not possible.

Nonobstructed TAPVR patients are usually in stable condition. Therefore, these infants rarely need significant preoperative intervention or pharmacologic support. Nonetheless, the current practice in most centers is to perform elective corrective surgery within a few days of diagnosis irrespective of the patient's age or weight. The impetus for early surgery has been a heightened awareness of the extremely high mortality associated with uncorrected TAPVR as well as the possibility of subclinical venous obstruction and the substantial risk of progression to irreversible cardiac and pulmonary vascular disease while awaiting delayed repair.

Surgical Technique

Preparation for and Management of Extracorporeal Circulation

The following descriptions apply specifically to TAPVR repair in neonates and young infants, with the understanding that older children and even adults also infrequently undergo corrective surgery for TAPVR. Central venous and umbilical arterial pressure catheters are usually placed during the preoperative resuscitation period. High-dose fentanyl is an ideal agent for the induction of anesthesia because it reduces stress-induced increases in pulmonary vascular resistance. Access to the heart and mediastinal structures is gained via a median sternotomy. After partial thymectomy, creation of a pericardial well, and systemic heparinization, an arterial cannula is placed in the ascending aorta. A single venous cannula is then positioned in the right atrial appendage. Alternatively, bicaval cannulation may be used to decrease the time of necessary circulatory arrest. Care is taken to avoid any undue manipulation of the heart until the institution of CPB because the myocardium of hypoxic, acidotic neonates with obstructed TAPVR is especially irritable and prone to ventricular fibrillation. Immediately after the commencement of CPB, the patent ductus arteriosus is dissected and ligated. During core cooling to a nasopharyngeal temperature of 18°C, the posterior pericardium is incised where it directly overlies the common pulmonary venous trunk. Once a core temperature of 18°C is achieved, the aorta is cross-clamped and antegrade cold cardioplegic solution is infused into the aortic root. Topical cold is also applied to the heart. Blood is then drained from the patient into the oxygenator to initiate the period of deep hypothermic circulatory arrest. At this point, the single atrial venous cannula may be removed to facilitate repair. If bicaval cannulation is used, periods of CPB can be alternated with periods of circulatory arrest, as needed. Circulatory arrest is a valuable adjunct to surgical repair of supracardiac, infracardiac, and mixed types of TAPVR because it provides a completely bloodless and quiescent operative field for fashioning of as large a pulmonary venous-to-left atrial anastomosis as possible. However, for the repair of TAPVR to the coronary sinus, standard CPB with two venous cannulas and moderate hypothermia (28°C) often suffices.

Supracardiac Type

Ligation of the vertical vein may be accomplished just after commencement of CPB or after completion of the repair. It is best to ligate the vein in its extrapericardial portion to avoid narrowing the ostium of the upper pulmonary veins. Because the condition of TAPVR renders the heart untethered by the pulmonary veins, excellent exposure of the posterior-lying common pulmonary venous confluence is afforded by retraction of the heart to the left or by lifting the apex of the heart to the right shoulder by using a pledgeted suture placed on the apex. However, one must be careful not to distort the cardiac and pulmonary venous anatomy during these maneuvers as distortion can result in kinking of the anastomosis when the heart is replaced to its natural position.

A longitudinal right atriotomy is performed and the left atrium is visualized through the septal defect. An incision is then performed on the posterior wall of the left atrium extending it superiorly toward the left atrial appendage for construction of a wide anastomosis. The particular site of the incision is chosen to optimize the apposition between the posterior wall of the left atrium and the pulmonary venous confluence and vertical vein without causing distortion (Fig. 97.8). A generous corresponding transverse incision is made in the common pulmonary venous confluence and vertical vein, where they lie in direct apposition to the posterior wall of the left atrium and left atrial appendage. Division of the vertical vein may be performed to use the distal end of the vein as part of a wide anastomosis. It is important not to extend the incision to include any individual pulmonary venous ostia. A continuous 7-0 or 8-0 polypropylene suture is used for the pulmonary venous-to-left atrial anastomosis using small and precise bites. The foramen ovale is then closed with the use of an autologous

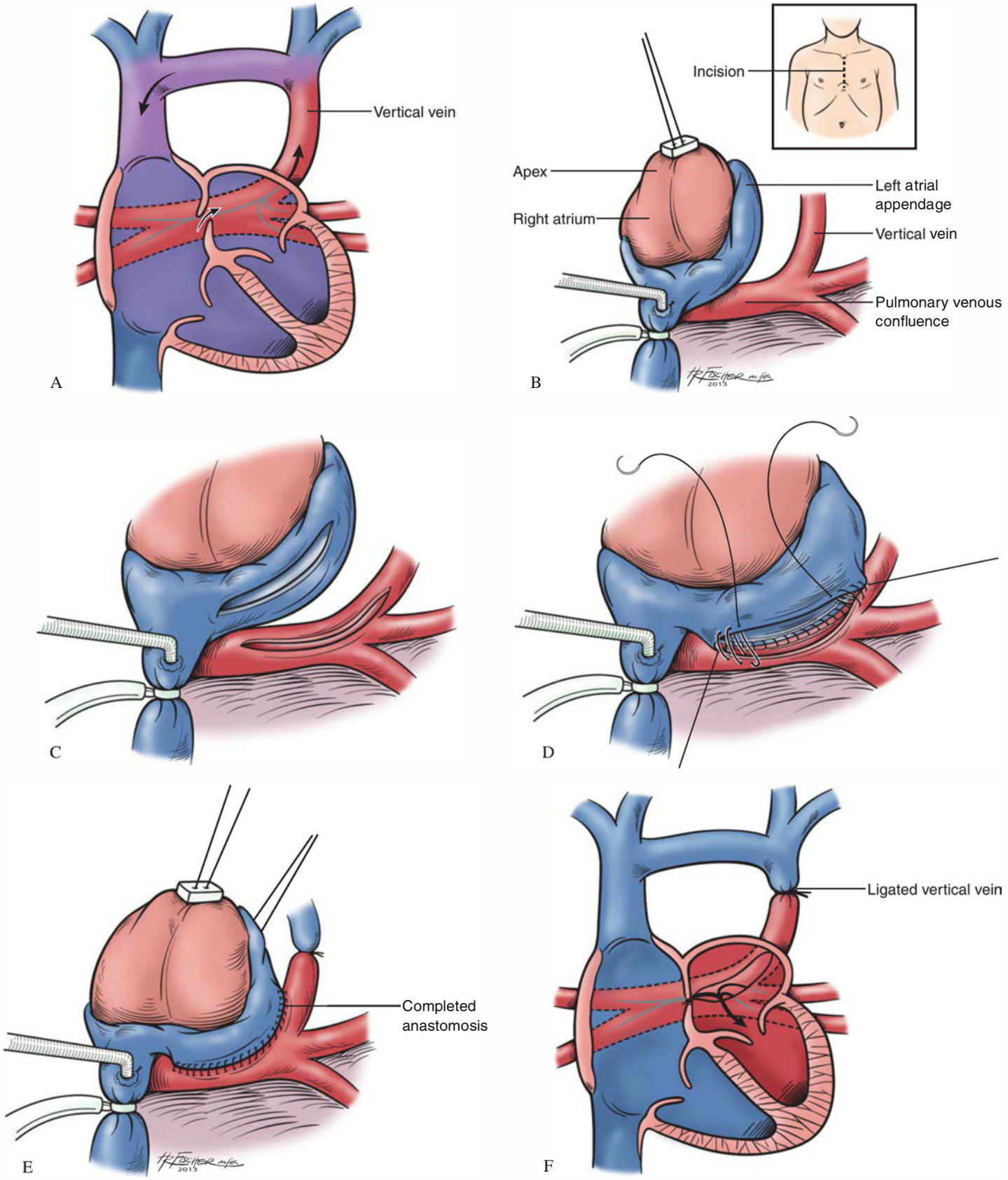


Fig. 97.8. Technique for repair of supracardiac-type total anomalous pulmonary venous return via a vertical vein (**A**). (**B**) A median sternotomy is performed. The pulmonary venous confluence is visualized by lifting the apex of the heart to the right shoulder with the use of a pledgeted stitch placed on the apex. (**C**) An incision is performed on the posterior wall of the left atrium extending it into the left atrial appendage to correspond with an incision performed on the pulmonary venous confluence and the vertical vein. (**D**) The anastomosis is performed between both incisions and particular care is placed on not distorting the anastomosis. Panel (**E**) shows the completed anastomosis. Panel (**F**) shows the flow from the pulmonary veins into the left atrium.

pericardial patch with continuous 6-0 polypropylene suture through the right atrium; direct suture closure of the foramen may jeopardize the patency of the common pulmonary vein-to-left atrium anastomosis. The right atriotomy is then closed with a continuous 6-0 polypropylene suture.

After completion of the repair, the heart is filled with saline, air is vented through the cardioplegia catheter site, and the venous cannula is reinserted into the right atrium. CPB is resumed, with systemic rewarming to a core temperature of 34 to 35°C. During rewarming, atrial and ventricular pacing wires are placed. A right atrial catheter may be placed for intravenous access and a left atrial catheter may be placed for postoperative monitoring.

Cardiac Type

Repair of TAPVR to the coronary sinus is approached through a longitudinal right atriotomy (Fig. 97.9). After performance of a median sternotomy and institution of CPB with bicaval cannulation, the patient is cooled to 28°C. A segment of atrial septal tissue is excised between the dilated coronary sinus ostium and the PFO; the valve of the foramen ovale is removed in the process. An unroofing incision is then made in the superior wall of the coronary sinus such that drainage of pulmonary venous and coronary sinus blood is redirected to the left atrium. An autologous pericardial patch is sutured with continuous 6-0 polypropylene to the perimeters of the foramen ovale and the coronary sinus ostium to close the resultant interatrial communication.

TAPVR directly to the right atrium is also approached through a right atriotomy. The pulmonary venous ostia are visualized as they open into the posterior aspect of the right atrium, usually by the way of a single venous trunk from each lung. A large ASD is created by excision of a portion of the atrial septum surrounding the foramen ovale. An autologous pericardial patch is then sutured over the resultant defect to include the pulmonary venous ostia. The functional results of such a repair are that the interatrial communication is closed and pulmonary venous drainage is baffled to the left atrium.

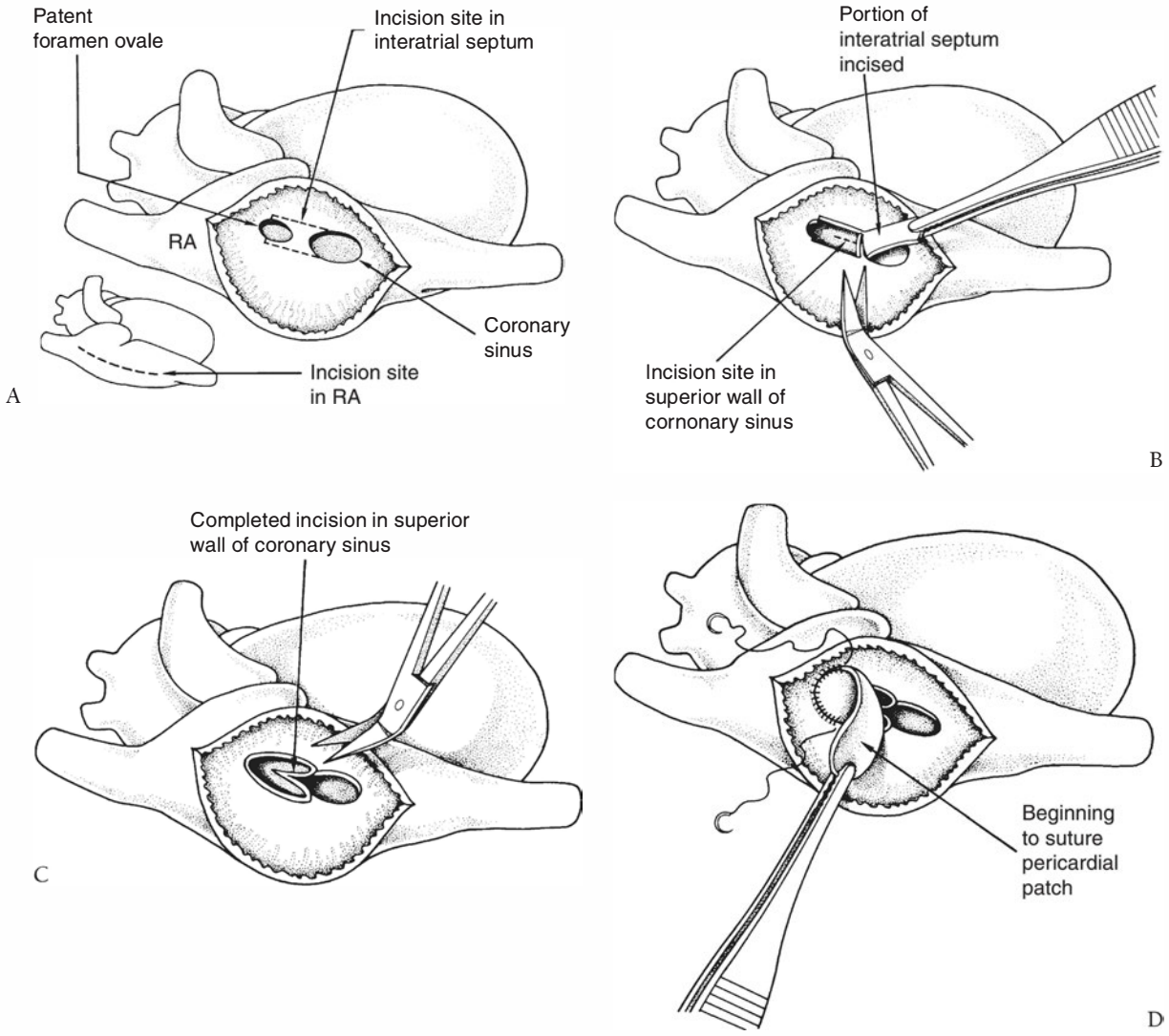


Fig. 97.9. Technique for repair of cardiac-type total anomalous pulmonary venous return via the coronary sinus. (**A**) Inset shows the site of the longitudinal incision in the right atrium. A portion of atrial septum is excised between the coronary sinus and foramen ovale, removing the valve of the latter. (**B**) Site of the incision in the superior wall of the coronary sinus. (**C**) An unroofing incision is made in the coronary sinus, allowing redirection of coronary sinus blood flow into the left atrium. (**D**) A pericardial patch is sutured around the perimeters of the coronary sinus and foramen ovale such that pulmonary venous blood returning to the former is diverted underneath the patch, through the patent foramen ovale, and into the left atrium. RA, right atrium.

Infracardiac Type

TAPVR below the diaphragm is obstructed in the vast majority of patients; thus, patients diagnosed with this anomaly usually undergo an emergency operation in the neonatal period. A midline sternotomy, CPB, and deep hypothermic circulatory arrest are used as described earlier. During the period of systemic cooling, the apex of the heart is retracted anteriorly to facilitate dissection and long mobilization of the descending vertical vein. The anomalous vertical vein is routinely ligated. Division of the vein may allow further mobility to allow the confluence to reach the left atrium without tension. As opposed to the supracardiac variant, the common pulmonary venous confluence in infra diaphragmatic TAPVR lies predominantly in a vertical orientation. Hence, a lengthy vertical incision in the vein is often required, extending from the superior aspect of the confluence into the descending vertical vein. A longitudinal right atriotomy is performed and an incision is created in the posterior wall of the left atrium such that it parallels the pulmonary venous confluence. Superior extension into the left atrial appendage may be required for a wider anastomosis. The common pulmonary venous-to-left atrial anastomosis is fashioned in a manner similar to that used for the repair of supracardiac lesions, using a continuous 7-0 or 8-0 polypropylene suture. Pericardial patch closure of the foramen ovale and continuous suture closure of the right atriotomy are performed in an identical manner to that described for supracardiac repair.

Mixed Type

Mixed patterns of pulmonary venous drainage, in which one or more pulmonary veins do not enter into the common pulmonary venous confluence but instead connect with the systemic venous circulation independently, are treated by a combination of the techniques described in the preceding paragraphs. Direct suturing of pulmonary veins to the left atrium is discouraged due to the higher risk of delayed pulmonary vein stenosis. In the event that only a single pulmonary vein from one lobe drains separately, it may be left uncorrected with minimal adverse physiologic sequelae.

Postoperative Management

In those infants who present initially with an obstructed subtype of TAPVR, pulmonary pressures are elevated due to high pulmonary vascular resistance secondary to preoperative injury and the inflammatory effects of CPB. Persistent pulmonary hypertension is a major cause of early morbidity and mortality after repair of obstructed TAPVR. It is not uncommon to see significant pulmonary hemodynamic lability and pulmonary hypertensive crises in the postoperative period in these patients. Although pulmonary artery-monitoring catheters are not a standard requirement for the management during the first 24 to 48 postoperative hours, it is essential to institute prophylactic measures to maintain an acceptably low pulmonary vascular resistance. Such strategies include hyperventilation-induced respiratory alkalosis and careful titration of the inspired oxygen concentration to minimize pulmonary vasoconstriction. If pulmonary pressures remain elevated beyond two-thirds or more of systemic pressure, right ventricular end-diastolic pressures may rise higher than left atrial pressure, thereby reducing stroke volume. In these cases, acidemia may ensue and should be vigorously treated pharmacologically as well as with ventilator management.

A continuous narcotic infusion (usually fentanyl) is useful for sedation, providing postoperative analgesia and blunting stress-induced increases in pulmonary vascular resistance. A paralytic agent to inhibit skeletal muscle activity with the intent of obtaining complete control of ventilation, and minimizing oxygen consumption may be useful in the acute postoperative period. Milrinone is an ideal agent when inotropic support is required because it also produces pulmonary artery vasodilation. Rarely, a sodium nitroprusside infusion may be required for the management of difficult cases of pulmonary hypertension. Nitric oxide is often a useful agent in cases of difficult-to-manage pulmonary hypertension. ECMO has been infrequently used when maximal combinations of the foregoing noninvasive measures fail to improve pulmonary vasospasm and right heart function. Before ECMO is initiated, mechanical causes of refractory pulmonary hypertension, such as pulmonary venous obstruction or anastomotic stricture, should be ruled out with echocardiography.

Usually, a progressive decrease in pulmonary vascular tone permits weaning of ventilation, inotropes, and pulmonary vasodilators within 24 to 48 hours after surgery. Pulmonary edema is a frequent complication after TAPVR repair, especially in nonobstructed cases in which there is long-standing preoperative right ventricular volume overload and in the presence of a diminutive left ventricle. Postoperative fluid management is critical, as is the judicious administration of diuretic agents such as furosemide. Due to the relatively small left-sided structures on TAPVR, the heart is very sensitive to small increases in preload. Measurement of left atrial pressures can be useful to guide fluid and inotropic management. If there is deviation from the expected postoperative course or the patient is refractory to the usual interventions, an echocardiogram should promptly be obtained to investigate the possibility of pulmonary venous obstruction or anastomotic stricture.

Results

Historically, the surgical mortality for TAPVR repair had been extremely high, as evidenced by a 65% to 85% rate of early death for infants undergoing repair in the 1960s. With the advent of deep hypothermic circulatory arrest in the 1970s, the 30-day operative mortality decreased markedly to 12% from 18%. Further advances in noninvasive diagnosis, surgical technique, cardiac anesthesia, and perioperative cardiopulmonary support, as well as adoption of the routine practice of early surgical intervention, have resulted in mortality rates of $<5\%$ in many recently reported series. Although young age at operation, preoperative pulmonary venous obstruction, infracardiac type of TAPVR, and emergent operation have all been previously indicted as risk factors for early death after isolated TAPVR repair, management practices of this lesion have virtually eliminated most identifiable preoperative risk factors for adverse outcome in the current era. Associated cardiac anomalies such as pulmonary atresia or the need for a systemic-to-pulmonary shunt may increase perioperative risk. Refractory pulmonary hypertension and cardiac failure continue to constitute the majority of intraoperative and early postoperative deaths in most series.

The most significant cause of late morbidity and mortality after corrective surgery for TAPVR remains recurrent pulmonary venous obstruction, which develops in 5% to 15% of patients within the first 6 to 12 postoperative months. The predominant pathologic manifestations of this complication are anastomotic fibrotic stricture, discrete stenoses of the individual pulmonary venous ostia, and a poorly understood diffuse fibrotic process that may involve the entire length of a pulmonary vein. There have been no technical factors that have reliably predicted or prevented the more insidious occurrence of diffuse, fibrotic stenosis of one or more pulmonary veins. This process accounts for a large proportion of the 5% to 10% late mortality rate after TAPVR repair. Various surgical techniques for managing postoperative pulmonary venous obstruction have been proposed, including revision of the common

pulmonary vein-to-left atrium anastomosis, patch angioplasty of stenotic pulmonary veins, creation of a sutureless pericardial well, and suturing of individual pulmonary veins directly to the left atrium. Pulmonary venous stents have been used with mixed results. Lung transplantation recently has been proposed for treating patients who have no other surgical alternatives.

Junctional rhythms and various degrees of heart block can complicate TAPVR repair of the cardiac type. The internodal tracts and the atrioventricular node itself may be disrupted during suturing around the coronary sinus, particularly around its anterior margin. Although some patients do sustain spontaneous conversion to a normal sinus rhythm after a few days of external pacing in the early postoperative period, others may require implantation of a permanent pacemaker as a result of a persistent postsurgical rhythm disturbance.

PARTIAL ANOMALOUS PULMONARY VENOUS RETURN

PAPVR is a congenital cardiac anomaly in which one or more, but not all, of the pulmonary veins connect with the right atrium or one of its systemic tributaries. By definition, at least one pulmonary vein must drain normally into the left atrium. Whereas the lesion is reported to occur in up to 0.7% of autopsy specimens, the frequency of antemortem diagnoses is somewhat lower as a result of a relatively high number of asymptomatic cases. The initial pathologic description was presented by Winslow in 1739, but it was not until 1949 that the first antemortem diagnosis was reported by Dotter and colleagues using angiocardiography.

Pathologic Anatomy

PAPVR results from involution of the right or left portion of the common pulmonary vein at a stage in embryonic development when primitive pulmonary-to-systemic venous anastomoses still exist. In approximately 80% of cases, the anomalous vein or veins arise from the right lung, whereas in only 10% of cases does the left lung represent the sole source of PAPVR. Most commonly the pulmonary veins from the right upper and middle lobes drain anomalously into the right superior vena cava or, less frequently, connect directly with the right atrium. The innominate vein, coronary sinus, azygos vein, portal vein, and inferior vena cava represent uncommon sites of systemic venous connection. The anomalous pulmonary veins may enter the systemic circulation as a common trunk or as multiple individual connections. An associated ASD is identified in at least 80% of patients with PAPVR. Although a high-lying sinus venosus defect is the most frequent coexisting cardiac malformation, a secundum ASD is also seen. In addition, a host of associated complex cardiac anomalies has been described in patients with PAPVR; in such cases, these complex defects dominate the clinical picture.

Scimitar Syndrome

By definition, the scimitar syndrome involves PAPVR from the right lung to the inferior vena cava. Some or all of the pulmonary veins unite in a common trunk to descend in a gentle curve alongside the right heart border en route to the inferior vena cava, giving the radiographic appearance of a scimitar (Turkish sword). The conjoined anomalous veins may enter the inferior vena cava near the cavoatrial junction or in its subdiaphragmatic portion. The right pulmonary artery and lung are hypoplastic in more than one-half of cases. Other associated anomalies include systemic arterial supply to the right lung (especially the right lower lobe) from aortopulmonary collaterals, abnormal lobation and bronchial distribution, secundum ASD, pulmonary vein stenosis, and a variety of other complex cardiac malformations.

Pathophysiology

The hemodynamic consequences of PAPVR are very similar to those of a large, isolated ASD. Pulmonary blood flow is substantially increased, but pulmonary vascular disease per se usually does not supervene for several decades with the lesion. The chronic right ventricular volume overload eventually yields right ventricular hypertrophy, dilation, and failure. The onset of pulmonary hypertension heralds an end-stage manifestation of a long-standing, uncorrected left-to-right atrial shunt. For unclear reasons, the scimitar syndrome is associated with pulmonary hypertension at a younger age and more frequently than other types of PAPVR.

Diagnosis

The majority of patients with PAPVR remain asymptomatic throughout early childhood. Those who eventually develop symptoms usually do so in the third or fourth decade of life, with the most common complaints being easy fatigability and mild exercise intolerance. Younger patients come to medical attention as a result of an incidentally discovered cardiac murmur, abnormalities on a chest X-ray, or recurrent pulmonary infections. Patients with associated major cardiac anomalies present in infancy with hemodynamic features referable to these complex lesions; in most such cases PAPVR is an incidental finding.

The mainstay in the diagnosis of PAPVR has become two-dimensional echocardiography with Doppler color-flow mapping. As in TAPVR, this modality is highly accurate in delineating the anomalous pulmonary veins and ASD. CT and MRI have proven to be useful in recent years to further define the anatomy and the relationship between the anomalous vessels and the right atrium or SVC. Cardiac catheterization is reserved for cases in which there are associated complex lesions or pulmonary hypertension is believed to exist. The pulmonary-to-systemic blood flow ratio (Q_p/Q_s) is usually found to be $>1.5:1$.

Surgical Management

To avert the onset of the potentially irreversible complications of right heart failure and pulmonary hypertension, elective surgical repair should be undertaken in all patients diagnosed with PAPVR. In general, the surgical principles of PAPVR repair include separation of the systemic and pulmonary venous circulations, avoidance of creating obstruction to superior vena cava or pulmonary venous blood flow, complete closure of the ASD, and preservation of sinoatrial (SA) node function.

Partial Anomalous Pulmonary Venous Return to the Low Superior Vena Cava or Right Atrium

We use a combination of techniques depending on the anatomy of the anomalous veins. For PAPVR with veins returning into the right atrium or low SVC, we tend to use a technique very similar to the simple pericardial baffle procedure originally described by Kirklin and associates in 1956 (Fig. 97.10). The heart and mediastinal structures are approached through a median sternotomy. The right pleural space may be entered to verify the origin of the anomalous veins from the right lung. The superior vena cava is completely dissected from the cavoatrial junction to the level of the innominate vein to clearly define the presence of any pulmonary veins returning to the SVC. With care taken to avoid obstructing access to the anomalous pulmonary venous ostia, the superior vena cava is cannulated with an angulated venous cannula placed above the anomalous pulmonary veins. If the anomalous veins enter very high in the SVC, direct cannulation of the innominate vein may be used. A standard venous cannula is used for

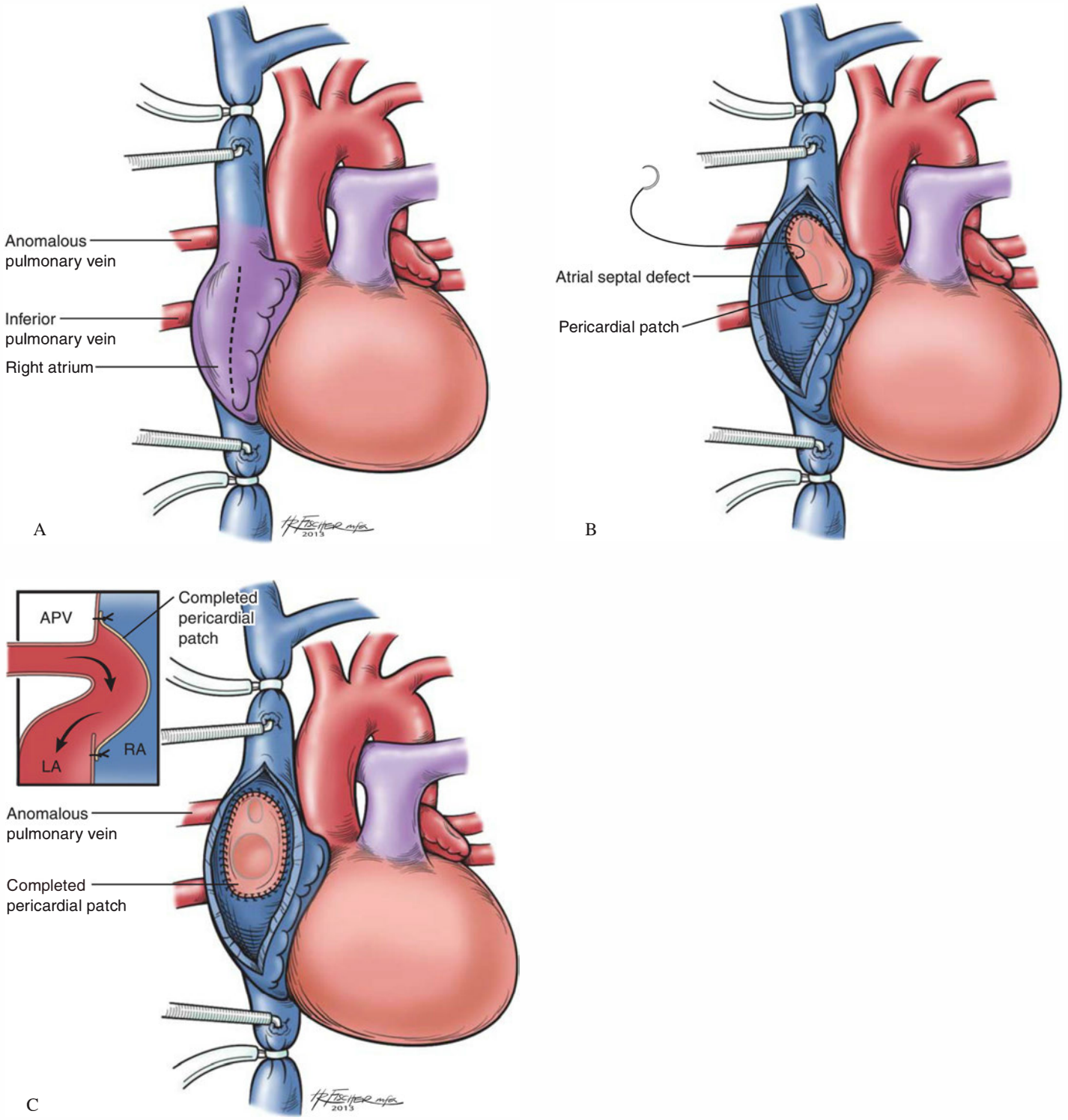


Fig. 97.10. Technique for repair of partial anomalous pulmonary venous return (PAPVR) to the right atrium. Panel (**A**) illustrates the location of an oblique right atriotomy after dissection of the superior vena cava and anomalous pulmonary veins. (**B**) After intracardiac visualization of all pulmonary veins, a piece of autologous pericardium is used to baffle the orifice of the anomalous vein through the atrial septal defect and into the left atrium. Panel (**C**) illustrates the completed baffle. Care must be made not to allow the baffle to obstruct the entrance of the superior vena cava.

inferior vena caval drainage. CPB is initiated with cooling to moderate systemic hypothermia (28 to 32°C). Occasionally, in our experience, cooler temperatures may be used, allowing temporary low flow to provide a bloodless field. The aorta is then cross-clamped, and cold potassium cardioplegic solution is infused in the aortic root.

A vertical incision is made in the right atrium lateral to the right atrial appendage extending upward to a level below the SVC/right atrial junction to avoid injury to the sinus node artery. Such an incision affords excellent exposure to all pulmonary venous ostia and the ASD and avoids injury to the SA node. If the ASD is narrow or the atrial septum is intact, a surgical atrial septostomy is created. A baffle of autologous pericardium is sutured into the lumen of the superior vena cava and the right atrium such that blood flow from the anomalous pulmonary veins is redirected from the superior vena cava, across the ASD, and into the left atrium. Care is taken to avoid narrowing the lumen

of the superior vena cava or obstructing the pulmonary venous ostia with the pericardial baffle. The incision in the right atrium and superior vena cava may be closed with a pericardial patch to eliminate the possibility of superior vena caval stenosis.

Partial Anomalous Pulmonary Venous Return to the High Superior Vena Cava

Even though the simple pericardial baffle technique may be used for PAPVR to the high superior vena cava, we favor the use of the Warden procedure when the pulmonary veins drain 1 cm or higher above the cavoatrial junction (Fig. 97.11). We believe that this procedure, described by Warden et al. in 1984, decreases the risk of postoperative obstruction of the SVC, pulmonary venous obstruction, residual atrial shunts,

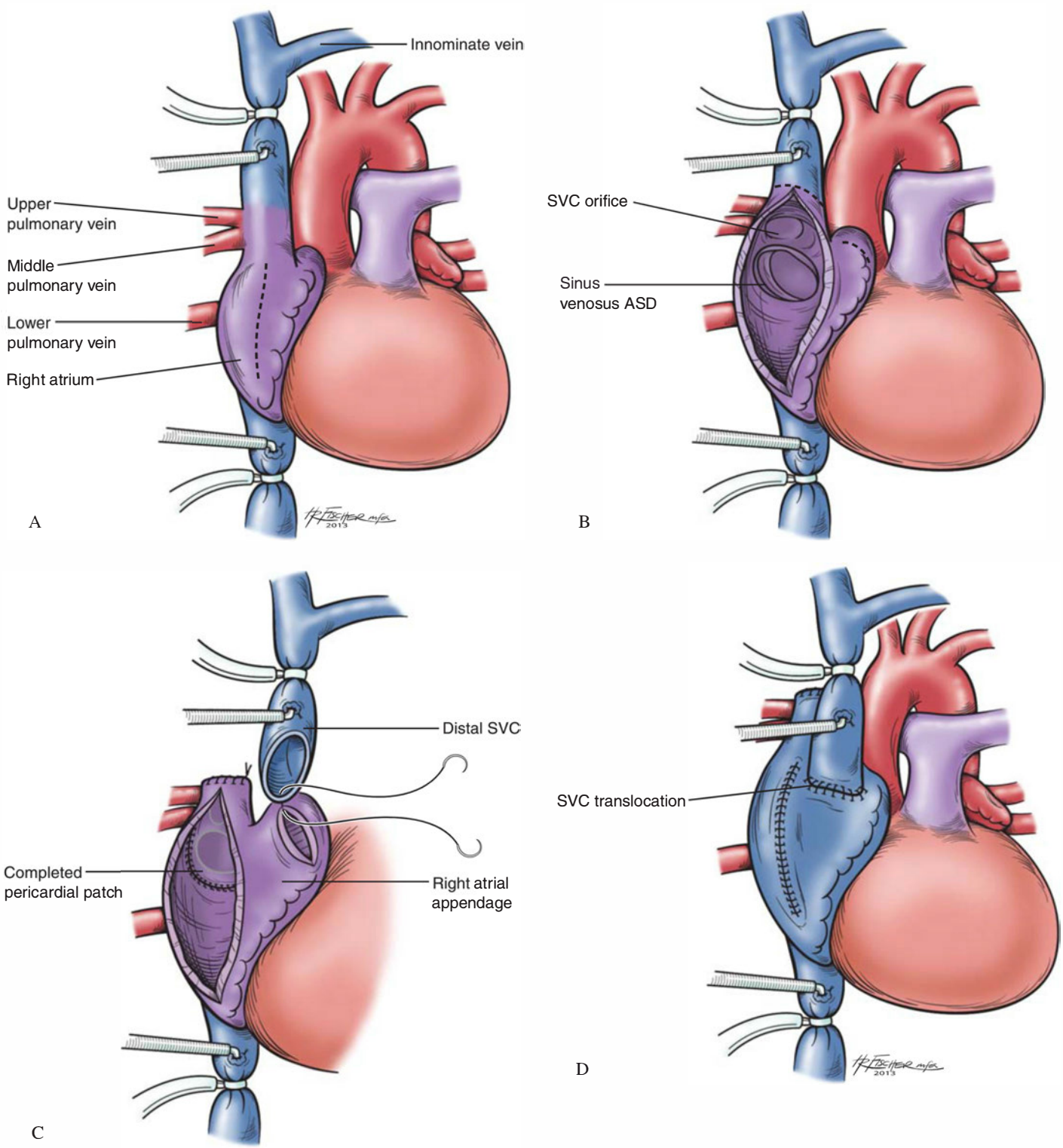


Fig. 97.11. Technique for repair of partial anomalous pulmonary venous return (PAPVR) to the superior vena cava using the Warden procedure. (**A**) After institution of cardiopulmonary bypass and cardioplegic arrest, a right atriotomy is performed. (**B**) The intracardiac anatomy is examined. The dashed lines indicate the location of incisions to be performed on the right atrium. (**C**) The superior vena cava is divided above the level of the lowest anomalous pulmonary vein. The cardiac end of the superior vena cava is sutured closed and the superior vena cava orifice is baffled through the atrial septal defect into the left atrium. An incision is made on the right atrial appendage and the distal superior vena cava is anastomosed to the appendage. Panel (**D**) shows the completed anastomosis.

and SA node dysfunction. Cannulation and CPB are conducted similar to the simple pericardial baffle technique. A longitudinal right atriotomy is performed and the ASD inspected. It may be necessary to enlarge the ASD superiorly, especially in cases of a secundum ASD, in order to allow adequate baffling of the SVC to the left atrium. The SVC is transected distal to the uppermost pulmonary vein. Depending on the anatomy, the azygos vein may need to be ligated and divided. The proximal SVC, containing the drainage from the pulmonary veins is then sutured closed, either primarily or with the use of an autologous pericardial patch. A pericardial patch is used to baffle blood from the cavoatrial junction (now containing only pulmonary venous drainage) through the ASD and into the left atrium. The tip of the right atrial appendage is divided and anastomosed to the distal SVC, therefore, directing the superior systemic venous drainage into the right atrium.

Scimitar Syndrome

In general, pulmonary resection rather than vascular reconstruction is recommended for the treatment of PAPVR in the scimitar syndrome if the right lung has been destroyed by recurrent infections, when it does not contribute appreciably to gas exchange, or in patients in whom an anomalous pulmonary vein arises from only a single lobe. Conversely, a variety of vascular reconstructive procedures have been proposed to reroute the pulmonary venous drainage to the left atrium in patients with sufficient right lung function and adequate pulmonary arterial blood supply. The anomalous pulmonary venous trunk is often too short for direct reimplantation into the left atrium; therefore, most procedures involve redirecting the anomalous drainage from the right atrium, across an ASD, and into the left atrial cavity via a pericardial tunnel. Aberrant aortopulmonary collaterals may be successfully treated by either surgical ligation or transarterial embolization.

Results

The surgical mortality for PAPVR repair is <1%. However, the higher mortality after repair of PAPVR in patients with the scimitar syndrome is a reflection of the frequently associated pulmonary hypertension and multiple complex cardiovascular anomalies. Although the reported incidences of late superior vena caval obstruction, pulmonary venous obstruction, residual atrial shunts, and rhythm disturbances is low after P APVR repair, no single surgical technique has demonstrated a distinct advantage in further reducing these complications.

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EDITOR'S COMMENTS

The surgical results of complete repair of TAPVR have dramatically improved over the last two decades. Currently, with the techniques described in this chapter, operative mortality for these defects is <5% in most major centers. Isolated patients still do not survive operation primarily because of severe obstruction of the pulmonary venous confluence or restrictive ASD. These infants often present in extremis with multisystem organ failure before operative intervention. Thus, early recognition of this congenital lesion with prompt medical stabilization and urgent surgical therapy may result in a marked improvement in overall survival.

We use the same approaches to supracardiac and intracardiac forms of TAPVR as described by the authors. The biatrial transverse incision allows a very wide exposure of the back of the left atrium and the widest possible anastomosis between the common pulmonary venous confluence and the left atrium. In most cases, we incise into the individual pulmonary veins to create the widest possible anastomosis and open beyond potential constrictive tissue, which may occur at the entrance points of the pulmonary veins to the common pulmonary venous confluence. In some cases, the pulmonary venous confluence is sutured to the back of the atrium such that the confluence extends beyond the atrial septum, and in this case, the atrial septum is recreated with a pericardial or homograft patch. In some cases, the right atrium is also enlarged during closure with a patch.

Because of the difficulties with the development of late obstruction in patients with single ventricle and total anomalous pulmonary venous return, we have more recently used "sutureless" techniques for primary repair of total anomalous pulmonary venous return in these patients and are extending this technique to patients with more standard isolated total anomalous pulmonary venous return. The advantages of a sutureless repair technique are that the venous confluence can be widely incised out into each of the branch veins and then the pericardium or perivascular adventitial tissue can be sewn to a wide opening in the back of the left atrium. With this technique in a primary repair situation, there is no need to suture at any point to the actual pulmonary vein wall, which may decrease the risk of scarring and late development of stenosis. Although sutureless techniques for repair of total anomalous pulmonary venous return have become increasingly popular, it is still not clear that there is an overall decrease in the incidence of late pulmonary venous obstruction with this approach.

An area of controversy in repair of total anomalous pulmonary venous return is the issue of leaving the vertical vein open after repair. In several experimental studies and clinical series, it appears that leaving a vertical vein open after repair

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EDITOR'S COMMENTS (*continued*)

seems to improve the postoperative stability of the patients and maintains cardiac output better than if the vertical vein is ligated and the vein used in the repair as described by the authors of this chapter. Initially, it was considered somewhat illogical that leaving a vertical vein open would significantly improve postoperative cardiac output since any decompression of the vertical vein into the right side of the heart would then be re-presented to the left atrium. It appears that perhaps the salient benefit of this approach is permitting a larger capacitance to the left atrium by essentially allowing the venous capacitance to be part of the left atrial capacitance and limiting left atrial hypertension, which may improve cardiac output even at the expense of what is a left-to-right shunt. If the vertical vein is left open there has been a reported significant incidence of development of a late left-to-right shunt that may need to be addressed surgically, or by interventional catheterization procedures.

The mixed types of TAPVR may represent technical problems in very small infants because anastomotic techniques for individual pulmonary veins may lead to significant late stenosis. In addition, patients who have all four pulmonary veins entering separately into the right atrium (although simple to repair by excising the atrial septum and baffling the veins to the left atrium) have a very high incidence of late development of pulmonary venous obstruction of the individual veins and severe pulmonary hypertension. These patients must have continued surveillance postoperatively, and if pulmonary venous obstruction occurs, prompt lung transplantation is the optimal therapy.

Controversy continues regarding the relative contributions of anastomotic strictures to the late development of pulmonary venous obstruction in TAPVR. It is our impression that most pulmonary venous obstruction is not related to an inadequate anastomosis, but instead to involution of tissue of the common pulmonary vein or the individual pulmonary venous entrances into the common pulmonary venous channel, which occurs in the first 2 months after birth. Thus, a widely patent anastomosis may become restrictive with time. In situations in which the common pulmonary venous channel and individual veins are dilated with a restriction at the level of the entrance into the heart, surgical revision can be undertaken using sutureless techniques with good success rates. This appears to be most common in patients who have cardiac forms of TAPVR, in whom a wide anastomosis to the left atrium is created by unroofing the coronary sinus. In some of these patients, continued involution of the common pulmonary venous tissue can cause restriction of this anastomosis and require revision. Where individual pulmonary veins are stenotic and the proximal vein *not* dilated, operative intervention has virtually no role in resolving severe pulmonary hypertension. These patients appear to have a diffuse process in the pulmonary vein extending back into the hilum of the lung, and even sutureless techniques of vein repair do not result in a significant drop in the pulmonary resistance in most cases. If there is mixed dilation of some veins and not others, then sutureless repair techniques may be attempted before consideration for lung transplantation. Nevertheless, these patients have not responded to interventional catheterization techniques or surgical therapy in most cases and should be referred early for consideration of lung transplantation. Very high mortality with conservative measures in these patients and the inability to stent veins to relieve obstruction in the more proximal vessels have been noted. The use of stents in pulmonary veins after repair of total anomalous pulmonary venous return or in patients with congenital pulmonary vein stenosis has been associated with recurrent obstruction very rapidly following stent implantation or balloon dilation of the pulmonary veins. Thus, stent implantation probably has no role in the chronic treatment of pulmonary vein obstruction but may have a role as temporary decompression in patients who have been listed for definitive therapy with lung transplantation. Even with consideration of lung transplantation, these patients develop severe pulmonary hypertension and often have a progressive downhill course before a lung donor becomes available.

An additional problematic group comprises patients with scimitar syndrome. Although the authors recommend that all patients with PAPVR and significant left-to-right shunts undergo operative repair, scimitar syndrome repair has been associated with a high incidence of pulmonary venous obstruction despite an initial successful baffling of the venous return to the left atrium. These patients then develop pulmonary venous obstruction and often hemoptysis from the right lung, which has little pulmonary blood flow other than bronchial supply. In addition, the aberrant aortopulmonary collaterals that enter the lung through the diaphragm and inferior pulmonary ligament may cause sources of bleeding; recurrent infection is not uncommon. In these patients, pneumonectomy of the affected lung can often resolve the hemoptysis and improve overall ventilation perfusion match.

Patients with scimitar syndrome presenting in infancy are a very difficult subgroup of patients with this anatomy. Often these children have hypoplasia of the right lung with a significant shunt from the aortopulmonary collateral vessels. In addition, associated congenital heart lesions are common. Even with complete correction of the cardiac defects, persistent pulmonary hypertension is common, and the mortality for operative intervention in these infants is significant. For these reasons, some authors advocate lung removal in these patients or medical management alone if possible. Some infants who have significant pulmonary hypertension in association with scimitar syndrome in infancy, often associated with other intracardiac shunts, may be palliated by interventional occlusion of significant aortopulmonary collateral vessels into the right lung coming through the diaphragm. Elimination of this additional source of left-to-right shunting may, in fact, improve pulmonary hypertension and failure to thrive in some of these children and delay the need for later surgical reconstruction of the scimitar vein.

The significant incidence of pulmonary venous obstruction after cardiac repair in isolated scimitar syndrome has led other authors to suggest that these patients do not undergo operative therapy. Reimplantation of the pulmonary veins into

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EDITOR'S COMMENTS (*continued*)

the back of the right atrium with baffling to the left atrium or direct anastomosis into the left atrium may be complicated by kinking of the pulmonary veins when the lung expands. Dr. John Brown and his colleagues have shown good success with direct anastomosis of the scimitar vein into the left atrium using a right thoracotomy approach with a low incidence of any turbulence or potential obstruction at intermediate follow-up. The encouraging results with this technique have led many centers, including ours, to preferentially use reimplantation through a thoracotomy approach if the patient does not have a significant additional intracardiac defect that would require cardiopulmonary bypass or a sternotomy approach such as an ASD. Most recently, we have used a technique similar to that described by Brown and associates for reimplanting the scimitar vein directly into the left atrium in patients who have a reasonable size right lung and significant left-to-right shunt, but who do not have an associated atrial or ventricular level defect that would require the use of cardiopulmonary bypass. We have divided the scimitar vein at its entrance into the inferior vena cava and, then used a short segment of PTFE graft to anastomose the vein to the left atrium. The graft functions as a stent of the anastomosis to prevent late stricture formation which has been the most frequent complication after direct anastomosis of pulmonary veins to the heart. Long baffles created from the entrance of the pulmonary veins at the level of the diaphragm along the right atrium into the left atrium have been associated with progressive stenosis. This is primarily because the veins take a right-angle course as they enter the inferior vena cava, and a baffle vertically into the atrium creates an obstructive junction at this site that can progressively occlude. We, therefore, incise the right atrial wall between the pulmonary veins and right atrium and suture the opening in the vein and atrium together, enlarging the pulmonary venous entrance point into the inferior vena cava up into the right atrium. In this manner, the veins enter in a wider orifice, which therefore can be baffled to the left atrium without creating a right-angle juncture. In a short follow-up, this approach has resulted in good patency of the pulmonary venous confluence.

Repair of PAPVR remains a surgical challenge because of the potential for pulmonary venous anastomoses in the atrium to develop obstruction. Vigilance in assessing the results of operative intervention and catheter intervention to maintain patency of the anastomoses may be necessary in some forms of anomalous venous return to ensure good long-term outcomes. For PAPVR associated with a sinus venosus ASD, we make incisions in the right atrium without extending the incision across the junction of the superior vena cava and right atrium. In this way, the sinus node artery is generally preserved. In most cases, with a cannula placed very high in the superior vena cava, one can examine the pulmonary venous entrance into the superior vena cava from below, and with gentle retraction on the superior vena cava, the suture line of the pericardial baffle can be accurately placed in the superior vena cava without actually opening the vena cava directly. This technique is useful if there is significant dilation of the superior vena cava-to-right atrial junction due to the anomalous venous return. In these cases, the floor of the superior vena cava can be patched closed, baffling the veins to the left atrium without creation of significant SVC obstruction. However, if there is only modest dilation of the vena cava, then a more elaborate approach may be necessary, including the use of the Warden procedure (dividing the superior vena cava above the entrance of the anomalous pulmonary veins, baffling the orifice of the superior vena cava and right atrium across the atrial septum into the left atrium, and then reconnecting the remaining superior vena cava to the right atrial appendage to reestablish flow to the right atrium). These techniques have resulted in very low complication rates with excellent results. It may be advantageous to leave the azygous vein open if possible, rather than ligation and division of the azygous vein routinely, because if any stenosis of the superior vena cava-to-right atrial anastomosis occurs, some decompression through the azygous system can be permitted with this technique.

TLS

98 Coronary Artery Anomalies in Children

Julie A. Brothers and J. William Gaynor

INTRODUCTION

This chapter focuses on managing coronary artery anomalies in patients without other congenital heart defects. Most coronary artery anomalies in number, origin, and distribution are of intellectual interest only. However, there are a few that are clinically significant and may result in myocardial ischemia, left ventricular dysfunction, and sudden death. This chapter will discuss anomalous origin of a coronary artery from the pulmonary artery, anomalous coronary artery that runs between the aorta and the pulmonary artery, and coronary artery fistula.

Normally, two-coronary arteries originate from separate ostia in the right and left aortic sinuses of Valsalva. The left main coronary artery (LMCA) arises from the left aortic sinus and usually bifurcates into the left anterior descending coronary artery (LAD) and left circumflex coronary artery. The LAD courses in the anterior interventricular groove while the left circumflex coronary artery runs in the left atrioventricular groove. The right coronary artery (RCA) arises anteriorly from the right aortic sinus and courses in the right atrioventricular groove. The RCA usually gives rise to the posterior descending artery at its terminus.

In most people, each coronary artery ostia is centrally located in the appropriate sinus of Valsalva. However, in some individuals the ostium may be located eccentrically close to a valve commissure. One or both coronary ostia may arise above the sinotubular junction, which is usually a benign finding but becomes significant if an aortotomy is necessary for aortic valve replacement or for another indication. If this anatomy is not recognized prior to the operation, the coronary artery could be transected. Another type of anomaly occurs when both coronary arteries arise from the same aortic sinus with either a single ostium or two separate ostia (Table 98.1). If the aberrant vessel courses posterior to the aorta or anterior to the pulmonary artery, this is believed to be benign but it becomes clinically significant if either the aberrant LMCA or RCA courses intramurally between the two great vessels as this has the potential for myocardial ischemia and sudden death.

ANOMALOUS ORIGIN OF A CORONARY ARTERY FROM THE PULMONARY ARTERY

Anomalous origin of a coronary artery from the pulmonary artery is a rare congenital anomaly that is almost always fatal if not diagnosed and treated. While anomalous origin of the LMCA from the pulmonary artery (ALCAPA) is the most common, other coronary arteries may also arise from the pulmonary artery. The RCA may arise from the pulmonary artery and is associated with ischemia and sudden cardiac death but is approximately 10 times less common than ALCAPA. Extremely rare are instances where the LAD, the circumflex, or both the left and right coronary arteries arise from the pulmonary artery; these variants are almost uniformly fatal.

The most important congenital coronary artery anomaly in this class is ALCAPA. It is a rare lesion with an incidence of 1 in 30,000 to 1 in 300,000 people. If left untreated, there is an extremely high mortality rate of 90% by age 1 year. It is the most common cause of myocardial infarction in childhood. ALCAPA is also known as the Bland–White–Garland syndrome after Bland and colleagues reported on both the clinical and autopsy findings in an infant with this anomaly in 1933.

Anatomy

In ALCAPA, the LMCA usually arises from the main pulmonary artery (MPA) but occasionally it will arise from the right pulmonary artery. When it arises from the MPA, it usually originates from the rightward aspect of the posterior (facing) sinus of the MPA (Figs. 98.1 and 98.2). It may also originate from the leftward aspect of the posterior (facing) and rarely from the anterior (nonfacing) sinus of the MPA (Fig. 98.2). An anomalous RCA usually arises from the anterior portion of the pulmonary artery.

ALCAPA usually occurs in isolation but there may be other associated defects, such as patent ductus arteriosus (PDA), ventricular septal defect (VSD), coarctation of the aorta, and tetralogy of Fallot.

Pathophysiology

Symptoms usually begin during infancy after ductal closure and the subsequent fall in the pulmonary vascular resistance. During fetal life, the systemic and pulmonary vascular resistances are similar and myocardial perfusion remains intact since the pulmonary arterial pressure is systemic. After the child is born but before ductal closure, the pulmonary artery pressure remains elevated, and the anomalous coronary artery remains perfused. Because of this, ALCAPA is rarely diagnosed in the first few days of life. After ductal closure, the clinical course is largely determined by the presence or absence of collaterals from the RCA to the left coronary system. If there is inadequate collateral circulation, myocardial ischemia and ventricular dysfunction will result from inadequate myocardial perfusion because the pulmonary artery pressure is now much lower than the systemic pressure and the left ventricle is being perfused with desaturated blood at a low pressure. If the pulmonary artery pressure remains elevated due to the presence of a PDA or VSD, then the left ventricular perfusion pressure may be adequate and ischemia may not occur. If ALCAPA is unknown prior to the closure of these defects, it will become apparent shortly after with the subsequent drop in pulmonary arterial pressure, usually with fatal outcomes.

If there is adequate collateralization, then the perfusion of the left coronary system is maintained. However, as the pulmonary vascular resistance falls, a left-to-right shunt develops from the RCA to the pulmonary artery. There is progressive dilation of the RCA and left coronary artery

Table 98.1 Origin of Both Coronary Arteries from One Sinus

1. Left main coronary artery originates from the right sinus of Valsalva (either from the right coronary artery or separate ostia)
   - Left main coronary artery courses anterior to pulmonary artery
   - Left main coronary artery courses through interventricular septum
   - Left main coronary artery courses between aorta and pulmonary artery
   - Left main coronary artery courses posterior to aorta
   - Rarely, the left anterior descending coronary or the left circumflex coronary artery alone may originate from the right sinus
2. Single left main coronary artery arises from the left sinus and bifurcates into the left anterior descending coronary and the left circumflex coronary arteries. The left circumflex coronary artery crosses the crux and continues as the right coronary artery
3. Single right coronary artery from the right sinus, which crosses the crux, continues as the left anterior descending coronary artery and the left circumflex coronary artery
4. Right coronary artery originates from the left sinus of Valsalva (either from the left main coronary artery or as separate ostium)
   - Right coronary artery courses posterior to aorta
   - Right coronary artery courses anterior to pulmonary artery
   - Right coronary artery courses between aorta and pulmonary artery

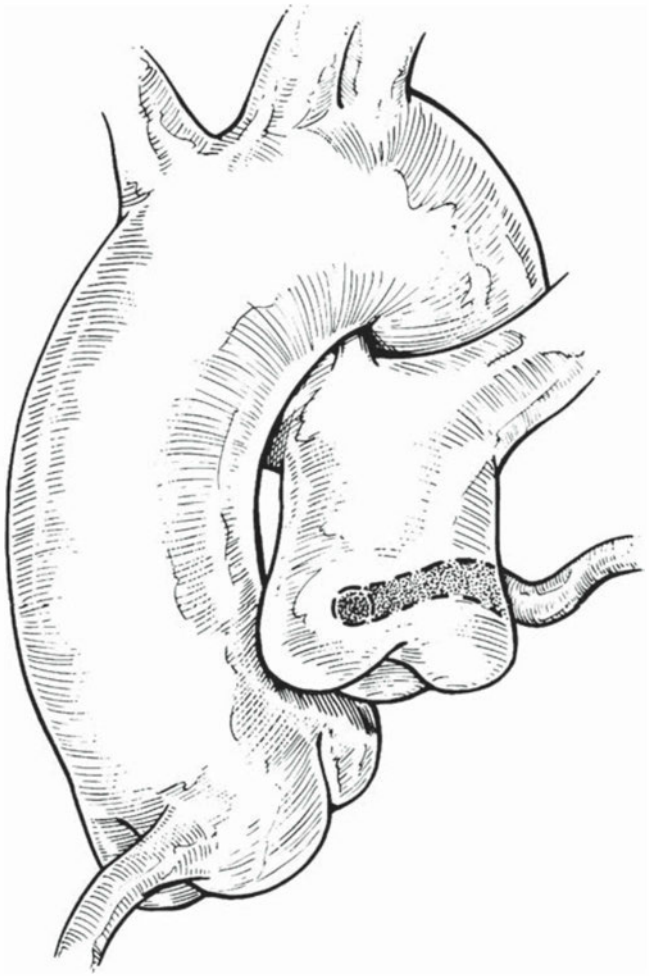


Fig. 98.1. The aorta and pulmonary artery showing anomalous origin of the left main coronary artery from the posterior-facing sinus of the pulmonary artery with the anomalous artery coursing behind the pulmonary artery.

systems, with reversal of flow in the left coronary leading to a pulmonary-coronary steal. The left-to-right shunt is relatively small related to overall cardiac output but is significant considering coronary blood flow. Children with significant collateral circulation may survive past infancy; however, there is usually progressive left ventricular dysfunction. In a small percentage of children, the collateral vessels are enough to maintain adequate myocardial perfusion at rest and sometimes even during exertion allowing these patients to not present until adulthood.

Clinical Presentation

Clinical presentation is usually between 4 and 6 weeks of age after the pulmonary vascular resistance has fallen. However, infants may not present until closer to 2 to 3 months of age when symptoms have increased in severity. Presenting signs and symptoms are those of congestive heart failure, including sweating and discomfort with feeding, tachypnea, poor weight gain, and pallor. The discomfort with feeding likely represents myocardial ischemia. Children who do not present as infants may be diagnosed at several months of age due to a loud murmur of mitral regurgitation (MR) secondary to papillary muscle dysfunction and ventricular dilation. Rarely, older children, adolescents, and adults may remain asymptomatic yet others may come to clinical attention because of exertional chest pain, presyncope, or syncope. There have been reports of sudden death with exercise in these older patients. The symptoms associated with anomalous origin of the RCA from the pulmonary artery are less severe and presentation may be at a later age during childhood but myocardial ischemia and death can still occur.

Physical examination of the infant with ALCAPA may demonstrate signs of congestive heart failure, including tachypnea, tachycardia, and hepatomegaly. Left ventricular dysfunction due to ALCAPA can be difficult to distinguish from dilated cardiomyopathy, for which it is often confused. The left heart is usually enlarged, often with associated MR and a gallop rhythm. If the left ventricular failure has resulted in pulmonary hypertension, then there may also be evidence of right heart enlargement and an accentuated pulmonary component of the second heart sound on examination.

Infants with ALCAPA will typically have an enlarged cardiac silhouette on

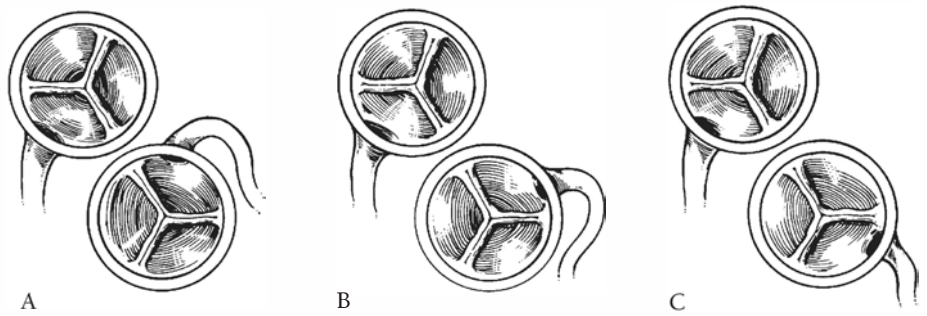


Fig. 98.2. (**A**) Anomalous origin of left main coronary artery from the rightward aspect of the posterior-facing sinus of the pulmonary artery. (**B**) Anomalous origin of the left main coronary artery from the leftward aspect of the posterior-facing sinus. (**C**) Anomalous origin of the left main coronary artery from the nonfacing sinus of the pulmonary artery.

chest X-ray, essentially due to the enlarged left atrium and left ventricle. It will look similar to dilated cardiomyopathy. In infants presenting with congestive heart failure, the electrocardiogram can be a useful diagnostic tool. Classically, these patients have developed lateral or anterolateral wall infarction with Q waves and ST segment elevation in leads I, aVL, and V4 to V6. While this electrocardiographic pattern can be found in other causes of myocardial infarction or cardiomyopathy, if this is seen in an infant in congestive heart failure, the diagnosis of ALCAPA needs to be strongly considered. As well, any infant presenting with dilated cardiomyopathy must be extensively evaluated to rule out ALCAPA. This diagnosis should also be considered in older children and adolescents with dilated cardiomyopathy because occasionally patients survive past infancy.

Diagnostic Imaging

Imaging by echocardiography with Doppler color flow often shows a dilated left ventricle with significant MR. The MR seen with ALCAPA is due to infarction of the mitral valve posterior leaflet and ensuing poor movement of the leaflet; fibrosis and fibroelastosis of the papillary muscle may also be present. Visualization of both coronary arteries is usually possible with echocardiography. An enlarged RCA is almost always present and should increase suspicion of this diagnosis. The origins of both coronary arteries should be identified, including the abnormal origin of the LMCA to the pulmonary artery. If visualization of the anomalous vessel is unclear, color flow Doppler may be useful to demonstrate retrograde flow from the coronary artery to the pulmonary artery. If there is uncertainty about visualization of both coronary ostia, then cardiac catheterization is mandatory to rule out ALCAPA.

Cardiac catheterization with angiography traditionally was utilized in diagnosing ALCAPA but it is now performed usually only if noninvasive imaging cannot adequately establish the diagnosis. When infants present with ALCAPA, elevated filling and pulmonary arterial pressures and a low cardiac output are noted utilizing cardiac catheterization. In older asymptomatic patients, only mildly elevated pulmonary arterial pressures, normal filling pressures, and cardiac output may be seen. A small left–right shunt may be present. A single dilated RCA arising normally from the aorta will be demonstrated using an aortogram. If significant collaterals are present, aortic root angiography will demonstrate the collaterals providing late, retrograde filling of the LCA with a blush of contrast subsequently filling the MPA. A step-up in oxygen saturation may be noted in the MPA if there is a large left–right shunt from the collaterals. If doubt remains regarding the diagnosis, a main pulmonary arteriogram with distal balloon occlusion may be helpful in demonstrating the anomalous LCA.

Magnetic resonance imaging (MRI) has emerged as a useful noninvasive diagnostic tool for defining congenital coronary anomalies. There have been case reports of using this method in diagnosing ALCAPA during infancy but no case series with this anomaly have been reported. However, studies have shown that magnetic resonance angiography has a similar sensitivity and specificity when compared with coronary angiography and may be helpful in delineating the proximal course of anomalous coronary arteries. Computed tomography (CT) scan has been used extensively for coronary artery delineation in adults. While there are many advantages of CT, including rapid acquisition time and high resolution, it is not useful in infants due to the radiation exposure and need for a slower heart rate with ECG gating.

Surgical Management

Indications

Surgical repair is indicated in all patients with ALCAPA. In infants who present with congestive heart failure, surgery should occur within the first few days of diagnosis as risk of continuing myocardial ischemia and death is very high. In an infant who presents with severe heart failure, surgery will likely need to be delayed for at least 24 hours to stabilize the patient using mechanical ventilation, inotropic support, and vasodilators, when necessary. Even in the sickest infants, Del Nido and colleagues reported that a two-vessel surgical repair is possible if left ventricular assist devices (LVAD) are utilized postoperatively. Other critically ill infants may need extracorporeal membrane oxygenation (ECMO). Since these infants are often quite ill, it is important that centers performing ALCAPA surgery have options such as LVAD and ECMO available or else the child should be transferred to a hospital with these capabilities. In older asymptomatic patients, surgery can be performed on an elective basis.

As the goal of surgery is restoration of a two-coronary system, simple ligation of the anomalous coronary should not be performed. Revascularization is possible even in those infants with severe left ventricular dysfunction and mitral insufficiency because significant recovery of function usually occurs. Even if severe MR is present, left ventricular aneurysmectomy and mitral valve repair or replacement are rarely indicated at the time of the initial procedure because the severity of MR almost always improves after revascularization.

Surgical Techniques

History of Surgical Techniques

The first successful operation for the correction of ALCAPA was surgical ligation of the anomalous artery at the pulmonary artery. Ligating the anomalous vessel prevents the left-to-right shunt, thus allowing the left ventricle to be perfused through collaterals from the RCA. However, due to the increased risk of late death after ligation, those children who have had a simple ligation of the anomalous coronary, establishment of a dual coronary artery system

should be considered. Because of the high rate of early mortality and increased risk of late sudden death with this procedure, a variety of techniques were developed to create a dual coronary artery system, including coronary artery bypass grafting (CABG), aortopulmonary window, and direct reimplantation.

CABG was accomplished using the left subclavian artery, the internal mammary artery (IMA), and saphenous vein. The first successful left subclavian artery-to-left coronary bypass was reported by Meyer and colleagues in 1968. Unfortunately, the results of bypass grafting, notably those with saphenous vein grafts, have been disappointing. Takeuchi and colleagues subsequently described the creation of an aortopulmonary window and intrapulmonary artery baffle using a pulmonary artery flap to direct blood flow from the aorta to the anomalous coronary artery. Finally, the procedure of choice in many institutions has become direct reimplantation of the anomalous coronary to the aorta as experience with the arterial switch operation for transposition of the great vessels has increased.

Coronary Artery Bypass Grafting

Left Subclavian-to-Left Coronary Artery Anastomosis. In the current era, CABG is rarely utilized in patients with ALCAPA. It may be used, however, to create a dual coronary artery system after previous ligation or due to stenosis or occlusion after a previous surgical repair. Left subclavian-to-left coronary artery anastomosis may be performed via a median sternotomy using cardiopulmonary bypass or via a left posterolateral thoracotomy without cardiopulmonary bypass. Using cardiopulmonary bypass may be necessary for the stabilization of critically ill infants but subclavian artery mobilization may be difficult through a median sternotomy. When a left thoracotomy is used, heparin is administered followed by mobilization of the subclavian artery. The subclavian artery is then divided distally. After the pericardium is opened, the anomalous coronary is mobilized. A partial occlusion clamp is placed and the anomalous coronary ostium is excised using a small button of pulmonary artery. An end-to-end anastomosis is created between the subclavian and coronary arteries with 7-0 Prolene. Repair of the pulmonary artery may be accomplished either primarily or with a patch of autologous pericardium. A different surgical approach is ligation of the anomalous coronary at the take-off from the pulmonary artery and creation of an end-to-side anastomosis between the subclavian and coronary arteries. The main reason the left subclavian-to-left coronary artery anastomosis is not used frequently is due to the risk of anastomotic stenosis or occlusion.

Left Internal Mammary Artery Grafting. The IMA is the conduit of choice in CABG. Due to the risk of occlusion and poor long-term results, the saphenous vein should not be utilized unless it is the only conduit available. The IMA can be successfully used for bypass grafting even in neonates and infants with some evidence for growth of the IMA after bypass grafting in children.

Direct Reimplantation

In most patients with ALCAPA, direct reimplantation of the anomalous coronary artery onto the aorta can be performed (Fig. 98.3) and is the procedure of choice. When the anomalous coronary ostium is located in the posterior-facing sinus, the procedure is fairly straightforward. Direct implantation is possible even if the ostium is located in the nonfacing sinus by excising a large button of pulmonary artery to extend the coronary artery. However, if the anomalous left coronary originates far leftward in the posterior-facing sinus or on the anterior nonfacing sinus, direct reimplantation may not be possible.

After induction of anesthesia and placement of monitoring lines, a median sternotomy is performed. The thymus is resected, the pericardium is opened and suspended in stay sutures. There is a risk of ventricular fibrillation due to myocardial ischemia and left ventricular dysfunction, so contact with the myocardium should be kept at a minimum until the patient is placed on cardiopulmonary bypass. This operation may be performed using either continuous low-flow bypass with moderate hypothermia (25°C to 28°C) or deep hypothermic circulatory arrest (18°C) in very small infants. Prior to cannulization, an aortic purse-string suture is placed distally near the innominate artery and another is placed in the right atrial appendage for a single venous cannula. Heparin is administered, the aortic and right atrial cannulas are inserted, and cardiopulmonary bypass is established. The left ventricle should be decompressed by the placement of a left ventricular vent via the right superior pulmonary vein. The pulmonary artery and epicardial course of the left coronary artery are visualized.

The aorta and both pulmonary arteries are fully mobilized. The ductus (or ligamentum) arteriosus is ligated to improve the mobility of the pulmonary artery. Tourniquets are placed around both the right and left branch pulmonary arteries



Fig. 98.3. After institution of cardiopulmonary bypass and induction of cardioplegia, the pulmonary artery is transected above the sinotubular junction and the anomalous coronary ostium excised with a generous button of pulmonary artery wall.

to occlude the branch pulmonary arteries and prevent run-off of cardioplegic solution into the lungs. Another way to prevent run-off is compression of the origin of the coronary artery from the pulmonary artery during administration of cardioplegic solution. A cannula is inserted in the ascending aorta for administration of cardioplegia solution. The aorta is then cross-clamped and cold cardioplegia is administered via the aortic root. If circulatory arrest is utilized, the head vessels are then occluded with tourniquets, the circulation is arrested, venous blood is drained into the reservoir, and the cannulae are removed. After adequate arrest, the pulmonary artery is transversely opened just above the sinotubular junction (Fig. 98.3). The anomalous coronary orifice is identified. The pulmonary artery is divided and the coronary ostium is excised from the pulmonary artery using a generous button of arterial wall, similar to the procedure used in the arterial switch operation. The segment of the pulmonary wall that is excised extends the proximal end of the coronary artery, which allows the aortic anastomosis to be accomplished without tension. The aortic commissure may need to be taken down to excise the coronary button if the coronary ostium is located near a commissure. If the coronary artery arises anteriorly from the pulmonary artery or from a branch pulmonary artery, the coronary artery can be extended using a tube constructed from pulmonary artery wall to allow reimplantation (Fig. 98.4). Using cautery, the proximal portion of the coronary artery is mobilized cautiously to avoid any small branches. Similar to the arterial switch operation, the aorta is then opened transversely just above the sinotubular junction and the incision is carried posteriorly above the left posterior sinus (Fig. 98.5). The sinus is then incised vertically to accept the coronary button. The coronary button is carefully aligned with the aortic incision to avoid twisting or kinking. Using a continuous suture of 7-0 polypropylene (Prolene), the anastomosis is started at the most inferior aspect of the coronary button, which is attached to the most inferior aspect of the incision in the sinus. The suture line is carried to the top of the incision anteriorly and posteriorly. The aorta is closed using a continuous suture of 7-0 Prolene, which is tied to the coronary button suture as the anastomosis is completed (Fig. 98.5). After the aorta has been closed, cardioplegia solution is administered and the anastomotic site is inspected for adequate filling of the coronary and hemostasis.

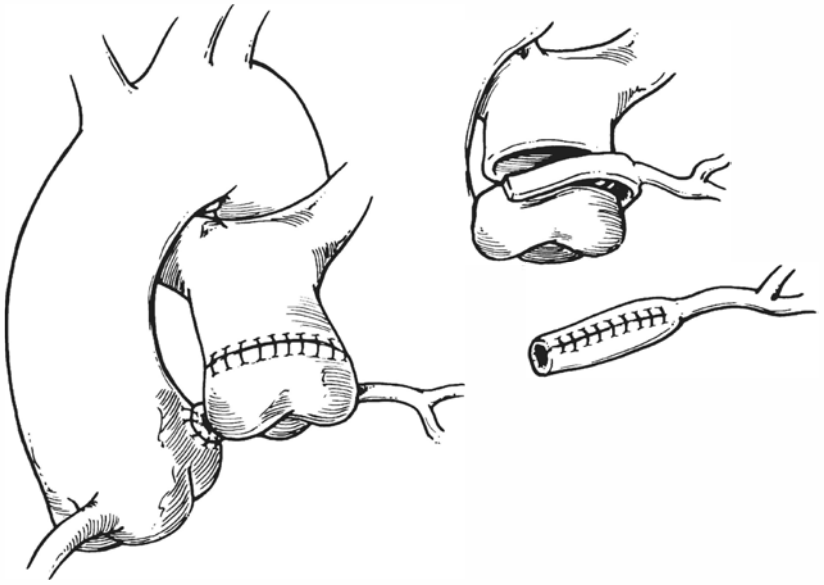


Fig. 98.4. Occasionally, when the coronary artery arises from the leftward or anterior aspect of the pulmonary artery, direct reimplantation may not be possible. In these situations, a tube can be constructed from a segment of pulmonary artery to lengthen the coronary and to allow reimplantation on the aorta.

In most cases, the pulmonary artery can be repaired primarily with a continuous suture of 7-0 Prolene (Fig. 98.6). The ductus (or ligamentum) should be divided to improve mobility of the pulmonary artery confluence and to allow reconstruction without tension. If there is tension or narrowing, the pulmonary artery should be repaired with a patch of autologous pericardium (Fig. 98.6). If a commissure was taken down during excision of the coronary button, the pulmonary artery should be reconstructed with pericardium and the commissure resuspended.

The patient is rewarmed and the aortic cross-clamp is removed. To minimize

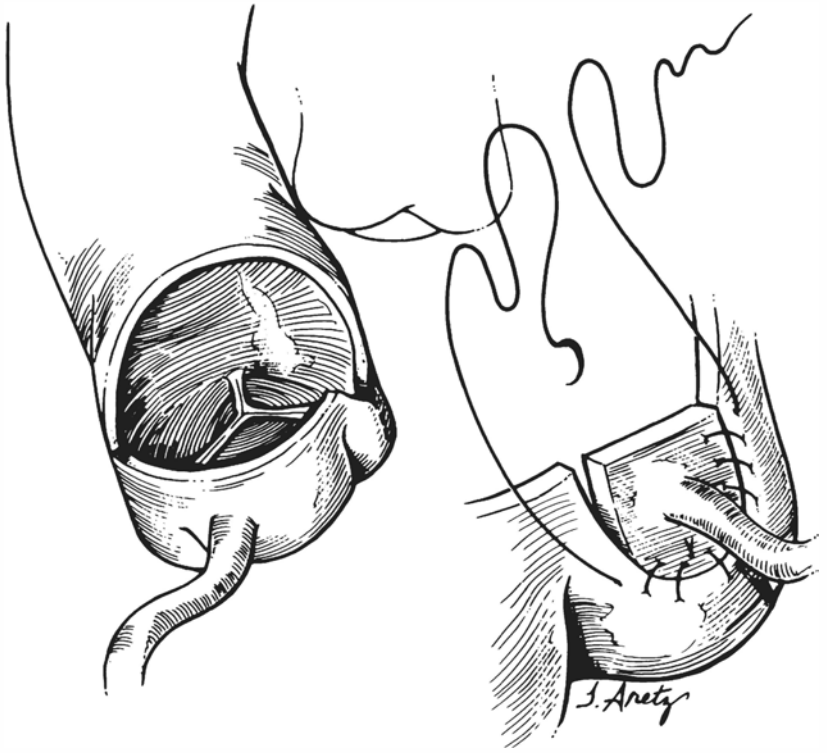


Fig. 98.5. After the anomalous coronary artery is mobilized, the aorta is opened transversely above the sinotubular junction and a vertical incision is made in the left posterior sinus to accept the reimplanted coronary.

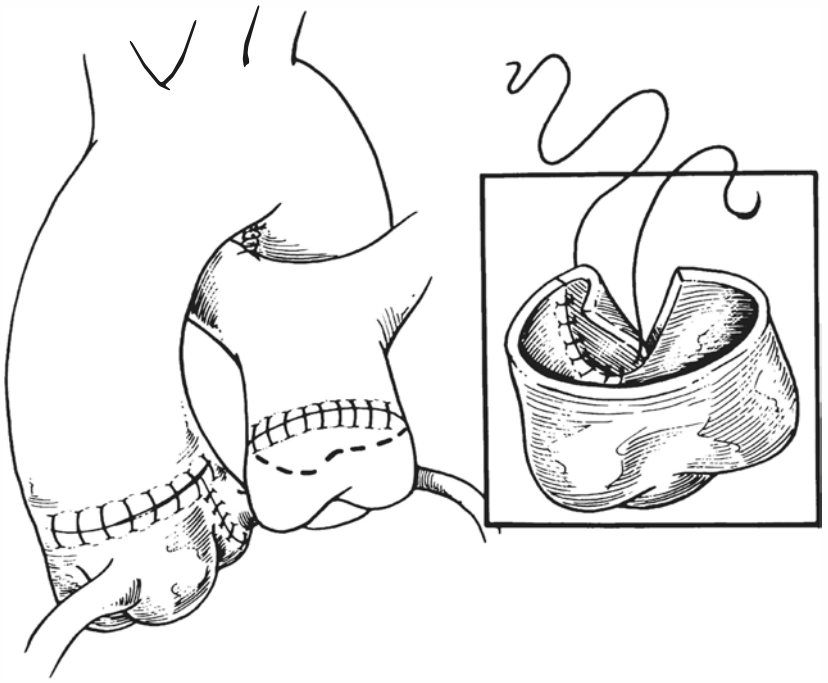


Fig. 98.6. After the coronary is reimplanted, the aorta is closed primarily. The pulmonary artery may frequently be closed primarily. Ligation and division of the ligamentum arteriosus improves the mobility of the pulmonary artery. Occasionally, patch repair of the defect in the pulmonary artery with autologous pericardium may be necessary (*inset*).

The procedure may be performed with either continuous low-flow cardiopulmonary bypass (25°C to 28°C) or deep hypothermic circulatory arrest (18°C). Cannulation is performed as for direct reimplantation. After induction of cardioplegia, a longitudinal incision is made in the anterior portion of the pulmonary artery (Fig. 98.7) and the ostium of the anomalous coronary is identified. Using a punch, a 5 mm diameter opening is made on the leftward aspect of the aorta above the sinotubular junction (Fig. 98.8). If there is any question regarding the placement of the aortic opening, then an anterior aortotomy should be performed, and the incision directly visualized to avoid damage to the aortic valve. Creating the aortopulmonary window above the sinotubular junction allows a downward angle of the baffle into the sinus if the ostium is located deep within a sinus. After a similar incision is made in the pulmonary artery directly opposite the opening in the aorta, these are anastomosed using a continuous suture of 7-0 Prolene, thereby creating an aortopulmonary window (Fig. 98.8).

ischemia time, the cross-clamp may be removed prior to the pulmonary artery reconstruction. The left ventricle is inspected for adequate perfusion and function and the suture lines are inspected for hemostasis. Right and left atrial lines are placed to adequately monitor pressure and for drug administration. Atrial and ventricular pacing wires are also placed. After complete warming, the patient is separated from cardiopulmonary bypass. Careful attention to the electrocardiogram during reperfusion and after separation from bypass is necessary to evaluate for ischemia. Inotropic support may be temporarily needed due to preoperative left ventricular dysfunction.

Modified Takeuchi Operation

The Takeuchi operation, or intrapulmonary artery tunnel, is an alternative surgical repair strategy for ALCAPA. Takeuchi and colleagues originally described the creation of an aortopulmonary window using a portion of anterior pulmonary artery wall to form a baffle directing blood from the aorta to the anomalous coronary artery ostium. In the modified repair, the baffle is constructed using a polytetrafluoroethylene (PTFE, Gore-Tex) patch. If the ostium is located near a commissure or arises from a branch pulmonary artery, then creating a baffle may not be possible.

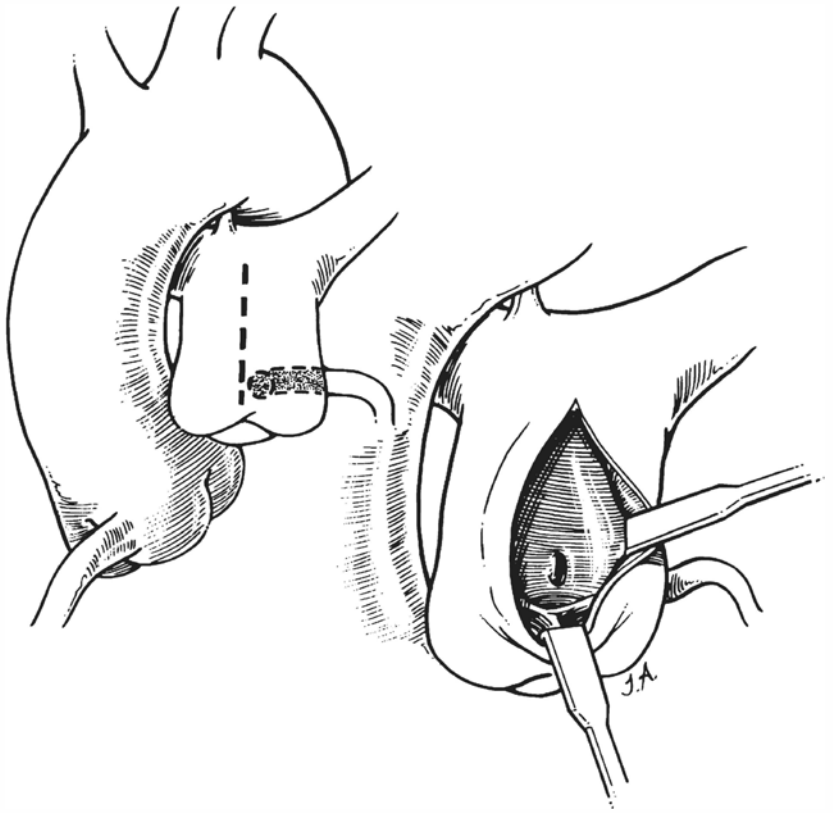


Fig. 98.7. After institution of cardiopulmonary bypass and induction of cardioplegia, a longitudinal incision is made in the main pulmonary artery and the ostium of the abnormal coronary is identified.

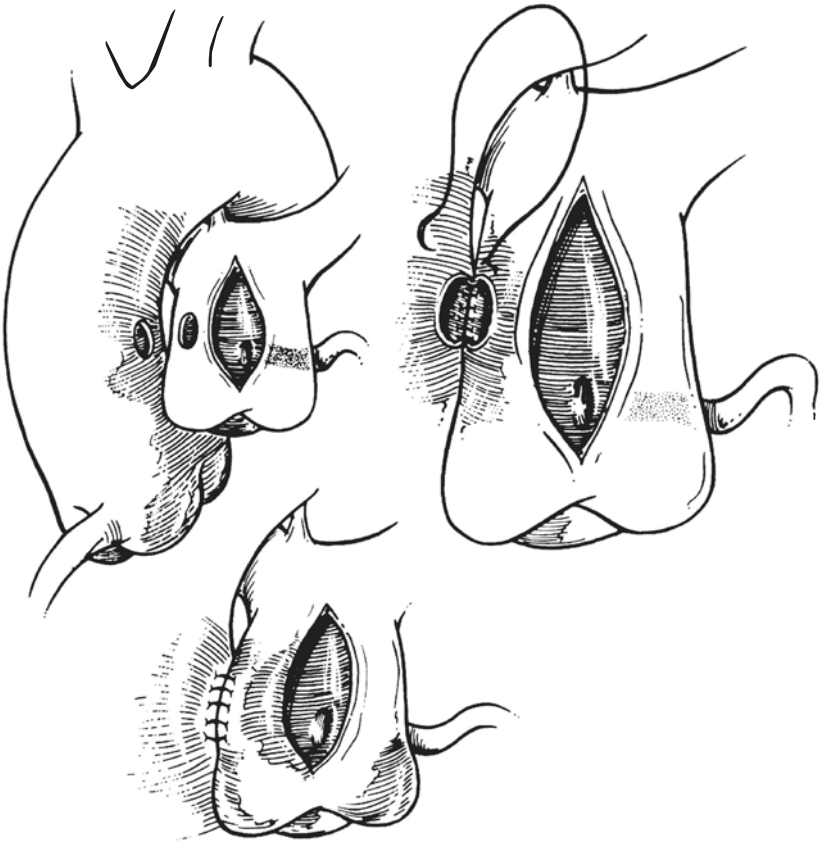


Fig. 98.8. Using a punch, a 5 mm opening is made in the aorta on the leftward aspect above the sinotubular junction. A similar opening is made in the pulmonary artery at the same level, and these are anastomosed to create an aortopulmonary window.

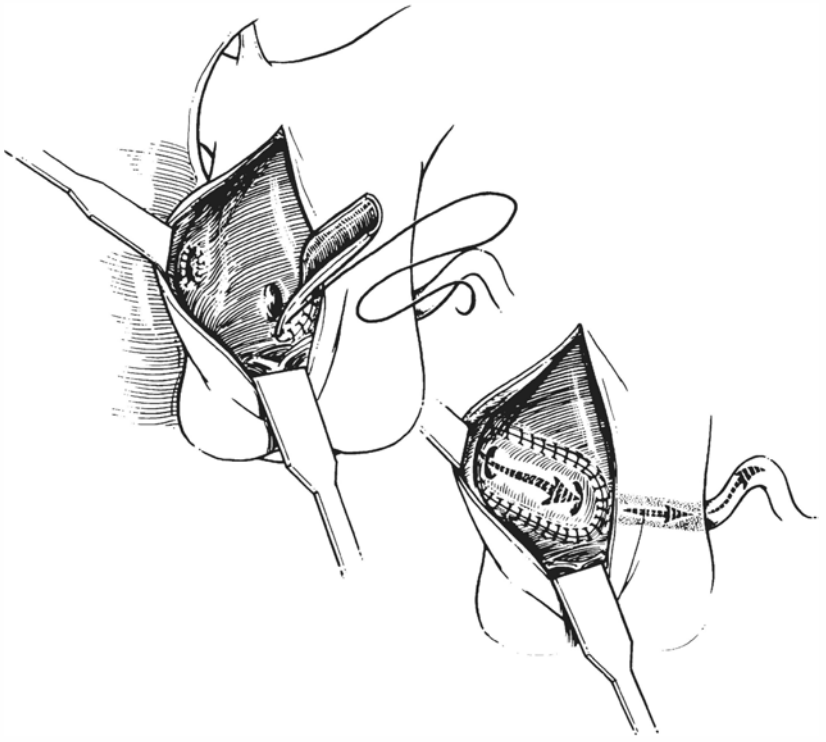


Fig. 98.9. A segment of 4 mm polytetrafluoroethylene (Gore-Tex) graft is opened longitudinally and used to fashion a baffle that directs blood flow from the aortopulmonary window to the anomalous coronary ostium.

A 4 mm PTFE tube graft is split longitudinally and customized to an appropriate length (Fig. 98.9). This graft acts as an intrapulmonary artery tunnel, baffling blood from the aortopulmonary window to the anomalous coronary ostium. The suture line starts at the anomalous coronary and is continued inferiorly along the pulmonary artery wall to the aortopulmonary window. The suture line is completed by returning to the coronary artery and finishing the superior aspect of the baffle. After the baffle is created, the pulmonary artery can be repaired using a prosthetic patch or autologous pericardium to avoid supravalvar right ventricular outflow tract obstruction (Fig. 98.10).

The main complications of the modified Takeuchi operation include baffle leak, baffle occlusion, and supravalvar right ventricular outflow tract obstruction.

Postoperative Management

Regardless of surgical technique, the most common postoperative issues are generally related to the infant's preoperative state: low cardiac output, left ventricular dysfunction, and hypotension. Weaning off of cardiopulmonary bypass can be difficult due to these issues. Optimizing the patient's hemoglobin, electrolytes, acid–base status, fluid status, and providing adequate inotropic support are very important. In infants and children with severe preoperative cardiac dysfunction, temporary support with an LVAD or ECMO may be necessary postoperatively. The second most common issue postoperatively is bleeding and is more commonly seen in small infants and those who require mechanical support. In these cases, aprotinin has been used intraoperatively to assist with coagulation and is often continued as an infusion for up to 12 hours postoperatively. Platelets and fresh-frozen plasma should be used aggressively to replace ongoing loss. Patients with low cardiac output and/or bleeding issues may be candidates for delayed sternal closure for 2 to 3 days.

Results

Simple ligation of ALCAPA has been shown to have unacceptable early and late mortality rates. In general, survival after establishment of a dual coronary system is excellent. Bunton and colleagues in 1987 reported on 24 patients with ALCAPA. There were 11 cases of coronary ligation or ostial closure and 11 cases that underwent a Takeuchi repair; 2 cases had other procedures. There was a 27% early mortality and a 25% late mortality over an average of 10.5-year

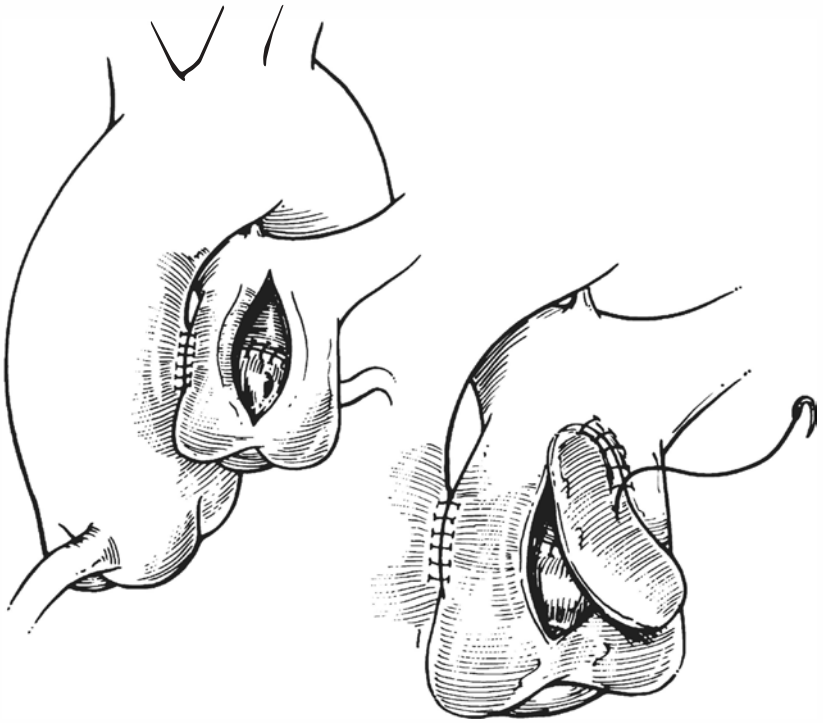


Fig. 98.10. After the baffle is completed, the pulmonary arteriotomy is repaired with a patch to avoid creation of supravalvar right ventricular outflow tract obstruction.

follow-up period in those who underwent coronary ligation or ostial closure. In those who underwent a Takeuchi procedure, there were no early or late deaths in over an 18.5-month follow-up period; however, two of these patients developed right ventricular outflow tract obstruction (one required a second operation) and one patient was found to have baffle occlusion. Backer and colleagues reported the follow-up surgical results of 20 patients who underwent different procedures. Of the nine patients who underwent ligation, there were two early deaths and one late death. In the 10 patients who underwent creation of a dual coronary artery system or the one patient who had cardiac transplantation, there were no deaths reported. Five patients underwent left subclavian-to-left coronary anastomosis and two developed significant anastomotic stenosis. Vouhe and associates from France reported on reimplantation of the anomalous coronary in 31 consecutive children. There were three hospital deaths and two additional deaths within the first 3 months. There were no late deaths. The one risk factor identified for early mortality was a shortening fraction of $<20\%$. Twenty-three survivors were studied more than 1 year after repair. All patients had normal left ventricular function, and in five of seven patients who had severe MR preoperatively, the severity had decreased to mild or none. The reimplanted coronary artery was patent in all patients. A report by Lange and colleagues in 2007 reviewed the long-term results of 56 patients with ALCAPA who underwent either subclavian artery anastomosis or coronary artery transfer. Patients who had a Takeuchi procedure or a left mammary artery graft were excluded. There were similar early mortality rates of 14% for both surgical groups but no mortality in those undergoing repair in the more recent era. Late mortality in each group was also similar with one patient in each group. At final follow-up (mean 14.5 years for subclavian artery group and 8.7 years for coronary transfer group), 95% of patients had normal left ventricular function and 84% had MR less than grade 2. In 2011, Imamura and colleagues evaluated long-term outcomes in children who required ECMO after ALCAPA repair compared with those who did not. They reported on 26 consecutive patients who underwent surgical repair with 21 patients in the not requiring ECMO and 5 patients needing ECMO postoperatively. Twenty-three patients underwent coronary reimplantation, two patients had coronary artery ligation, and one had a left subclavian artery-to-left coronary artery bypass graft. There was no early or late mortality in either study group. One patient in each group required cardiac transplantation. In the ECMO group, all five patients needed reoperation: one underwent heart transplantation, three required mitral valve replacement (one underwent this procedure twice) for severe MR, and one underwent mitral valvuloplasty initially and then required atrial septal defect closure and pulmonary stenosis repair. At a mean follow-up of 7.8 ± 5.9 years, nearly all patients had normal left ventricular function and no more than mild valve regurgitation, excluding those with mitral valve replacement.

Special Considerations

Mitral Regurgitation

The ideal approach for managing MR during initial repair remains controversial. In general, MR is present at the initial presentation, often to at least a moderate degree. Even in patients with severe MR, in the majority MR improves significantly after coronary reperfusion alone. This is likely due to improvement in left ventricular dysfunction and decrease in left ventricular size postoperatively. Because of this, most centers have avoided mitral valve repair at the time of ALCAPA repair. Further, valve repair at the time of initial surgery can result in longer ischemic times and can be technically more difficult to perform in an infant heart compared with a later time when the child is bigger. However, some authors do recommend routine annuloplasty at the time of initial repair, whereas others propose annuloplasty or mitral valve replacement only with severe MR. In summary, we believe that early surgical repair of the mitral valve is generally not necessary. If valve repair in the setting of severe MR is needed later, it is likely to be more successful and easier to do technically. However, if severe MR persists late postoperatively, causes of ongoing myocardial ischemia should be investigated.

Late Presentation in the Adult

As discussed previously, it is rare for adults to present with ALCAPA. Surgery is indicated in all cases but generally can be done electively. The surgical choice is coronary bypass grafting using the left internal thoracic artery; this is generally a low-risk procedure that all adult cardiothoracic surgeons can perform with a low mortality rate.

ANOMALOUS CORONARY ARTERY COURSE BETWEEN AORTA AND PULMONARY ARTERY

Anatomy

A coronary artery that courses intramurally in the wall of the aorta between the aorta and pulmonary artery can result in myocardial ischemia and sudden death, notably in

children and young adults. When there are two separate ostia, either the RCA or LMCA may arise from the inappropriate sinus of Valsalva and subsequently course between the great vessels (Fig. 98.11). When the two ostia are in the same sinus, the ostium of the anomalous coronary artery is frequently small and elliptical (or "slit-like") with an acute-angle take-off from the aorta. A similar situation may be found if there is a single coronary artery arising from the right aortic sinus and the LMCA or LAD runs between the aorta and pulmonary artery or if the single coronary arises from the left aortic sinus and the RCA courses between the great vessels. Alternatively, an LMCA that arises from the right aortic sinus and courses through the infundibular septum does not appear to carry the same increased risk of sudden death.

Pathophysiology

When the anomalous coronary courses intramurally between the aorta and pulmonary artery, it is associated with increased incidence of sudden death. The greatest danger of sudden death is during or just after maximal exertion. Both anomalous LMCA from the right sinus and RCA from the left sinus are associated with sudden death but the former carries a higher risk. Based on autopsy studies of anomalous aortic origin of a coronary artery (AAOCA) patients, sudden cardiac death is hypothesized to occur from decreased anomalous coronary blood flow resulting in myocardial ischemia and/or ventricular tachyarrhythmias. This diminished blood flow is likely due to an anatomical malformation of the anomalous vessel, which may include one or more of the following: acute-angle take-off from the aorta creating a "slit-like" orifice that easily collapses, presence of an ostial ridge, and/or proximal intramural course that gets compressed within the aortic wall between the great arteries. Because of these malformations, the risk of ischemia increases with vigorous exercise when there is a significantly greater cardiac output and oxygen demand placed on the heart. It is unlikely that ischemia occurs every time the patient exercises; rather, it appears due to the collective effect of several smaller ischemic events over time that may lead to an unstable myocardium more prone to lethal tachyarrhythmias.

Clinical Presentation

The true prevalence of an anomalous left or right coronary artery between the aorta and pulmonary artery is unknown but estimates range from 0.1% to 0.3% of the general population. Making this diagnosis challenging is that there are no characteristic physical findings in patients with this coronary anomaly. The physical examination is almost always normal. There may be an innocent murmur in children. This is often what prompts the referral to a cardiologist where an echocardiogram is obtained, leading to the diagnosis. Many patients with this anomaly are asymptomatic and may remain so throughout their life while, in others, the initial presentation may be sudden death. When symptoms are present, they most commonly include chest pain, palpitations, dizziness, presyncope, or syncope during or just after with exertion. Because of this, the diagnosis must be considered in any patient with exercise-induced complaints suggestive of myocardial ischemia or in those presenting with aborted sudden death or sudden death.

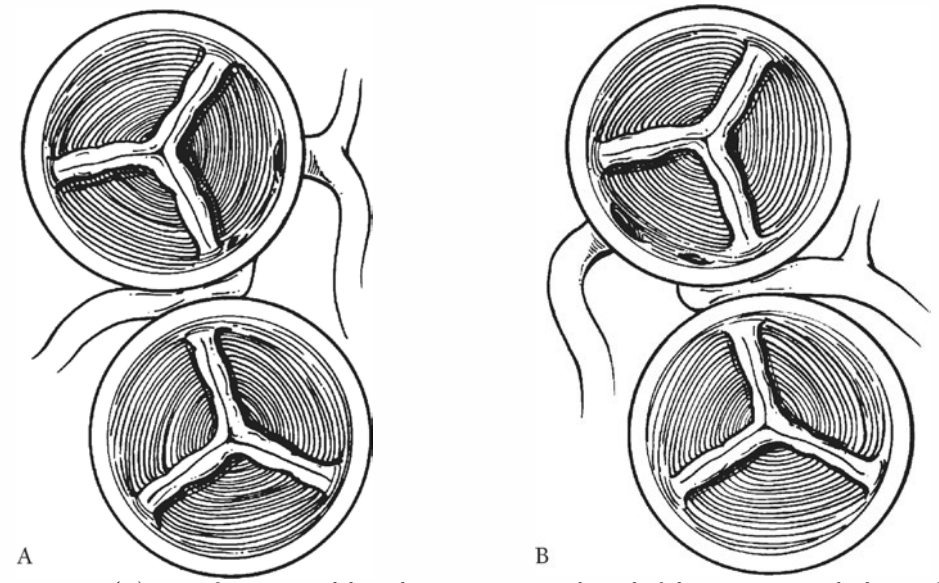


Fig. 98.11. (**A**) Anomalous origin of the right coronary artery from the left aortic sinus with abnormal coursing of the right coronary artery between the aorta and pulmonary artery. (**B**) Abnormal origin of the left main coronary artery from the right aortic sinus with abnormal coursing of the left main coronary artery between the aorta and pulmonary artery.

Diagnostic Imaging

Anyone presenting with presyncope, syncope, or chest pain during or just after exertion should undergo further evaluation. A resting electrocardiogram should be obtained to evaluate for ventricular hypertrophy, arrhythmias, and evidence of previous myocardial infarction.

Echocardiography with color Doppler should be performed to confirm normal intracardiac anatomy and to evaluate heart function, especially focusing on areas of abnormal wall motion signifying possible history of ischemia. Close attention should be paid to the proximal coronary anatomy and the coronary artery origins. Identification of both coronary artery origins is often possible in many patients by two-dimensional echocardiography, especially when combined with color Doppler flow mapping to identify the anomalous origin. In patients in whom the anomalous vessel courses intramurally within the aortic wall, the anomalous vessel may appear to arise normally where it exits the aorta using traditional two-dimensional imaging alone; color Doppler imaging can be very useful in helping show the direction of blood flow within the aortic wall, thereby delineating an anomalous vessel from one that arises normally.

When the coronary artery origins cannot be adequately delineated or to confirm the diagnosis, then other noninvasive techniques, such as MRI or CT scan, are frequently used. Occasionally, transesophageal echocardiography will be utilized to visualize the coronary anatomy; however, this is a more invasive options and is usually not the first choice in children. While cardiac catheterization with coronary angiography still remains the gold standard for detecting anomalous coronary arteries, it is being replaced by noninvasive imaging as described above. Coronary angiography may be necessary in adult patients to first evaluate for other coronary disease prior to undergoing surgical intervention.

After diagnosis, most patients will undergo further evaluation for myocardial

ischemia at rest and stress. This usually includes an exercise stress test often accompanied by some form of imaging, such as nuclear perfusion imaging and/or stress echocardiography. However, because ischemia is intermittent in patients with AAOCA, using a single exercise test on which to base management decisions may not be reliable. In a study performed at our institution evaluating children with anomalous coronary artery, 9 of 16 patients who had a preoperative exercise test had presented with cardiovascular symptoms but only 1 had an abnormal exercise test. Further, there have been reports of patients with normal exercise tests who have subsequently experienced sudden death.

Surgical Management

Indications for Surgery

Surgical intervention is indicated in any patient with this diagnosis who has signs and/or symptoms of myocardial ischemia or ventricular arrhythmias. Further, most would agree that surgery is indicated in asymptomatic patients with anomalous left coronary artery from the opposite sinus of Valsalva because of the high risk of sudden death. The management of asymptomatic patients with anomalous RCA has not been defined; this controversy is even greater in the management of children and young adults who appear to have a higher risk of sudden death than those identified later in adulthood.

Unroofing Procedure

This has become the procedure of choice for patients with anomalous coronary artery with anomalous coronary artery from the wrong sinus of Valsalva with an interarterial and intramural course. After the median sternotomy is performed, the pericardium is opened, and the anatomy is inspected. The aorta is cannulated close to the innominate artery and a two-stage venous cannula is inserted via the right atrial appendage. After cardiopulmonary bypass with moderate hypothermia is established, a left ventricular vent is placed via the right superior pulmonary vein. The ascending aorta is cannulated for the administration of cardioplegic solution. The aorta is cross-clamped and cardioplegic solution is administered. After adequate arrest, a transverse aortotomy is performed and the coronary ostia are identified. If the anomalous coronary ostium arises close to the aortic valve commissure, it may be necessary to take down the commissure (Fig. 98.12). The slit-like ostium is opened longitudinally starting at the anomalous coronary ostium and continued into the correct sinus. A segment of the common wall between the aorta and coronary is excised and the intimal surfaces are approximated with interrupted sutures using 8-0 Prolene. If the aortic commissure was taken down, it is resuspended with a pledgeted suture (Fig. 98.13). The aortotomy is then repaired and the cross-clamp removed after deairing. As the patient is rewarmed, evidence of myocardial ischemia should be evaluated using the electrocardiogram. The patient is then separated from cardiopulmonary bypass in the usual manner.

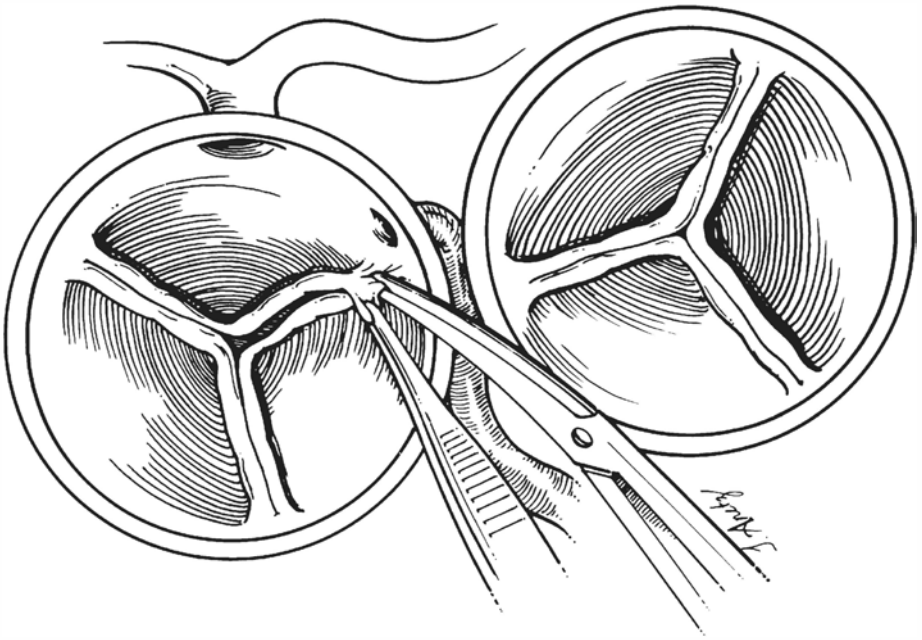


Fig. 98.12. When the coronary arises anomalously from the opposite sinus of Valsalva and courses between the pulmonary arteries, the ostium is frequently abnormal. Repair may be undertaken by remodeling of the ostium. The abnormal ostium is usually located near a commissure, and this commissure must be initially taken down to allow remodeling of the ostium.

Creation of a Neo-Ostium

An alternate technique for when the anomalous coronary passes intramurally below the aortic valve commissure is to create a neo-ostium in the correct sinus (Fig. 98.14). A probe is passed through the intramural segment of the anomalous coronary into the correct sinus. The coronary is opened at the location where it exits the aorta and a neo-ostium is created. The intima is sewn to the aortic wall with interrupted sutures. This technique avoids the takedown and resuspension of the commissure.

Other Techniques

When there are two separate coronary ostia with an interarterial but not an intramural course, coronary artery translocation with reimplantation has been utilized. This is similar to reimplantation done for the arterial switch procedure and was described above for the ALCAPA repair. The coronary artery is excised with a button of aortic tissue and usually reimplanted above the commissure in the correct sinus. Another technique for the potential prevention of compression of the coronary artery between the aorta and pulmonary artery was translocating the MPA to the left pulmonary artery or translocating the right pulmonary artery and bifurcation anterior to the aorta, thereby leaving the coronary circulation intact. This technique may be utilized when the anomalous coronary course is not intramural but the ostia are close together. Combining coronary artery patch angioplasty using pericardium in addition to translocating the MPA to the left pulmonary artery has also recently been proposed. CABG utilizing saphenous vein or IMA grafts may be appropriate in the older adult; however, concern about long-term graft patency in the child, adolescent, or young adult makes this a less desirable surgical option.

Surgical Results

No data exist regarding long-term neo-ostia patency rates in the unroofing procedure. However, short- to mid-term results have overall been reassuring with no reports of ostial stenosis by echocardiography. One study by Romp and colleagues described a patient who developed severe aortic

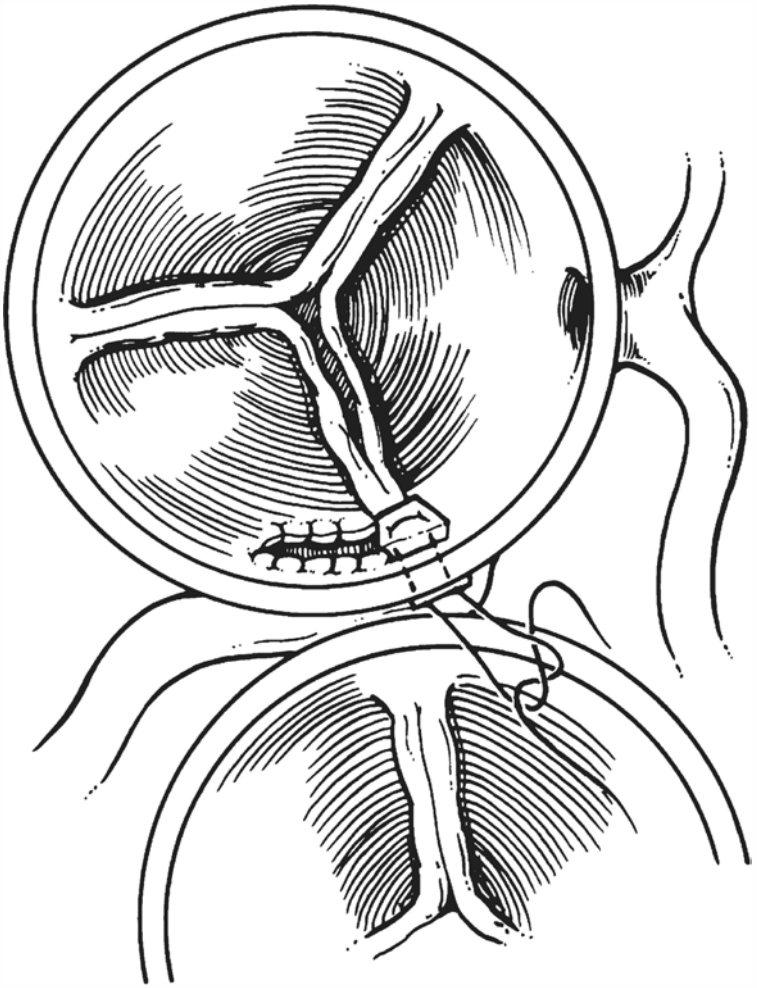


Fig. 98.13. The walls of the coronary artery and aorta are anastomosed with interrupted sutures of 8-0 polypropylene (Prolene), enlarging the ostium and preventing compression by the great vessels. The commissure of the aortic valve is resuspended with a pledgeted suture.

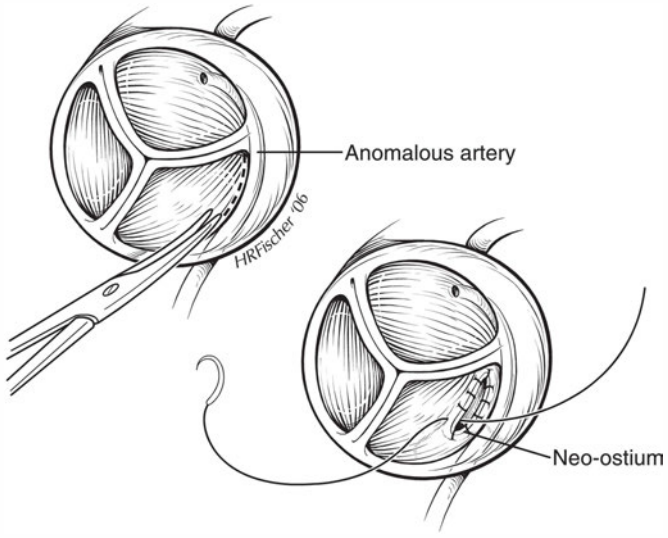


Fig. 98.14. A neo-ostium is created by opening the anomalous artery in the correct sinus as it exits the aortic wall. The intima is sewn to the aortic wall with interrupted sutures.

insufficiency after commissural takedown and resuspension, ultimately requiring aortic valve replacement 44 months after the initial operation. In a study by Brothers and colleagues at our institution that evaluated children in the short- to mid-term after the unroofing procedure, subclinical evidence of postoperative myocardial ischemia was found in one-half of the patients with anomalous RCA and one of eight patients with anomalous left coronary artery. This was despite patent neocoronary ostia by echocardiography and with the patients remaining asymptomatic during testing. The clinical implications of these findings in the long-term are unknown, and further evaluations are needed over the next several years. In a recent study, Mainwaring and colleagues from Stanford University reported on mid-term surgical results on a series of 50 patients who underwent repair of anomalous coronary artery. The different repairs included unroofing, unroofing plus coronary bypass, coronary artery reimplantation (those without intramural component), and pulmonary artery translocation (those with a single coronary ostium without an intramural component). There was no operative or late mortality at an average of 5.5 years of follow-up. All patients who were symptomatic prior to surgery were now symptom-free and most had a normal, unrestricted activity level.

While coronary artery reimplantation has been utilized by some, one study by Rinaldi and colleagues reported that two patients required emergency bypass grafting. In an analogous procedure utilized for transposition of the great arteries, long-term follow-up studies have demonstrated coronary obstruction in up to 8% of patients with a recent report by Pedra and associates noting proximal eccentric intimal thickening by intravascular ultrasound in a majority of patients evaluated at least 5 years after the original surgery.

CORONARY ARTERY FISTULA

A coronary artery fistula is defined as an abnormal communication between a coronary artery and a cardiac chamber or any of the great vessels (e.g., coronary sinus, vena cavae, pulmonary artery, or a pulmonary vein). Coronary fistulas that end in right-sided structures, such as the right atrium, right ventricle, or pulmonary artery, are described as coronary arteriovenous fistulas while those that terminate in left-sided structures are called arterio-arterial fistulas.

Coronary artery fistulas are usually congenital. They make up 0.2% to 0.4% of congenital heart defects but account for

nearly half of all congenital coronary anomalies. They can be in isolation or together with other congenital heart disease. Acquired fistulas are the result of cardiac surgery, cardiac catheterization (including coronary angioplasty, other transcatheter procedure, endomyocardial biopsy), or from a complication due to Kawasaki disease.

Anatomy

Coronary artery fistulas may arise from either the right or the left coronary artery with occasionally both coronary arteries affected. Most arise from a coronary artery with an otherwise normal distribution. The fistulous connection may arise in the midportion of the coronary artery with a normal vessel continuing beyond the fistula or as an end artery at the termination of the vessel. In the portion of the vessel proximal to the fistula, the coronary is dilated and elongated, usually in proportion to the size of the shunt across the fistulous connection. The portion of the vessel distal to the fistula most often returns to a small diameter.

The most common sites of termination are the right ventricle and the right atrium. Fernandes and colleagues described 93 patients with coronary artery fistulas. A single fistula was present in 83 patients with the other 10 having multiple fistulas. The most common site of origin was the RCA with the right ventricle being the most common termination site. Fifty-six patients had isolated coronary fistulas and the others had additional cardiac lesions. In a review of 286 patients, Lowe and colleagues found the RCA to be the site of origin in just over half of the patients and the left coronary artery system in approximately one-third. The right ventricle was the most common site of drainage (39%), with the right atrium (including the coronary sinus and superior vena cava) and the pulmonary artery as the drainage sites in 33% and 20%, respectively. In the remaining 8%, the site of drainage was the left atrium or left ventricle.

Pathophysiology

Coronary fistulas result in a left-to-right or left-to-left shunt. When they drain to the right side of the circulation, there is usually a small-to-moderate sized shunt. However, when it drains into a left-sided chamber (i.e., left-to-left shunt), an aortic run-off develops with similar physiology as aortic regurgitation. The effects of the shunt are related to the amount of blood flow from the fistula to the chamber and into which chamber the fistula drains. It can also be affected by myocardial ischemia resulting from "steal" from the coronary circulation.

Clinical Presentation

In general, patients with coronary artery fistula are asymptomatic and are therefore rarely diagnosed in infancy or childhood. Many patients are diagnosed serendipitously during a murmur evaluation. When symptomatic, the most common complaints are shortness of breath with exercise and fatigue. Angina is uncommon, even though there may be coronary steal. Ventricular dysfunction and congestive heart failure may occasionally be present and is much more common in the adult. In an older patient, atrial fibrillation may be caused by a coronary artery fistula to the right atrium, leading to right atrial enlargement.

On examination, a continuous murmur is often auscultated. The murmur may be confused with that of a PDA but is heard lower on the sternal border than would be typical for a PDA. Cardiomegaly may be present as well. Similar to aortic regurgitation, a wide pulse pressure may be appreciated in large left–left shunts.

Natural History

In congenital coronary arterial fistulas, the fistula is likely present early in life and gradually increases in size over several years. Despite progressive enlargement of the coronary arteries, spontaneous rupture is rare and is usually due to aneurysmal dilatation and weakening of the vessel wall due to a congenital defect or atherosclerosis. Bacterial endocarditis may occur secondary to turbulent flow. The spontaneous closure of small fistulas has been reported.

Diagnostic Imaging

A normal electrocardiogram is found in about one-half of patients or may show evidence of ventricular volume overload. Atrial fibrillation may be noted in older adult patients who have right atrial fistulous connections. If coronary steal is present, evidence of myocardial ischemia in the affected region may be noted.

Chest X-ray films are usually normal but may show cardiomegaly or evidence of congestive heart failure. Giant aneurysms of the involved coronary artery can sometimes be noted.

Two-dimensional echocardiography is useful to demonstrate the enlarged coronary artery, the origin of the fistula, the chamber into which it drains, and any cardiac chamber enlargement and/or hypertrophy. The actual fistula may be demonstrated best by color Doppler.

MRI is an additional noninvasive imaging technique to diagnose and provide detailed anatomy of the coronary arterial fistula; in time, it may take the place of cardiac catheterization, but currently it is still used as an adjunctive imaging modality.

Coronary catheterization with selective coronary angiography remains the gold standard to define the coronary anatomy and the hemodynamic significance of the fistula. These results help in planning the surgical repair. Often, an experienced interventional cardiologist can successfully coil embolize the coronary artery fistula without the morbidities associated with cardiopulmonary bypass and sternotomy. However, the indications for coil embolization have not been well-defined so most patients undergo surgical closure as the preferred therapy.

Surgical Management

Indications for Surgery

All patients with symptomatic fistulas should undergo closure. Patients with very small fistulas may not require surgical closure; however, because the natural progression of the fistula is to enlarge, these patients should be closely followed. Asymptomatic patients with moderate-to-large fistulas should undergo elective surgical closure.

Surgical Techniques

Coronary artery anatomy must be clearly defined by coronary angiography before surgical closure. Each operation should be individualized based on the anatomy. Often, the fistula can be ligated or oversewn at its origin or termination without the use of cardiopulmonary bypass; however, cardiopulmonary bypass should always be available.

Through a median sternotomy, the coronary anatomy is visualized and carefully inspected, noting the site of the enlarged vessel. The fistula may be ligated without the use of cardiopulmonary bypass when it is located at the distal end of the coronary artery and there is no viable myocardium distal to the fistula (Fig. 98.15). This is performed by placing a ligature around the coronary artery immediately proximal to the fistula, thus temporarily occluding the fistula. The heart is observed for signs of ischemia and the electrocardiogram is

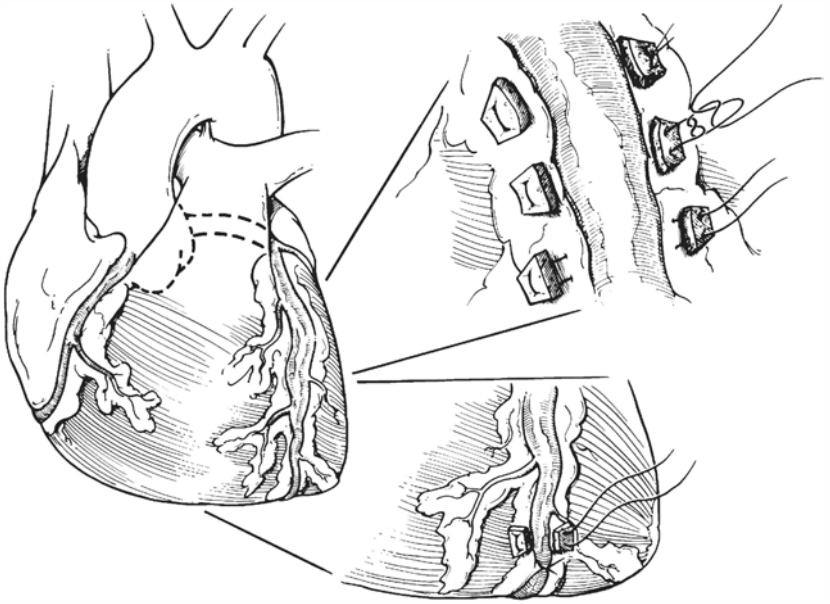


Fig. 98.15. If the termination site of the fistula is at the distal aspect of the coronary and no significant myocardium is in jeopardy, the coronary may be ligated proximal to the termination site. If the fistula terminates in the mid-portion of the left anterior descending coronary artery, the fistulous communication may be closed with multiple pledgeted sutures placed underneath the coronary artery so as not to impair distal perfusion.

monitored. If there are no signs of ischemia and there is adequate myocardial perfusion, the ligature is tied. To ensure complete closure, a second suture ligature should also be placed. Intraoperative transesophageal echocardiography is useful to verify whether the fistula is closed.

Cardiopulmonary bypass is indicated for several reasons, including: the fistula arises from the middle of a coronary artery, the fistula is inaccessible, another cardiac lesion needs to be repaired simultaneously, or if the complete coronary course is inadequately defined. The aorta and both cavae are cannulated and bypass is initiated. If cardioplegic arrest is necessary, the fistula should be temporarily compressed during administration of the cardioplegia to prevent run-off through the fistula into the heart. If adequate arrest is not possible because of flow through the fistula, retrograde administration of cardioplegia solution may be necessary.

There are a variety of techniques that may be used to close the fistula. If the fistula terminates in the mid-portion of the LAD, the fistulous communication can be closed by placing multiple pledgeted sutures beneath the coronary artery, while avoiding distal perfusion compromise (Fig. 98.15). If distal perfusion of the coronary bed is affected fistula closure, coronary bypass grafting may be necessary. Another technique that can be used when the fistula arises from the mid-portion of the coronary is to longitudinally open the coronary on the epicardial surface and oversew the origin of the fistula from within the coronary artery (Fig. 98.16). The coronary artery can then be closed primarily.

If the fistula terminates in the right atrium or right ventricle, it may be closed directly from within the cardiac chamber (Fig. 98.17). A right atriotomy is performed after cardiopulmonary bypass and the termination site of the fistula is identified from within the chamber. The use of cardioplegia may be helpful for localization. The termination site may be closed primarily or with a pericardial patch (Fig. 98.18).

Surgical Results

The operative mortality for coronary artery fistula repair is quite low with outstanding late results. Few patients appear to have recurrence of the fistula. Lowe and colleagues reported on 22 patients after surgical closure of coronary artery fistulas. In 14 patients, the fistula was closed using sutures without cardiopulmonary bypass. The termination of a fistula within a cardiac chamber was closed using cardiopulmonary bypass in six patients. In the remaining two patients, saphenous vein bypass grafts were necessary to maintain distal perfusion after fistula closure. The operative and long-term mortality rate was 0 at an average follow-up time of 10 years. While a small residual fistula remained in one patient, there were no recurrences. Fernandes and colleagues reported on 56 patients who underwent closure of isolated coronary artery fistula. There was no early or late mortality but two patients had perioperative myocardial infarctions. Finally, Mavroudis and colleagues reported on 17 pediatric patients with an average age of 5.5 years. Eight underwent fistula closure utilizing cardiopulmonary bypass with one of these patients requiring distal IMA bypass graft. There was complete closure in all with no recurrences without any operative or late deaths reported.

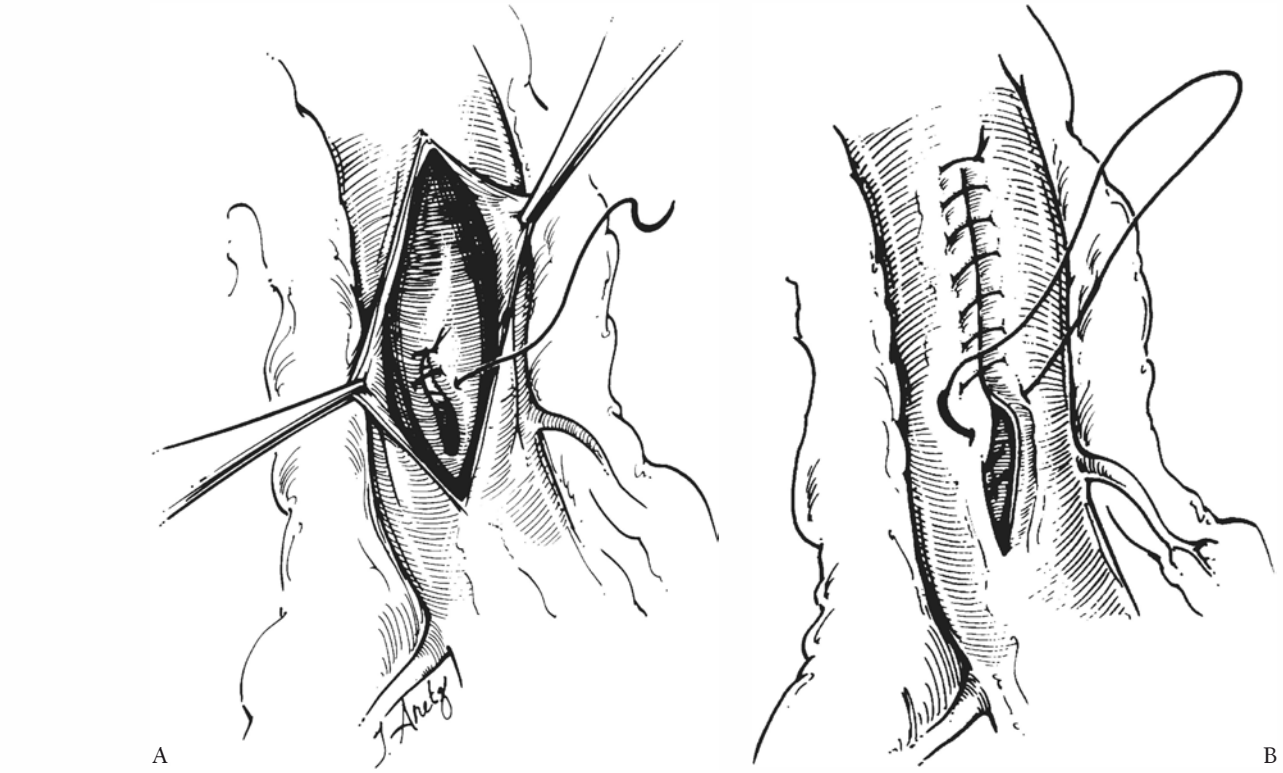
CONGENITAL OSTIAL ATRESIA OF THE LEFT MAIN CORONARY ARTERY

Anatomy

Congenital atresia of the LMCA ostium is a rare congenital coronary anomaly with <50 cases reported in the literature. As defined by Musiani et al., in this disease, there is no LMCA ostium; instead, the LAD and circumflex coronary arteries, which are located anatomically in the correct position, end blindly, and receive blood flow solely retrograde through the RCA, usually through at least one collateral vessel. These collateral vessels are generally inadequate to perfuse the left side of the heart and these patients are almost always symptomatic. While there is no coronary orifice from the left sinus of Valsalva, there may be a dimple on the inner aortic surface, a blind pouch, or no marking at all. This lesion is not the same as a single RCA; in the single coronary artery, blood flow occurs in a centrifugal, or antegrade, manner and most patients with this condition are asymptomatic, unless there is underlying atherosclerosis or congenital heart defects. Left main ostial atresia usually occurs alone; however, there has been associations noted with supravalvar aortic stenosis, VSD with pulmonic stenosis, right coronary ostial stenosis, and PDA and aortic regurgitation.

Pathophysiology

The main pathophysiology underlying congenital left main ostial atresia is collateral vessel flow from the RCA. There is blood flow from the RCA to the left coronary artery system through collateral vessels that are smaller in size than the left-sided coronary arteries. Therefore, there is not enough blood flow through these small



A B

Fig. 98.16. (**A**) When the fistulous communication arises from the mid-portion of the dilated coronary, the coronary may be opened longitudinally and the origin of the fistula oversewn from within the coronary. (**B**) The coronary artery is closed primarily.

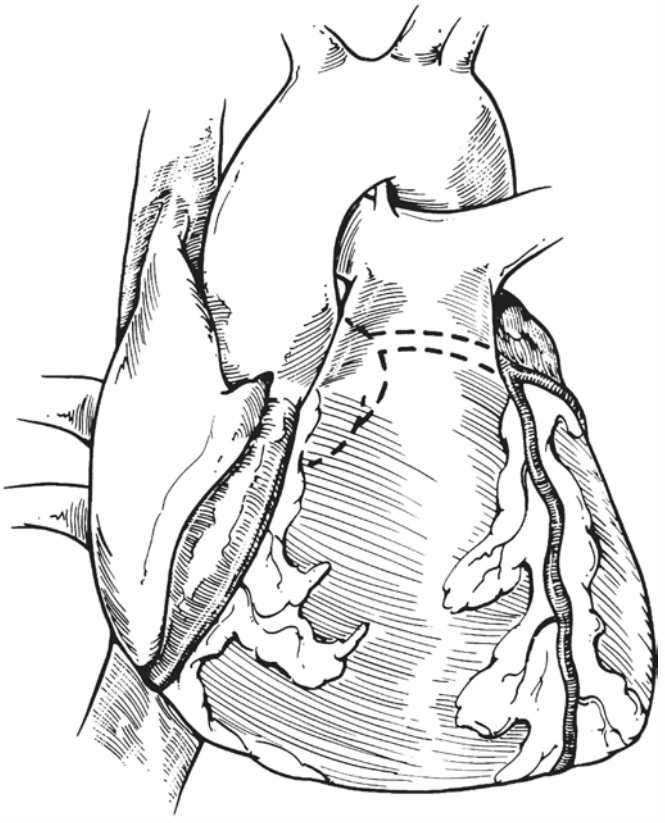


Fig. 98.17. A coronary arterial venous fistula arising from the mid-portion of the right coronary artery and terminating in the right atrium.

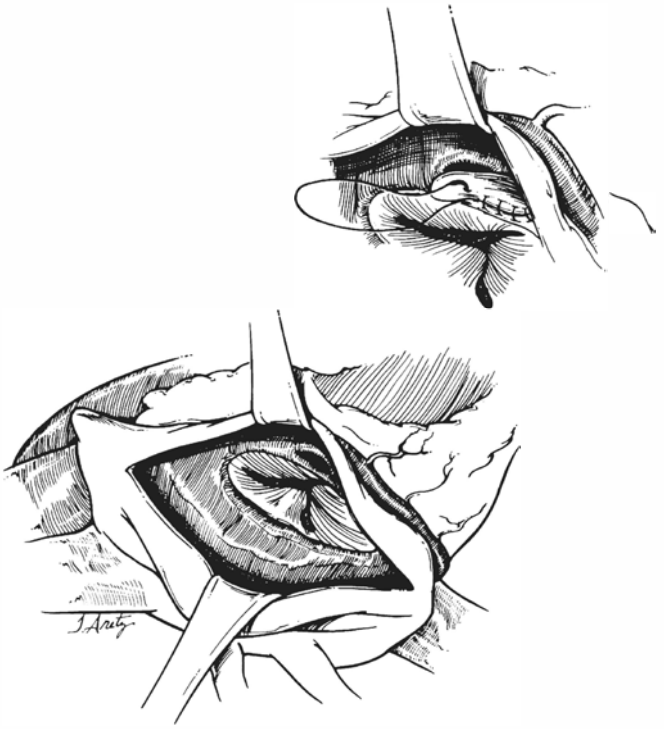


Fig. 98.18. After institution of cardiopulmonary bypass and induction of cardioplegia, the right atrium is opened and the termination site of the fistula identified from within the right atrium. This may be closed primarily or with a patch of pericardium.

vessels to perfuse the myocardium, which leads to ischemia and the potential for sudden death.

Clinical Presentation

Clinically, there appears to be early-, middle- and late-onset presentations with congenital atresia of the LMCA ostium. However, no matter what age the presentation, nearly every patient reported in the literature was symptomatic. The difference in age at presentation likely has to do with the extent of the collateral circulation as well as the presence or absence of other congenital heart defects. The early-onset patients, generally infants and young children, present with clinical signs and symptoms similar to ALCAPA or dilated cardiomyopathy. These patients generally present with feeding difficulty, failure to thrive, emesis, and dyspnea. In one patient, a holosystolic murmur was present due to severe MR from infarcted papillary muscles. They are also more likely to have an associated heart defect, suggesting that there is increased ischemia due to other congenital lesions that may cause increased oxygen demand. The older children and adolescents tend to present with syncope, dyspnea, angina, and ventricular tachyarrhythmias. The older adults are likely to have dyspnea and angina. Sudden death as the first presentation is present at all ages.

Diagnostic Imaging

Because symptoms are nonspecific and can be due to other reasons, diagnostic imaging is imperative in this diagnosis. In infants and children, chest radiography may reveal cardiomegaly with pulmonary congestion due to heart failure; in adults, the radiograph may be normal. The 12-lead electrocardiogram may show evidence of anterolateral Q waves, lateral T wave inversion, right bundle branch block, or ventricular tachycardia. A transthoracic echocardiogram should be performed and is likely to show a dilated left ventricle with poor function and MR. This is similar to the echocardiographic findings in ALCAPA. Close attention should be paid to the coronary artery ostia. Doppler color flow can be used to demonstrate retrograde flow from the RCA to the left coronary artery system without filling of the pulmonary artery. However, this may be difficult to demonstrate with echocardiography alone. A cardiac catheterization with coronary angiography is the gold standard and should be performed on any patient with the suspicion of ALCAPA or congenital atresia of the left main coronary ostium. Angiography should note direction of flow from the RCA to the left coronary system and attempt to delineate whether or not the PA is filled retrograde. If there is a question with diagnosis, selective RCA angiography should be performed, if possible

Surgical Management

Because of the high risk of sudden death with this ostial atresia of the LMCA, surgery should be undertaken immediately after diagnosis. The majority of the cases reported in the literature have been treated with CABG. This includes both children and adults. While CABG in adults may be the procedure of choice, the use of this procedure in children is questionable as the long-term results of CABG in children, especially using saphenous vein grafts, is questionable. More recently, there have been reports of surgical revascularization that creates a dual coronary artery system, with the belief that, like patients with ALCAPA, long-term outcomes should be improved.

Varghese and colleagues described the use of an autologous pericardial patch to attain surgical revascularization in a single case of left main coronary ostial atresia. Under cardiopulmonary bypass, the aorta was transected and a dimple was noted where the normal left coronary ostium should be. The aorta was then incised vertically to the location of the ostium and the incision was extended down the LMCA and ended before the bifurcation into the LAD and circumflex coronary arteries. If there is an atretic membrane, it is removed. An autologous pericardial patch was then used to reconstruct the atretic LMCA ostium.

Bonnet and colleagues described the use of surgical revascularization of the left main coronary arteries, with two of the patients having LMCA atresia. In their technique, after standard cardiopulmonary bypass, hot-induction blood cardioplegia, followed by cold blood cardioplegia, and warm reperfusion were used to help attain myocardial preservation. The main PA was transected to visualize the aortic root and left coronary artery system. The aortic incision began on the anterior portion of the aortic root, extending toward the coronary orifice. In those lesions that were atretic or occlude, such as in LMCA ostial atresia, the LMCA was incised beyond the atretic portion. Then, the two incisions (aortic and coronary) were connected using an onlay patch, consisting of saphenous vein, autologous pericardium, or PTFE. This patch not only enlarges the LMCA but also extends onto the portion of the aorta that was incised, creating a "funnel-shaped" neo-ostium.

Finally, Kaczorowski and colleagues recently described the use of homograft patch ostioplasty in the treatment of LMCA ostial atresia in three children (Fig. 98.19). In all cases, bicaval cannulation was used and cardiopulmonary bypass was established. Two of the 3 children, who were

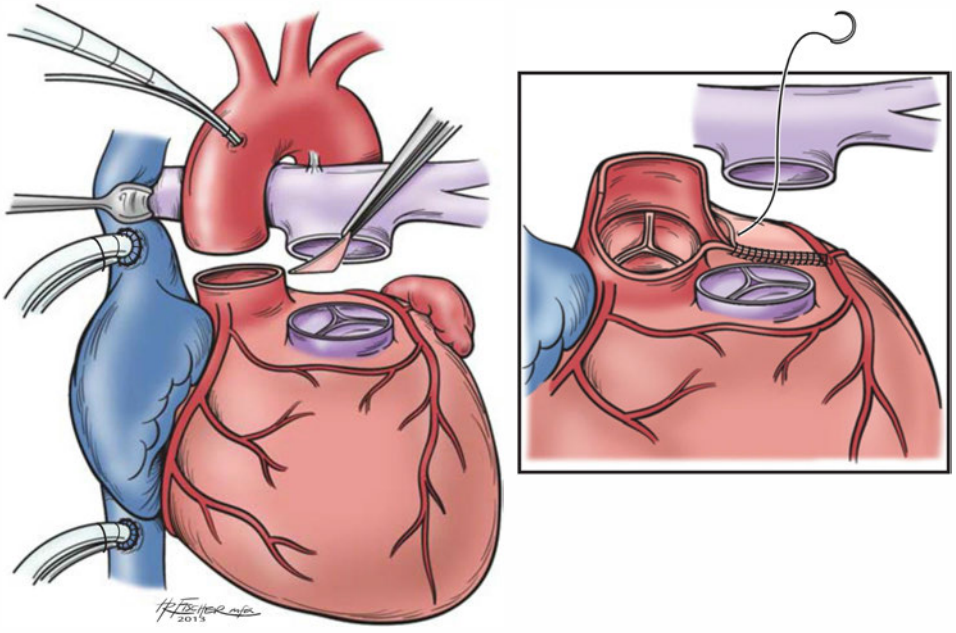


Fig. 98.19. Technique for ostioplasty of the left coronary ostium with homograft patch. Left coronary ostial atresia before intervention (*main illustration*) and with homograft patch augmentation of the ostium and left main coronary artery (*inset*).

initially thought to have ALCAPA, had their PA opened to evaluate for a coronary ostium; when none was found, an aortotomy was performed. Exploration in the aorta revealed no coronary ostium. In two patients, after identifying the blind-ending LMCA on the surface of the heart, an incision was made in the aortic wall, directed inferiorly to the aortic sinus and an incision was made in the LMCA until its division into LAD and circumflex coronary arteries. The ostium was enlarged using a pulmonary homograft patch that also joined the aortic sinus to the proximal LMCA. In one patient, the blind-ending LMCA was sewn onto the posterior aortic wall with anterior augmentation. Confirmation of coronary patency was achieved using coronary probes and with evidence of back bleeding. The aortotomy (and pulmonary artery in two cases) was closed and the patients were removed from bypass.

Surgical Outcomes

Long-term outcome data are missing in this population. Long-term results from CABG used for other coronary arterial abnormalities (e.g., ALCAPA), remain uncertain, although some reports using IMA grafts have shown good results. Postoperative mortality, at least in the short term, appears to be related to the extent of myocardial damage at the time of diagnosis and operation. It appears that if LMCA ostial atresia is recognized early and the patient has enough collateral vessels, then short-term results are encouraging after surgical revascularization, notably with those procedures that establish a dual coronary system.

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EDITOR'S COMMENTS

Multiple operations have been devised for the treatment of anomalous left coronary artery in children and are well outlined by Dr. Gaynor in this chapter. Although each technique may be applicable in certain circumstances, we believe that reimplantation of the coronary is the most ideal technique and establishes a two-coronary system with the least risk of late stenosis or occlusion. Past experience with the Takeuchi operation has also been favorable; however, we have seen cases in which the baffle dehisced in the pulmonary artery and created a left-to-right shunt, which in addition allowed decompression of the coronary artery. Thus, the reimplantation technique prevents the possibility of any left-to-right shunt through the coronary, and in the worst-case scenario, if the coronary occludes, the patient would have a situation similar to coronary ligation but at least would not have an additional volume load on the ventricle.

As described by Dr. Gaynor, we have taken the approach of reimplanting the coronary ostium in the left coronary sinus of Valsalva in a more anatomic location. Other authors have used a punch to create an opening in the aorta for reimplantation of the coronary. However, when this is used, the opening must be made more distally on the aorta to avoid blind damage to the commissural attachments of the aortic valve. Thus, we believe that it is simplest to open the aorta transversely, make an incision down into the left coronary sinus, and use the same suture line to close the aortotomy incision. In this way, damage to the aortic valve can be avoided and excellent exposure is obtained. We have used absorbable suture for the suture lines in young infants to encourage growth.

An additional controversy with anomalous left coronary artery is the management of significant mitral valve regurgitation in these patients. As noted by Dr. Gaynor, in the majority of cases, mitral regurgitation resolves when a two-coronary system is reestablished with resolution of distal myocardial ischemia and ventricular dysfunction. However, there are very rare cases of infants who have significant myocardial damage at the time of presentation and situations in which the papillary muscles themselves have undergone infarction with progressive elongation and fibrosis. In these patients, mitral valve repair can be undertaken with good results, often with papillary muscle shortening. Valve replacement for this condition should be extremely rare. One technical note at the time of operation in patients with anomalous left coronary artery is the fact that there is often a thin-walled venous plexus in the area between the aorta and pulmonary artery that has to be mobilized for the exposure to the coronary for reimplantation. This troublesome area of venous collaterals may require meticulous cauterization to avoid significant bleeding after the reimplantation is performed.

Coronary arterial fistulas are rare, but the majority of fistulas should be closed. The transcatheter coil embolization or occlusion device techniques have become increasingly popular, and these fistulas can be closed in the catheterization laboratory. However, in patients with a broad-based fistula proximal in the coronary artery, occluder devices will potentially compromise coronary flow and, therefore, surgical treatment is the procedure of choice. As noted by Dr. Gaynor, in the majority of cases, either direct ligation in the epicardial course of the fistula or suture obliteration beneath the coronary artery as the fistula dives into the myocardium can be performed with the patient off cardiopulmonary bypass, with excellent closure rates. More distal end artery fistulas at the distal ends of coronary arteries can often be difficult to close because the exact distal extent of the origin of the fistula is sometimes difficult to determine from the epicardial surface. Ligation and even suture obliteration of these distal coronary arteries may be the most satisfactory technique for ensuring that there is no recurrence of the fistula or residual flow. Even when from an epicardial inspection the fistula appears to be completely closed, intraoperative echocardiography is valuable because minor amounts of flow may still be present and complete obliteration is the goal in all cases. Not uncommonly, we have seen residual flow that requires additional sutures or ligatures for complete obliteration.

The situation of anomalous origin of the left coronary from the right or the right coronary from the left passing between the great vessels is becoming an increasingly recognized entity. Although in the past, these coronary anomalies were most commonly seen at catheterization, more recently, the techniques of echocardiography and ultrafast computed tomography have identified these coronary anomalies more frequently. They are especially frequently identified in children who are undergoing echocardiography for participation in sports or for some other nonassociated vague symptomatology. This has led to a situation in which many of these patients are being identified who are totally asymptomatic and brings up the question of whether all patients with these anatomies should undergo surgical treatment. The overall incidence in the general population is not completely known. These lesions have been associated with sudden death in athletes, especially when there is a slit-like origin of the anomalous coronary. We have, therefore, found it difficult not to recommend surgical intervention in patients with a slit-like orifice of these anomalous coronaries because it is unclear which patients will have sudden cardiac symptoms and potentially even death from coronary compression.

It is now apparent from increasing experience with the anomalous aortic origin of the coronary artery from the inappropriate aortic sinus that the risk of sudden death, especially in patients with anomalous right coronary from the left coronary sinus is extremely low. Although clearly symptomatic patients with signs of ischemia with this lesion should undergo surgical therapy, it is difficult to justify surgical intervention in patients who are completely asymptomatic and have this lesion found on a routine screening echocardiogram. Unfortunately in many cases, patients with this anomaly will be restricted from active athletics, which then significantly compromises the patient's quality of life. In some cases, repair of these defects has to be performed simply to allow the patient to have a more normal life style. However, as has been noted in the chapter, a study at The Children's Hospital of Philadelphia has shown that even with complete repair of these defects subclinical ischemia can be identified in some cases. Therefore, the true value of surgical intervention for this condition is not well-defined.

Patients who have a single coronary artery with a branch crossing between

(*continued*)

EDITOR'S COMMENTS (*continued*)

the great vessels have an unknown risk for ischemia and sudden death. In most of these cases, there is no mechanism for the compression of the anomalous course of the vessel, unless there is a slit-like orifice. Certainly, in patients in whom the vessel passes intramyocardially through the septum, there has not been an incidence of sudden death, and most of these situations are benign variations in anatomy.

The value of translocating the pulmonary artery to the left to separate the great vessels is somewhat controversial, since the annulus of the aortic valve and the infundibulum of the pulmonary outflow tract is fixed and moving the distal vessel causes relatively little variation in the anatomy at the level of the coronary artery.

As imaging modalities are becoming more and more accurate, more abnormal coronary arteries are being picked up in childhood.

Some believe most anomalous origins of the right coronary from the left coronary sinus should be considered a normal variant and surgery should not be recommended.

The use of CABG and coronary reimplantation is less desirable than unroofing procedures. Bypass of an anomalous coronary, even with an internal mammary artery, may be associated with closure or stenosis of the bypass graft due to competitive flow through what is in most circumstances a situation of normal coronary flow. Thus, in the absence of proximal stenosis, bypass grafting should not be a primary therapy.

TLS

99 Pediatric Heart Transplantation

Charles B. Huddleston

The first pediatric heart transplant was performed by Dr. Adrian Kantrowitz on a small infant with tricuspid atresia in 1967. The child died soon after the procedure but this ushered in the concept of heart replacement for unreconstructable congenital heart disease. There were very few transplants in children from the late 1960s into the early 1980s. From the mid-1980s onward there was a steady increase in the number of pediatric transplants related in large part to the development of cyclosporine as an immunosuppressant as well as the successes of the group at Loma Linda University led by Dr. Leonard Bailey in children and that of Norman Shumway at Stanford University. Approximately 400 to 450 heart transplants are performed in children per year throughout the world. This number has been relatively static over the past 15 years. Survival in children is similar to that seen in adults at 5 to 10 years post-transplant but improves at 15 years (48% vs. 34%). A number of changes have occurred in pediatric heart transplantation over the past 20 years. Transplantation as primary therapy for any congenital heart disease (hypoplastic left heart syndrome in particular) is unusual. There has been a steady increase in the use of ventricular assist for children with end-stage heart failure, usually due to cardiomyopathy. This chapter will reflect some of these changes. Although "pediatric heart transplantation" should be limited by age, I will also include transplantation in adults with congenital diagnoses.

INDICATIONS

Heart transplantation in children has two basic indications—cardiomyopathy and congenital heart disease. On balance, these indications are approximately evenly split, although the congenital diagnosis predominates in younger patients and cardiomyopathy accounts for the majority of patients in the teenage group. The technical aspects of the procedure in cardiomyopathy are no different than that used in adults. The technical challenges lie in replacing the heart in a child with disordered anatomic arrangements of the great vessels (arteries and veins) and with anatomic derangements made so by prior corrective or palliative procedures. Hypoplastic left heart syndrome was a major indication for heart transplantation in the early 1990s in large part due to the poor survival following the Norwood procedure. Current survival with reconstructive first-stage palliation has been approximately 80% for the participants in the congenital registry of the Society of Thoracic Surgeons. In addition to this, transplant waiting time mortality may be as much as 20% to 25% in this group of neonates. It is impractical to consider transplantation for even half of infants with hypoplastic left heart syndrome in that at least 500 infants are born each year with this congenital lesion in the United States where less than 100 donors in this age group are available. Among those with congenital heart disease as the primary diagnosis, most have single-ventricle anatomy and may be at various stages of the reconstructive pathway from diagnosis to Fontan. Whereas a better understanding of the implications of early palliation for these patients has led to more successful outcomes with the Fontan procedure, the survival curves for those patients clearly declines at a much faster pace than the normal population or even the overall survival curve for children with congenital heart disease. These patients present with need for special consideration as far as the technical aspects of the transplant, the presence of significant elevation in pulmonary vascular resistance, the presence of preformed antibodies, and the presence of multiple small aortopulmonary collaterals. There remain a significant number of young adults who survived atrial switch procedures for transposition of the great arteries. Many (all?) of those patients will ultimately develop heart failure. This group will be out of the age range for pediatrics, but a clear understanding of the anatomy involved is necessary for a technically successful outcome.

CONTRAINDICATIONS

Absolute contraindications to heart transplantation include ongoing malignancy, multisystem organ failure refractory to intensive treatment for heart failure, uncontrolled infection, and significant psychosocial issues. The presence of renal or hepatic dysfunction depends in large part on the degree and whether the patient might be candidate for combined or staged transplantation of either the liver or kidney. Elevated bilirubin has consistently been a marker for poor outcomes in heart transplantation. Renal dysfunction may be particularly problematic as both cyclosporine and tacrolimus (the primary drugs of immunosuppressant regimens posttransplant) have significant nephrotoxic side effects. The availability of effective mechanical support in children has broadened the recipient pool to some degree as those patients with significant organ dysfunction can be stabilized with ventricular assist devices and made better candidates.

Elevated pulmonary vascular resistance has been a classic risk factor for heart transplantation. The availability of multiple drugs to treat this in both the acute and chronic setting has altered the approach to this. As a baseline, the pulmonary vascular resistance should be $<$6 to 8 Woods units and the transpulmonary gradient should be $<$15 mmHg. It is often difficult to obtain accurate values of one or both of these because of congenital or acquired anatomy, resulting in multiple sources of pulmonary blood flow and difficulty accessing the pulmonary vasculature at the time of the cardiac catheterization. Patients deemed to be at high risk due to elevated pulmonary vascular resistance should be extensively studied to evaluate their response to a variety of pulmonary vasodilators. A lack of response acutely should be followed by long-term treatment with pulmonary vasodilators such as bosentan or prostacyclin in combination with inotropic support and then re-studied. Occasionally, an open lung biopsy may be of use to evaluate the presence of coexisting pulmonary parenchymal

disease. Patients with a very high systemic ventricular end-diastolic pressure may benefit from ventricular assist device implantation for several weeks and then reevaluated. Failing these measures, heart–lung transplantation would be an alternative, recognizing that the long-term prognosis following this transplant procedure is significantly worse than isolated heart transplantation.

DONOR ASSESSMENT/ MANAGEMENT

After the usual criteria for donor acceptance for organ acceptance have been met (blood-type compatibility, absence of transmissible disease), size match and organ function are then considered. Size match is of particular concern for small infants as very small donors are unusual. Accepting a heart from a donor three times the weight of the recipient will generally work out well, realizing that it may be necessary to open the left pleural space, remove some pericardium, and leave the sternum open for a few days posttransplant. For older children and teenagers, the range is usually 20% above and below the recipient weight. Many surgeons are of the opinion that a larger donor is preferred for patients with borderline elevated pulmonary vascular resistance; there are little data to support that position. Blood-type compatibility has been challenged in infants up to 1 year of age with results that mimic earlier results, albeit with more a complex immunosuppressive regimen.

The assessment of donor heart function is typically done with echocardiography only. The donor should be on only a modest degree of inotropic support with satisfactory blood pressure and evidence of good cardiac output clinically. Evaluation of the cardiac markers of ischemia (myocardial fraction of creatine phosphokinase and cardiac troponin I) should be routine. Donor hearts with borderline systolic function may be resuscitated using intravenous infusion of triiodothyronine. The basis of this is evidence that brain death is associated with reduction in cortisol and thyroid hormone production. Vasopressin is often necessary in the treatment of diabetes insipidus; the use of this drug will often allow a reduction in inotropic support. There are no absolute guidelines for the upper limit of inotropic support allowable for a donor heart to avoid posttransplant primary graft dysfunction, but generally one should avoid those requiring high doses of two or more.

POST-TRANSPLANT COMPLICATIONS

Graft Failure

This is the most common cause of early posttransplant deaths, especially in those transplanted at <1 year of age. Graft failure may be related to poor preservation, poor donor selection, early rejection, pulmonary hypertension, or technical issues. Support for graft failure posttransplant is usually ECMO. Intra-aortic balloon counterpulsation is generally ineffective in children with fast heart rates and a compliant aorta. Ventricular assist is feasible with current small devices available. Pulmonary hypertension as a cause of early graft failure usually presents with elevated central venous pressure, tricuspid valve regurgitation, and low cardiac output. The pulmonary artery and right ventricular pressure may not be elevated because the donor right ventricle may not be able to generate high pressures. The usual measures of treatment would be sedation with neuromuscular paralysis, inhaled nitric oxide, and inotropic support. Occasionally, mechanical support is necessary. One must be certain that there are no technical problems with the pulmonary artery anatomy or anastomosis, particularly in the setting of prior palliative operations involving the pulmonary arteries.

Rejection

The incidence of acute rejection is lower in infants than in older children and teenagers. Nonetheless, surveillance is necessary in all age groups. A high index of suspicion should be maintained in patients with preformed antibodies going into the transplant procedure. The diagnosis of cell-mediated rejection is relatively straightforward using endomyocardial biopsy material. Antibody-mediated rejection, however, is significantly more difficult to diagnose. Although biopsy material can be stained for complement factors (Cd4 being the most common), this is not completely reliable. Evidence of poor cardiac function clinically and by echocardiography with a biopsy that is negative for cellular rejection should prompt this diagnosis. Treatment for acute cellular rejection is high-dose steroids (10 mg/kg methylprednisolone intravenous) daily for 3 days. Antibody-mediated rejection requires plasmapheresis as well as drugs aimed at reducing the production of antibodies.

Bleeding

These patients often come to transplant having had multiple prior operations with the anticipated adhesions. In addition, they may be taking anticoagulants or have disordered hemostatic mechanism due to long-standing heart failure. Additional use of blood products is anticipated.

Other Complications

Infection is a constant risk for these patients due to the immunosuppression necessary. Prophylaxis against *Pneumocystis jirovecii* is necessary using either sulfamethoxazole/trimethoprim daily or inhaled pentamidine monthly. Renal insufficiency related to marginal pretransplant renal function and calcineurin inhibitors posttransplant is relatively common to some degree and may occasionally require temporary dialysis. Seizures may occur in 5% to 10% of patients due to posterior reversible encephalopathy syndrome. Posttransplant coronary vasculopathy is a long-term complication, occurring in 35% of patients by 10 years posttransplant; this is much more common in adults, present in more than 50% at 10 years posttransplant. Lymphoproliferative disorder and other malignancies are present in approximately 10% of patients in long-term follow-up.

OPERATIVE TECHNIQUES

General Comments

As mentioned above, the technique of transplantation for patients with cardiomyopathy is the same as with adult transplantation and will not be presented further in this chapter. The focus will be on issues that are unique to congenital heart disease. The number of different combinations of congenital anomalies and their anatomic nuances preclude an encyclopedic description of each method of recipient preparation and donor implant. The principles presented for the conditions described can be adapted for each individual situation. The patients with single-ventricle anomalies provide the greatest challenges primarily because of the abnormalities in situs and venous anatomy as well as the obligatory pulmonary artery anomalies associated with the palliative procedures that these children have undergone in the past. Careful planning of the procedure by reviewing prior operative notes, cardiac catheterizations, and other imaging studies is crucial to conducting a safe operation. A computed tomography (CT) scan with contrast is particularly

helpful in providing landmarks for careful sternal re-entry. It is important to obtain sufficient donor tissue for whatever reconstruction is necessary, usually the superior vena cava and branch pulmonary arteries. In the setting of multiorgan retrieval where lungs in particular are being retrieved, additional length of aorta should be obtained to use if pulmonary artery reconstruction is anticipated. Alternatively, some of the native tissue that would otherwise be discarded with the recipient heart may be suitable for patches. Caval anastomoses (as opposed to an atrial anastomosis) have become the standard for transplantation with the exception of small infants where the risk of anastomotic narrowing of the superior vena cava is relatively high.

Hypoplastic Left Heart Syndrome

Although reconstructive surgery has generally supplanted transplantation as primary therapy for HLHS, there are circumstances where the risks of the Norwood procedure are prohibitive and the balance shifts toward transplantation. These risk factors are pulmonary valve stenosis or regurgitation, severe right ventricular dysfunction, severe tricuspid valve regurgitation, or syndromic infants (e.g., Turner syndrome). Small size (<2.5 kg) is another risk factor for the Norwood procedure, but finding a suitably small donor is problematic.

Donor Procurement

The major concern is to acquire all of the aortic arch and the proximal descending thoracic aorta to the level of the ligamentum arteriosum. This provides sufficient donor aorta for the arch reconstruction. At the time of donor operation, the innominate vein may be ligated to get better access to the arch vessels. After aortic clamping and cardioplegia administration, the innominate, left carotid, and left subclavian arteries are divided at their origins. The descending aorta is taken just beyond the ligamentum arteriosum. The rest of the procurement proceeds as per the usual technique.

Recipient Operation

The ductus arteriosus, branch pulmonary arteries, and aortic arch with its branches are dissected out extensively. When dissecting out the ductus arteriosus and during the distal arch reconstruction, care must be taken to avoid injury to the recurrent laryngeal nerve. Cannulation for arterial inflow may be performed by one of the three methods: via the main pulmonary artery with control of the branch pulmonary arteries, directly into the ductus arteriosus with ligation of the pulmonary artery end of the ductus, or via the innominate artery usually through a small graft sewn to the innominate. The last method provides the greatest flexibility for cannulation during the organ implant and potentially allows for regional perfusion during arch reconstruction so that no period of circulatory arrest is necessary. Bicaval cannulation is generally preferred and the patient is cooled to 18°C. While the cooling is proceeding, the donor heart is prepared by removing the cephalad aspect of the aortic arch leaving a "tongue" of aorta to reconstruct the arch (Fig. 99.1A). The recipient heart is excised, ligating the ascending aorta with a silk tie (Fig. 99.1B). The left atrial anastomosis is performed first. I prefer to perform

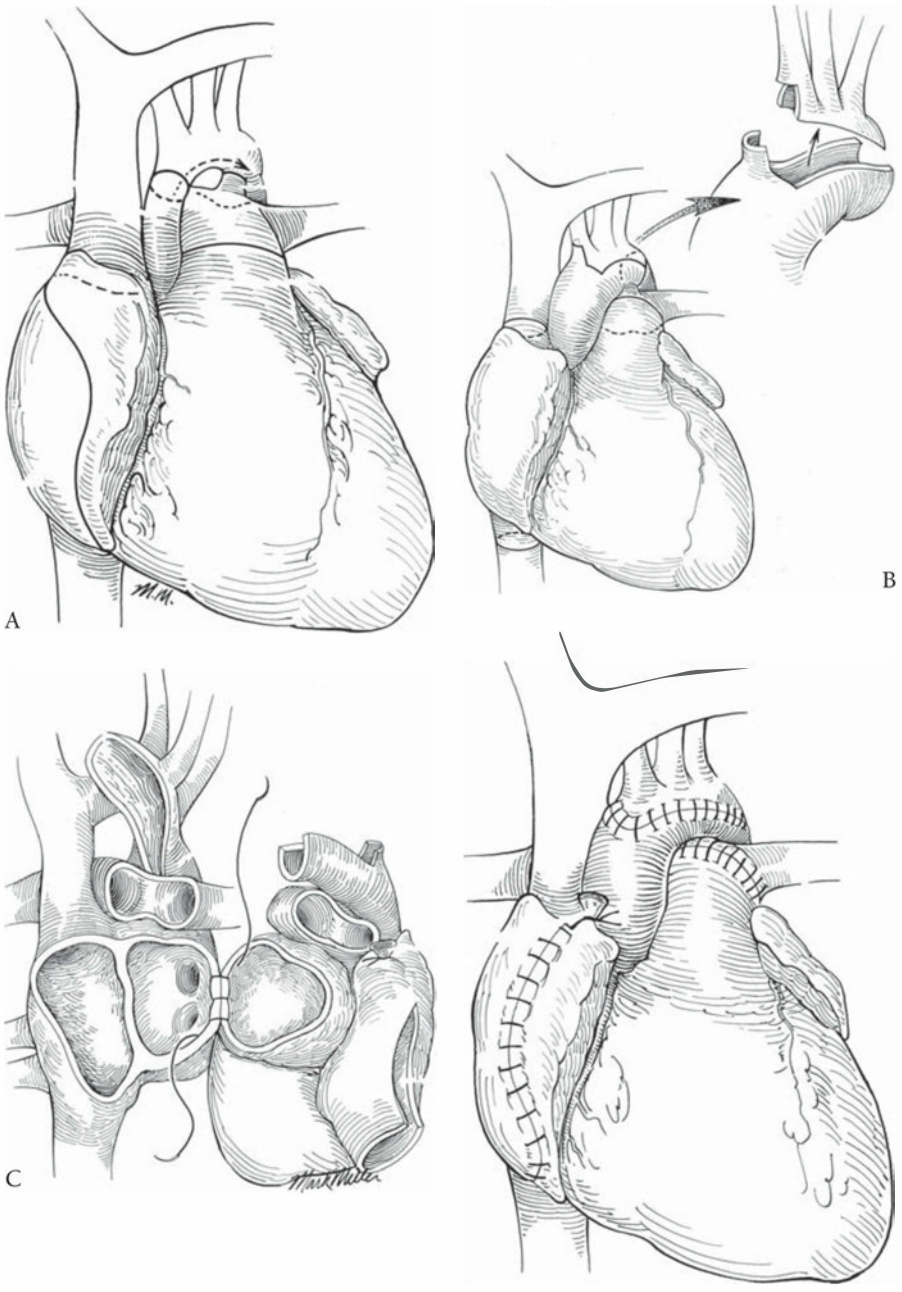


Fig. 99.1. Transplantation for hypoplastic left heart syndrome. **(A)** The recipient heart is removed leaving right and left atrial cuffs and dividing the main pulmonary artery between the bifurcation and the sinotubular ridge of the pulmonary valve. The ascending aorta is transected below the innominate artery and the underside of the aorta is opened beyond the insertion of the ductus arteriosus. **(B)** The donor heart should be retrieved with a segment of aortic arch all the way to the ligamentum arteriosus of the donor. The arch branches are removed leaving a "tongue" of aortic tissue for the arch reconstruction. **(C)** The transplant is performed with the left atrial anastomosis first, followed by the posterior right atrial anastomosis and then the arch reconstruction. **(D)** The pulmonary artery and anterior right atrial anastomosis are completed with the cross-clamp removed.

a portion of the right atrial anastomosis now because visualization will compromise coronary sinus blood return once the aortic reconstruction is completed and the clamp removed. At this point, either circulatory arrest or regional cerebral perfusion is initiated. Regional perfusion requires occlusion of the innominate artery proximal to the inflow graft sewn there and occlusion of the other arch branches as well as clamping the descending thoracic aorta 1 to 2 cm beyond the insertion of the ductus arteriosus. All the ductal tissues are excised and an incision is carried down into the aorta beyond the distal extent of ductal tissue. The remainder of the aortic arch is opened all the way to the distal ascending aorta (Fig. 99.1C). The aortic anastomosis is performed beginning distally and bringing the suture line all the way around the entire aortic arch. Following completion of this, the pulmonary artery anastomosis is performed in an end-to-end manner; the recipient MPA is often much larger than the donor. Finally, the transplant is completed with the anterior portion of the right atrial anastomosis (Fig. 99.1D).

Approximately 15% of all infants with HLHS have bilateral superior venae cavae (Fig. 99.2A). There are two methods of handling this. One is to take the donor innominate vein and sew the end of this to the end of the left superior vena cava. In this case, the right SVC will have to be sewn to the donor SVC. Another (preferred by me) is to perform the recipient cardiectomy in such a manner as to leave the coronary sinus attached to the left atrial posterior wall, emptying into the recipient right atrium (Fig. 99.2B and 99.2C). The coronary vein branches have to be ligated or included in the left atrial suture line to control bleeding. This avoids an anastomosis to small, thin-walled vessels which would be at risk for stenosis.

The posttransplant management of these infants is similar to the care required for any infant undergoing major open heart surgery with the exception of a higher risk of pulmonary hypertension. Most of these infants have been waiting for a prolonged period of time with a significant left-to-right shunt imposed by the circulation associated with HLHS. Given the high risk, it is reasonable to preemptively treat with nitric oxide and neuromuscular blockade.

"Congenitally Corrected" Transposition of the Great Arteries

These patients have discordant connection on both the ventriculoarterial level and atrioventricular level, resulting in no net shunt or cyanosis, but an anatomic right ventricle serving as the systemic ventricle and the anatomic left ventricle serving as the pulmonary ventricle. The natural history of this lesion varies considerably, in part related to the associated cardiac anomalies. However, it is typical for the anatomic right ventricular function to begin to deteriorate as patients approach adult years. Tricuspid valve regurgitation is a complicating factor in this clinical deterioration. A typical patient will have had a prior operation—closure of ventricular septal defect with or without a conduit from the pulmonary ventricle to the main pulmonary artery. A pacemaker is often in place as well. Some patients will have had an "anatomic correction"—arterial switch + atrial switch. Although that operation restores "normal anatomy," some patients will have progressive poor ventricular function of the anatomic left ventricle.

Donor Operation

This is the same as a typical cardiac harvest. It is helpful to retrieve as much length of the ascending aorta as possible.

Recipient Operation

The aorta is generally off to the left of the midline and anterior (Fig. 99.3A).

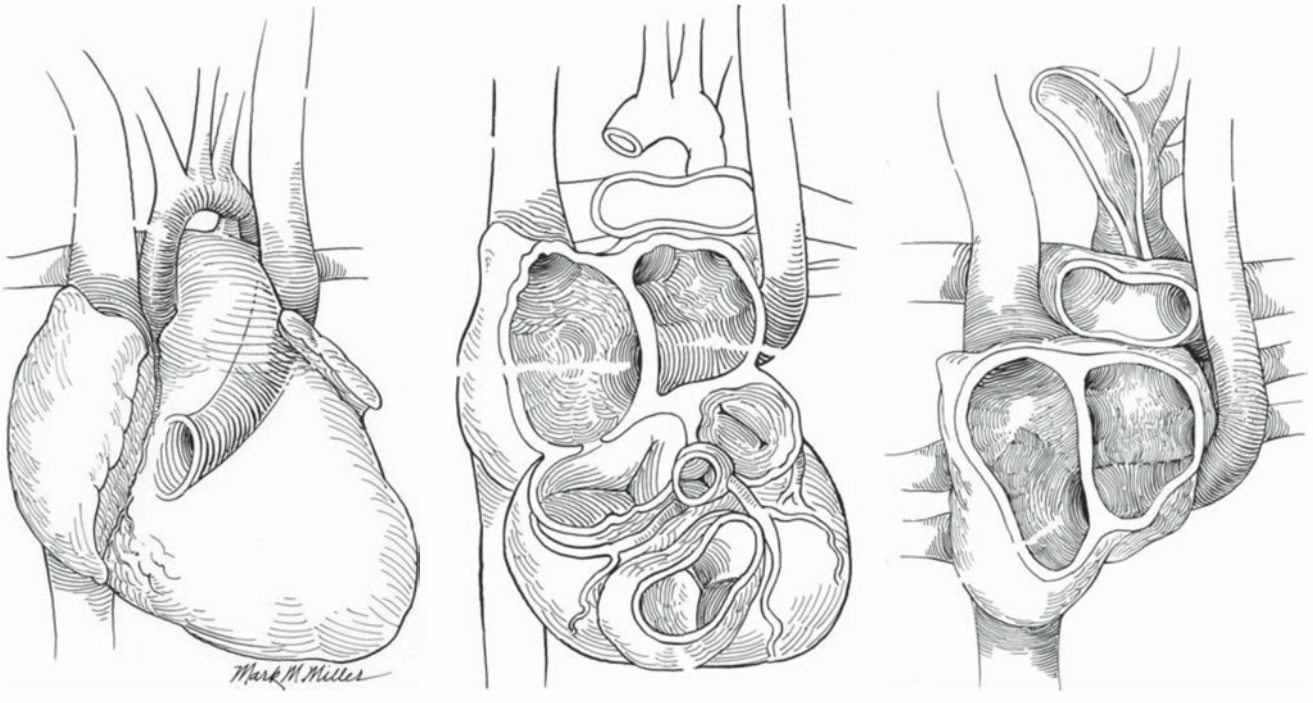


Fig. 99.2. Transplantation for HLHS with bilateral superior venae cavae. (**A**) The left SVC drains via the coronary sinus into the right atrium and there is no "bridging" innominate vein. (**B**) The recipient cardiectomy involves removing the heart paying particular attention to the left atrial cuff. The posterolateral portion of this is right on the atrioventricular groove leaving the coronary sinus intact. (**C**) The recipient cardiectomy is completed and the left superior vena cava and coronary sinus remain in situ. Some vein branches will need either individual ligation or inclusion in the left atrial suture line.

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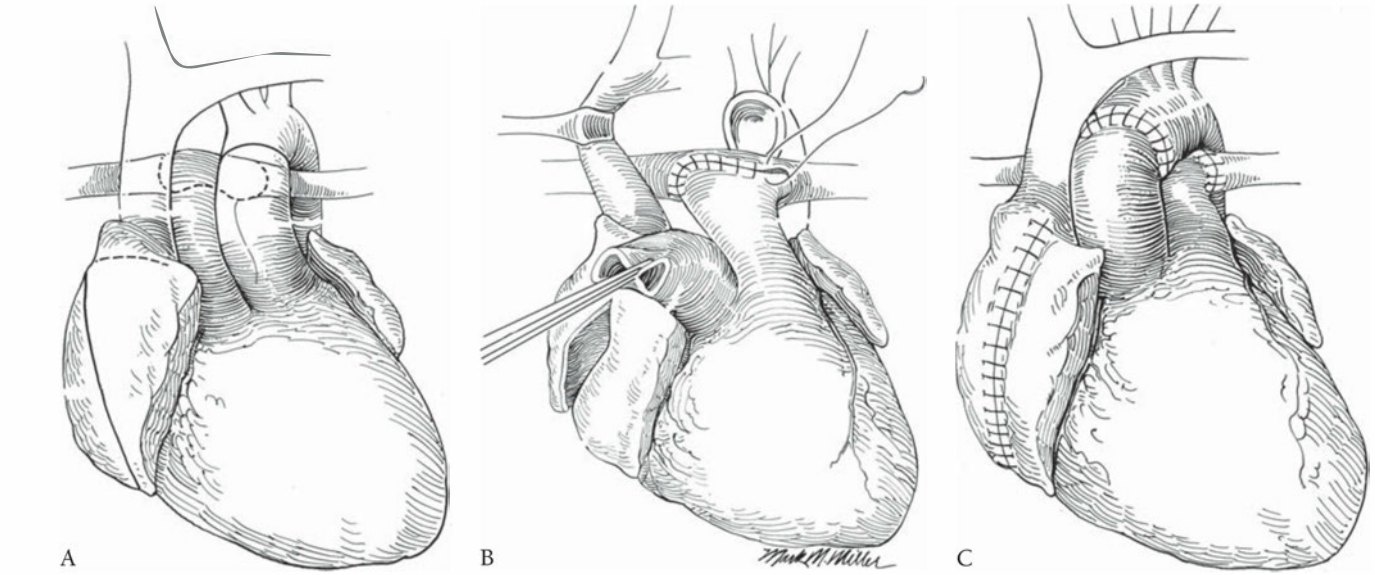


Fig. 99.3. Transplantation for "congenitally corrected" transposition of the great arteries. (**A**) The aorta is positioned to the left and anterior; occasionally, it is quite far out into the left side of the mediastinum. The main pulmonary artery is to the right and posterior to the aorta. (**B**) The recipient cardiectomy has been performed; the aorta is divided distally, in a more medial position. The pulmonary artery anastomosis is placed partially into the left pulmonary artery, rather than directly into the recipient main pulmonary artery. (**C**) The completed transplant demonstrating the aorta now in a more normal position to the right of the pulmonary bifurcation.

Cardiopulmonary bypass is initiated per the usual techniques. The aorta is transected relatively cephalad so that it is closer to the midline. The branch pulmonary arteries are mobilized so that the main pulmonary artery can be placed off to the patient's left. It may be necessary to open the left pulmonary artery and close a portion of the right side of the main pulmonary artery to place this anastomosis in the appropriate position. The pulmonary artery anastomosis should be performed prior to the aortic anastomosis so that visualization is optimized (Fig. 99.3B). The remainder of the transplant procedure is conducted in the usual manner. Again, a long length of donor aorta may facilitate this connection.

Transposition of the Great Arteries—Status Post-Senning or Mustard Procedure

Most centers stopped performing the atrial switch procedures for transposition of the great arteries in the mid-1980s. Thus, the youngest patient presenting for transplantation with this anatomy will be approximately 25 years old. Nonetheless, these are transplants that should be performed by a surgeon familiar with congenital heart disease. It is often difficult to recognize deterioration in these patients based upon objective imaging data. The echocardiogram typically shows a dilated anatomic right ventricle with diminished systolic function for many years. Functional data such as $VO_{2\ max}$ may be useful. Symptoms of progressive heart failure are most important and indicative of impending problems. Cardiac catheterization is important to evaluate for pulmonary hypertension and any evidence of baffle stenosis, particularly in the patients post-Mustard repair. This will need to be relieved prior to transplantation if present, usually by balloon dilatation with or without stenting. Many of these patients will have pacemakers in place.

Donor Operation

No special alterations in the donor harvest are necessary. Additional length on the aorta and main pulmonary artery should be obtained.

Recipient Operation

Careful sternal re-entry is important as with any patient undergoing redo cardiac surgery. In this case, the right ventricle and/or the pulmonary venous atrium are very dilated and often pressed up against the back of the sternum. Injury to this will likely result in significant hemodynamic deterioration. The recipient cardiectomy must be performed recognizing that the pulmonary venous atrium is now in a very anterior position (basically where the right atrium usually lies). Opening into this allows the surgeon to visualize the baffle so that it can be excised in its entirety. For the post-Mustard patients, this involves simply resecting the prosthetic patch (Fig. 99.4A). Occasionally, this is pericardium. This may make it difficult to identify borders as it is sewn to the interior of the atrial mass. Once this baffle is removed, it is obvious how best to proceed to provide a site for the left atrial anastomosis. The SVC and IVC will need to be cut off the atrial mass with as much tissue as feasible to allow for wide open anastomoses. Any stenosis in the SVC can be handled by either taking the anastomosis up higher or patching it. For the post-Senning patients, there are three layers to enter to get to the posterior left atrial wall (Fig. 99.4B and 99.4C). First, the anterior "right" atrial wall, entering the anterior pulmonary venous confluence. Next the anterior wall of the systemic venous pathway is in view. After penetrating that, one encounters the barrier created by the atrial septum sewn anterior to the left pulmonary venous orifices. The SVC and IVC should be removed with a cuff of atrial tissue followed by the removal of all barriers to the posterior atrial wall to get to the original left atrium so that the left atrial anastomosis is performed without any intervening tissue between the pulmonary venous orifices and the donor mitral valve. In the case of both the post-Senning and post-Mustard transplants, there will be a large amount of atrial tissue to trim away to prepare for the left atrial anastomosis. As with "corrected transposition," the pulmonary artery anastomosis may need to be moved to the

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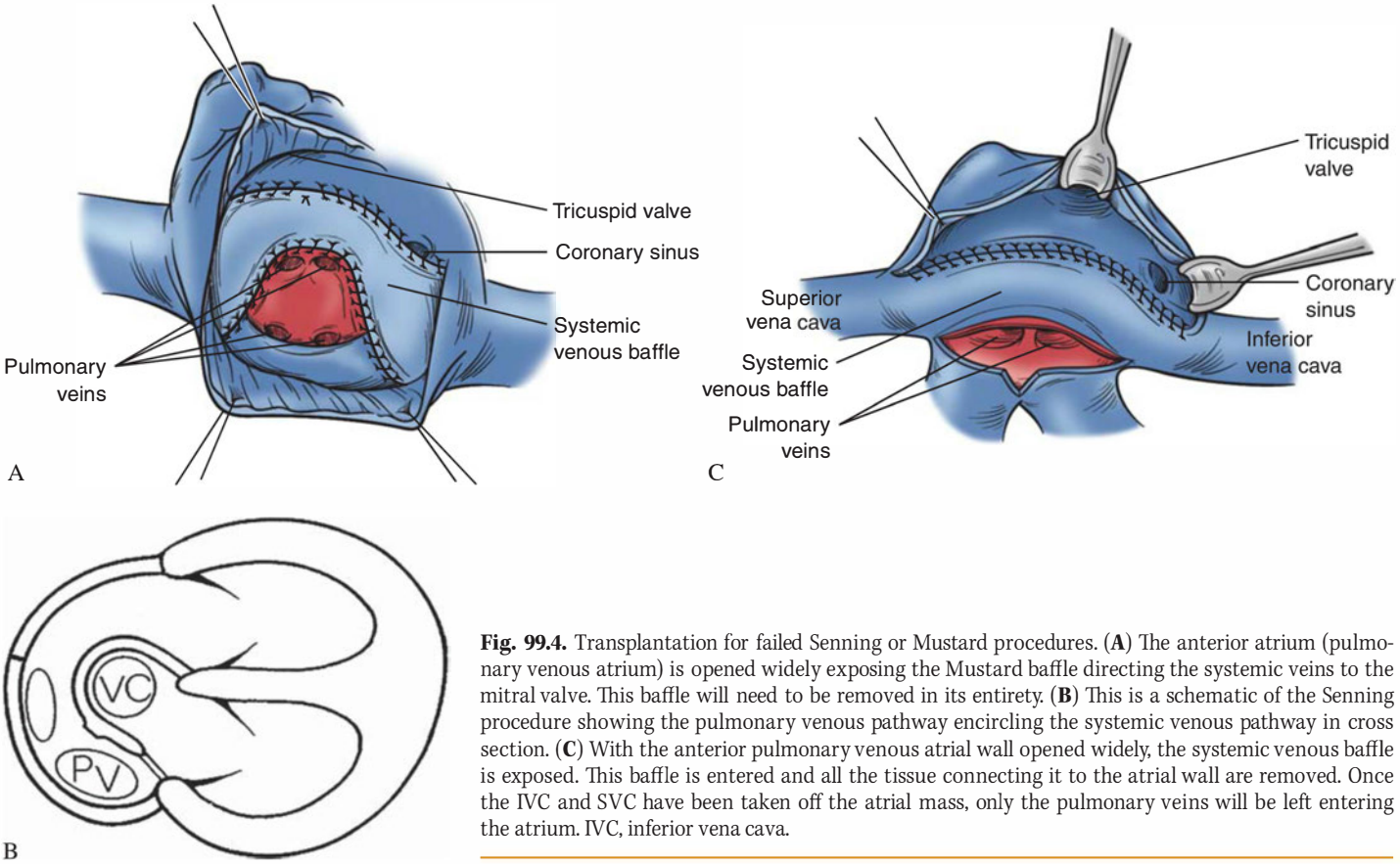


Fig. 99.4. Transplantation for failed Senning or Mustard procedures. (**A**) The anterior atrium (pulmonary venous atrium) is opened widely exposing the Mustard baffle directing the systemic veins to the mitral valve. This baffle will need to be removed in its entirety. (**B**) This is a schematic of the Senning procedure showing the pulmonary venous pathway encircling the systemic venous pathway in cross section. (**C**) With the anterior pulmonary venous atrial wall opened widely, the systemic venous baffle is exposed. This baffle is entered and all the tissue connecting it to the atrial wall are removed. Once the IVC and SVC have been taken off the atrial mass, only the pulmonary veins will be left entering the atrium. IVC, inferior vena cava.

left by incising from the orifice of the MPA into the LPA while closing off some of the right side of the MPA.

Failed Fontan

This is an increasing indication for heart transplantation in pediatrics. With improved survival of both early neonatal palliation of single-ventricle anatomy and the Fontan procedure itself, there are more children and young adults living with this circulation, some of whom develop complications requiring transplantation. Heart failure and protein-losing enteropathy are the two most common indications in this group.

Donor Operation

Pulmonary artery reconstruction or patch angioplasty and dealing with anomalies of the systemic venous return are the two anatomic challenges to transplantation in this group. The harvest should include as much of the superior vena cava as possible—at least up to the entry of the innominate vein—and branch pulmonary arteries. If the lungs are being harvested for transplantation, the pulmonary artery length will be limited. Taking some of the descending thoracic aorta will add to the flexibility of the transplant procedure.

Recipient Operation

This transplant procedure is perhaps the most challenging of what might be considered "standard" transplant procedures for congenital heart disease. It is challenging because of the multiple prior sternotomies, the native anatomy, the constructed anatomy, and the consequences of long-term cyanosis and of the Fontan circulation (chronically elevated central venous pressure). The cardiovascular structures may be densely adherent to the back of the sternum, creating potential havoc with sternal re-entry. The aortopulmonary collaterals complicate management of cardiopulmonary bypass as well as visualization of the anatomy with the heart opened and removed. Cannulation of the IVC may be difficult because of the location and nature of the conduit used to baffle the IVC to the pulmonary artery. There is generally no main pulmonary artery to identify, but rather a long confluent pulmonary artery between the hilar regions. The Fontan connections to the pulmonary arteries will have to be patched or taken down in such a way as to avoid creating stenosis there. Finally, these patients usually come to transplantation with very poor nutritional status related to protein losing enteropathy, some degree of liver dysfunction, and presensitized on an immunologic basis.

The illustrations for this show a post-Fontan recipient with associated bilateral superior venae cavae and thus bilateral Glenn shunts (Fig. 99.5A–99.5E). In this instance, the IVC is baffled to the pulmonary artery via an intra-atrial approach. Cannulation is performed high in the SVCs and as low as possible in the IVC. It may be possible to cannulate only the right SVC as there is an extensive network of venous connections in the facial and intracranial venous systems. In this case, the left SVC is snared to simplify the procedure. It is advisable to check the venous pressure above the snare to be sure that venous hypertension will not be an issue. The recipient cardiectomy is performed removing most of the right atrium and the entire cavopulmonary baffle. A cuff of right atrial tissue should be left on the orifice of the IVC to provide a bit more length for this anastomosis. The bilateral SVCs may be handled in couple of ways. Each of these will be presented separately.

The first method involves removing each SVC from its site of connection to the pulmonary artery with a plan of attachment to the donor innominate vein and right SVC. In most cases, the pulmonary artery defects left will require patching with appropriate material rather than directly oversewn. The transplant procedure is modified to allow

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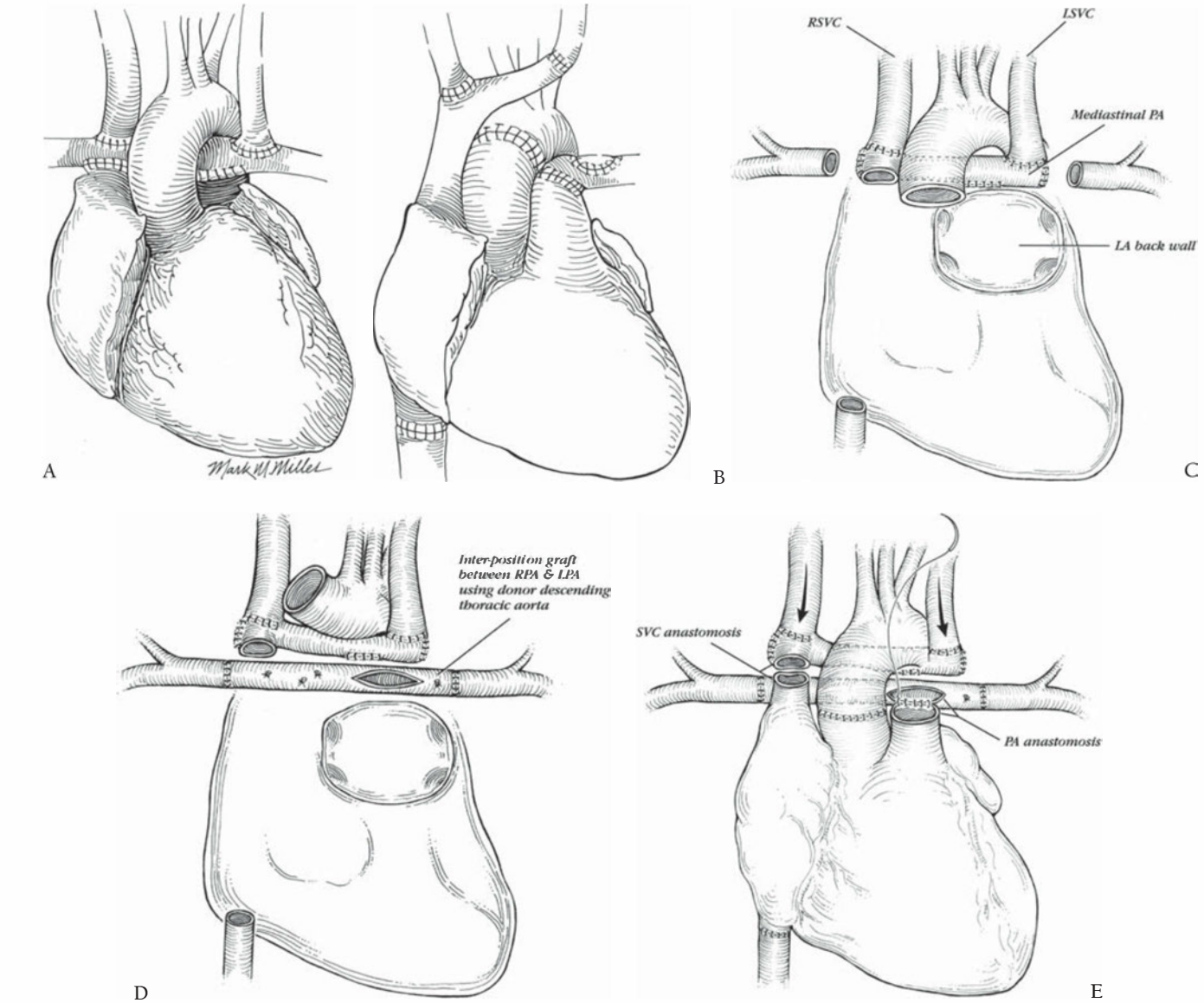


Fig. 99.5. Transplantation for "failed" Fontan. (**A**) This illustrates the heart of a patient with bilateral superior venae cavae and an intra-atrial lateral tunnel type of Fontan. Both superior venae cavae (SVCs) are anastomosed directly to the pulmonary artery. The IVC blood is directed via the intra-atrial baffle to the cardiac end of the SVC which is anastomosed to the underside of the right pulmonary artery. (**B**) In this illustration, the right and left SVCs have been disconnected from the respective pulmonary arteries which are then patched. A long segment of innominate vein obtained from the donor harvest is attached to the left SVC and the right SVC is then attached to the junction of the donor SVC and innominate vein. The underside of the right pulmonary artery is patched and the inferior vena cave (IVC) is anastomosed directly to the donor IVC. The illustration shows the innominate vein draped over the aorta; this is usually better placed posterior to the ascending aorta. (**C**) An alternative is to use the mediastinal pulmonary artery segment connecting the SVCs as an "innominate vein." The right and left pulmonary arteries are transected just lateral to the insertions of the SVCs on each side. The left lateral end is either patched or oversewn. The lateral opening on the right side will be used to connect to the donor SVC. (**D**) If the entire mediastinal branch pulmonary arteries are not available with the donor heart, an interposition graft of the donor descending thoracic aorta (taken at the time of harvest) or a synthetic graft is placed between the distal right and left pulmonary arteries. An opening in this graft is created at the proposed site for the donor pulmonary artery anastomosis. (**E**) The transplant procedure is then performed connecting the donor main pulmonary artery to the opening created in the interposition graft. The donor SVC is connected to the segment of recipient pulmonary artery between the SVCs and the IVC is anastomosed directly to the donor IVC.

for an anastomosis between the innominate vein and the left SVC. This donor innominate vein is usually best positioned behind the ascending aorta rather than draped across the top of the aorta. The left SVC is in a somewhat posterior position as well. The right SVC is anastomosed directly to the donor SVC and the recipient IVC is attached to the donor IVC end-to-end. The donor main pulmonary artery is connected to the mediastinal pulmonary artery at an appropriate position from hilum to hilum. This is facilitated if there is a long segment of donor pulmonary artery available.

An alternative method is to leave the SVCs attached to the recipient pulmonary artery, as illustrated in Figure 99.5 series. This intervening segment of pulmonary artery between the SVCs becomes the "innominate vein" as it is detached from the more distal right and left pulmonary

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arteries proximal to their hilar continuation. The new pulmonary artery segment is then reconstructed either by using the harvested donor branch pulmonary arteries left in continuity with the main pulmonary artery or with an intervening vascular conduit between the hilar regions. This conduit may be donor descending thoracic aorta or a synthetic tube graft. The advantage of leaving the SVCs connected to the recipient pulmonary artery is that it avoids the tedious nature of an anastomosis of a very thin-walled, fragile vein to another, risking suture line stricture. The disadvantage is that the reconstruction involves anastomoses to both branch pulmonary arteries in the hilar regions. If the donor aortic segment or synthetic tube graft is used for the pulmonary artery reconstruction, this can be done with the recipient heart out of the operative field simplifying this procedure. Circulatory arrest may also facilitate any of these steps in the reconstruction to avoid the field being obscured by pulmonary venous return from the extensive aortopulmonary collaterals generally present. The donor pulmonary artery anastomosis is then performed in an appropriate location in the mediastinal pulmonary artery segment. The donor SVC is connected to the underside of the new "innominate vein," and the IVC anastomosis is to the donor IVC. The order of connections is usually the left atrium, followed by the pulmonary artery reconstruction, the inferior vena cava, and finally the aorta. The cross-clamp can be removed at this point and the SVC anastomosis performed while rewarming.

In the presence of a "classic" Glenn shunt (where the pulmonary arteries are rendered discontinuous), the pulmonary artery reconstruction will nearly always require an additional segment of donor vascular tissue for an interposition graft. If the prior Fontan procedure was an atriopulmonary type, the most difficult part of the procedure may be sternal re-entry. An imaging study is crucial in these patients to identify what is lying directly behind the sternum. The right atrium in this situation is often enormous and stuck directly to the back side of the sternum. It may be necessary to begin cardiopulmonary bypass via femoral access prior to the sternotomy to avoid inadvertent entry into the right atrium which is much more difficult to control than even aortic entry. This leads to extremely long cardiopulmonary bypass times but is obviously preferable to the alternative.

Situs Inversus

These patients often have complex associated congenital heart disease, usually of a single-ventricle variety and will have had multiple prior procedures. Occasionally, this is associated with an isolated cardiomyopathy, but that is less common. In either instance, this entity is a significant technical challenge for transplantation. The likelihood of finding a suitable donor who happens to have situs inversus and an otherwise normal heart is extraordinarily unlikely given that it is estimated to occur at a rate of 2 per 10,000 in the general population. I have never encountered this in over 20 years of heart transplantation experience. Thus, techniques designed to modify the recipient anatomy to accommodate placement of a normal heart is necessary.

Donor Operation

Donor procurement depends on how the surgeon specifically plans to handle the systemic and pulmonary venous connections as well as the needs based upon associated anomalies and prior palliative procedures. To allow for the greatest flexibility, the harvest should include all the SVC and a long segment of innominate vein. If the patient has had a prior Glenn shunt or Fontan procedure, one should harvest as much mediastinal pulmonary artery or descending thoracic aorta as possible.

Recipient Operation

Cannulation directly into the SVC and IVC at points as far distant from the right atrium is necessary. The aorta should be cannulated in a distal location as well. The incision in the right atrium is near the atrioventricular groove, leaving sufficient amount of tissue for the modifications necessary for the implantation of the donor heart. The incision in the left atrium is also near the atrioventricular groove. As much of the atrial septum as possible should be left behind. The pulmonary artery is transected close to the bifurcation. The aorta is transected distally to move the anastomosis closer to the midline.

One acceptable technique for implantation of the donor heart is reminiscent of the Senning procedure for transposition of the great arteries (Fig. 99.6A–99.6D). The atrial septum of the donor is mobilized by dividing it at its caudad and cephalad portions. It is then reattached to the free wall of the left atrium anterior to the right-sided pulmonary veins. When an atrial septal defect is present, pericardium or prosthetic material may be used in addition to or in place of the atrial septal tissue. This directs the pulmonary venous return from the right lung to the left across the midline. The interatrial groove is then mobilized extensively. An atriotomy is placed anterior to the left-sided pulmonary veins. The left atrial anastomosis is thus placed on the left side of the mediastinum. It is usually necessary to perform the right atrial anastomosis next. A portion of the left superior aspect of the new right atriotomy can be closed primarily to move more of this anastomosis to the right and to better match the size of the donor right atrial cuff. The recipient pulmonary artery is usually positioned to the patient's right. This can be effectively moved to the left by mobilizing the branches or by extending the arteriotomy out onto the left pulmonary artery while partially closing the right side. The pulmonary artery anastomosis should be performed before the aortic anastomosis so that the pulmonary artery can be accurately seen. The aortic anastomosis is performed in the usual manner, but perhaps more distally so that it is closer to the midline.

There are other techniques that may be applied and may serve as better options, depending on the associated anomalies. The first of these is a modification of that described previously in which the atrial groove is dissected extensively and split so that two separate atria result. Frequently, the anterior portion of the atrial septum is too thin to precisely split; in that case, the septum should be devoted to the pulmonary venous atrium. These two atrial orifices are then transposed, moving the systemic (left-sided) atrium anterior and to the right and moving the pulmonary (right-sided) atrium posterior and to the left. The SVC and IVC need to be extensively mobilized—dividing the azygos vein above and mobilizing the subdiaphragmatic veins below. The atrial and arterial connections are then performed as described previously.

Another modification is based on the principle of devoting the atrial mass to the pulmonary venous connection and reestablishing systemic venous flow with bicaval anastomoses. This may be the preferred technique when the patient has undergone palliation for associated single-ventricle anatomy. Cannulation in the SVC must be at or above the entry of the innominate vein and in the inferior vena cava below the diaphragm. The left-sided SVC is divided at its junction with the heart and just below the entry of the innominate vein. The cardiac end of the SVC is oversewn. The donor SVC is connected to the right side of

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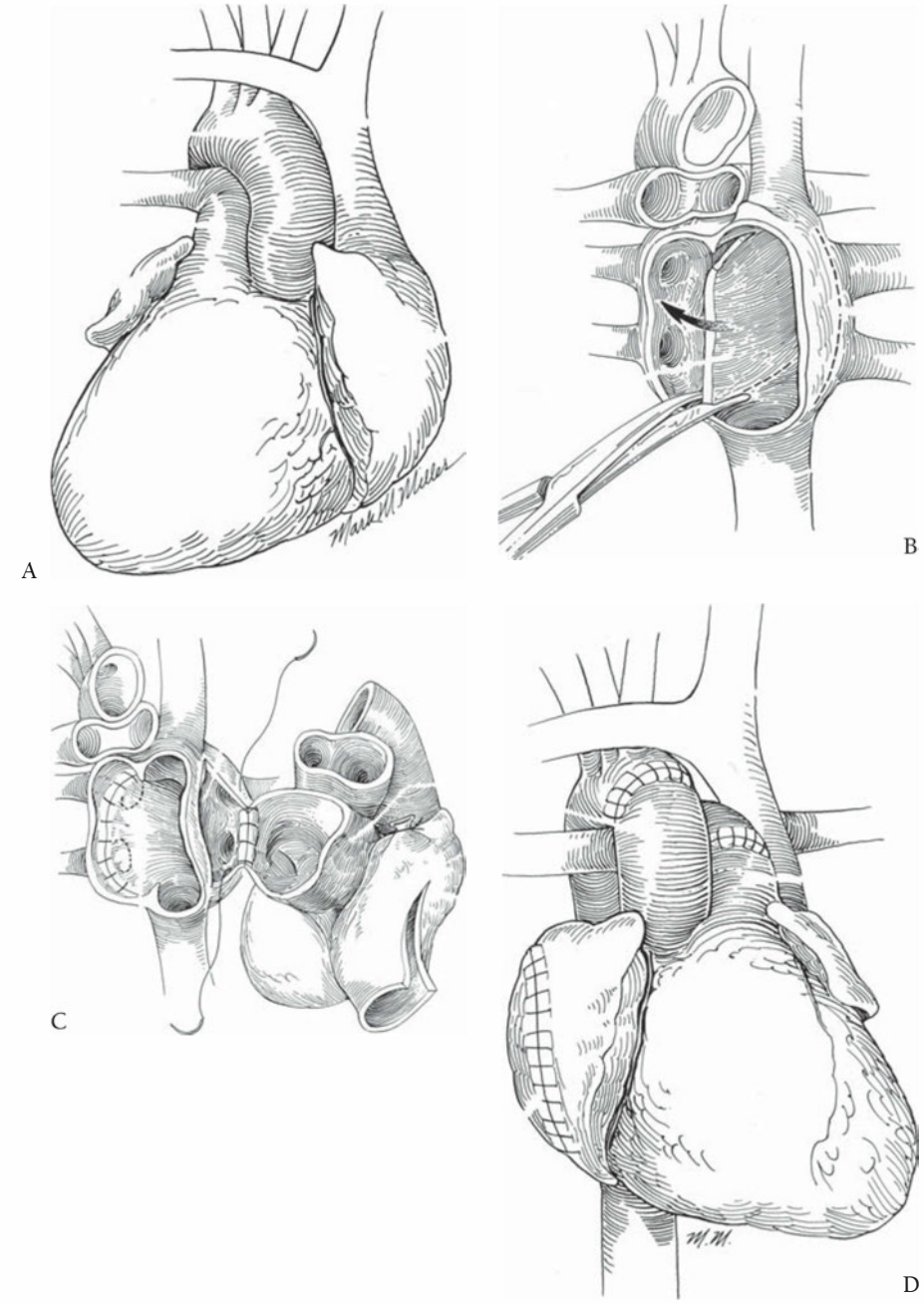


Fig. 99.6. Transplantation for situs inversus. (**A**) This illustrates a patient with "simple" situs inversus—no associated congenital heart disease. The SVC and IVC enter the right atrium on the patient's left side and the left atrium is on the patient's right side. (**B**) The recipient cardiectomy has been performed leaving a large amount of atrial wall and septum behind. The atrial septum is mobilized by cutting the caudad and cephalad portions back. The dashed line is the proposed site for the left atrial anastomosis. (**C**) The cut edge of the atrial septum is sewn to the free wall of the left atrium anterior to the entry of the right pulmonary veins. The atrial groove on the left side is extensively dissected out, and an atriotomy is placed just anterior to the entry of the left pulmonary veins. The donor heart is then sewn in place starting with the left atrial anastomosis on the patient's left side. The right atrial anastomosis is placed to the new right atrial opening into which both venae cavae have been directed. The pulmonary artery anastomosis will be moved out onto the left pulmonary artery somewhat because the recipient main pulmonary artery is often too far to the right. (**D**) The completed transplant.

the recipient innominate vein, placing this on the patient's right side. The recipient IVC will have to be moved as far to the right as possible. The diaphragm may be divided on the right side to assist with this. Leaving a flap of right atrial tissue attached to the IVC may be of assistance. Splitting the orifice to the IVC on the right side and sewing it to the pericardium to the right of it also moves the orifice closer to an appropriate anatomic location. The left atrial anastomosis is performed to a portion of the systemic venous atrium on the patient's left side. The remainder of the transplant procedure is carried as per usual techniques.

It may be necessary to carry out the transplant procedure with a combination of these techniques to accommodate the anatomy with which the patient presents. In any event, it is a formidable challenge and one should be aware of the potential for obstruction in the various pathways created so that they may be dealt with either by interventional cardiac catheterization procedures or by reoperation.

SUMMARY

There are no anatomic contraindications to heart transplantation. Any anomaly has a suitable means of implantation of the donor heart, although it may be necessary to alter the recipient anatomy and donor procurement to meet the individual needs. In addition, heart transplantation can be successfully performed in the presence of situs inversus totalis. It is essential that a full anatomic evaluation of the recipient be performed in addition to a careful review of prior operative reports so that appropriate plans can be made for donor procurement as well as the transplant procedure itself. The surgeon should have thorough understanding of congenital heart disease and experience in the various palliative and corrective procedures employed therein.

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EDITOR'S COMMENTS

As greater experience has been gained in transplantation of the heart in pediatric patients, especially those with congenital heart disease, it is apparent that virtually all anatomic malformations are amenable to orthotopic implantation of a donor heart. Abnormalities of systemic and pulmonary venous return can generally be addressed either by relocating the vessels or adding additional length from donor tissues. In addition, experienced congenital heart surgeons can use prosthetic materials to reconstruct most venous connections so that orthotopic implantation of the heart can be performed. One remaining concern exists in patients who have dextrocardia associated with either normal situs or situs inversus. Although successful orthotopic cardiac transplantation has been performed in patients with situs inversus, patients with dextrocardia and normal situs represent a situation in which the implantation of the heart in the pericardial space may produce rotation of the apex toward the right and can distort the septal position and tricuspid valve. This is less of an issue if there is cardiac enlargement and the pericardial space is large enough that the apex can assume a more normal location after transplantation. However, in children with a relatively normal heart size in whom transplantation is performed with situs inversus or dextrocardia, it may be necessary to excise a portion of anterior pericardium on the left to allow the apex of the heart to sit in the left chest.

As cardiac transplantation becomes more commonly applied in children with congenital heart disease, the need for donor organs increases. The success of reconstructive procedures for congenital heart disease has created a fairly large population of children and young adults with complex cardiac reconstructions, a significant proportion of whom may ultimately require cardiac replacement. Thus, children, particularly those in the early teenage group, often present for cardiac replacement at a time when donor organs of suitable size for adult recipients are becoming increasingly infrequent. The use of assist devices such as implantable left ventricular or right ventricular assist pumps and total cardiac replacements needs to be extended to the pediatric population to permit survival while these patients are awaiting a donor organ. In most pediatric heart transplant centers, the average waiting time for donor organs has progressively increased, and many patients coming to transplant are on some form of assist.

ECMO has been the primary method of ventricular assistance in patients awaiting organ transplantation in the pediatric population until recently. Now, various pulsatile cardiac assist devices are becoming available for the pediatric population. Although the Thoratec and Heart Mate devices can be used in older children and adolescents, there is still a limitation on available devices in the United States for small infants and children. The new DeBakey Child Heart may permit fully implantable devices in smaller children, but it is not currently useful for infants and neonates. The availability of the Berlin Heart assist device in the United States has made it possible to bridge even neonates and small infants to heart transplantation over waiting times of several months. These devices have the advantage of multiple different sizes for the potential population of recipients. Nevertheless, all assist devices still have significant complication rates, with thrombosis, thromboembolism, and problems of anticoagulation being the primary issues.

Whereas 1-year survival after heart transplantation in children is approximately 80%, the results of the international heart and lung transplant registries suggest that 10-year survival of adult recipients will be approximately 30% to 40%. Thus, the long-term survival of patients after heart transplantation is still significantly lower than that of an age-matched population. It is, therefore, logical to attempt to preserve the native heart as long as possible if an adequate functional result is obtained in any patient with congenital heart disease. Cardiac transplantation can then be used as an additional palliative therapy after earlier palliative interventions have failed.

The approach to cardiac transplantation in infancy for children with hypoplastic left heart syndrome remains controversial. As more centers have achieved excellent results with staged palliative reconstructive operations, such as the Norwood procedure and ultimately Fontan reconstruction, it becomes increasingly difficult to justify the use of scarce donor organs for patients who have other surgical options. For these reasons, most centers, including ours, have abandoned cardiac transplantation as primary therapy for hypoplastic left heart syndrome. Reconstructive surgery with the Norwood operation has been extended to even infants of very low birth weight, and there has been no incremental increase in mortality with patients who have aortic and mitral atresia and an extremely diminutive ascending aorta. The early mortality of approximately 20% is comparable with the mortality on the waiting list for most centers that do primary transplantation for patients with hypoplastic left heart syndrome. Although the percentage of patients who will ultimately require transplantation after Fontan reconstruction is unknown, it is anticipated that the majority of transplants will not be required until patients reach the teenage years at which time better availability of organs may occur. A particular concern in centers doing high-volume infant cardiac surgery is the need for cardiac transplantation in patients who failed an early reconstructive operation and require ECMO support for postoperative survival. These patients may be salvageable if a donor heart were to become available within a reasonably short time frame. Because of the limited availability of infant organs, such salvage of these critically ill patients is not currently likely. It is perhaps in these patients that the limited available infant organs should be preferentially used. Several centers have shown satisfactory results if transplantation can be performed in a timely manner even in patients on ECMO support before transplant.

Because of the concerns of increasing waiting times for infants and neonates who may be unstable, there is increasing interest in the use of ABO-incompatible transplantation for these patients. The ability to use a non-ABO-compatible donor heart in infants extends the potential donors available and can potentially decrease the waiting time for some of the sickest patients. The early results with these techniques have been quite acceptable, with no increase in rejection episodes or early morbidity and mortality. The exact age at which ABO-incompatible transplantation can no longer be accomplished is not clearly delineated, but after 1 year of age, measurement of antibodies titer to other blood groups would appear to be appropriate.

One difficulty with transplantation of patients who have undergone staged reconstructive surgery for hypoplastic

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EDITOR'S COMMENTS (*continued*)

left heart syndrome and have developed contraindications to continuing down the single-ventricle repair pathway is that often allograft tissue is necessary for reconstruction of the aortic arch and pulmonary arteries. It has been demonstrated that implantation of allograft tissue can result in significant elevations of panel reactive antibodies (PRA), which can increase the difficulty in cross-matching donor heart to a recipient and increase the risk of early rejection or chronic rejection episodes. Complex strategies including plasmapheresis and exchange transfusions may be necessary to permit transplantation with elevated PRA; clearly an increased incidence of rejection is a potential complication. The use of decellularized allograft material may decrease the risk of this complication; however, the routine use of the decellularized allografts has not been advocated for staged reconstruction due to the friability of this tissue at second- and third-stage reconstruction, making the surgery more difficult at the time of reoperation.

The optimal implantation technique for orthotopic heart transplantation remains controversial. Caval anastomotic techniques have been associated with a lower incidence of tricuspid regurgitation and better atrial function after transplant and therefore may be preferred in most cases. The late incidence of caval stenosis in children has not been identified but potentially can be treated with balloon dilation or stenting if it occurs. The use of separate pulmonary venous anastomoses has been raised in adult cardiac transplantation in the hope of improving left atrial and mitral valve function; however, the use of separate pulmonary venous cuff anastomoses may be associated with greater risk of arrhythmia.

The immunosuppression regimen after pediatric cardiac transplantation is variable from one institution to another. There is no clearly accepted superior technique for immunosuppression. The majority of centers use a triple-drug technique of azathioprine or mycophenolate mofetil, cyclosporine or tacrolimus, and steroids. Steroid-free regimens have been promoted in the hope of increasing late linear growth. In most centers, attempts are made to wean infant patients from steroids 6 months after the transplant, beyond the period when most early rejection and infection episodes occur. The use of tacrolimus for primary immunosuppressive therapy rather than cyclosporine in infants and very young children decreases the risk of hirsutism and gingival hyperplasia, which can impede feeding.

Controversy remains regarding the level of pulmonary hypertension at which orthotopic cardiac transplantation cannot succeed. Patients with significant transpulmonary gradients and elevations of pulmonary vascular resistance may have satisfactory results from orthotopic cardiac transplantation if the pulmonary resistance is reversible. Hemodynamic evaluation is important to assess in each patient before listing for heart transplantation. In some patients with an elevated transpulmonary gradient but in whom primary left heart dysfunction is present, heterotopic heart transplantation can permit the use of a heart transplant rather than a heart and lung transplant with acceptable outcome. The number of heterotopic heart transplants performed in children, however, is so few as to preclude any evaluation of long-term effectiveness of this therapy.

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100 Heart and Lung Transplantation in Children

Stephanie Fuller and Thomas L. Spray

Clinical heart–lung transplantation began with Cooley in the late 1960s when the first heart–lung transplant was performed in an infant with atrioventricular canal defect and pulmonary hypertension. The child died of pulmonary insufficiency 14 hours after the operation. The first clinical success was not achieved until 1981 when Reitz performed a heart–lung transplant in a 45-year-old patient with pulmonary hypertension. As success was achieved in adult patients, heart–lung transplantation was performed in an increasing number of pediatric patients, from only a few in 1984 to as many as 40 in 1988. Successful clinical use of isolated lung transplantation in an adult by Cooper in 1984 established the possibility of lung transplantation alone for certain forms of cardiopulmonary disease. These techniques were ultimately extended to the pediatric population such that the use of heart–lung transplantation in children has gradually been restricted to children with irreparable cardiac defects associated with pulmonary disease while isolated lung transplantation (either single or bilateral) has become an accepted modality of therapy for children with pulmonary vascular disease or primary pulmonary disease with reparable cardiac defects. By 2012, approximately 1,200 children, the majority adolescent, have undergone single-lung or bilateral-lung transplantation. The number of heart–lung transplantations in the pediatric population has declined significantly and is rare in the United States.

INDICATIONS FOR TRANSPLANT

General indications for heart–lung or lung transplantation in pediatric recipients are similar to those in adults. End-stage restrictive or obstructive parenchymal pulmonary disease or primary or secondary pulmonary vascular disease associated with correctable congenital heart defects can be considered for cardiac repair and lung transplantation if the cardiac repair is durable and not associated with significant inherent mortality. The presence of significant left ventricular dysfunction or cardiopulmonary defects that are uncorrectable are best treated by heart–lung transplantation.

Although general indications for lung transplantation are similar in adults and children, the types of pulmonary disease seen in children are different from those in adults. Obstructive pulmonary disease is extremely unusual in children as is pulmonary fibrotic disease. The great majority of lung transplants in children >5 years old are for cystic fibrosis. If present, fibrotic disease other than cystic fibrosis is often due to radiation fibrosis as a consequence of the treatment for leukemia and lymphoma. Younger children, under the age of 6, are transplanted for idiopathic pulmonary arterial disease and congenital heart disease with pulmonary venous stenosis. Neonates are only usually referred for significant surfactant abnormalities such as surfactant B or C deficiency for which there is no other treatment.

Children with cystic fibrosis represent the single largest group of pediatric patients who require lung or heart–lung transplantation. Most of these children are teenagers because the majority of cystic fibrosis patients survive to adulthood with intensive medical management and suffer from a slow decline. Indications for consideration of pulmonary transplantation in cystic fibrosis patients include increasing hospitalization for infection, progressive weight loss in older patients or lack of weight gain in younger patients despite nutritional supplementation, and increase in oxygen dependence or hypercarbia with gradual deterioration of pulmonary function. In general, a forced expiratory volume in 1 second (FEV_1) of <30% of predicted values is a relative indication for transplantation.

Cardiopulmonary diseases requiring transplantation in infancy are rare but include congenital diaphragmatic hernia, surfactant protein deficiencies, pulmonary vein stenosis or veno-occlusive diseases, and primary pulmonary hypoplasia.

The indications for transplantation in children with pulmonary hypertension are somewhat subjective because of the poorly defined natural history of pulmonary hypertension, either primary or secondary, in this age group. Generally, indications for consideration include progressive exercise intolerance, the onset of syncope, hemoptysis, angina pectoris, and significant right ventricular failure. These symptoms are often correlated with hemodynamic abnormalities, with elevations of right atrial pressure to >8 mmHg with a decreased cardiac index and a total pulmonary vascular resistance of >20 Woods units/m^2. When the product of right atrial mean pressure and pulmonary vascular resistance index is >360, poor survival is expected and transplantation is considered. Some children with significant pulmonary vascular disease may respond to vasodilators, including prostacyclin, and the use of chronic prostacyclin infusion may improve the stability of patients while they await suitable donor organs. The availability of increasingly novel therapies for pulmonary hypertension has decreased the need for lung transplantation in a significant number of patients. Most patients can now at least have significant palliation of their disease by medical management with transplantation reserved only for those patients who develop right heart failure or other major complications. In patients with Eisenmenger syndrome and a right-to-left cardiac shunt, the onset of severe polycythemia in association with hemoptysis, right ventricular failure, or progressive exercise intolerance may be considered a relative indication for transplantation.

A particularly difficult subgroup of patients comprises those with pulmonary vein stenosis and pulmonary veno-occlusive disease. These children are often extremely unstable with severe pulmonary hypertension. Because delivery of blood to the left ventricle is limited in these

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patients, there has been a high incidence of death while awaiting organs, and therefore these children should be listed early in the course of their disease. It is important to recognize patients who have pulmonary vein stenosis or veno-occlusive disease that is not amenable to further surgical therapy. Despite the increased enthusiasm for the use of sutureless pulmonary vein repair in patients with pulmonary venous stenosis, there are no significant data that this approach has decreased the incidence of recurrent obstruction and in most of these patients the process appears to be a progressive disease. The lack of dilation of the pulmonary veins in the hilum of the lung in the presence of pulmonary vein stenosis is a marker that there are abnormalities of the veins that will not respond to surgical intervention. In addition, very small veins at the time of repair of total anomalous pulmonary venous return are associated with very poor long-term outcome and these patients may also be considered for transplantation. Patients who have stenosis of only one or two pulmonary veins or occlusion of one vein with other veins not obstructed yet in the presence of significant pulmonary hypertension are a very poor subset of patients for any surgical intervention. These patients who will not respond to surgical therapy should be considered for lung transplantation. Stenting of stenotic pulmonary veins may be palliative in some cases as a bridge to pulmonary or cardiopulmonary transplantation. Although it is possible to use chronic ventilation, nitric oxide therapy and even extracorporeal membrane oxygenation (ECMO) as a bridge to lung or cardiopulmonary transplantation in children, patients with pulmonary vein stenosis or pulmonary vascular disease are often unable to be adequately resuscitated with chest compressions while ECMO support is initiated because of the inability to get adequate blood flow to the left ventricle to provide cerebral blood flow during cardiac arrest. Heart–lung transplantation provides an option for patients who have uncorrectable congenital heart disease and pulmonary vascular disease. Typically, these patients become candidates once palliation is no longer successful, once exhibiting severe heart failure requiring inotropic support, or severe pulmonary hypertension requiring inhaled nitric oxide or infused prostaglandin.

Infectious complications are more common after cardiopulmonary and pulmonary transplantation than after solid-organ transplants. Of particular importance in pediatric recipients is the common occurrence of viral infections of the transplanted lungs. Children continue to be exposed to a wide range of viruses, including adenovirus and respiratory syncytial virus in addition to the influenza viruses. In addition, cytomegalovirus (CMV) infection is very common after pediatric lung transplantation because many children have not yet been exposed to CMV infection and active immunity is not present. An additional concern in children is the presence of Ebstein–Barr virus (EBV). There is a higher incidence of seronegativity for this viral pathogen in children than in adults, and EBV infection can be quite subtle in early childhood, making diagnosis difficult. A significantly increased risk of associated lymphoproliferative disease in patients who have sustained an EBV infection after transplant has been reported and may progress despite decreased immunosuppression in these patients.

CONTRAINDICATIONS TO TRANSPLANTATION

Contraindications to transplantation are malignancy, multisystem organ failure, and sepsis. Other contraindications are primarily mechanical. Patients with severe scoliosis or restrictive chest wall mechanics may have chronic hypoventilation even if normal lungs are implanted. Significant associated metabolic diseases including renal insufficiency or uncontrolled diabetes are relative contraindications to transplantation, and patients with portal hypertension and biliary cirrhosis may also be considered poor candidates for cardiopulmonary transplantation alone. Patients who have had multiple prior surgical procedures with involvement of the pleural spaces require more complicated operations for implantation of donor organs. This is a particular problem in patients with chronic cyanosis, who may have multiple and extensive collateral vessels in the adhesions from previous surgeries that may be difficult or impossible to control at the time of transplant. Severe and even fatal bleeding complications have been noted in these patients in our series. High-dose steroid dependence is a relative contraindication to transplantation because of poor wound healing or sepsis. However, moderate doses of steroids have not been considered a contraindication in our experience. A long-standing history of noncompliance with medical interventions by the patient or family may be considered a relative contraindication to undertaking a procedure of such magnitude. Uncontrolled collagen vascular disease is also a contraindication to consideration for transplantation.

SELECTION OF OPERATIVE PROCEDURE

Historically, the majority of transplantations in children have been by the heart–lung en bloc technique. Whereas it is apparent that comparable results can be obtained with heart–lung or lung transplantation in many children, the need to maximize the availability of scarce donor organs and use the heart for other patients has produced a gradual decrease in the use of combined heart–lung transplantation for primary pulmonary diseases. Most heart–lung transplantation is now reserved for patients with uncorrectable congenital heart defects associated with severe pulmonary vascular disease. Heterotopic heart transplantation may be used in some children with elevated pulmonary resistance in whom improvement in pulmonary resistance can be anticipated with improvement in cardiac output. We use heart–lung transplantation for only those children with congenital heart disease who have a poor chance of long-term correction or those children with severe left or biventricular dysfunction. Pulmonary transplantation with preservation of the native heart is preferred in children with primary or secondary pulmonary vascular disease associated with normal ventricular function and a relatively simple or correctable congenital heart defect. Even children with severe right ventricular dysfunction are considered candidates for pulmonary transplantation alone if the right ventricular ejection fraction is $>10\%$ and tricuspid valve insufficiency is graded less than severe. Patients with repairable cardiac defects can receive cardiac repair and lung transplantation alone, and our experience includes children with atrial septal defect, ventricular septal defect (VSD), patent ductus arteriosus, vascular rings, atrioventricular canal defects, pulmonary vein stenosis, peripheral pulmonary arterial stenosis, and pulmonary atresia with nonconfluent pulmonary arteries in addition to children with anomalies of pulmonary venous return.

An additional consideration in pediatric patients is the preference of single-lung versus bilateral sequential lung transplantation. In children with cystic fibrosis, bilateral lung transplantation is preferred to remove the infected lungs and decrease

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the sources of potential sepsis. A similar consideration is given to patients with chronic bronchiectasis who may be best served by bilateral sequential transplantation and removal of infection sources in the lungs. Single-lung transplantation is a possibility in patients with pulmonary fibrosis and pulmonary vascular disease. Although there have been successful series of single-lung transplantations for primary and secondary pulmonary hypertension, these reports have suggested that the postoperative course is more complicated in such patients. The entire cardiac output is delivered to the transplanted lung if single-lung transplantation is used in the presence of severe pulmonary vascular disease. Thus, patients may be unstable in the postoperative period and have additional instability when infection, rejection, or bronchiolitis obliterans occurs in the transplanted lung. We, therefore, prefer bilateral sequential transplantation in most children with pulmonary vascular diseases to improve postoperative stability and the distribution of pulmonary blood flow. In addition, the use of bilateral sequential transplantation in younger children and infants allows for maximum possible growth and development of the lungs. Single-lung transplantation is still considered in children with primary pulmonary hypertension in whom there is a relative contraindication to entering one pleural space, such as those children who have had multiple previous thoracotomies with cyanosis.

DONOR SELECTION

Donors for cardiopulmonary or pulmonary transplantation are sparse compared with donors for other organs. Only 10% to 15% of cardiac donors may be suitable or donation of heart–lung blocks or lungs. This rarity reflects the damage often done to the lungs during gastric aspiration either at trauma or with a sudden neurologic event. In addition, severe pulmonary edema from either neurogenic or cardiac cause can affect oxygen exchange of the potential donor lungs. General criteria for pulmonary donors includes blood type compatibility, normal gas exchange with an arterial partial pressure of oxygen (PaO_2) of >300 mmHg on 100% oxygen, and 5 cm H_2O positive end-expiratory pressure. A clear chest X-ray film showing no infiltrates or contusions, age younger than 50 years with no history of pulmonary disease and a <20 pack-year smoking history, and normal findings on electrocardiograms and echocardiograms are criteria for cardiopulmonary and pulmonary donors. Bronchoscopy should confirm normal airway anatomy and easily cleared secretions with no ready accumulation of further secretions. A history of pulmonary disease or prolonged smoking or asthma is a relative contraindication for donation. In addition, demonstrated aspiration of gastric contents, contamination of the tracheobronchial tree, or severe lung contusion is considered contraindication to donor use. In some cardiopulmonary donors, the requirement of very high-dose inotropic drugs in the face of suitable fluid management or significant ventricular hypertrophy or dysfunction on an echocardiogram is considered a contraindication to using the combined heart–lung block.

As in all organ donation, the presence of human immunodeficiency virus or hepatitis A or B is a contraindication for use of donor organs. Hepatitis C organs may be used in hepatitis C positive recipient and in some cases may be used if the severity of the condition of the recipient warrants the risk of hepatitis C transmission. Although it is generally advisable to match CMV-seropositive donors with CMV-positive recipients, successful transplantation is not precluded by CMV mismatching. CMV prophylaxis is routinely used after transplantation and CMV infection is usually adequately treated if it occurs. Occasionally lungs that initially are deemed inappropriate for transplant may, with aggressive donor management, be rendered usable. Careful evaluation of each potential donor is therefore important to maximize the availability of suitable donor organs. Size matching between donor and recipient is important in pediatric lung and heart–lung transplantation. For heart–lung transplantation, it is desirable to have the donor weight within 20% to 30% of the weight of the recipient. Although it is possible in cardiac transplantation to use hearts of donors several times the body weight of the recipient, the fact that excessive lung size may compress the heart limits the size discrepancy that is acceptable for combined heart–lung transplantation in children. Donor–recipient size matching is more liberal when double-lung or single-lung transplant is contemplated. Because the lungs have the capacity to expand to fill chest cavities of significantly larger recipients, it is possible to use smaller donor lungs. In addition, it is possible to use large donor lungs and use lobes or trim portions of the parenchyma on the lungs to allow them to fill the chest cavity without impinging on cardiac function. Bronchial size match between donor and recipient is better correlated with height and age than with weight. Thus, most patients are listed for lung transplantation with a size range of 3 to 4 inches above and below the size of the recipient. However, heights of twice the size of the recipient can be considered if lobes are to be used from larger donors.

ORGAN PROCUREMENT

Flexible bronchoscopy is preferable at the time of lung procurement. The presence of direct trauma to the lungs or pulmonary contusions should be evaluated by direct inspection before the heart–lung or lung block is removed. The donor receives methylprednisolone and antibiotics and is heparinized before organ procurement. The technique of organ procurement from pediatric donors is similar to that of adult donors except that the volumes of cardioplegic and pulmonoplegic solutions are adjusted for the weight of the donor. Cardioplegic solution is given for a total dose of 30 cm^3/kg of donor weight and pulmonoplegia for 50 cm^3/kg donor weight. Antegrade crystalloid cardioplegic solution and Perfadex (extracellular low potassium dextran) pulmonoplegic solution has been used in most centers for organ preservation. Prostaglandin E_1 (500 μg) is injected directly into the main pulmonary artery at the time of cross-clamp. Lungs are topically cooled with ice slush solution with both pleural spaces widely opened.

When a heart–lung block is harvested for a single recipient, the cardioplegic solution is administered into the aorta and pulmonoplegic solution directly into the pulmonary artery with venting of the heart by division of the left atrial appendage. Division of the inferior vena cava at the diaphragm permits evacuation of cardioplegia without distention of the ventricle. The trachea is mobilized above the level of the carina and minimal dissection of the carina and lateral trachea is done. The lungs are then gently inflated to a pressure of approximately 20 mmHg and the trachea stapled. The superior vena cava is ligated and divided and the esophagus mobilized in the superior mediastinum and stapled and divided. The aorta is transected at the level of the innominate artery, and the distal aorta in the posterior pericardial space is mobilized and ligated and also divided. If additional aortic length is necessary, the aorta is not transected, but the arch vessels are divided individually. With incision of the pleura at the paraspinal region, bilaterally, the entire heart–lung block is then excised and the esophagus then removed from the specimen. If the heart and lung are to be harvested separately, cardioplegic and pulmonoplegic solutions are administered as in the combined heart–lung

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technique; however, the interatrial groove is developed and then the heart is eased by the division of the aorta and pulmonary artery, leaving the bifurcation of the pulmonary artery for the lung. The left atrium is then excised with a limited left atrial cuff, leaving as much as possible of the pulmonary venous confluence bilaterally for the lung implantation. The superior and inferior venae cavae are divided at the pericardial reflection. Many choose to use retrograde flush for the lungs at a dose of 5 to 10 ml/kg per pulmonary vein. The lung block is then excised as is the heart–lung block by the division of the trachea with gentle inflation of the lungs. The organs are then put in iced saline solution in sterile bags and transported to the recipient center on ice. Although some groups have used cardiopulmonary bypass for cooling of the entire heart–lung block, this technique is not widely used in the United States. Preservation times of >9 hours for lung transplantation have been successfully achieved with the use of these techniques.

RECIPIENT OPERATION

It is particularly important to take care in the removal of the recipient organs for heart–lung or lung transplantation to obtain absolute hemostasis in the mediastinum and pleural spaces and to avoid injury to the thoracic nerves including the phrenic, recurrent laryngeal, and vagus nerves. The bilateral thoracosternotomy or clamshell incision (Fig. 100.1) through the fourth intercostal space allows excellent exposure of the pleural spaces for takedown of adhesions and aids in obtaining hemostasis before implantation of the donor organs. When the clamshell incision is made, the incision needs to be carried as superiorly as possible in the midline so as to transect the sternum above the xiphoid to add stability of the sternal fragments when they are rewired after the procedure. Entrance in the fourth intercostal space is preferred; however, it is generally better to enter the chest more superiorly than inferiorly because the opening of the clamshell incision allows great exposure in the superior mediastinum. Many centers are now doing bilateral thoracotomies with separate implantation of each lung in patients who have adequate stability to undergo lung transplantation without the use of cardiopulmonary bypass. In implantation of heart–lung blocks, the sternotomy incision is generally preferred.

HEART–LUNG TRANSPLANTATION

The technical aspects of heart–lung implantation in children are similar to those used in adults. Cardiopulmonary bypass is used with bicaval and aortic cannulation while the lungs are excised bilaterally. Caval tapes are lowered around the cavae to eliminate venous return to the right atrium. The phrenic nerves are carefully protected on a pedicle of pericardium, which is mobilized before the organs are removed. The lungs are excised individually with the division of the mainstem bronchus, pulmonary artery and pulmonary veins after ligation, and division of the vessels. After the lungs have been removed, the heart is excised, leaving a cuff of right atrium and distal aorta for implantation of the new organs. A portion of the wall of the left pulmonary artery can be left in situ to avoid dissection around the recurrent laryngeal nerve on the left. After excision of the heart and each lung, the trachea is mobilized in the mediastinum behind the stump of aorta and divided approximately two rings above the carina. After meticulous hemostasis is obtained, the donor heart–lung block is placed in the mediastinum and the lungs passed behind

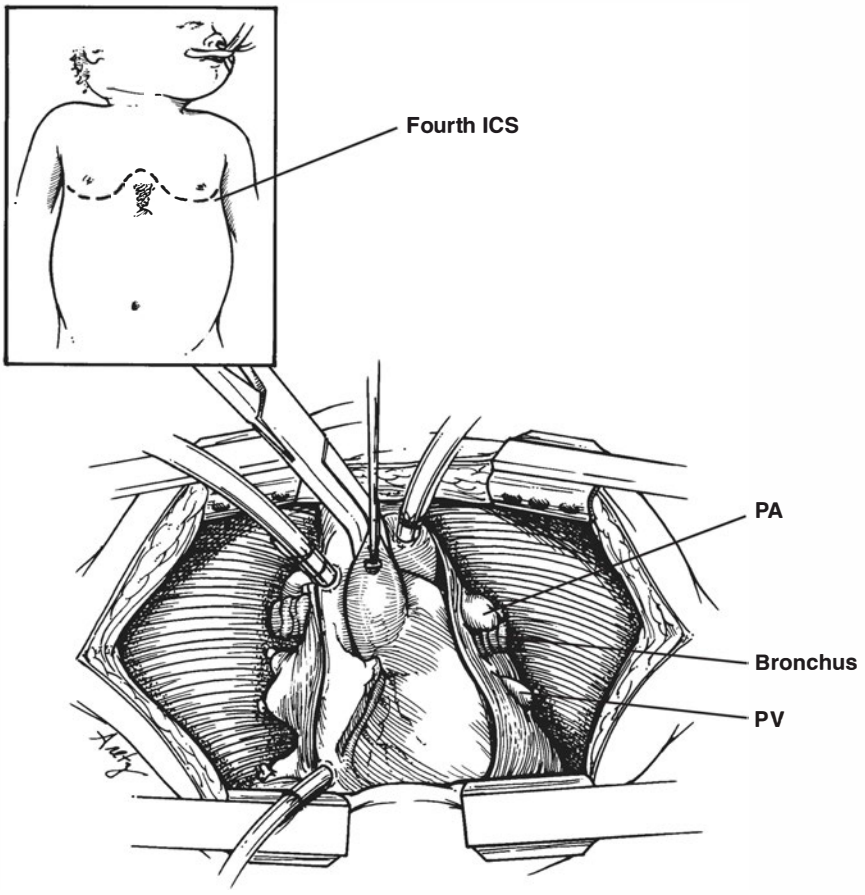


Fig. 100.1. The right and left lungs are removed sequentially with ligation of the branches of the pulmonary arteries and veins and stapling and division of the bronchus. The bronchial stump is left stapled, the bronchus is divided distal to the staple line, and the lungs are excised. The patient is on cardiopulmonary bypass, allowing removal of both lungs with the support of the circulation. As is illustrated, a cross-clamp can be applied, cardioplegia induced, and the cardiac repair performed before lung implantation. (**Inset**) The clamshell thoracosternotomy incision. The incision is carried transversely in the midline as high as possible to allow adequate sternum for sternal stability after rewiring with chest closure. The fourth intercostal space (ICS) is opened bilaterally and the incision carried out into the axillae. The incision in the chest can be carried more inferiorly because the trap door-like opening of the clamshell incision allows greater superior mobility than inferior mobility. It is, therefore, imperative not to enter the chest too high or dissection of the lungs will be impeded near the pulmonary veins and inferior pulmonary ligaments. PA, pulmonary artery; PV, pulmonary vein.

the pedicles of pericardium containing the phrenic nerves. Anastomoses between the donor and recipient trachea, aorta, and right atrium are then accomplished with the patient on cardiopulmonary bypass. In older children, the tracheal anastomosis can be accomplished with a running suture; however, our preference is to perform a running anastomosis of the membranous portion of the trachea and interrupted anastomosis of the cartilaginous portion anteriorly. We have used absorbable sutures in children in hopes of permitting better growth of the anastomoses. The aorta and right atrium are both anastomosed with running suture. In most cases, separate superior vena caval and inferior vena caval anastomoses are not used, and the right atrium of the recipient is excised with the heart at the time of recipient preparation. In very small children, however, caval anastomoses may lead to purse stringing of the suture line and obstruction and, therefore, atrial anastomoses are used in small infants. To prevent distention of the heart during reperfusion and while weaning from cardiopulmonary bypass, a vent is inserted via the amputated left atrial appendage into the heart.

BILATERAL SEQUENTIAL LUNG TRANSPLANTATION

The child is positioned supine and the shoulders elevated on a rolled towel. With this positioning, the entire chest can be prepared down to the axillae and draped as a sterile field (Fig. 100.1, inset). The transverse thoracosternotomy (clamshell incision) is made in the fourth intercostal space. The internal mammary pedicles are divided bilaterally. Chest retractors are placed in each thoracotomy incision, and the thymus is divided in the midline or resected completely if exposure of the superior mediastinum is poor. The pericardium is opened and the heart is suspended with pericardial stay sutures. After heparinization, the aorta and right atrium are cannulated for bypass. We prefer to use cardiopulmonary bypass in all lung transplant procedures in children because of the ease with which bypass permits removal of the recipient lungs and the fact that after removal of both lungs, the airway can be irrigated with antibiotic solution if necessary to decrease septic secretions. In addition, the removal of both the lungs before implantation decreases the ischemia time on the second implanted lung.

Separate caval cannulation is used if heart–lung transplantation or cardiac repair is contemplated in addition to lung transplantation. With the child on cardiopulmonary bypass, the lungs are removed bilaterally by ligation of the pulmonary artery branches, pulmonary venous drainage, and stapling of the right and left mainstem bronchi. Care is taken to dissect around the bronchial stumps as minimally as possible. The donor lung block is then brought to the field, and the right and left lungs are separated from the combined lung block by the division of the bronchus approximately two rings from the take-off of the upper lobe orifices bilaterally. The pulmonary arteries are trimmed to appropriate length for suturing to the recipient and the pulmonary venous confluence is divided by dividing the remnant of the left atrium between the right and left pulmonary veins. The left lung is usually implanted first because traction on the heart may be necessary to aid in exposure and once the lung is implanted, the heart can then recover from any injury while the right lung is implanted. The bronchial stump on the left is cut as short as practical; however, generally the length of the left mainstem bronchus at implantation is longer than the right mainstem bronchus. The bronchial anastomosis between the donor and recipient is then created with running absorbable suture for the membranous bronchus and interrupted simple absorbable sutures for the cartilaginous bronchus. An end-to-end anastomosis of the bronchus without telescoping of one bronchus into the other is used to prevent stenosis or malacia of the anastomosis. If a significant size discrepancy between the donor and recipient bronchus is present, then minor telescoping of the bronchial anastomosis is accepted. The pulmonary arterial anastomosis is then created with a partial occlusion clamp on the pulmonary artery of the recipient and a running suture is used to complete the anastomosis. The venous anastomosis is created in a similar fashion. Just before completion of the venous anastomosis, venous return is permitted to the heart, the arterial clamp is released, and air is aspirated from the veins before opening of the lungs for reperfusion. The same technique is then used for implantation of the right lung. On the right, the stump of the right mainstem bronchus is generally cut within two or three rings of the carina to have the shortest distance for revascularization of the anastomosis. Protection of the bronchial suture lines is accomplished with peribronchial tissues tacked over the anastomosis with absorbable suture if such tissue is available. However, with suitable trimming of the recipient bronchus, the suture line will be retracted into the mediastinal tissues such that good revascularization and protection of the anastomosis is available. We have abandoned the use of pericardial pedicle wraps or omentum for bronchial wraps in children, and the incidence of bronchial complications has not increased by using an unprotected anastomotic technique.

If cardiac repair is to be performed in association with bilateral sequential lung transplantation, the patient is cannulated for bypass, the lungs are removed as earlier noted, and the aorta is cross-clamped and cardioplegic solution administered. Closure of the intracardiac defects is performed through the right atrium if possible to prevent ventricular incisions in patients with significant pulmonary hypertension in whom right ventricular function may be depressed. After completion of the cardiac repair, reperfusion of the myocardium is permitted while the sequential lung implantation is performed.

SINGLE-LUNG TRANSPLANTATION

In children in whom only a single lung is required, the operation is performed either through a posterolateral thoracotomy on the transplant side or, more commonly, with the patient in a supine position and via a partial thoracosternotomy (Fig. 100.2). An anterior thoracotomy can be used, or the sternum can be crossed partially in the midline, allowing better access to the heart for cannulation and cardiopulmonary bypass. Closure of atrial septal and VSD can be readily accomplished with this technique. The patient is placed on cardiopulmonary bypass with caval cannulation through the right chest and direct aortic cannulation. With cardioplegia, the right atrium can be opened and the atrial septum patched or primarily closed and VSD approached across the tricuspid valve (Fig. 100.2). Explantation of the lung and implantation of the donor lung then proceeds as in bilateral sequential lung transplantation.

SPECIAL SITUATIONS

Congential pulmonary vein stenosis is an extremely rare condition in children but is associated with high mortality and is usually not amenable to direct surgical repair. These children often present with severe pulmonary hypertension and it is not uncommon for transcatheter attempts at stabilization, including balloon dilation and stent implantation in the pulmonary veins, to have been performed prior to

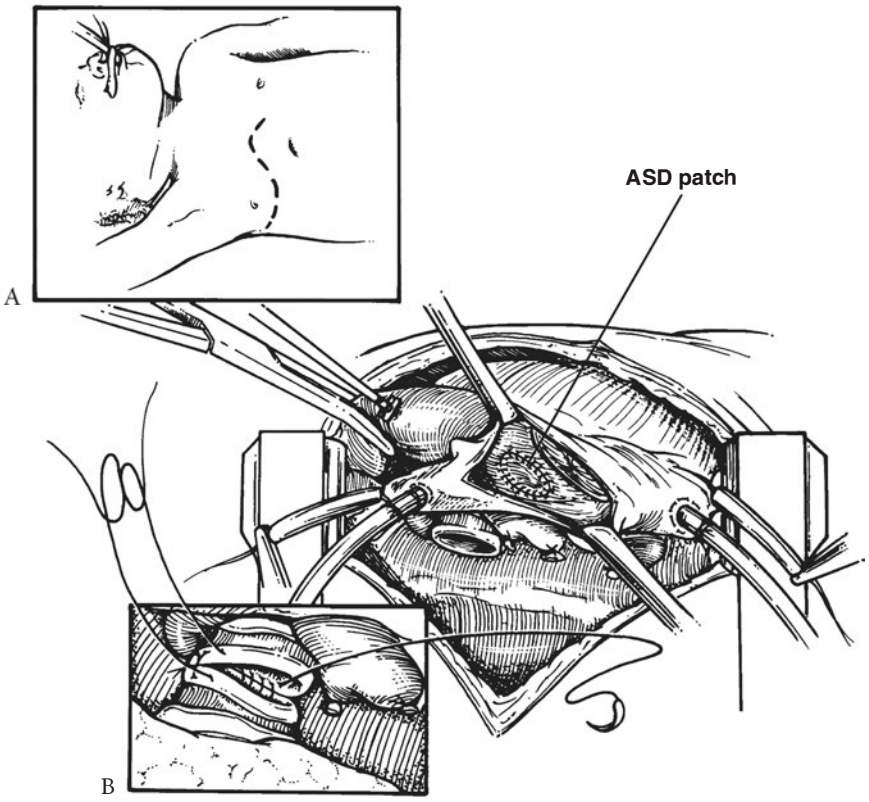


Fig. 100.2. The aorta is cross-clamped and bicaval cannulation with caval tapes is used. With cardioplegia or fibrillatory arrest, the right atrium can be opened and an atrial septal defect (ASD) closed primarily or patched. Ventricular septal defects can be approached across the tricuspid valve through this incision and closed with a polyester (Dacron) patch. The atrium is then closed, and the heart is allowed to reperfuse during implantation of the lung. (**Inset A**) For single-lung transplantation and repair of intracardiac defects of the atrial or ventricular septum, a right anterior thoracotomy crossing the sternum can be used. Again, the sternum is transected as high as possible to aid in sternal stability postoperatively. The fourth intercostal space or fifth intercostal space is then opened and the pleura entered. (**Inset B**) The bronchial anastomosis is created using a running absorbable suture for the membranous portion of the bronchus and interrupted simple sutures for the cartilaginous bronchus without telescoping. In this manner, an end-to-end anastomosis is created that allows the least chance for stenosis and granulation tissue at the suture line. No bronchial wrap is used, but peribronchial tissues may be tacked over the anastomosis if they are available.

referral for lung transplantation. In addition, interventional techniques have been used to stabilize patients while they are awaiting donor organs because this subgroup of children has the highest mortality under such circumstances. Because these children have often had stents implanted in the pulmonary veins, the implantation of the donor lungs is altered. At the time of lung implantation on cardiopulmonary bypass, it is necessary to excise the pulmonary venous confluence bilaterally including the excision of the stents, which often extend into the left atrium. This can be accomplished in either of two ways: (1) by implanting the right and left lungs with bronchial and pulmonary arterial anastomoses and then subjecting the patient to a period of aortic cross-clamping and cardioplegia while the pulmonary venous confluence is excised bilaterally and the pulmonary venous confluence of the donor lung is implanted directly into the left atrium with a running suture line bilaterally or (2) if possible by implanting both lungs and completing all anastomoses under a single period of cardioplegia. The anastomoses in lung transplantation in very small children are simple and often only require 15 to 20 minutes per lung implant, and therefore the total period of cardioplegia may not exceed 40 to 50 minutes for the entire procedure. We have used both of these techniques for implantation in patients with pulmonary vein stenosis with equal success.

In children in whom a patent ductus arteriosus is associated with severe pulmonary hypertension, it is advisable to place the patient on cardiopulmonary bypass and to divide and oversew the ends of the ductus arteriosus rather than ligate the ductus in situ. The elevation in pressure during ligation may result in enough pressure on the suture line to allow recanalization and division of the ductus ensures that recanalization will not occur.

A particular concern after cardiac repair at lung transplantation is the development of dynamic right ventricular outflow tract obstruction after the procedure. Many patients with elevated right ventricular pressure in the presence of shunt lesions such as VSD or a large patent ductus arteriosus may have severe right ventricular hypertrophy that creates dynamic outflow tract obstruction when the right ventricular pressure drops after successful lung transplantation. In severe cases, these patients may best be treated by patch augmentation of the right ventricular outflow tract, but in the majority of patients the avoidance of inotropic drugs postoperatively or use of beta-blocking agents may be sufficient to relieve the dynamic obstruction in the early postoperative period. In the majority of patients, this dynamic obstruction will resolve spontaneously after several days.

An unusual subgroup of children who may require lung transplantation comprises those with pulmonary atresia with VSD and nonconfluent pulmonary arteries. These children often have undergone multiple previous attempts at unifocalization operations to recreate a distal pulmonary vascular bed. Thus, many children will have had previous thoracotomies or median sternotomy incisions. In these children, typically the ascending aorta is markedly dilated and somewhat posteriorly located. Thus, the space available in the mediastinum for the placement of the pulmonary arteries is limited and placement behind the ascending aorta can result in compression of the right pulmonary artery by the dilated ascending aortic root. The operation is performed through the thoracosternotomy incision, as in bilateral sequential lung transplantation described earlier. The aorta is cross-clamped with the patient on bypass, and cardioplegia is administered. In this case, an incision is made in the right ventricular outflow tract to gain exposure to the VSD, which is closed with a polyester (Dacron) patch (Fig. 100.3). A pulmonary homograft conduit is then sutured to the superior margin of the ventriculotomy incision and augmented with a gusset of

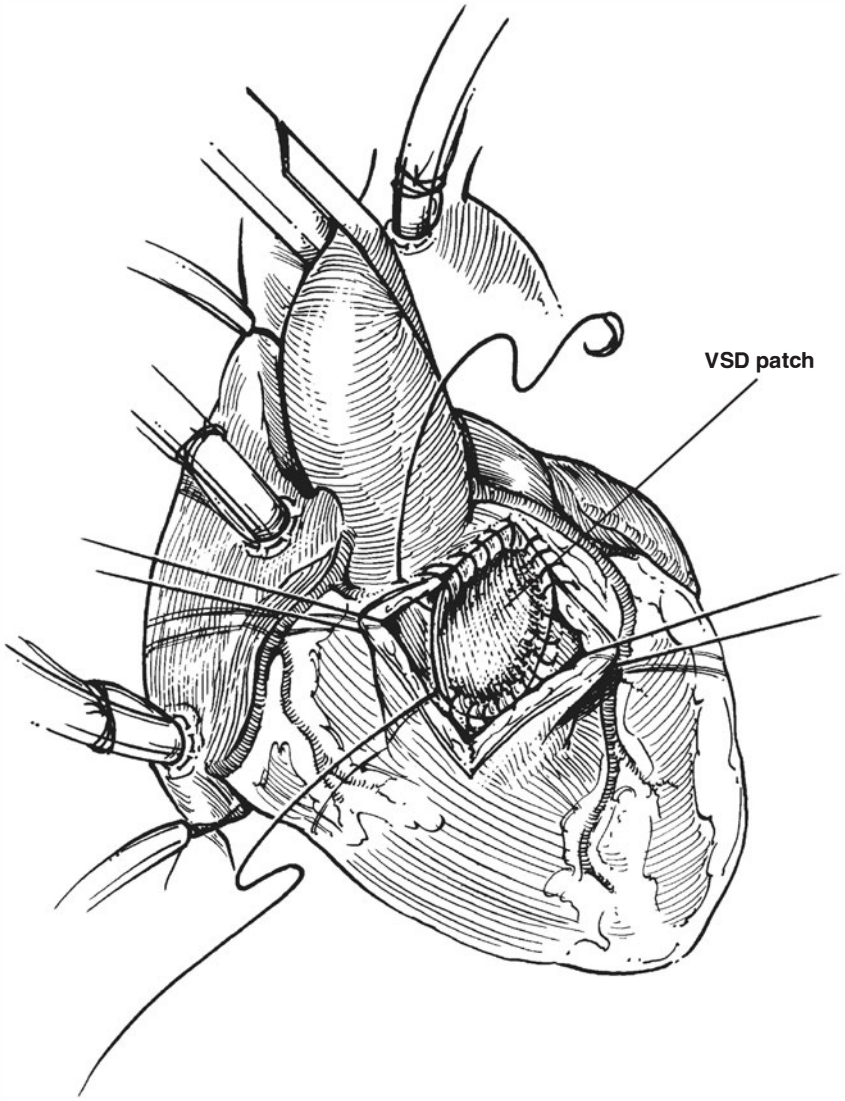


Fig. 100.3. Technique for cardiac repair and lung transplantation for pulmonary atresia with ventricular septal defect (VSD) and nonconfluent pulmonary arteries. With the heart on cardiopulmonary bypass and under diastolic cardioplegia, a right ventriculotomy incision is made vertically, avoiding major epicardial coronary branches. Through this defect, the VSD is exposed and patched with a polyester (Dacron) patch. Running or interrupted mattress sutures can be used for the placement of the patch, and in the posteroinferior aspect, care must be taken to avoid the conducting tissue if the defect is in the usually conoventricular location. Superiorly, the patch can be secured to the free margin of the ventriculotomy incision or to the conal septum. Care must be taken to ensure complete closure of the defect because reoperation for residual cardiac defects will be difficult in a patient on chronic immunosuppression.

polytetrafluoroethylene (PTFE) to allow a gentle take-off of the newly reconstructed right ventricular outflow tract from the right ventricle (Fig. 100.4). The remainder of the operation can then be performed with the heart reperfused and fibrillating or under continued cardioplegia. The lungs are then implanted sequentially as described earlier; however, the pulmonary artery anastomoses are performed last. After the bronchial and pulmonary venous anastomoses are completed, the pulmonary arteries are brought to the midline, either anterior or posterior to the ascending aorta. When brought anterior to the aorta, the pulmonary confluence is recreated in the middle with an absorbable suture posteriorly (Fig. 100.5). The reconstructed right ventricular outflow tract is then reconnected to the recreated pulmonary bifurcation with nonabsorbable suture because the homograft tissue is nonviable and growth is therefore not an issue (Fig. 100.6). After reconstruction, venous flow is returned to the lungs and air is evacuated through a vent in the left ventricle or atrium and from the ascending aorta to fully deair the heart before the patient is weaned from cardiopulmonary bypass.

LOBAR LUNG TRANSPLANTATION

Because suitably sized donor lungs are often not readily available, techniques have been devised to use lung grafts of reduced size (lobes) for smaller children. Cadaveric and living donor lobar transplants have now been performed with satisfactory results. The use of living donors for lobar lung transplantation has made donor organs more readily available for children with very unstable conditions who might not survive waiting for suitable cadaveric organs to become available. In addition, the use of cadaveric lobes from larger donors has expanded the donor pool for infants and children for whom identical-sized donors are rare. Because lung growth can continue to late childhood, we generally prefer to use lobes from pediatric cadaveric donors for infants and very small children in hopes that additional lung growth will continue. In older children, lobar transplants from adults may provide quite adequate alveolar volume even for continued growth despite the theoretical disadvantage of lack of additional donor lung alveolar growth. The indications for lobar lung transplantation and donor evaluation are similar to those for whole-lung transplant. Living donors, however, add additional concerns of donor evaluation. Both psychosocial and physiologic factors must be considered in these patients because the harvest of the lobe must result in the least possible morbidity to the donor. If bilateral lung transplantation is contemplated, the larger donor is generally preferred for donation of the right lower lobe because the total lung volume is lower in this lobe than the left lower lobe.

Technique of Donor Lobectomy

If lobes are used from larger cadaveric lungs, variations in lobar anatomy can be addressed at the time of dissection away from the operative field and the most suitable lobes identified for implantation into the recipient. In general, we prefer to use lower lobes for implantation because of the geometric advantages of the shape of the lower lobes bilaterally and the relatively consistent bronchial and vascular anatomy. Often the right upper lobe has multiple lobar arterial branches that require reconstruction to create a single pulmonary artery for anastomosis to the recipient.

The right middle lobe is a relatively small amount of lung tissue for implantation and has variable venous drainage, which may complicate venous anastomosis and dissection. The use of a middle lobe from an adult to a young child or infant

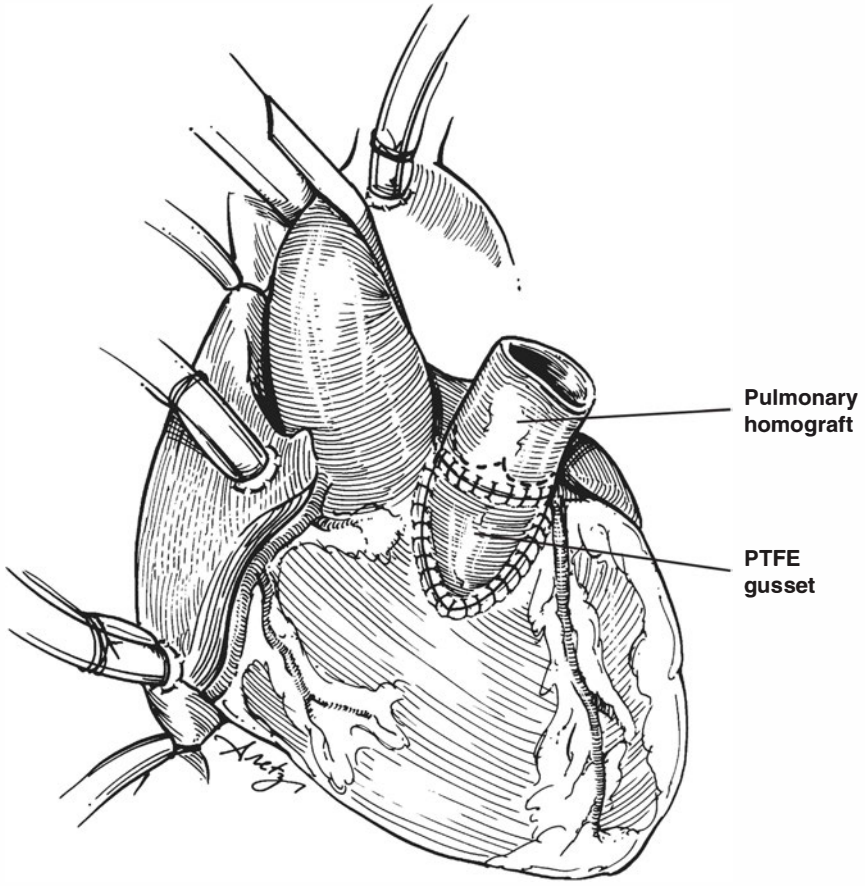


Fig. 100.4. After closure of the ventricular septal defect, a pulmonary homograft of the largest size that will fit in the chest cavity to allow for adequate growth of the recipient is selected and secured to the superior margin of the ventriculotomy incision with nonabsorbable suture. A gusset of polytetrafluoroethylene (PTFE) is used to create a gentle take-off of the conduit from the right ventricle. Care must be taken to obtain adequate hemostasis of this suture line to avoid late pseudoaneurysm formation.

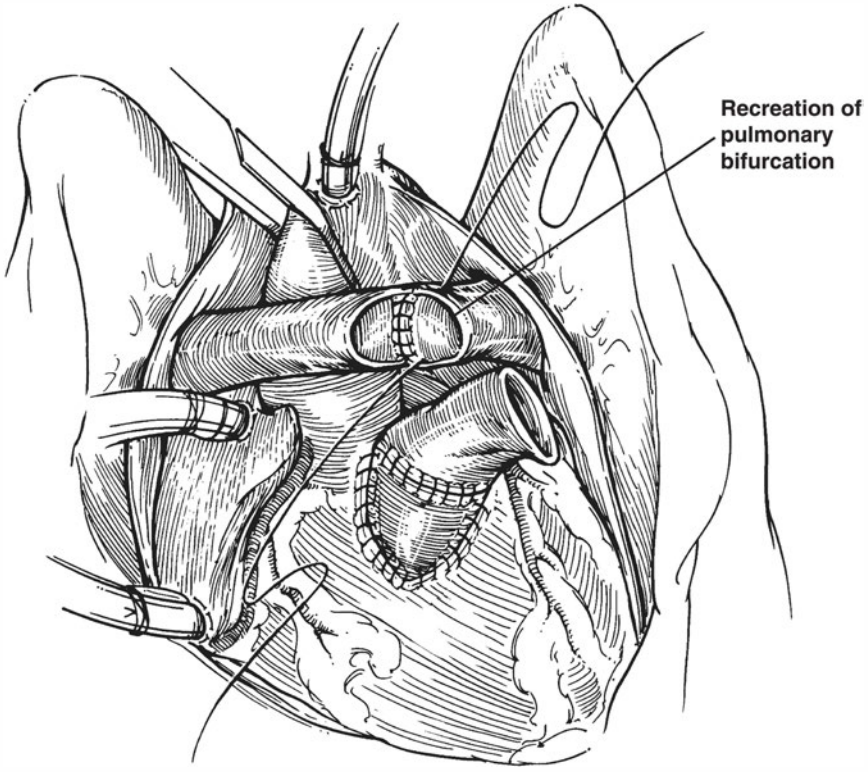


Fig. 100.5. If the aorta is large and posteriorly located, the pulmonary arteries are brought anterior to the aorta for reconstruction. The lungs are implanted bilaterally with bronchial and venous anastomoses and then the pulmonary arteries are brought anterior or posterior to the phrenic nerves to the ascending aorta and reapproximated in the midline.

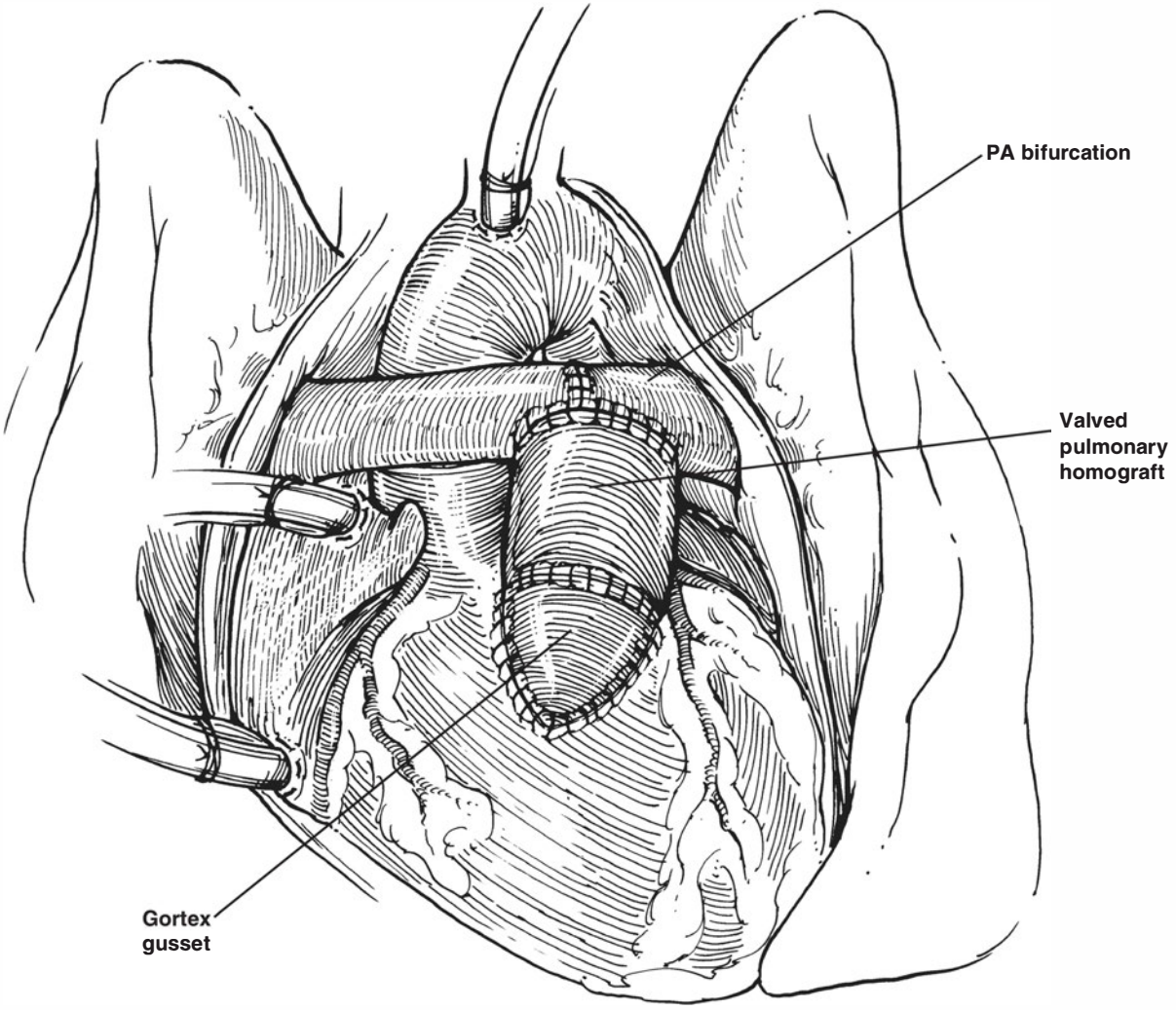


Fig. 100.6. The reconstruction is completed by anastomosis of the pulmonary homograft reconstruction of the right ventricular outflow tract to the newly reconstructed pulmonary confluence anterior to the aorta. PA, pulmonary artery.

might be associated with poor lung growth because the addition of alveolar number is unlikely to occur from an adult lung. For these reasons, we prefer to use lower lobes in most patients.

The technique of donor right lower lobectomy is shown in Figure 100.7. A posterolateral thoracotomy is performed and the inferior pulmonary ligament divided with electrocautery. Dissection in the fissure between the upper and lower lobes is created to identify the branches of the pulmonary artery to the right lower lobe and to define take-off of the right middle lobe pulmonary artery branch. The pericardium is opened around the inferior pulmonary vein and the venous drainage from the middle lobe identified to ensure that it does not enter the inferior pulmonary vein directly. After heparinization, a vascular clamp can be placed below the take-off of the right middle lobe pulmonary artery, leaving a stump of pulmonary artery adequate for anastomosis to the recipient. A vascular clamp can also then be placed on the inferior pulmonary vein at its entrance to the left atrium with dissection in the interatrial groove to allow a suitable stump of tissue for oversewing of the venous entrance in the donor and leaving an adequate stump of pulmonary venous confluence for anastomosis in the recipient. The vessels are then divided, and the remaining fissures are divided with the stapler. After division of the pulmonary artery, the bronchus to the right lower lobe is dissected as minimally as possible to preserve blood supply to the lobe. The middle lobe bronchus is identified, and the bronchus is divided with a scalpel. The donor lobe is then removed and taken away from the operative field for preparation and preservation. The pulmonary artery and left atrium are closed with running nonabsorbable suture, and the bronchus is closed with a stapler if adequate length is available or with simple nonabsorbable sutures. A pleural flap can be used to cover the bronchial stump if necessary.

The technique of donor left lower lobectomy is illustrated in Figure 100.8. Again, the inferior pulmonary ligament is divided and the pulmonary artery is dissected within the fissure to identify the lingular and lower lobe branches. The pericardium is opened around the inferior pulmonary vein and fissures are completed with a stapler. With the patient heparinized, a clamp is placed on the pulmonary artery proximal to the take-off of the superior segment of the lower lobe. Depending on the number and location of the lingular branches one or more may need to be sacrificed in order to have sufficient length of donor pulmonary artery to facilitate the anastomosis in the recipient. The pulmonary vein is clamped and divided, and then the bronchus to the left lower lobe is exposed behind the pulmonary artery. Once the lingular bronchus is identified, the

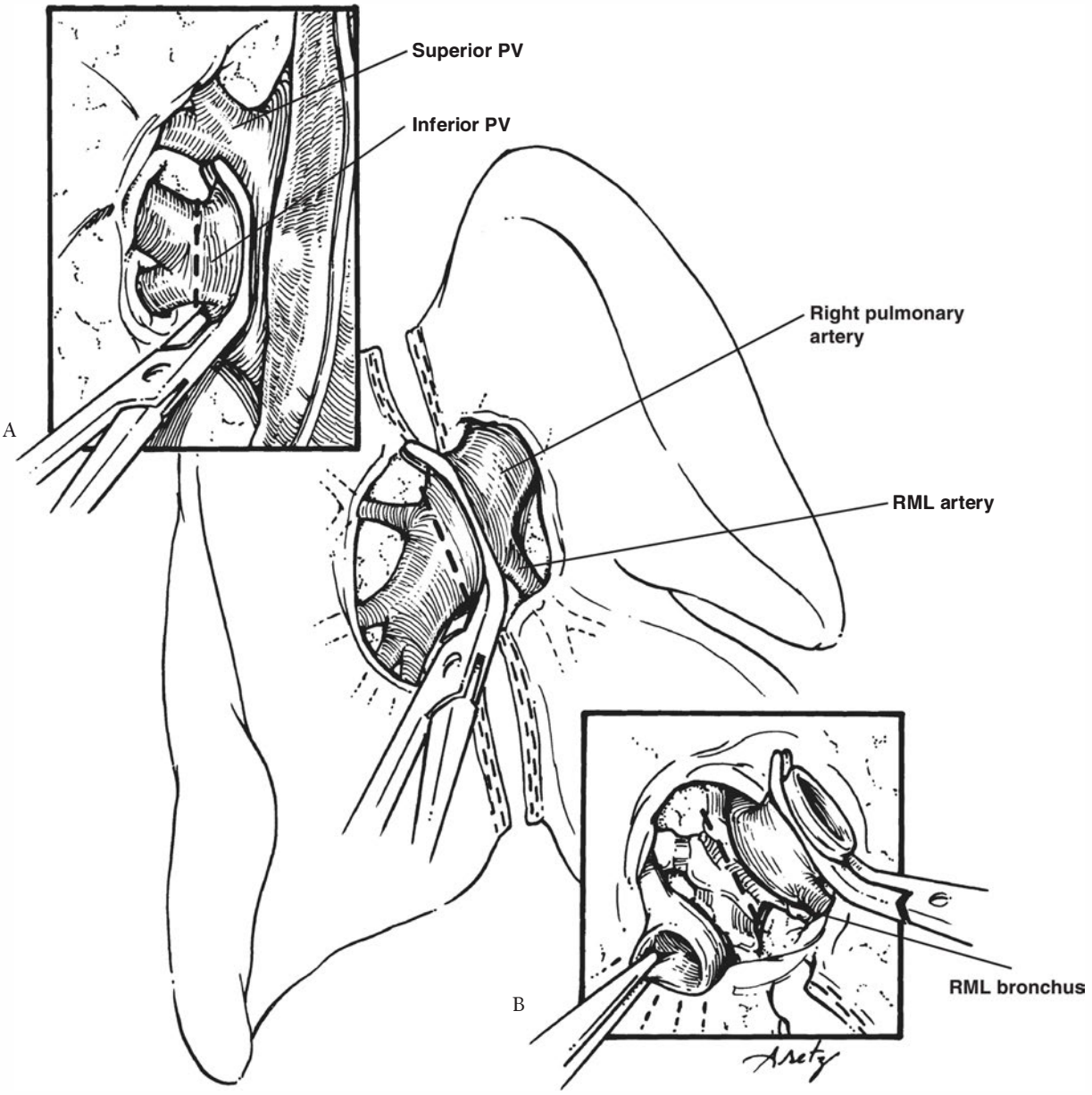


Fig. 100.7. Technique for dissection and excision of the right lower lobe for living donor lobectomy. The pulmonary artery is exposed in the fissure, and dissection is completed to allow identification of the right middle lobe (RML) pulmonary artery. Mobilization of the pulmonary artery is then performed so that a vascular clamp can be applied just distal to the take-off of the middle lobe vessel. The fissures are completed with staplers and before placement of clamps. (**Inset A**) The pulmonary venous dissection is created by takedown of the pericardial reflection and isolation of the right lower lobe pulmonary venous confluence. Care must be taken to identify the middle lobe vein entrance into the upper lobe bifurcation or the superior aspect of the lower lobe and to protect this venous drainage if at all possible. Heparinization of the donor is then performed, a clamp is placed on the pulmonary artery and pulmonary vein confluence, and the artery and vein are divided. (**Inset B**) With retraction of the stumps of the pulmonary artery, the bronchus is identified and divided just distal to the take-off of the middle lobe bronchus. The donor lobe is then removed for preservation, and the vessels and bronchus are oversewn. PV, pulmonary veins.

main bronchus is transected obliquely with care taken to include the superior segment bronchus in the donor lobe, leaving a 2 to 3 mm rim of bronchus above the take-off of the superior segmental lobar branch. Often the takeoff of the superior segmental bronchus is quite proximal and care must be taken to divide the bronchus so that the superior segmental bronchus is not separated from the basal trunk. The pulmonary artery is then oversewn with nonabsorbable suture, and the bronchial stump is closed with interrupted nonabsorbable suture and covered as in the right donor lobectomy as necessary.

Preservation of the living donor lobes is performed by infusion of pulmonoplegic solution in the pulmonary artery until the venous effluent is clear. Gentle inflation can be used during flushing to aid in evacuation of any residual blood. Prostaglandin E_1 may either be added to the pulmonoplegic solution or infused in the donor before lobectomy. Because of the anticipated short ischemia time on donor lobes, gentle inflation and clamping of the bronchus during transport can be used, or the lungs can be left uninflated during the short period of transportation to the recipient.

Recipient Operation

The recipient operation for implantation of donor lobes is similar to the complete lung implantation technique. The short length

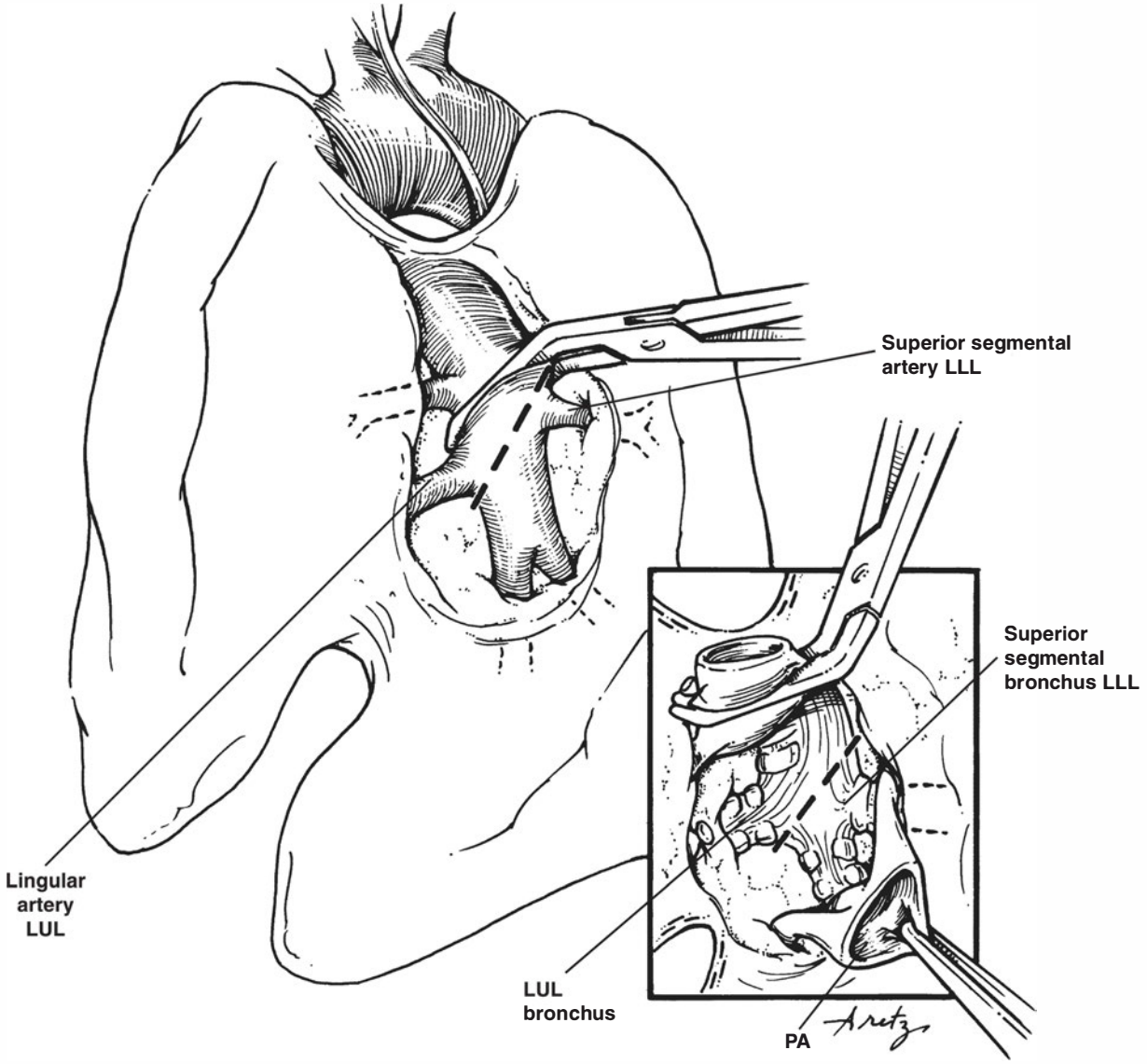


Fig. 100.8. Technique for the preparation of left lower lobe (LLL) living donor lobectomy. The left pulmonary artery (PA) is identified in the fissure and mobilized, and the take-offs of the superior segmental branch of the lower lobe and the lingular branches are identified. After completion of the fissures with staplers and dissection of the pulmonary veins, the patient is heparinized, and a vascular clamp is placed on the PA above the take-off of the superior segmental branch of the lower lobe. Small lingular branches may be ligated or, as illustrated, left in situ and an oblique incision made just distal to the take-off of the lingular branch and obliquely carried superior to the take-off of the superior segmental branch. The PA can then be patched or oversewn with care taken to maintain perfusion to the lingular branch of the PA. The bronchus is then exposed behind the PA and an oblique incision made, keeping the origin of the superior segmental bronchus with the donor lobe and the lingular bronchus with the upper lobe. LUL, left upper lobe.

of the pulmonary venous confluence from single lobes makes it valuable to leave longer stumps of pulmonary veins in the recipient to gain length for the anastomosis. If proper organization and timing of donor and recipient operations is performed, it is possible to limit the ischemia time on each donor lobe to <45 minutes.

POSTOPERATIVE MANAGEMENT

Postoperative management and restriction of postoperative fluid administration are important after heart–lung and lung transplantation to attempt to attenuate the magnitude of reperfusion edema of the transplanted organs. Immediate postoperative issues are graft function, control of bleeding, and hemodynamic stability. Isoproterenol is often used in heart–lung transplant recipients to maintain the cardiac rate from 110 to 150 beats per minute. The isoproterenol may also lower pulmonary vascular resistance post transplant. The effects of immunosuppression agents such as cyclosporine and tacrolimus on renal function may be minimized by the use of diuretics and low-dose dopamine. Weaning of the fraction of inspired oxygen (FiO_2) is performed to maintain a PaO_2 of >70 mmHg and low amounts of positive end-expiratory pressure are used to prevent water accumulation in the lungs and atelectasis. We generally perform a fiberoptic bronchoscopic examination of the anastomosis 12 to 24 hours after lung transplantation to assess the blood supply and suction of any residual secretions before extubation of the patient. In addition, a thoracic epidural catheter for analgesia is placed prior to extubation.

It is generally advisable to keep children with severe pulmonary hypertension and congenital heart disease sedated and paralyzed for 12 to 24 hours after transplant

to minimize hemodynamic instability. Patients who are chronically debilitated or have been chronically ventilated, pretransplant may require prolonged weaning after successful transplant procedures. If a patient had a tracheostomy preoperatively, this is usually replaced within 12 hours of surgery. Early reperfusion damage of the transplanted lungs, which is generally manifested by a diffuse pulmonary infiltrate on chest X-ray films, usually resolves after 5 to 7 days post transplant. Often, copious serous drainage from chest cavities may occur representing transudative fluid loss across the visceral pleura or exudative fluid loss from takedown of extensive intrapleural adhesions. Early mobilization is encouraged to aid in re-expansion of the transplanted lungs and surveillance cultures of sputum along with bronchoalveolar lavage cultures are performed as indicated. Immunologic staining of donor and recipient lung samples for common viruses has been performed to identify early potential viral infections of the transplanted lung. In our experience, most of the early graft dysfunction after lung transplantation is associated with positive viral cultures of the donor organs, which are particularly common in pediatric donors. It has been noted that patients after lung transplantation have a significant incidence of gastroesophageal reflux and aspiration, which may damage the donor lungs and increase the risk of late development of bronchiolitis obliterans. Therefore, in some centers, routine gastroesophageal reflux procedures are performed in the postoperative period to limit the risk of chronic aspiration.

The immunosuppressive regimen used for heart–lung and lung transplantation is similar to that used for pediatric heart transplants. No standardized regimen has been developed. We have primarily used tacrolimus and mycophenolate mofetil with steroids in our most recent pediatric experience. For lung transplant specifically, anti-thymocyte globulin has been used intraoperatively prior to reperfusion as induction immunosuppression with favorable results decreasing the incidence of acute cellular rejection. For heart–lung and lung transplants, ganciclovir administered for a 6-week period post transplant is used to prevent early CMV infection. Oral nystatin is used to reduce the likelihood of oral candida while on immunosuppression. It is yet unclear if there is any significant superiority of one immunosuppression protocol over another. However, we have used tacrolimus to make dosing and administration easier for children who may require multiple doses of cyclosporine to maintain adequate drug levels and for cystic fibrosis patients, who may have difficulty with gastrointestinal absorption. Common complications of cyclosporine include hirsutism, seizures and gum hyperplasia in addition to renal dysfunction. These complications are somewhat lessened by the use of tacrolimus. Steroids have been associated with the onset of diabetes but we have not noted an increased incidence of infection with steroid use. Although weight gain has been appropriate in small children who have undergone lung or heart–lung transplantation, bone growth may be somewhat decreased by the use of immunosuppressive regimens that include steroids.

REJECTION SURVEILLANCE AND MANAGEMENT

Bronchoscopy is used even in small infants and children for rejection and infection surveillance. Bronchoalveolar lavage fluid is sent for culture on a routine basis, and biopsy specimens are obtained either through a fiberoptic bronchoscope or though a suction catheter positioned in the distal airways under fluoroscopic guidance if the airways are too small to allow an adequate sized fiberoptic bronchoscope to pass into the distal airways. Significant documented rejection is treated by pulse steroids and increases in dosage of tacrolimus or cyclosporine as necessary.

Refractory rejection can be treated with cytolytic agents such as OKT3 if the patient does not rapidly respond to an increase in steroid dose. Echocardiography is used to evaluate cardiac function in children after heart–lung transplantation. Differential rejection of heart and lungs is well documented; however, cardiac rejection appears to be rare in the absence of associated pulmonary rejection. In addition, the incidence of cardiac rejection in cardiopulmonary transplant is lower than in cardiac transplant alone. Thus, in the absence of evidence for pulmonary rejection, cardiac biopsies are not routinely performed except for when echocardiographic findings are concerning for rejection.

RESULTS

Early postoperative complications are uncommon. The incidence of airway stenosis is approximately 10% and can be treated conservatively with serial balloon dilation. True airway dehiscence occurs rarely and is managed emergently. Nerve injury resulting from the explantation of the native lungs is more frequent with phrenic nerve injury occurring approximately 15% of the time. It is managed conservatively in most cases and often resolves within 6 months of surgery. Recurrent laryngeal nerve injury occurs 10% of the time, rarely resulting in respiratory difficulty but potentially augmenting the possibility of aspiration.

Relatively few results of cardiopulmonary or pulmonary transplantation have been reported in children. However, the overall reported rates to the Registry of the International Society of Heart and Lung Transplantation show a significant increase in the number of pediatric lung transplants performed over the last decade, particularly in the 12 to 17 age range while there have been significantly fewer heart–lung transplants. For heart–lung transplant, there has been no increase in survival over the past 30 years. The average half-life survival in heart–lung recipients is approximately 3 years for those patients >1 year of age. For infants <1 year of age, however, only half of the patients survive >2 years. Excluding those patients who die within the first year post transplant, average survival is approximately 6 years. Survival is poorest in those patients transplanted because of complex congenital heart disease compared with pulmonary hypertension. Lung transplantation survival statistics in pediatric populations have improved significantly, particularly due to improved early survival. After the first year post transplant, survival in pediatric lung transplant has not changed significantly in decades. The half-life is best in older children at 12 to 17 year range surviving to 4.3 years. Early 1- and 5-year survival rates for recipients transplanted in the recent decade are 83% and 50%. As expected, the initial operative mortality is higher in children requiring pulmonary transplantation and cardiac repair and in patients with pulmonary hypertension. The long-term results are improved after initial operative mortality is excluded from the analysis.

The major obstacle to long-term survival after lung transplantation in children is the development of bronchiolitis obliterans, which may be a manifestation of chronic rejection. It is most commonly seen in older patients and in patients >5 years from transplant. The onset of bronchiolitis obliterans is associated with progressive dyspnea and reduction of oxygen saturation on room air plus a documented decrease in FEV_1 or in forced expiratory flow over mid-expiration ($FEF_{25–75}$). The clinical diagnosis of bronchiolitis obliterans may be considered if there is a sudden decrease in FEV_1 of 20% or greater from the maximum post transplant baseline level, unassociated with infection, acute rejection, or bronchial complication. Although some patients have had stabilization of the level of bronchiolitis obliterans with increased immunosuppression with

addition of an antilymphocytotoxic agent, such as OKT3, in other children a progressive decline in lung function has occurred that either required retransplantation or resulted in late mortality. In heart–lung transplant recipients, the onset of bronchiolitis obliterans may occur independent of the development of coronary graft atherosclerosis or chronic rejection of the transplanted heart. The majority of malignancies seen in pediatric transplant patients is related to post transplant lymphoproliferative disorder (PTLD), secondary to EBV, occurring in approximately 15% of patients within 5 years of transplant. PTLD is treated with reduction of immunosuppression and Rituximab with additional chemotherapy as needed.

CONCLUSION

Lung and heart–lung transplantation can be performed safely and successfully in patients who have no other therapeutic options. Donor availability remains limited and waiting list mortality high. Although results are encouraging, obliterative bronchiolitis remains a serious late concern that must be addressed for the future success of cardiopulmonary transplantation in children. As increasing experience has been gained in cardiac repair and lung transplantation and the use of reduced-size donor grafts, the technical limitations to cardiopulmonary and pulmonary transplantation have been largely eliminated. The immunologic issues related to late development of chronic rejection and bronchiolitis obliterans, however, make cardiopulmonary and pulmonary transplantation a palliative procedure for the majority of recipients.

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EDITOR'S COMMENTS

Drs. Fuller and Spray have addressed heart and lung transplantation in children. Dr. Spray has had a significant experience in transplantation and certainly is an appropriate expert in this subject. The authors have clarified the major differences and indications for lung transplantation between adults and children. The CHOP group uses cardiopulmonary bypass for bilateral lung transplantation and often use cardioplegia to simplify the pulmonary venous anastomoses. I think they have clearly demonstrated the efficacy of using such adjuncts.

As they have stated, the difficulty is getting appropriate number of lungs for children for transplantation. Although living donor transplantation has been used successfully, it is difficult to find appropriate adult donors for bilateral lung transplantation. I think the future of this endeavor will be to make certain that every possible lung is available for transplantation including the use of deceased donors. EVLP has provided a method for conditioning damaged lungs. The Toronto group has popularized this technique and we have studied this extensively in the laboratory. There is no doubt in my mind that a day will come when virtually every potential donor lung will be conditioned and then used for transplantation. This will certainly help alleviate the organ donor shortage and increase the use of this very scarce resource.

ILK

101 Ebstein's Malformation of the Tricuspid Valve in Children

Sitaram M. Emani and Pedro J. del Nido

Ebstein's malformation refers to a specific congenital defect of the tricuspid valve (TV) in which displacement of the hinge points of the septal and posterior leaflets toward the apex results in "atrialized" portion of the right ventricle (RV) and varying degrees of tricuspid regurgitation. The functional RV beyond the tips of the abnormal leaflets may be small, thin-walled, and hypokinetic or dyskinetic. The preoperative assessment, timing of intervention, and techniques for TV repair are discussed. Although an "ebsteinoid tricuspid valve" may also accompany L-transposition of the great arteries, the management of this entity is not discussed in further detail.

PREOPERATIVE PLANNING

Severity of Leaflet Abnormality

Ebstein's malformation can be categorized according to the severity of apical displacement and hypoplasia of septal and anterior leaflets, as well as by the size of atrialized chamber. Echocardiography and magnetic resonance imaging (MRI) are typically used to categorize the malformation into types A–D, with type A describing very mild displacement of the septal leaflet, a large "sail-like" anterior leaflet, and a small atrialized chamber. Type D describes near absence of functional tricuspid leaflet tissue and severe apical displacement of hinge points of both anterior and septal leaflets, resulting in a massive atrialized chamber, and orientation of the inflow orifice toward the RV infundibulum. The categorization is useful in surgical planning, as patients with more severe forms of the malformation are more likely to require leaflet substitution with prosthetic material and RV unloading with cavopulmonary shunting.

Atrial Septal Defect

In patients with Ebstein's malformation and atrial septal defect, the direction of flow across the defect indicates relative compliance of the downstream ventricles. Gradual worsening of cyanosis heralds the decline in RV compliance and function. Catheter-based closure of the atrial septal defect in these patients may alleviate cyanosis but does not improve long-term prognosis related to ongoing RV volume overload and progressive RV failure.

Pulmonary Valve Stenosis or Atresia

Fetal or postnatal echocardiogram may demonstrate pulmonary atresia, but imaging may not reliably distinguish functional versus anatomic atresia in a fetus or infant with a patent ductus arteriosus (PDA) and significant left to right flow. The presence of pulmonary regurgitation suggests functional rather than anatomic atresia. The distinction is clinically relevant since the patient with anatomic atresia is dependent upon the PDA for pulmonary blood flow, whereas ductal closure may be the desired treatment for a neonate with functional atresia since interruption of the PDA restores antegrade flow across the pulmonary valve. Both functional and anatomic atresia are risk factors for mortality in neonates with Ebstein's malformation. Similarly, RV outflow tract obstruction is a risk factor for poor outcomes in non-neonates undergoing repair.

Right Ventricular Function and Mass

MRI can be used to estimate the volume and wall thickness of the functional RV in the evaluation of Ebstein's malformation beyond infancy. A dilated and thin-walled RV may not tolerate isolated TV repair, and unloading maneuvers, such as cavopulmonary shunting, may be necessary adjuncts. In the evaluation of the neonate or fetus, Doppler estimation of the RV pressures allows risk stratification. Although low-RV pressure is almost universally present in children and adults with Ebstein's malformation, estimated RV pressures by echocardiography of <20 mmHg may be a sign of inadequate RV function in neonates.

Inducible Atrial and Ventricular Arrhythmias

Preoperative electrophysiologic studies are helpful at identifying the presence of accessory pathways or substrate for atrial or ventricular arrhythmias. Although ablation of accessory pathway can be performed in the catheterization laboratory, the presence of inducible atrial arrhythmias is an indication for intraoperative cryoablation–MAZE in a patient undergoing TV repair. The presence of inducible ventricular arrhythmias should raise consideration for intraoperative epicardial defibrillator coil placement, thus avoiding subsequent need for transvenous lead placement across the TV.

SURGICAL INDICATIONS AND TIMING

Operative intervention for Ebstein's malformation typically follows a bimodal distribution, with initial phase occurring in neonates and infants, followed by a phase that occurs in adolescence and adulthood. The indications for operation, the type of procedure performed, and the operative mortality vary significantly for these two groups.

Neonates

Prenatal diagnosis of Ebstein's malformation has allowed improvements in immediate postnatal management of this disease. Risk stratification prenatally can be performed by the assessment of RV size and function as well as by the measurement of RV pressures and direction and degree of flow across the PDA and right ventricular outflow tract. In severe forms of Ebstein's malformation, the RV pressure estimated by Doppler velocity of tricuspid regurgitation jet is low (<20 mmHg), suggesting poor systolic function of the RV. Presence of anatomic pulmonary atresia is a risk factor for mortality in neonates with Ebstein's malformation. Antegrade pulmonary blood

flow across a patent pulmonary valve may not occur in the presence of a PDA, resulting in functional pulmonary atresia. In a subset of patients with functional atresia and adequate RV systolic function, antegrade flow may resume following ductal closure. A patient with PDA and pulmonary regurgitation may present with extreme circulatory failure, and expeditious ligation of the ductus arteriosus abrogates the circular shunt. A sternotomy approach for ligation of the PDA allows access for cardiopulmonary bypass in case of circulatory collapse from inadequate right heart structures. A neonate with Ebstein's malformation may present with circulatory failure or extreme cyanosis, warranting surgical intervention. In these patients, biventricular repair with TV repair alone carries a high mortality, and single-ventricle palliation with RV exclusion should be considered.

Adolescents and Adults

A patient who survives the neonatal period may remain relatively asymptomatic for years, even with significant tricuspid regurgitation. Indications for intervention include development of symptoms—specifically exercise intolerance, cyanosis, or arrhythmias. Timing of repair in the asymptomatic patient is controversial, but the presence of left ventricle compression, progressive right atrial (RA) or RV dilation by noninvasive imaging studies may herald onset of symptoms.

SURGICAL TECHNIQUES

Several techniques have been described for TV repair. Original procedures developed for this abnormality involved posterior tricuspid annuloplasty and transverse or longitudinal plication of the atrialized portion of the RV, thereby bringing the true annulus to the level of the functional annulus. A significant portion of these patients subsequently require TV replacement. The operation described by da Silva et al. termed "cone reconstruction" has been most recently adapted by many pediatric centers. The procedure bears similarities to the technique described in 1988 by Carpentier, but differs in the extent of mobilization and rotation of leaflet apparatus. The components of the tricuspid repair include leaflet reconstruction, RV reduction by plication or resection, RA reduction, and creation of atrial lesions to prevent arrhythmias. Augmentation of deficient leaflets and papillary muscle repositioning may be necessary as well. Adjunct procedures include maintenance of a patent foramen ovale, creation of superior cavopulmonary shunt, and replacement of the pulmonary valve.

"Cone" Reconstruction of Tricuspid Valve

The goal of leaflet reconstruction is to utilize redundant valvular tissue from the anterior and posterior leaflets of the TV to re-create a new septal leaflet, which is appropriately located at the true annulus of the TV. The TV in Ebstein's malformation may contain adequate surface area of leaflet material resulting from a large annular diameter and anterior leaflet circumference. Regurgitation results from maldistribution of leaflet material, such that the height of the posterior and septal leaflets is inadequate to coapt centrally with the anterior leaflet. The goal of the cone reconstruction is to redistribute existing leaflet tissue to the true annulus in the posterior and septal regions. Edge-to-edge reconstruction of leaflet material yields a circumferential leaflet of adequate height but smaller circumference, which allows for central coaptation. The leaflet reconstruction can be subdivided into discrete phases.

1. *Detachment of leaflets and delamination of leaflet material.* A circumferential incision is made in the anterior leaflet near its hinge point to the annulus, beginning at the anteroseptal commissure, extending clockwise onto the posterior leaflet and further onto the septal leaflet if present. Identification of the true annulus may be challenging due to tethering of leaflet tissue to the ventricle. In patients with type D malformation, there may be failure of delamination of the anterior leaflet tissue from underlying RV, giving an echocardiographic appearance of absent leaflet tissue. However, careful examination reveals distinct layer of leaflet tissue that can be delaminated from the RV mass. The distinction between leaflet and annulus is most obvious in the mid-portion of the anterior leaflet, and therefore, a circumferential incision is started at this point and carried in the clockwise and counterclockwise directions (Fig. 101.1). The hinge point to the annulus becomes easier to identify once delamination has been initiated on the anterior leaflet. Care must be exercised during delamination to avoid injury to the underlying myocardium or leaflet material itself. The leaflet detachment process is continued clockwise onto the posterior leaflet and septal leaflets, harvesting as much usable leaflet tissue as possible (Fig. 101.2). By delamination of fused leaflets, sufficient quantities of leaflet material can be frequently harvested to create a new TV without addition of prosthetic material.

2. *Division of tethering secondary chords and muscle bundles.* To allow adequate mobility of the leaflets for a cone-type reconstruction, the secondary chordal attachments and tethering muscle bundles must be mobilized down to the apex of the RV. These can be adequately visualized once the base of the leaflet has been detached from the tricuspid annulus (Fig. 101.3).

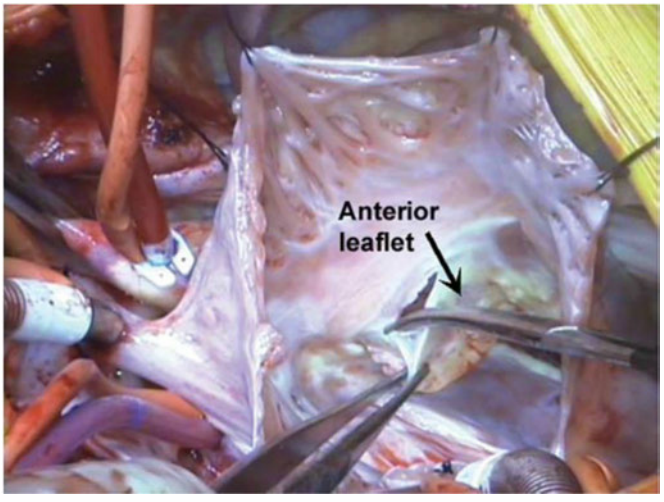


Fig. 101.1. Leaflet mobilization begins with detachment of the anterior leaflet at the level of the annulus. The circumferential incision at the base of the leaflet is carried counterclockwise to the level of the anteroseptal commissure.

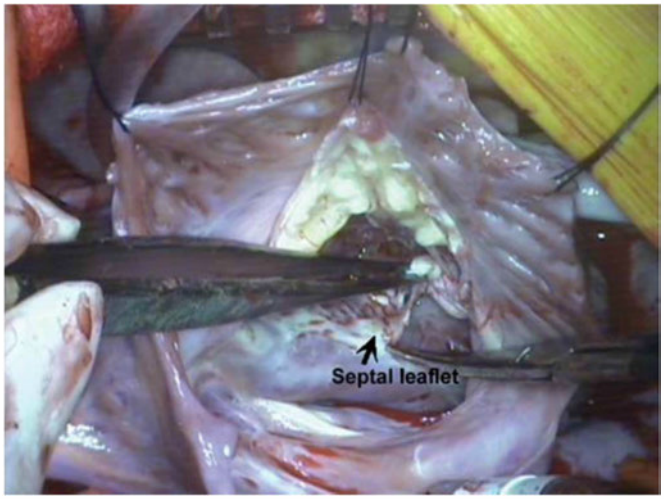


Fig. 101.2. The incision in the anterior leaflet is continued clockwise onto the posterior and septal leaflets. The septal leaflet may be fused to the underlying myocardium and require careful delamination.

Primary chordal attachments must be identified and preserved during this process, but inadequate excavation of tethered papillary muscles may result in residual regurgitation following repair. Repositioning the anterior leaflet papillary muscle toward the septum either by transection and re-implantation or suture approximation of the anterior and septal papillary muscles is an adjunct to papillary muscle mobilization, which prevents distraction of the tips of the reconstructed leaflets, improves coaptation, and may prevent dilation of the RV cavity.

3. *Leaflet lengthening.* Once leaflet material has been harvested, reconstruction of the septal and posterior leaflets is performed by approximation of cut edges of septal and posterior leaflets, thus "lengthening" this leaflet tissue while reducing the effective circumference. Sewing edges of posterior leaflet to septal leaflet reduces the circumference of the new valve while increasing the height of its leaflets (Fig. 101.4). Edge-to-edge approximation is continued until sufficient leaflet height is obtained to reach the true tricuspid annulus without tension. By reconfiguring existing surface area of leaflet material, adequate zone of coaptation is achieved without augmentation with additional prosthetic material. The septal leaflet is approximated to the anterior leaflet at the commissure to create support for both of the anterior and newly constructed septal leaflet. Since the anteroseptal commissure is maintained in its original position, edge-to-edge closure of the anterior leaflet and the new septal leaflet results in clockwise rotation of the entire valve apparatus, thus creating a funnel or "cone" configuration. The cone reconstruction converts a narrow but large-diameter TV into a smaller-diameter but taller-leaflet valve utilizing the same surface area of leaflet material. A subset of patients may have insufficient leaflet material to create a tension-free reconstruction up to the level of the true tricuspid annulus. Augmentation of septal leaflet with autologous pericardium or other leaflet substitute material may be necessary in these patients.

4. *Annular and ventricular remodeling.* Abnormally thin and dilated segments of RV must be excised or plicated during the reconstruction in order to optimize RV dynamics and prevent splaying of papillary muscles following cone reconstruction (Fig. 101.5). Identification of the right coronary artery and posterior descending coronary artery is critical to avoid injury to these structures during resection or plication of the RV. The section of RV that is usually remodeled includes the acute margin near the diaphragmatic surface. The goal of RV remodeling is to reduce the volume of the dilated ventricle as well as remove dyskinetic or akinetic regions of RV to improve ventricular efficiency. Reduction of RV volume also minimizes the distraction of papillary muscles and primary chordal attachments, which contribute to recurrent tricuspid regurgitation. The diaphragmatic aspect of the true tricuspid annulus is reduced by plication as an extension of RV plication or resection, as well as near the anterior leaflet (Fig. 101.6). The annulus is reduced to match the size of the newly reconstructed TV leaflets. Once the leaflets are attached to the neoannulus, reinforcement of the plicated annulus with a partial flexible ring prevents dehiscence, particularly in adolescents and adults (Fig. 101.7).

5. *Leaflet reattachment.* Following annular reduction and RV remodeling procedure, the reconstructed TV leaflets are attached to the neoannulus at the level of the true annulus. This reconstruction is reinforced by the placement of a prosthetic annuloplasty ring in the adolescent or adult patient. Injury to the atrioventricular (AV) node is avoided by the placement of a partial ring that spares the septal portion (Fig. 101.7).

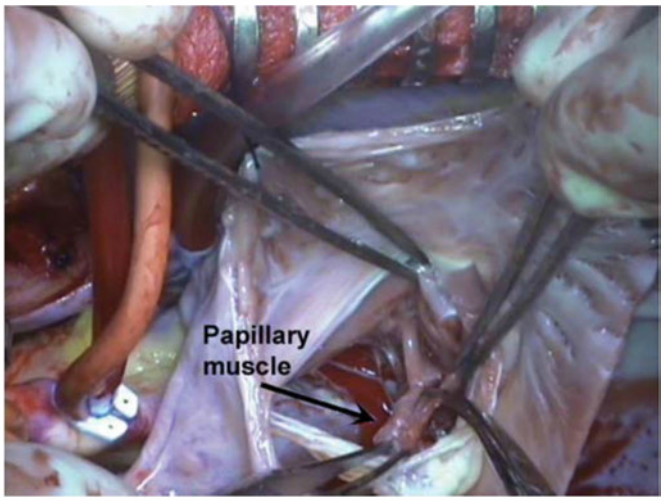


Fig. 101.3. Division of secondary chords and tethering muscle bundles maximizes mobility of the anterior leaflet. Failure to mobilize the papillary muscle may result in tethering of the leaflet edge and residual regurgitation. During mobilization of fused papillary muscles, care must be taken to avoid transection of primary attachments to the leaflet edge.

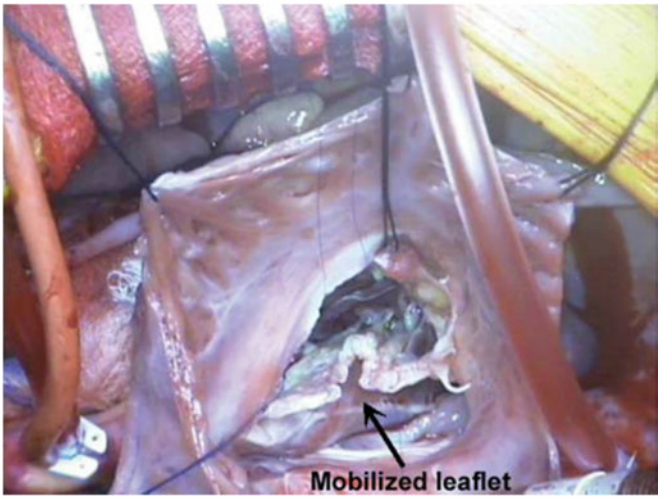


Fig. 101.4. The mobilized leaflet tissue is rotated clockwise to create a "cone" configuration. The edges of harvested septal and posterior leaflet are approximated to augment the height of the neoseptal leaflet. The valve complex is then approximated to the anterior leaflet along the anteroseptal commissure.

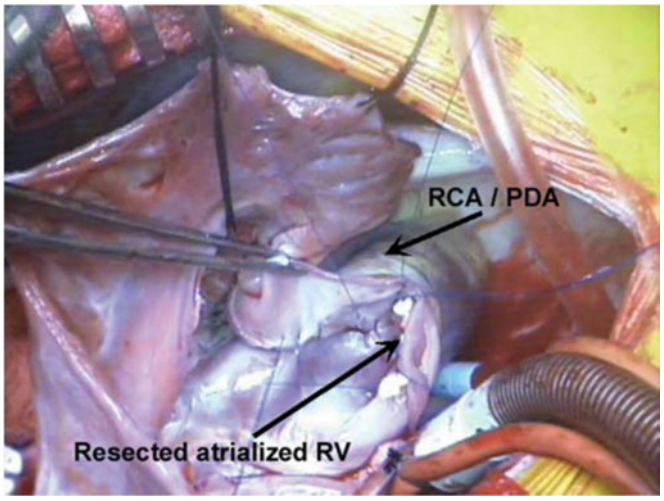


Fig. 101.5. Atrialized right ventricle (RV) is resected, with care taken to avoid injury to the right coronary artery (RCA) and posterior descending coronary artery (PDA). The ventricular margins are approximated in several layers from apex to base. An alternative to resection is plication of the atrialized portion of RV.

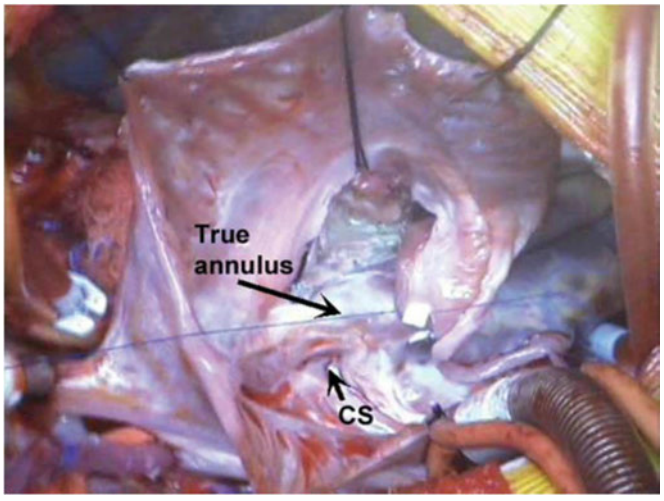


Fig. 101.6. The ventricular closure continues to the level of the true tricuspid annulus, effectively plicating the true annulus. The mobilized leaflets will be reattached at the level of the true annulus. CS, coronary sinus.

6. *Cryoablation for prevention and treatment of atrial arrhythmias.* A right-sided cryoablation–MAZE is performed for patients with documented paroxysmal atrial flutter and fibrillation or inducible atrial arrhythmia by preoperative electrophysiologic testing. However, we have expanded our indications for RA cryoablation–MAZE to prophylactic application in all patients undergoing repair. Patients with chronic atrial fibrillation may benefit from biatrial Cox-maze procedure. The lesion set most commonly utilized includes isthmus (1) between the inferior vena cava and the ostium of the coronary sinus; (2) between the ostium of the coronary sinus and the true TV annulus; and (3) between the anterior portion of the TV annulus and the edge of the atriotomy (Fig. 101.8). A fourth lesion between the foramen ovale and the atriotomy incision near the superior vena cava has been described, but in our experience increases the risk of postoperative sinus node dysfunction. If AV nodal conduction abnormality is present or suspected at the time of TV repair, placement of epicardial pacing leads is recommended to prevent subsequent lead placement through the reconstructed valve. Previously placed endocardial RV leads may be sutured within the annulus of the TV to avoid interference with TV function.

7. *Atrial septal defect closure.* A residual atrial level communication may be maintained as a mechanism for RV unloading. Superior cavopulmonary shunt is preferred if right heart failure is anticipated or encountered postoperatively (see below).

Palliative Shunt and Right Ventricle Exclusion

Neonates who demonstrate hemodynamic instability or severe cyanosis and require intervention are at high risk for mortality. They frequently demonstrate low-RV pressures suggestive of systolic dysfunction or have functional or anatomic pulmonary atresia. In these patients, a biventricular circulation is untenable not only because the RV is unable to support a full cardiac output but also due to compression of the LV from the massive RV. In these patients, exclusion and reduction of the RV along with an aortopulmonary shunt may provide stability for eventual single-ventricle palliation or biventricular conversion. Maintenance of the tricuspid valve tissue maintains the option for leaflet reconstruction, should the RV

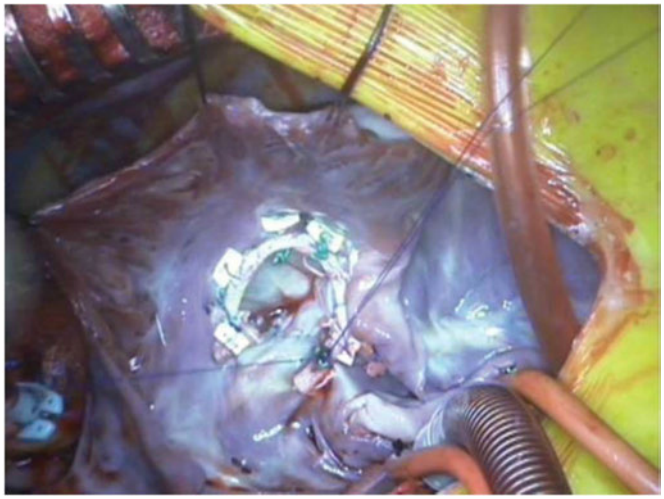


Fig. 101.7. The leaflet reattachment to the annulus is reinforced with the addition of a partial flexible annular ring.

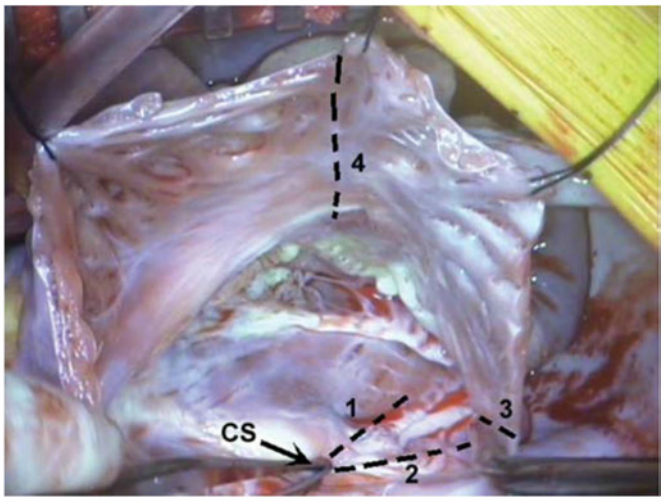


Fig. 101.8. The right atrial cryoablation–MAZE procedure involves placement of lesions between (1) coronary sinus (CS) and true tricuspid annulus, (2) CS and inferior vena cava, (3) inferior vena cava and right atriotomy, and (4) right atriotomy and anterior tricuspid annulus. An additional lesion between the foramen ovale and the right atriotomy near the superior vena cava may be performed (not shown). CS, coronary sinus.

eventually be deemed suitable for biventricular conversion.

Superior Cavopulmonary Anastomosis

A superior cavopulmonary anastomosis may be necessary as an adjunct to tricuspid valve repair in patients with severely dilated and thin-walled RV as a means of unloading the right heart. Qualitative preoperative assessment of RV mass and systolic function by noninvasive imaging studies help guide the decision. Alternatively, hemodynamics can be assessed following tricuspid valve repair. Evidence of right heart failure (elevated central venous pressures, decreased cardiac output) in the operating room following separation from cardiopulmonary bypass would indicate a need for cavopulmonary shunting.

Tricuspid Valve Replacement

Prosthetic tricuspid valve replacement is reserved for patients who fail attempt at tricuspid valve repair. A biological prosthesis is preferred over mechanical due to the risks of thrombosis in the latter. The options for biological prosthesis include porcine and pericardial tissue valves. Porcine bioprosthesis has been preferred to pericardial prosthesis since the porcine leaflet material is thinner than the pericardial tissue; thus theoretically, it is better suited to function with the lower opening and closing pressures associated with right heart circulation. There are, however, no data to support this bias. AV nodal injury is prevented by implantation of the valve in the suprannular position near the septal aspect, leaving the coronary sinus on the ventricular side of the valve.

OUTCOMES

Early (hospital mortality) for non-neonates undergoing repair is between 3% and 6%. Long-term data are available regarding repair techniques other than the cone reconstruction. Actuarial survival and freedom from reoperation at 10 years are approximately 90% and 77%, respectively. Long-term results are beginning to emerge regarding durability of the cone reconstruction suggesting favorable results. Moderate tricuspid regurgitation at discharge is a risk factor for reintervention, and consideration should be given to early reoperation in these children to avoid sequelae of right heart failure. Sudden death due to arrhythmias has prompted more aggressive postoperative monitoring and management with antiarrhythmic agents, catheter-based ablation, and defibrillator implantation.

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EDITOR'S COMMENTS

Until relatively recently, patients with Ebstein's malformation of the tricuspid valve with atrial septal defect who survived infancy were managed medically for many years before any surgical intervention was recommended. The primary reason for delay in operative intervention was the fact that these patients remained relatively stable despite severe cardiomegaly on chest X-ray films and cyanosis from right-to-left shunting at the atrial level. It was believed that the only operative intervention would be valve replacement, and a high incidence of heart block was associated with valve replacement in the earlier surgical eras. Thus, operative intervention was delayed until the patient became old enough to accept a satisfactory-sized adult prosthesis with probable epicardial pacemaker placement.

The markedly improving results with valve repair for Ebstein's malformation as pioneered by Danielson at the Mayo Clinic and recently by DaSilva in Brazil has changed the approach to patients with Ebstein's malformation significantly. Because most valves can be repaired (approximately 90% in most series), earlier intervention may be recommended for patients with severe cardiomegaly and cyanosis. With earlier repair and with valve repair rather than replacement, the biventricular function can be maintained, and elimination of atrial level shunting decreases the chronic effects of cyanosis and markedly improves exercise tolerance in these patients. With current techniques, a low incidence of complete heart block has been noted. Nevertheless, as emphasized by the authors, arrhythmias are common after repair of Ebstein's malformation of the tricuspid valve. Many of these patients may have accessory conduction pathways, and long-term antiarrhythmic therapy or ablation, either before valve repair or at the time of operation, may be necessary to control arrhythmias. Direct surgical approaches for controlling atrial arrhythmias at the time of valve repair have been advocated, and the results have been quite excellent.

The optimal valve replacement for patients who require replacement rather than repair is somewhat controversial. Mechanical valves in the right side of the heart require anticoagulation, and thrombosis may be more frequent than on the left side of the heart despite adequate warfarin therapy. Therefore, tissue valves have preferentially been used on the right side of the heart and have the additional advantage of permitting access for catheterization if necessary. Tissue valves, however, in young children have a significant incidence of degeneration even on the right side of the heart, and therefore valve repair should be undertaken if at all possible in all cases.

An infant with significant congestive heart failure with Ebstein's malformation represents a continuing therapeutic dilemma. These patients often have abnormal ventricular function or obstruction to the right ventricular outflow tract, which accounts for the severity of the hemodynamic instability early in life. Thus, simple valve repair may not be possible in most of these infants, and the operative mortality with surgical intervention has been high. If infants do not respond to medical management, then the optimal operative approach remains controversial. As described in this chapter, in some patients who have only mild right ventricular outflow tract obstruction from leaflet tissue rather than at the valvar level, it is occasionally possible to close the entrance into the right ventricle with a patch to control the magnitude of tricuspid regurgitation and convert the patient to single-ventricle physiology. In these patients, coronary sinus flow and Thebesian flow must still be ejected from the right ventricle, and therefore an unobstructed outflow tract to the pulmonary artery is necessary. Recognition of the distention of the small ventricle has led Starnes to modify his original procedure, creating a punch opening or fenestration in the center of the patch to allow decompression of the right ventricle. Even with these approaches, operative mortality is significant. As noted in this chapter, the most significant problem occurs in infants with Ebstein's malformation and pulmonary atresia or significant pulmonary stenosis. These patients often are ill immediately after birth, and the right ventricle is markedly dilated and can cause abnormal ventricular function on the left side of the heart. If such patients can be identified in utero, then in utero listing for heart or heart–lung transplantation may be the most appropriate therapy. Operative intervention after birth in these rare children has been often unsuccessful, and in our experience heart transplantation may be the best option. As reparative techniques improve, however, some of these infants may be suitable for operative intervention if the right ventricle has adequate volume and function.

An increasing experience with repair of Ebstein's malformation in neonates and infants has been reported by Knott-Craig. In his small series of patients who were unstable in the neonatal and infant period and did not respond to medical management, valve reparative techniques were successful in the majority of cases. Thus, it may be reasonable to attempt valve repair in the majority of these patients before converting them to single-ventricle physiology.

Attempts to ablate the right ventricle have been unsuccessful because of the amount of inflow into the ventricle from Thebesian veins, and therefore occlusion of the inflow into the ventricle without adequate outflow has not been successful. Attempts to reconstruct the right ventricular outflow tract and eliminate inflow into the ventricle have also not been successful because of the inability of the right ventricle to adequately decompress. Ventricular dysfunction is common in these patients with a thin-walled, poorly contractile right ventricle, probably secondary to in utero severe tricuspid regurgitation. Sano and his associates have been successful in complete excision of the right ventricular free wall in rare cases with pulmonary atresia and Ebstein's malformation with patch closure or oversewing of the tricuspid valve inlet. These radical approaches of RV exclusion have been successful in isolated cases and may represent a reasonable alternative in patients who have significant RV dilation and RV dysfunction and abnormal

(*continued*)

EDITOR'S COMMENTS (*continued*)

ventricular septum that can bulge into the left ventricular outflow tract and cause left ventricular outflow tract obstruction. In these cases, removal of the free wall of the right ventricle may allow septal repositioning and improvement in left ventricular outflow tract diameter.

Drs. Emani and del Nido have appropriately emphasized the fact that closure of atrial septal defect to alleviate cyanosis in patients with Ebstein's malformation may not actually improve long-term prognosis. It is a tempting in patients who present with increasing cyanosis to eliminate the right-to-left shunt with a catheter-based ASD closure. However, in the authors' and our own experience, closing the atrial septal defect simply unmasks the underlying ventricular dysfunction in these patients and often worsens the tricuspid regurgitation. An important thing to remember in Ebstein's malformation is that significant right-to-left shunting implies abnormalities of compliance of the ventricle rather than the magnitude of tricuspid regurgitation. Similarly, the authors have noted that patients presenting in the neonatal period with low-RV pressures may have a very poor prognosis from attempts at valve repair. If the RV pressure is low, these patients generally have had very abnormal ventricular function and compliance and these patients may represent the most severe form of the disease. Single-ventricle approaches are probably the best used in this subset of patients.

The most severe form (Type D) of Ebstein's malformation can often still undergo complete repair using the cone technique. Nevertheless, as emphasized by the authors it is important when mobilizing an anterior leaflet in this severe subgroup of patients, not to significantly damage the ventricular myocardium. In some cases, the leaflet is essentially created from the endocardial surface and significant dissection of myocardial tissue is necessary to mobilize the leaflet. In patients who present with cyanosis with this type of lesion, ventricular function may be severely abnormal postoperatively. This can be a very high-risk subset of patients and postoperative RV failure, which is difficult to treat, can be a complication.

The authors note that valve replacement in Ebstein's malformation is becoming increasingly less common. They suggest placement of the valve above the coronary sinus to avoid the area of the conducting tissue. If some valve leaflet tissue is allowed to remain in the area of the AV node, it is possible to place the tricuspid prosthesis in the normal anatomic location, which avoids subjecting the coronary sinus to right ventricular pressure.

Overall, the surgical approach to Ebstein's malformation remains a technical challenge. Although repair techniques are evolving and the use of the bidirectional Glenn or cavopulmonary shunt permits more drastic reduction of the tricuspid annulus to achieve better competence, the wide variation in ventricular function, valve morphology, and age at presentation make this patient population medically challenging.

TLS

102 Mitral Valve Repair in Children

Christian Brizard

INTRODUCTION/BACKGROUND

Mitral valve repair in children is guided by the same surgical rules than in adults but the anatomical substrate differs greatly. They have been set by Carpentier more than two decades ago. The technical difficulties vary according to the anatomy and to the size and age of the patient. The indications for surgery and the timing of the surgery have to take into account a large number of issues and are, therefore, more complex than in adults.

This chapter will cover mitral valve repair in children, congenital and acquired, excluding the mitral valve in atrioventricular discordance, the mitral valve in univentricular hearts, and the mitral valve of the hypoplastic left heart syndrome. We have added in this edition the surgical approach to the repair of the residual or recurrent regurgitation the left atrioventricular valve in complete atrioventricular septal defects (AVSDs).

ANATOMY AND EMBRYOLOGY

Anatomy

The normal anatomy of the mitral valve in children does not differ from the adult one and has been described. Precise knowledge of the normal anatomy is essential to read the echocardiographic study, to understand the pathological anatomy and to plan the repair.

Embryology

The leaflet and chordal tissue derive from the endocardial cushion tissue lying on the inner surface of the atrioventricular junction. The anterior leaflet originates from the superior and inferior cushions, whereas the posterior leaflet derives from an infolding of the lateral wall. As the cushion tissue elongates and grows toward the ventricular cavity, the leaflets shape progressively into a funnel-like structure totally attached to the myocardium while perforations appear in the valve leaflet edges. The perforations grow and form the chordae tendineae. The ventricular layer of the cushions will generate the fibrous part of the mitral valve and the chordae. Simultaneously, the development of the papillary muscle takes place: The anterior and posterior parts of a horseshoe ridge within the left ventricle lose contact progressively with the ventricular wall. They will form the papillary muscles, increasing their size while keeping contact with the cushion tissue at their tip.

PATHOLOGY

Congenital Anomalies of the Mitral Valve

Congenital valve stenosis and congenital mitral valve insufficiency are presented together because they have identical pathology and associated lesions. They are frequently associated in the same patient and require similar surgical techniques for the treatment.

Cleft Mitral Valve

Very often isolated, the cleft mitral valve can be easily differentiated from a left atrioventricular valve in a partial AVSD. The cleft is centered on the aortic commissure between the noncoronary cusp and the left coronary cusp, and there is no suspension apparatus on the edges of the defect. The papillary muscles are normal. Lack of valvular tissue can be seen and is secondary to the regurgitation through the cleft. The defect is not stenotic and may generate only little regurgitation for a long time.

Accessory Valve Tissue and Valvular Tags

In these anomalies, often found in association with other valvar anomalies, the spaces between the chordae are filled with a network of myxoid, valve-like tissue. When there is continuity between the anterior and the posterior leaflet, the accessory tissue may generate a gradient directly related to the size of the perforations in the accessory tissue. When the accessory valve tissue is entrapped in the left ventricular outflow tract, the mitral valve may become regurgitant due to the traction exerted by the accessory valvular tissue on the anterior leaflet, opening the valve in mid systole; however, in such cases, the left ventricular outflow tract obstruction is the predominant hemodynamic lesion and is the most frequent mode of diagnosis. The accessory mitral valve tissue in isolation often does not generate significant gradient or insufficiency.

Lesions Associated with Lack of Valvular Tissue

Three major anatomical types have been identified, although there is a continuum between them. Their recognition is useful for the planning of the repair. The functional lesion can be either predominantly regurgitant or predominantly stenotic, it can be both stenotic and regurgitant, or the valve can have a normal function.

Parachute Mitral Valve

The parachute mitral valve can be found in isolation. It can be integrated in a Shone syndrome. There is a dominant papillary muscle with the orifice of the mitral valve overriding the tip of the papillary muscle. There is a spectrum of lesions for the chordae ranging from complete absence and fusion of the tip of the papillary muscle to the free leaflet edge to relatively normal looking chordae with good mobility of the leaflet. An accessory papillary muscle, usually very small is devoted to a short segment of the free edge, or even to the under surface of the leaflet tissue with or without second orifice (double-orifice mitral valve). The functional anatomy depends on the interaction between the amount and mobility of leaflet tissue, size of the fenestrations and the presence, length, and quality of the chordae. The parachute mitral valve has almost always a stenotic component.

Papillary Muscle to Commissure Fusion

This syndrome is a spectrum. It ranges from papillary muscle tip fused to the

commissural area of the free edge to short, almost normal looking chordae. This anomaly can be limited to one papillary muscle only. The valve is generally more regurgitant than stenotic. When the papillary muscles are hypertrophied, the bulk of their mass is responsible for a valve predominantly stenotic.

Hammock Valve (Arcade Valve)
The suspension apparatus may have lost all resemblance to the normal anatomy. There is either no papillary muscle identifiable or multiple very small ones behind the posterior leaflet. The leaflets are suspended directly by a network of chordae directly attached to the posterior wall of the ventricle. This attachment is generally displaced toward the base of the heart with an excess of tension on the anterior leaflet and extreme limitation of posterior leaflet motion. The valve is most often predominantly regurgitant.

Regurgitant Mitral Valves with Normal Anatomy Associated with Congenital Cardiac Lesions

Isolated Annular Dilation; Isolated Elongation of the Chordae and/or the Papillary Muscle
There is no evidence of the congenital origin of these lesions. They are not found at birth unlike the previous anomalies described above. They are usually associated with significant volume loading of the left ventricle, that is large ventricular septal defect or large patent ductus arteriosus. Sometimes minor anomalies of the valvular tissue or the papillary muscles can give an indication toward a true congenital origin. The papillary muscle may have an ischemic aspect, and even rupture, this is mostly seen in neonates.

ALCAPA
The mitral regurgitation in patients with anomalous coronary artery from the pulmonary artery is of ischemic origin. The anatomy is normal. The functional classification is of systolic restriction of one of the segments of the posterior leaflet (Carpentier type IIIb).

Supravalvar Mitral Ring

Quoted as a common cause of congenital mitral valve stenosis, the supravalvar mitral ring is an acquired fibrous construction attached to the posterior annulus of the mitral valve and from both commissures to the mid-height of the anterior leaflet. The supravalvar mitral ring is secondary to turbulent flow through the mitral orifice. The primary lesion of the mitral valve responsible for the turbulent flow can be obvious, stenotic, or regurgitant or can be very discrete and difficult to identify. The supravalvar mitral ring is prone to reoccur after surgical resection, unless the underlying anatomical anomaly has been identified and corrected. The supravalvular ring can be encountered very early in life. It has to be suspected every time the transvalvular gradient increases during follow-up or when the Doppler gradient is greater than what the anatomy depicted with the echocardiographic study would suggest; sometimes, it is only found at operation.

Mitral Valve Disease with Excess Leaflet Tissue

They are Marfan syndrome, Loyes–Dietz syndrome, mitral valve prolapse, Barlow disease, Elher–Danlos syndrome, and mucopolysaccharidosis type I. All include elastic fibers alteration and myxomatous tissue proliferation of various degrees. Most are now well associated with chromosomal mutations.

Acquired Mitral Valve Disease

Rheumatic Heart Disease
Acute rheumatic fever (ARF) is an autoimmune disorder. The immune response to group A streptococcal M protein generates T cells and antibodies that cross-react with cardiac antigens. In some patients, the acute damage to the valves will induce chronic and evolving lesions secondary to the scarring process and/or the hemodynamic modifications. This is known as rheumatic heart disease.

Acute Lesions
Acute lesions are exclusively regurgitant. On inspection, the valvar tissue and the chordae are swollen but supple. Prolapse predominantly affects the anterior leaflet. This prolapse is usually related to large elongation of the marginal chordae that appeared stretched; chordal ruptures are rare. Multiple small nodules (2 to 3 mm diameter) can be seen on the free edge of either mitral leaflet. The annular dilation is secondary to the myocarditis.

Chronic Lesions
The healing of the spongiosa induces fusion of chordae as demonstrated by reduction in their number and increase of their thickness. The physiology of the regurgitation is always a combination of prolapse of the anterior leaflet, retraction of the posterior leaflet, and annular dilation. In the pediatric age group, the mitral valve is exclusively or predominantly regurgitant. The stenosis appears later and the age of apparition of the mitral stenosis varies greatly with the geographical origin of the population affected, suggesting different pattern of infection (i.e., age of first ARF episode) and influence of other factors (genetic mostly and alimentation).

Infective Endocarditis
Bacterial endocarditis of the mitral valve is rare. It is always a regurgitant lesion. At the Royal Children's Hospital, Melbourne, in the last decade most patients had normal native mitral valve. It is very important for the surgeon to be able to differentiate intact valvar tissue, supple thin and resistant from infected tissue, thickened edematous and friable.

The Left Atrioventricular Valve in Repaired Atrioventricular Septal Defect

Anatomy
The anatomy of the left atrioventricular valve after complete AVSD repair has standard features: The superior and inferior bridging leaflets are partitioned transversally at the level of the crest of the septum. Their mobility is very limited at that level; it increases further away from the partition point. The superior and inferior bridging leaflets face one another through the zone of apposition made of by rough surface. The quality of the suspension apparatus to the free edge varies greatly from patient to patient. The zone of apposition between these two leaflets has often been closed with continuous or interrupted sutures at the time of the primary repair. In the most common configuration, the left lateral leaflet (LLL) or posterior leaflet is normally developed and the superior and inferior bridging leaflets separate from one another and delineate a triangular orifice at the inferior aspect of the left AV valve annulus. This orifice is covered with the LLL. It is triangular; the base hinges at the posterior aspect of the annulus and the tip faces exactly where the superior and inferior leaflets separate. Between one-third and one-fifth of the circumference of the reconstructed left AV valve belongs to the LLL. Two papillary muscles make the suspension apparatus. The anterior papillary muscle underneath the zone of apposition between the superior bridging leaflet and the LLL and the posterior papillary muscle

underneath the zone of apposition between the inferior bridging leaflet and the LLL and their respective commissure. The leaflet has a broad base when it covers one-third of the annulus or a narrow base when it covers one-fifth. In that configuration, the LLL is tall and narrow and unstable.

In the least common configuration (6% to 10% of the complete AVSD), the LLL is diminutive or absent. Both superior and inferior bridging leaflets are supported by the anterior papillary muscle, while the posterior papillary muscle is commonly diminutive or more rarely absent. Both bridging leaflets reach the left lateral annulus where a true commissure can be seen. Most often, the diminutive posterior papillary muscle is supporting a second orifice in the inferior bridging leaflet.

Mechanism of the Regurgitation in Repaired Left Atrioventricular Valve

Valves with Normally Developed Left Lateral Leaflet

The regurgitation is related to the lack of apposition facing the tip of the LLL. The closure of the cleft does not restore a surface of apposition; in fact, the little apposition surface may be reduced and distorted by the cleft closure (Fig. 102.1). If the cleft closure has ruptured or partially ruptured, then the defect created and the doming of the combined bridging leaflets augments the regurgitation. If the regurgitation has been long-standing, the secondary lesion or dysplastic lesions on the edges of the cleft are severe with thickening, sometimes calcification and severe retraction of the leaflet tissue. However, it is almost the rule for the LLL to be thin and pliable with no secondary or dysplastic lesion. There is no restriction of leaflet motion and no prolapse.

Absent or Very Hypoplastic Left Lateral Leaflet

At the time of initial repair, the cleft closure is not performed or only partially executed in order to avoid the creation of a stenosis. Consequently, the regurgitation is always through the cleft, with retraction of the edges of the cleft and very few or no chordae attached to them (Fig. 102.2).

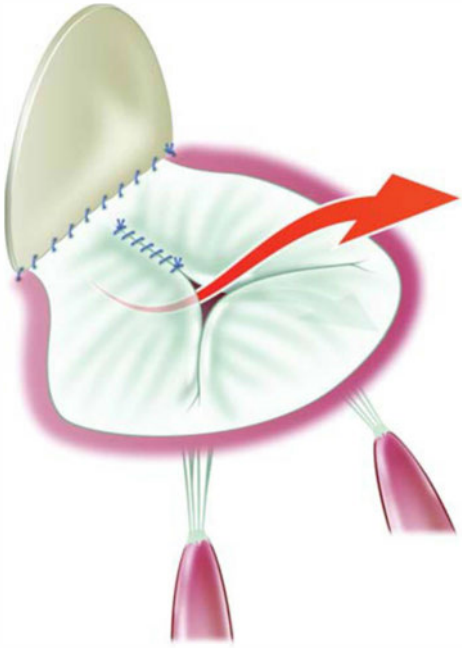


Fig. 102.1. The jet of regurgitation in the repaired left atrio ventricular valve in the setting of a normally developed left lateral leaflet. Most often, the lack of surface of apposition in front of the tip of the left lateral leaflet is the source of the regurgitation. The regurgitant jet has a characteristic direction toward the posterior wall of the left atrium and runs alongside the atrial surface of the left lateral leaflet.

INDICATION AND PLANNING OF THE REPAIR

Echocardiography

The long axis view obtained from the apex or from subcostal view of the transthoracic study is best to grade the regurgitation, provide an accurate estimation of transvalvular gradient and define the precise amplitude of any prolapse or restriction. The short axis view gives a direct evaluation of the area of the mitral orifice, a precise localization of the regurgitant jet. It allows an analysis of the papillary muscles (presence, size, location, and symmetry). The transeosophagal echo is superior for the anatomical details of the suspension apparatus, the evaluation of the functional classification in relation to the anatomy (response to the question: how much prolapse/restriction and where?), but it is less useful to grade the severity of the regurgitation. The transgastric position allows for a short axis cut with precise measurement of the shortening fraction and an en face view of the mitral valve.

For mitral stenoses, the peak instantaneous and mean gradients across the valve have to be interpreted according to the quality of the diastolic function of the heart and the associated lesions (mainly Q_p/Q_s, the presence of an intra-atrial shunting and gradient across the foramen ovale/atrial septal defect). The overall impact of the gradient on the surgical indication has to be weighted with the pulmonary artery pressure but mostly the clinical tolerance.

Functional Classification

Transthoracic and transoesophageal echocardiography allows classifying the malformations according to the motion of the leaflets in one of the three following types:

Type I: Normal leaflet motion. The regurgitation results from a lack of coaptation between the leaflets.

Type II: Leaflet prolapse. The free edge of one or the two leaflets overrides the plane of the orifice during systole.

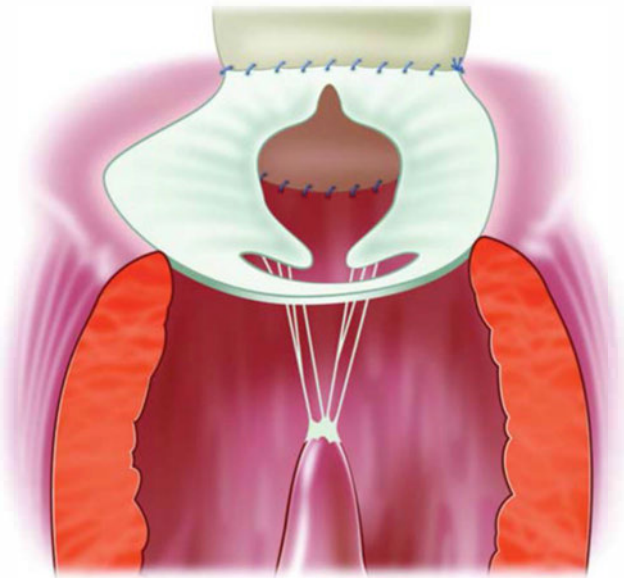


Fig. 102.2. The jet of regurgitation in the repaired left atrio ventricular valve in the setting of a diminutive left lateral leaflet. The coaptation defect between the superior and inferior bridging leaflets generates a vertical jet of regurgitation into the left atrium.

Type III: Restricted leaflet motion. The motion of one or the two leaflets is limited.

This can be secondary to short or stiff leaflet tissue or suspension apparatus (type IIIa) or the leaflet can be pulled away from the coaptation area by a paradoxical motion of the ventricular wall (type IIIb or systolic)

Other Investigations

Catheter study and angiography generate no additional information to the echocardiography and should not be performed.

Magnetic Resonance Imaging allows precise calculation of the ventricular volumes irrespective of the septal geometry; this may help for the decision-making with small left ventricle in mitral valve stenosis. The regurgitation fraction is measured accurately. Gradients and flows are demonstrated with MRI. In small patients, MRI does not help with the analysis of the valve anatomy.

Three-dimensional echocardiography is progressing rapidly with the exponential increase of computer power and miniaturization of the probes. The information generated are, however, of little use in small patients as the spatial resolution is still insufficient. Generally, the most obvious benefit is the ability to locate *precisely* the types II and III on the 3D en face view while they are quantified much more precisely on the 2D.

Indications

The indication for surgical intervention has to weigh several considerations.

According to the Mitral Valve Annulus

Large Mitral Valve Annulus (>30 mm in Female Patients and 32 mm in Males).

Using a wide range of mitral valve repair techniques, the probability of a successful repair of the valve is very high. A remodeling annuloplasty will not be outgrown and will not generate stenosis with the growth of the patient. The surgical indication is similar to the current indications in the adult population: The patients should be operated on as soon as the volume of the regurgitation is severe, irrespective of the severity of symptoms. The probability of repair is directly related to the experience of the surgical team but the repair of virtually all valves is an accessible goal.

Mitral Valve Annulus <18/20 mm

Biventricular repair should be considered only if the mitral valve annulus is not hypoplastic (Z value greater than −1.5). The repair is technically very challenging while the replacement is only possible with the use of surgical artifacts associated with significantly increased mortality. In these patients, the surgical indication should be differed as long as the patient can be managed with intense medical therapy, including transfusion. Aggressive medical therapy allows delaying the surgery for several months in some instances and can generate significantly more favorable operating conditions. However, the requirement for positive pressure ventilatory support more invasive than continuous nasal flow (either CPAP or other mode of pressure support) should trigger the surgical indication and so would a flat weight curve.

Intermediate Mitral Valve Annulus (>20 mm and Smaller than Adult Size)

In these patients, the mitral valve replacement can be safely performed in anatomical position. Therefore, the timing of the mitral valve repair does not need to be delayed for fear of replacement in supra-annular position. It is generally safe, however, to wait for a long time (up to several years) with a severe regurgitation, provided that adequate monitoring of the pulmonary artery pressure and ventricular function is achieved. Contrary to adult patients, in children, long-term ventricular function returns to normal in patients with decreased systolic function preoperatively.

According to Associated Lesions

Large Left-to-Right Shunts

These shunts can generate severe functional regurgitation that subsides with the treatment of the shunt. The mitral regurgitation should be addressed separately only if a prolapse of the anterior leaflet or a congenital valve can be identified. Similarly, modest mitral valve anomaly can generate severe gradient in this context. No or minimal intervention on the valve may only be required.

Aortic Valve Stenosis and Shone Syndrome

Whether to embark on a biventricular repair or a univentricular pathway in neonates with combined aortic and mitral valve anomalies is one of the most difficult problems in pediatric cardiology. The minimal size of the left ventricle required to survive a biventricular repair in isolated critical aortic stenosis is not a satisfactory criterion. The aim of a biventricular repair should be an excellent long-term functional result, which supposes normal ventricular function and normal pulmonary resistances. At the Royal Children's Hospital, Melbourne, VIC, we limit this surgery to patients who have (1) no endocardial fibroelastosis on the echocardiographic study and intraoperatively, (2) a normal or very close to normal size left ventricle, (3) a normal size mitral valve annulus, and (4) the mitral and aortic valves can satisfactorily be repaired. In the early postoperative period, any difficulty to wean from the ventilator or an absence or minimal pulmonary pressure decrease with persisting reactivity should indicate a revision of the surgical strategy.



SURGICAL TECHNIQUE

Bypass Technique

Cardiopulmonary bypass in Melbourne is conducted with moderate hypothermia 32°C, hemoglobin of 10 to 12 g/dl. Pump flow of 150 ml/min/kg or 1 l/min/m^2. Myocardial protection with intermittent warm blood cardioplegia is administered every 15 to 20 minutes. Venous cannulation should allow ample access to the interatrial groove. The left atrium is entered through the interatrial groove after some dissection of the groove is performed.

Exposure and Analysis

The exposure is enhanced with mattress sutures inserted into the posterior annulus pulling the valve upward and toward the operator. The snugger on the inferior vena cava is pulling upward and to the left. A self-retaining retractor for mitral surgery adapted to the size of the patient has to be used in all cases. Once satisfactory exposure of the mitral valve is gained, the valve is systematically analyzed confronting the observation with the preoperative echocardiographic study. The functional classification is confirmed but the extension of mitral valve prolapse or restriction is based on the echocardiographic study on the beating heart. Then the analysis of the anatomy takes place. The operator must evaluate the texture, aspect, and size of the mitral valve leaflets; the presence and location of jet lesions; the number, aspect, and distribution of the chordae; the presence of commissural tissue and dedicated suspension apparatus; and the present size, location, and morphology of the papillary muscles. The examination finishes with a careful check for accessory mitral valve tissue in the interchordal spaces. A supravalvular ring is confirmed or eliminated. The diameter of the annulus and of the opening of the mitral valve is compared with the reference one for the patient's body surface area. In Melbourne, we use a modification of the sizes provided by Kirklin.

Mitral Valve Repair

Adapted to the Functional Classification

Correction of Type I

An annuloplasty is mandatory in all mitral valve repairs for regurgitation with the exception of some isolated type I without annular dilation, mostly cleft mitral valve. Attempts to perform mitral valve repair without annuloplasty have resulted in recurrence. To accommodate an adult size device, or a larger size annulus than what would be indicated from the area of the anterior leaflet, leaflet enlargement with glutaraldehyde-treated pericardium of the posterior leaflet, the anterior leaflet, or both are used (Fig. 102.3). When no annuloplasty device is available for the size of the patient or when the device is thought to be too small to allow for growth without stenosis, then a custom annuloplasty limited to the posterior annulus is indicated. The annuloplasty has to incorporate both trigones. For that purpose, we like to use expanded PTFE sheet folded once or twice to provide rigidity. The mattress sutures should not be tied too tightly to avoid corrugating effect (Fig. 102.4). In very small patients, we have found that the most effective annuloplasty allowing for growth was a compression of the annulus by the way of isolated mattress sutures tied over them (Fig. 102.5). In our experience, other means to allow room for growth have provided unreliable support of the annulus.

Correction of Type II

Correction of type II is rarely necessary in mitral valves with abnormal anatomy but is common in other types of valves. Multiple

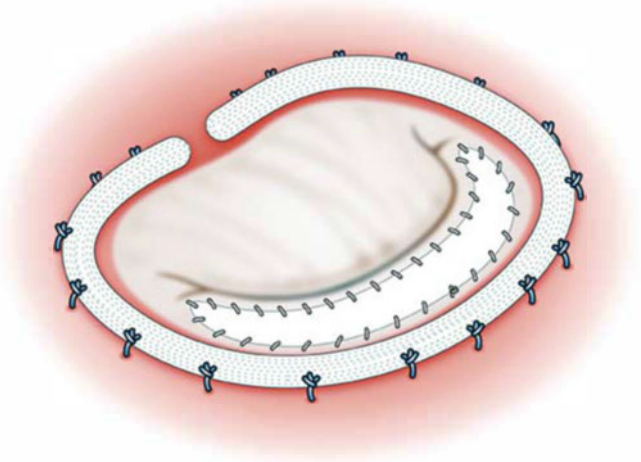


Fig. 102.3. Remodeling annuloplasty using a complete ring. The ring has an adult size. When the surface of leaflet tissue does not cover an adult annulus area, the leaflet tissue can be augmented with treated autologous pericardium.

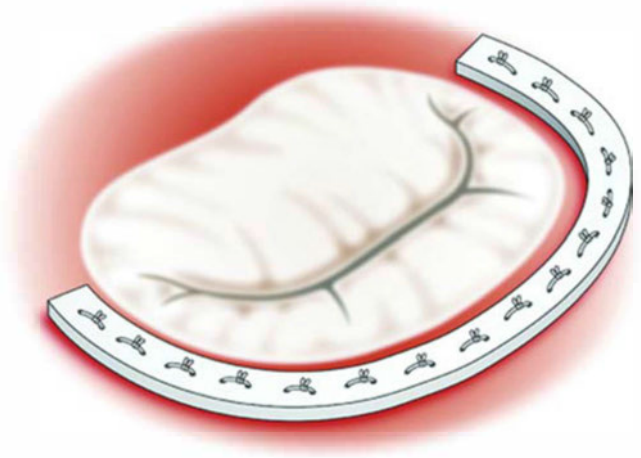


Fig. 102.4. Posterior annuloplasty in patients with less than adult size annulus. Band of polytetrafluoroethylene may be folded once for increased rigidity. It is secured from one trigone to the other.

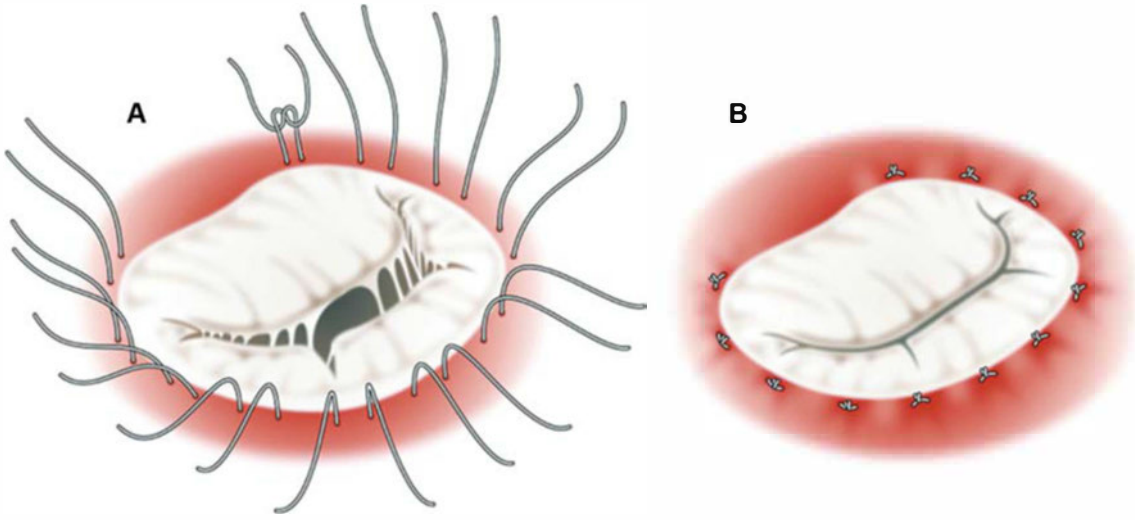


Fig. 102.5. Technique of posterior annuloplasty suitable for very small mitral annulus. (**A**) Mattress sutures are applied in the posterior annulus as for any mitral valve repair and (**B**) tied to compress the posterior annulus.

techniques are available, to be used in isolation or in combination depending on the width of the prolapse (evaluated intraoperatively or on 3D echo), the height of the prolapse (based on the 2D echocardiographic study), and the aspect of the chordae. All techniques are efficient and reliable providing that the repair is adequate (restores a large surface of apposition between anterior and posterior leaflets) and avoids overcorrection.

The chordal shortening requires thin and flexible chordae. The correction generates significant shortening of the chordae and is only utilized when significant length of shortening is necessary (Fig. 102.6).

The chordal transfer, mostly between secondary chordae and the free edge, corrects a localized prolapse (Fig. 102.7).

Wedge resection (Fig. 102.8) and sliding plasty (Fig. 102.9) generate different degrees of correction of prolapsus to multiple chordae. They are very well adapted to prolapsus extended to a large segment of the anterior leaflet.

Artificial chordae: The use of artificial chordae should be restricted to the absence of available chordae of appropriate strength and quality in the area of prolapse. The insertion requires a rigorous technique to avoid overcorrection and large knots at the free edge (Fig. 102.10).

Papillary muscle shortening: Papillary muscle shortening is used rarely in infants and young children suffering from Barlow disease. It can also be used in the correction of scarred elongated ischemic papillary muscle (Fig. 102.11).

Correction of Type III

Successful correction of restricted leaflet motion and insufficient leaflet tissue is the essence of congenital mitral anomalies, especially in the first year of life.

Access to the suspension apparatus is the key to adequate mobilization of the latter. It can be done through the mitral valve orifice when it is sufficient. In very small patients, the mitral orifice is very small and does not allow for good access to the suspension apparatus. In these situations, the detachment of the posterior leaflet generates a good view of the papillary muscles. Adequate thinning, mobilization from the posterior wall, splitting and fenestration of the papillary muscles can then be performed safely with good exposure (Fig. 102.12A and B). The posterior leaflet is afterward reconstructed with the enlargement of the valvar tissue (Fig. 102.12C) when necessary.

The augmentation of the valvular leaflet tissue can be limited to the posterior leaflet

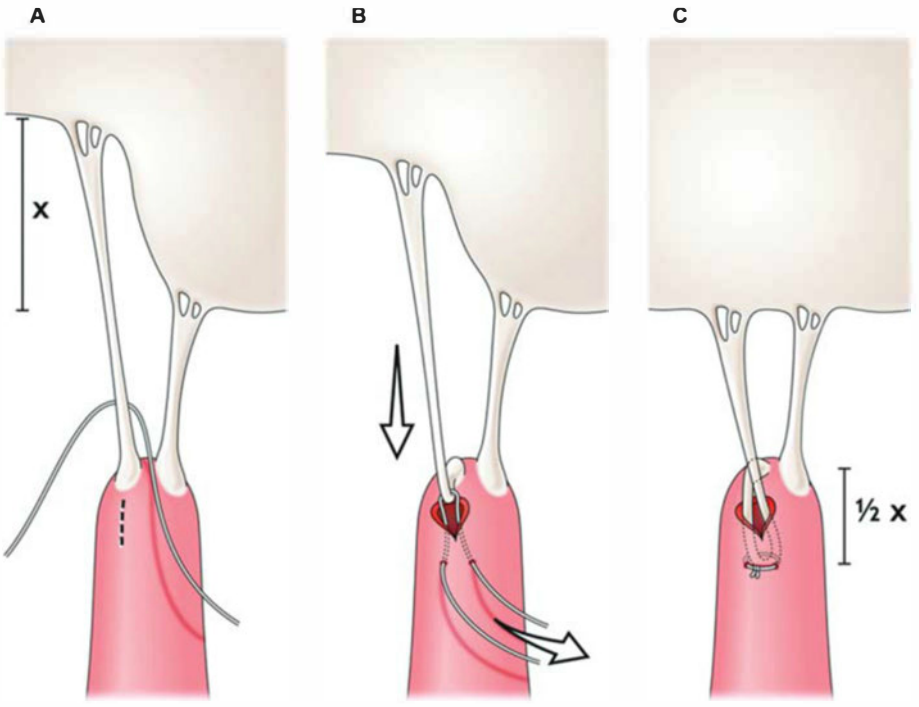


Fig. 102.6. Chordal shortening: note the extent of the shortening achieved.

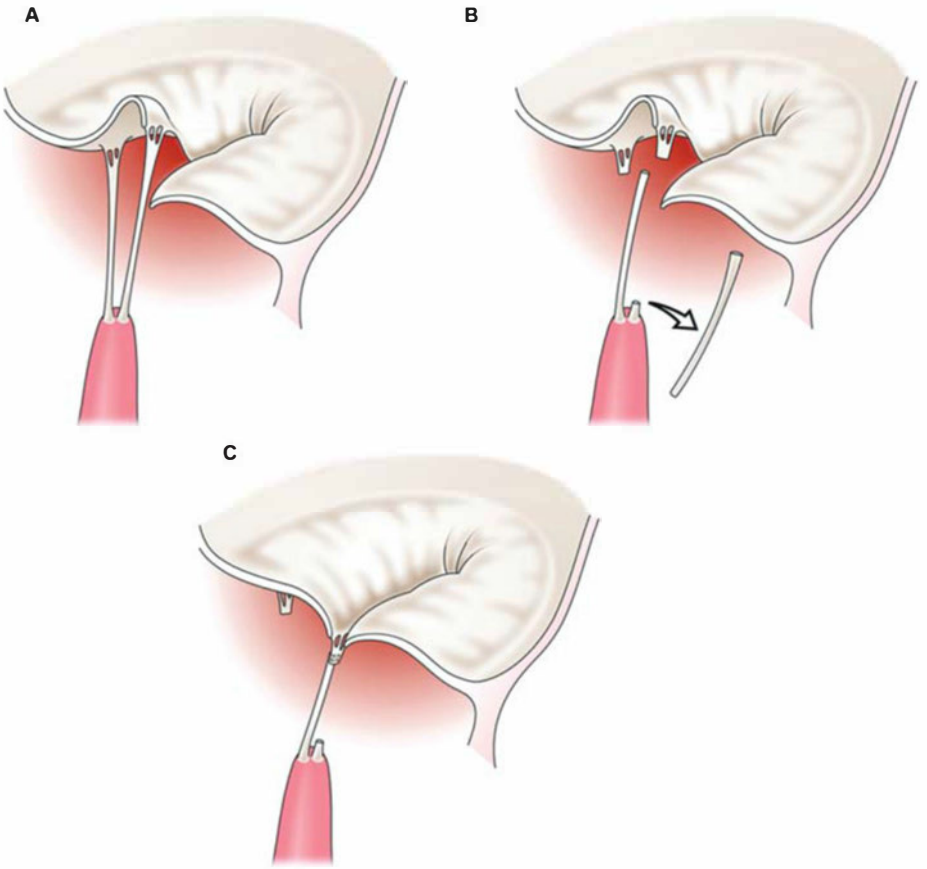


Fig. 102.7. Chordal transfer: only secondary chordae should be used and not the basal chordae.

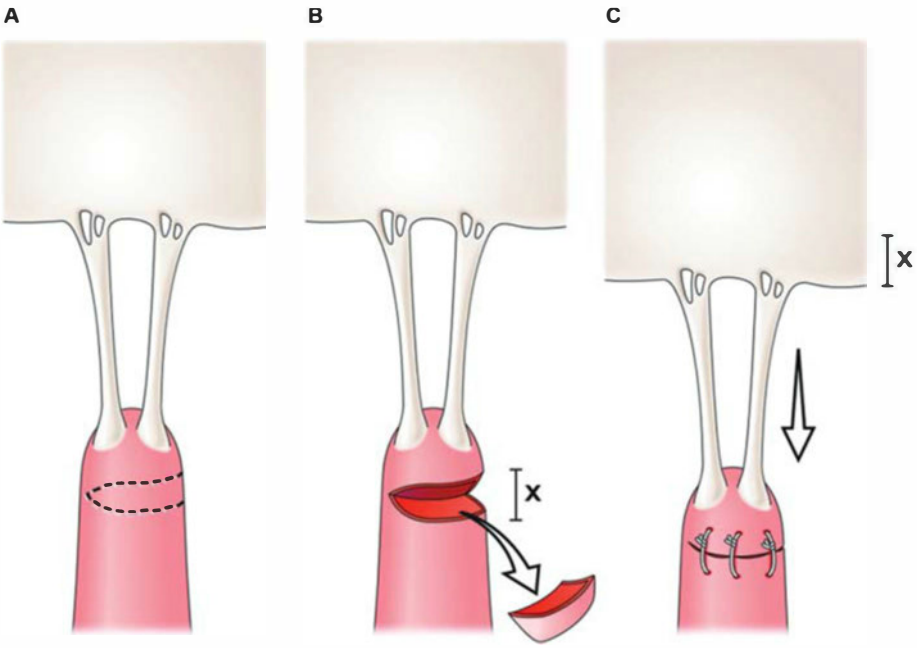


Fig. 102.8. Wedge resection: achieves limited shortening distributed to several chordae.

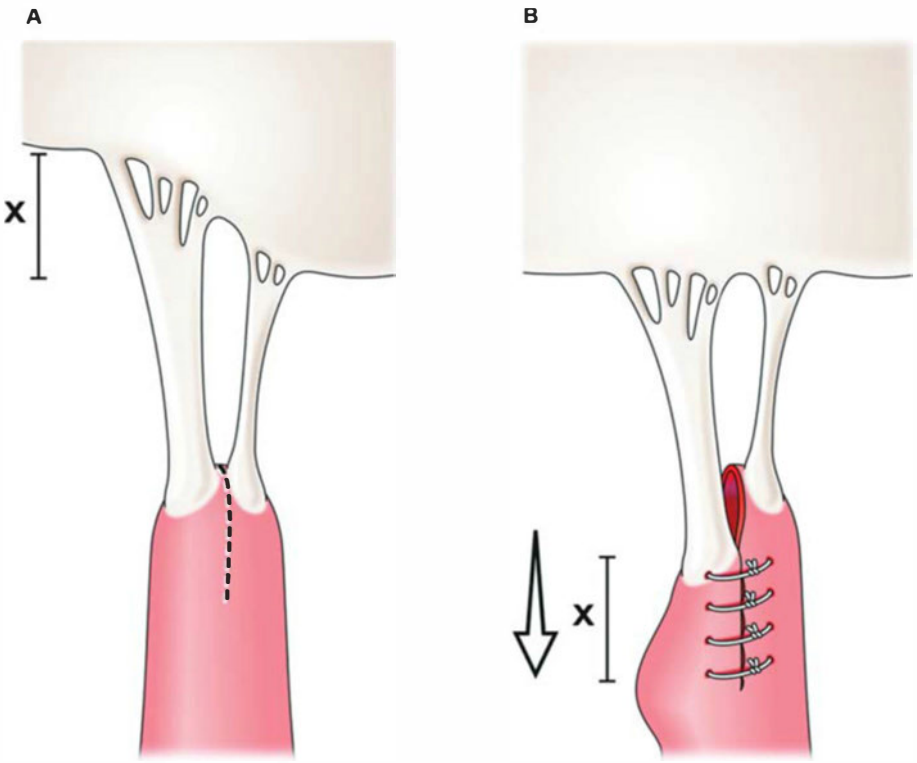


Fig. 102.9. Sliding plasty: achieves controlled shortening for thickened chordae.

or to the anterior leaflet (Fig. 102.13), or used in both. The extension of the posterior leaflet should be less than a half of the height of the leaflet. It can be limited to the area of the middle scallop; alternately, when the detachment is extended from one commissure to another, to allow for a large opening in diastole, the extension should either be a band or emulate the shape of the posterior leaflet with three scallops and two commissures but should not have a crescent shape. The extension of the anterior leaflet should be done in the body of the leaflet, leaving a strip of valvar tissue close to the hinge point to avoid mechanical stress at this level. The height of the extension should not be greater than two-fifth of the height of the leaflet, leaving the area close to the free edge intact to allow for supple and efficient surface of coaptation. It should be symmetrical from trigone to trigone.

Resection of Supravalvular Rings and Accessory Mitral Valve Tissue

Resection of supravalvar tissue requires an excellent exposure of the leaflet tissue. The supravalvar tissue can sometimes be peeled off the valvular tissue. More often, there will be the need for a careful cleavage with blunt dissection. Perforation to the anterior leaflet may occur and should be closed with simple figure-of-eight suture (Fig. 102.14).

Resection of Accessory Mitral Valve Tissue

This requires rigorous surgical technique to delineate perfectly the mitral valve chordae from what can be resected without compromising the integrity of the suspension apparatus (Fig. 102.15). Various approaches to the suspension apparatus may have to be combined.

Repair of the Left Atrioventricular Valve in Repaired Atrioventricular Septal Defects

Patients with Normally Developed Left Lateral Leaflet

The secondary lesions in the area of regurgitation are mildly excised to reach pliable leaflet tissue. The cleft is closed with a long and narrow patch of treated autologous pericardium. This patch extends into the ventricular cavity to build a zone of apposition facing the tip of the LLL. The extremity of the patch is suspended either to adjacent marginal chordae and/or with artificial PTFE chordae (Fig. 102.16).

Patients with Absent or Very Diminutive Left Lateral Leaflet

The repair relies on the construction of a large zone of apposition on each side of the cleft using the leaflet tissue itself. The free edge after minimal debridement is suspended to the predominant PM with artificial chordae using the technique described before. To build some height to the coaptation area, a compression annuloplasty of the posterior annulus in front of the cleft is applied and generates redundancy to the leaflet tissue. If required, the superior and/or the inferior bridging leaflets can be

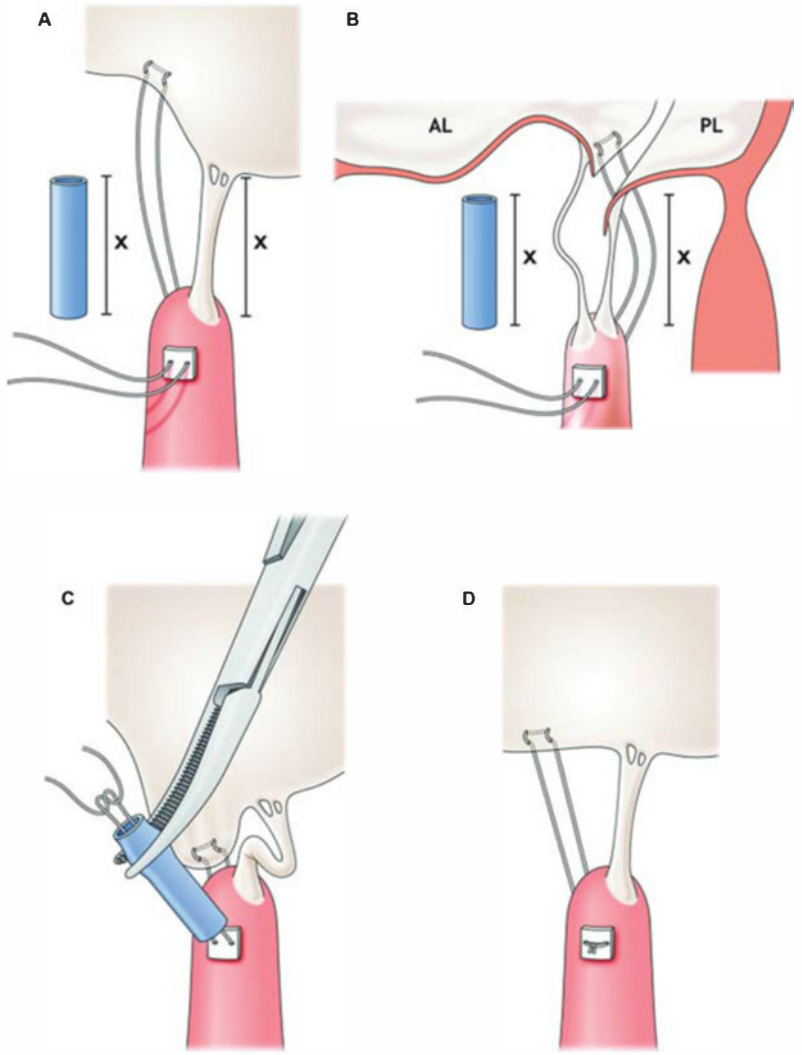


Fig. 102.10. Insertion technique for ePTFE chordae: (**A**) a template is made from a short plastic tube cut at the required length and slid over the distal part of the stitch. The reference is an adjacent normal chordae to the free edge. (**B**) Here the secondary chordae facing the prolapse area is the reference. (**C**) The free edge of the leaflet is lowered to the contact of the papillary muscle. The artificial chordae is tied while the template is clamped. (**D**) The template is removed and the mattress suture is pulled to bring the knot in contact with the papillary muscle.

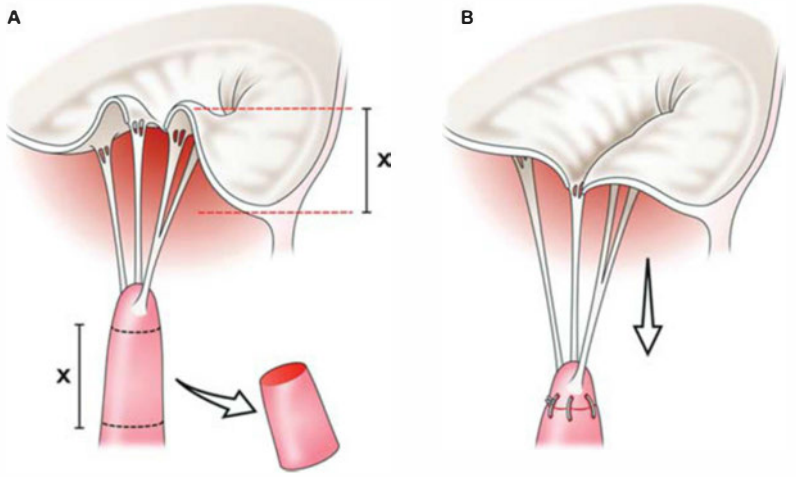


Fig. 102.11. Papillary muscle shortening can be used in combined anterior and posterior type II. It is useful when the papillary muscle as a whole has to be shortened, usually in chronic ischemic papillary muscle or in Barlow disease. (**A**) Section of papillary muscle is excised. (**B**) Apex of papillary muscle with attached chordae is sutured to the base of the muscle resulting in shortening.

augmented with a patch of treated autologous pericardium (Fig. 102.17).

RESULTS

The results have to be presented in two groups: the group of patients before 1 year of age and the one after 1 year of age, alternately with mitral annulus <18 mm or >18 mm. In the first group, the survival of the patient is the primary goal and it is best achieved through repair. After 1 year of age, the technical conditions approach the one of adults; the repair of all valves is an achievable goal and replacement of mitral valves is exceptional. Replacement does not influence long-term survival significantly with current anticoagulation protocols and mechanical prosthesis.

Congenital Mitral Valves in Neonates and Infants

At the Royal Children's Hospital in Melbourne, VIC, 13 patients <1 year underwent initial mitral valve repair for regurgitation between 1996 and 2006. There were three reoperations with two reoperations requiring valve replacement. There was one early death in a neonatal Marfan syndrome and one late hospital death related to inappropriate orientation to biventricular repair for a patient with Shone syndrome.

During the same time span (1996 to 2006), nine patients <10 months were operated on for congenital mitral stenosis. The median age was 5 months (range: 1 week to 10 months). All had severe failure to thrive and severe pulmonary hypertension. The mean preoperative gradient was 13 ± 2.3 mmHg. The malformations included papillary muscle to commissure fusion ($n = 6$), parachute mitral valve ($n = 2$), excess tissue ($n = 1$), and supravalvular ring ($n = 1$) of whom one patient had Shone's syndrome. Three patients had reoperations. The first patient had three reoperations, leading ultimately to MVR with a mechanical valve. The second patient (neonatal Shone's complex and interrupted aortic arch) underwent reoperation (resection of supravalvar membrane) 2 years following initial intervention. There were no deaths. Other teams have similar results in this age group justifying early operation in severely symptomatic patients.

Congenital Mitral Valves in Patients Older than 1 Year of Age

Mitral Regurgitation

From the recent series available, repair should be expected in more than 90% of the patients with a risk for hospital mortality

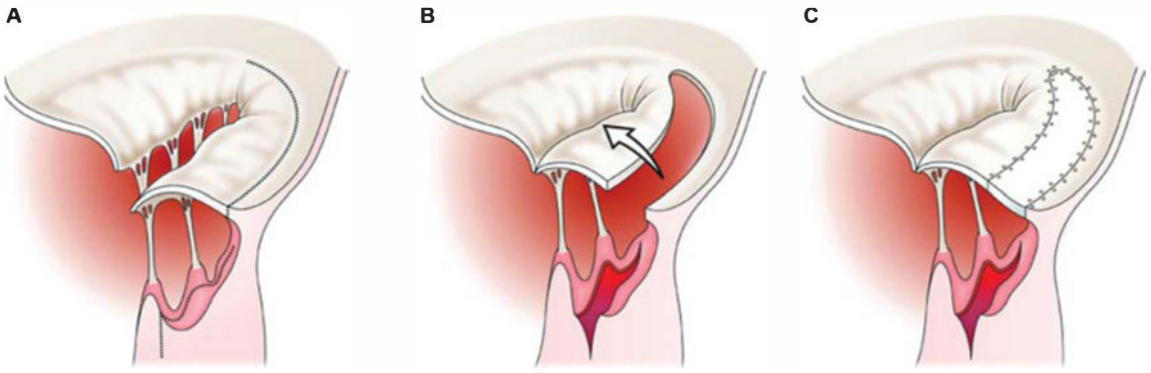


Fig. 102.12. (**A**) Detachment of the posterior leaflet allows (**B**) access to the suspension apparatus for mobilization (here in a hammock valve). (**C**) The patch enlargement of the posterior leaflet treats the type III and allows for a larger annuloplasty.

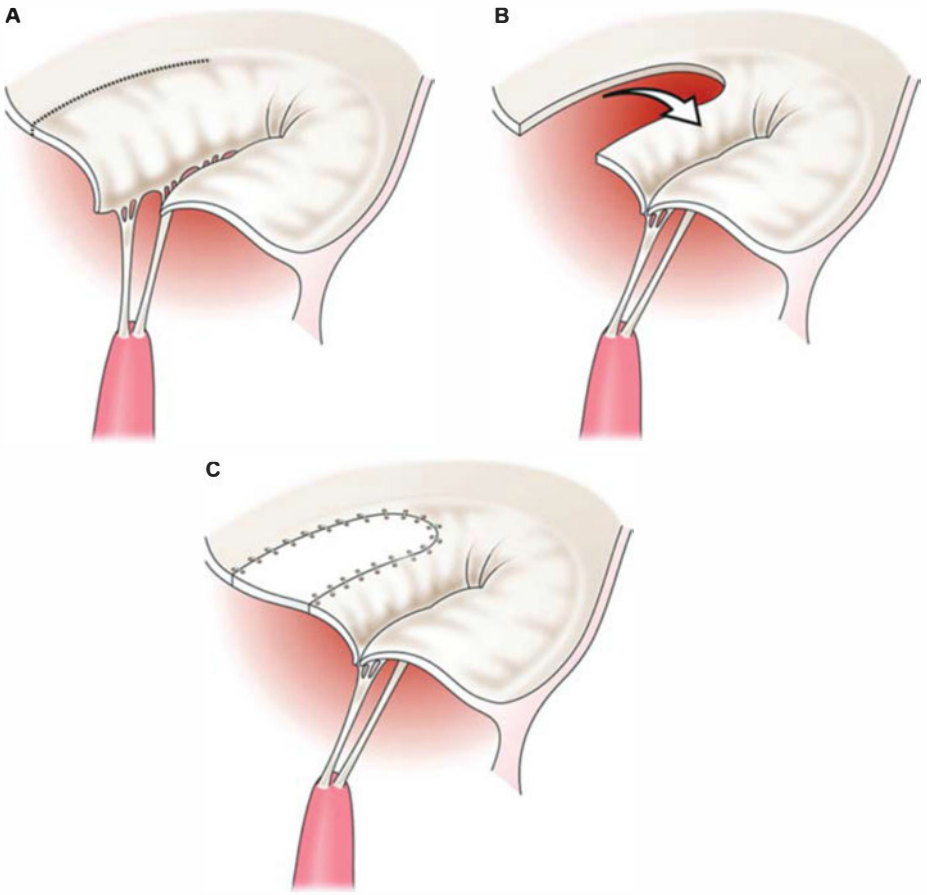


Fig. 102.13. Correction of type III with augmentation of the anterior leaflet. The patch augmentation treats the lack of valvular tissue in congenital valve or in rheumatic valve.

rate <10%. Results in the current era should be significantly improved compared with previous series stretching over a long time span. The expected reoperation rate should be <15% at 15 years.

Mitral Stenosis

The hospital mortality should be very low when the mitral stenosis is isolated. Mitral stenoses with associated cardiac lesions generate high mortality and reoperation rate but are rare in this age group. After mitral valve repair for mitral stenosis, residual gradient are frequent but often well tolerated and reoperations are indicated according to the level of pulmonary hypertension. Supravalvar mitral ring have a high recurrence rate.

Rheumatic Mitral Valves

Mitral valve repair in the pediatric age group in units with significant experience is achieved in >90% of the patients with hospital mortality lower than 2%. Large variations in the reoperation rate are reported (45% to <10% at 5 years). These variations could be attributed to the quality of the follow-up, to the regional and national specificities including whether access to surgery is free or not but most importantly to the compliance to secondary antibio prophylaxis. At the Royal Children's Hospital, Melbourne, VIC, between 1996 and 2005, 88 patients aged 6 to 24 years old had surgery for rheumatic mitral valve insufficiency. All had mitral valve repair initially. Freedom from reoperation at 70 months with 32 patients at risk is 78%. There were 15 reoperations in 13 patients of which 7 were replacements and 8 were further repairs. There was one early death and one late death.

CONCLUSION

In children with significant mitral valve disease, mitral valve repair in all is the objective. If mitral valve replacement is unavoidable, it should be delayed until it can be done at low risk. In that context, an imperfect repair represents a satisfactory palliation if the surgical indication cannot be delayed altogether.

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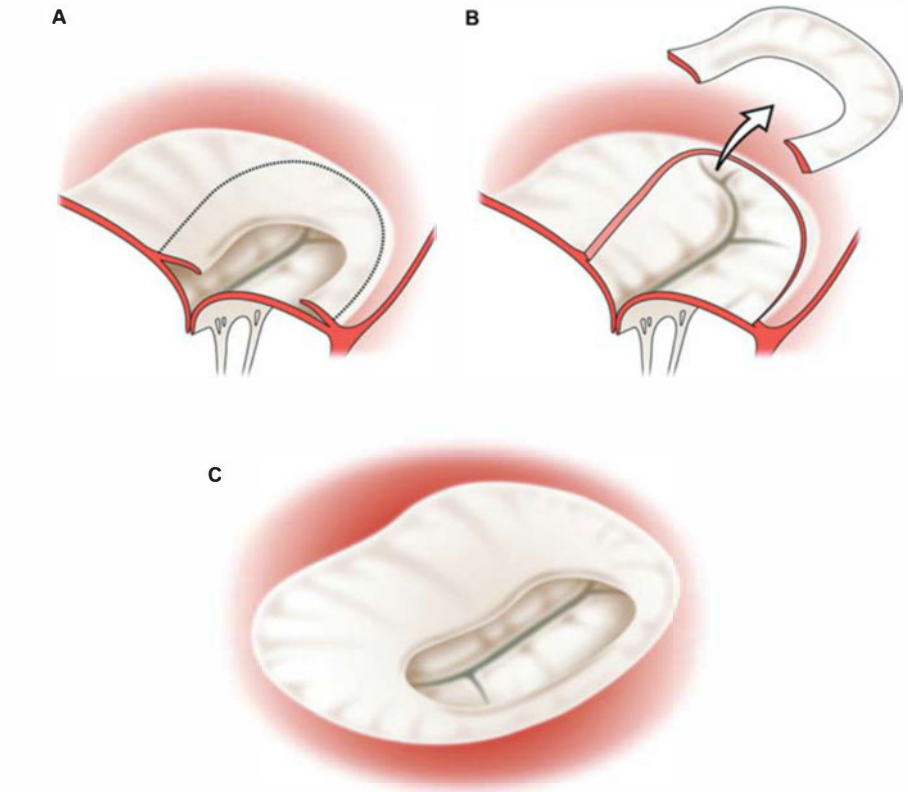


Fig. 102.14. Supravalvular ring. (**A**) The implantation of the supravalvar ring is high close to the annulus on the posterior leaflet side (*inferior side* of the picture), while it is at the mid-level of the anterior leaflet (*left side* of the picture). (**B**) The excision of the supravalvular ring is done with blunt dissection and peeling off the leaflet tissue. (**C**) completed result after excision.

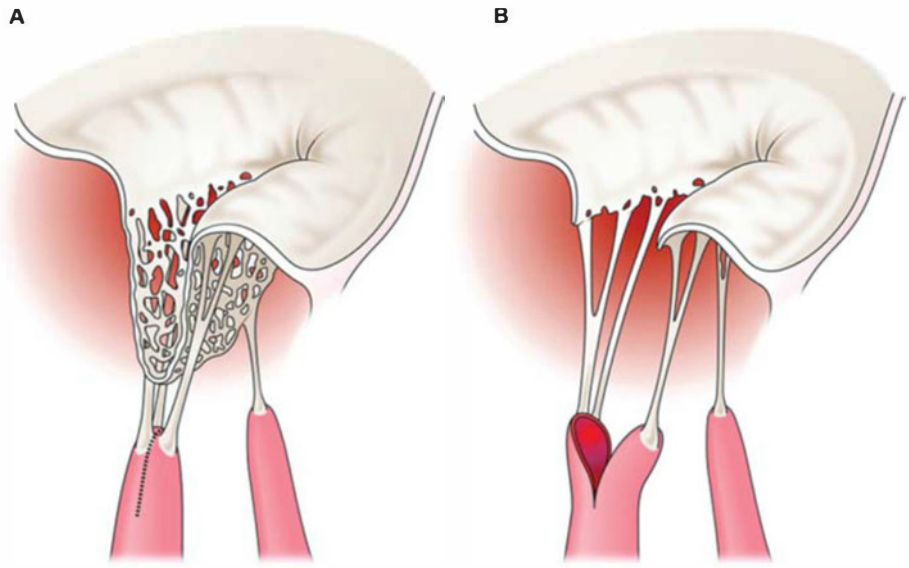


Fig. 102.15. (**A**) Excision of extra valvular tissue, here within a parachute mitral valve. (**B**) Great care must be taken to preserve the suspension apparatus.

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Fig. 102.16. (**A**) Normally developed left lateral leaflet: construction of a coaptation surface in front of the tip of the left lateral leaflet. (**B**) The strip of treated autologous pericardium is sutured to the edges of the cleft. (**C**) The free edge of the patch is suspended laterally to the thickened marginal chordae and/or using artificial chordae.

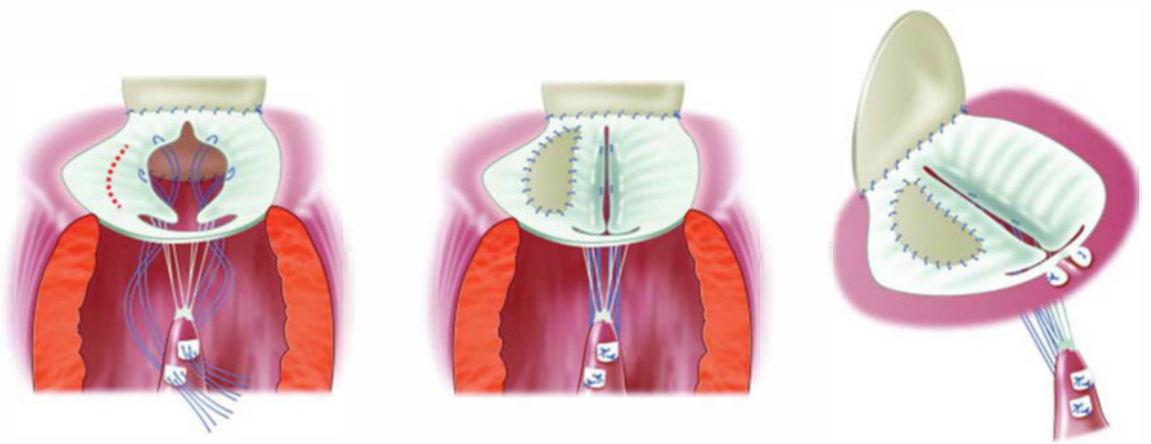


Fig. 102.17. Diminutive or absent left lateral leaflet: (**A**) suspension of the edges of the cleft with artificial chordae. (**B**) If necessary, a patch of treated autologous pericardium is used to increase the size of the superior bridging leaflet and provide redundancy allowing for a large zone of apposition (**C**).

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EDITOR'S COMMENT

Mitral valve repair continues to be a surgical challenge in pediatric cardiac surgery. The wide range of anatomic abnormalities and the often palliative nature of valve repair in association with the extreme rarity of congenital valvular abnormalities result in only relatively small reported series of operations for these abnormalities. The results of valve repair in the pediatric population for more common adult diseases (such as rheumatic heart disease) are much more reproducible, and significant series of valve repairs with good results have been published. Patients with mitral regurgitation from either a congenital or acquired cause may respond to valve reparative operations as eloquently described by Dr. Brizard; however, the surgical technique has to be individually tailored to each anatomy. The effect of newer technology, such as three-dimensional echocardiography, on the preoperative assessment of valvular function and anatomy for guiding surgical repair is unknown; however, with the development of these techniques and more real-time imaging, better assessment of valve function may lead to improved outcomes in the future.

Dr. Brizard and his colleagues have had excellent results with valve leaflet augmentation extension procedures of both the posterior and anterior leaflets. Although we have had very limited experience with these techniques at the Children's Hospital of Philadelphia, the results have not been quite as good as those presented by Dr. Brizard and his group. In situations in which a restricted leaflet motion is present, it is not clear that augmenting the leaflet surface in its superior margin with these extensions will always result in better coaptation because the free margin is still tethered by the restricted motion. In augmentation of the anterior leaflet also, the chordal attachments at the free margin may be as important as the overall surface area in creating accurate valve function. Nevertheless, Dr. Brizard makes a very good point that these leaflet augmentation methods need to be accurately tailored to the valve anatomy, and excessive patching may actually cause problems.

An area of controversy concerns the supravalvar mitral ring. There is often confusion in the reported series of repair of supravalvar mitral ring between the anatomic entity described by Dr. Brizard and the patient with cor triatriatum sinister. Membrane-like structures dividing the left atrium with a stenotic central orifice can be completely excised with a very low recurrence rate. The acquired form of supravalvar mitral ring such as described by Dr. Brizard is distinguishable from cor triatriatum by the attachment of the supravalvar ring to the mid-portion of the anterior mitral leaflet. This may well be an acquired structure as noted; however, the pathophysiology of development of the lesions is not known. Although recurrence is a possibility due to abnormal flow characteristics across the mitral valve, there have been only a few reported cases of recurrence of supravalvar mitral rings in the literature. Perhaps, this is related to the extreme rarity of the condition or the fact that the underlying stenotic or regurgitant process, which leads to the development of the ring, is addressed at the time of the resection of the ring itself. There appears to be a slightly greater incidence of supravalvar mitral obstruction when there is a left superior vena cava entering the coronary sinus, perhaps creating abnormal flow characteristics across the mitral valve.

Although infective endocarditis of the mitral valve is usually associated with mitral regurgitation, large vegetations on the mitral valve can lead to primary mitral stenosis. The most common situation is a combination of regurgitation and stenosis. If the vegetation is obstructive, resection of the vegetation and valve repair can be performed; however, regurgitation can develop as additional healing of the valve occurs. In most cases, if there has not been extensive destruction of leaflet tissue, valve reparative techniques are preferable to valve replacement in infective endocarditis with the goal of maintaining the native valve for as long possible.

Cardiac catheterization is rarely necessary in the current era for the evaluation of valvular lesions on the left side of the heart. Nevertheless, catheterization to measure right ventricular pressure and pulmonary pressure and resistance may be necessary in some cases to determine optimal timing for valve intervention. Interventional catheterization with balloon valvuloplasty of even significant congenital mitral stenosis may be an effective palliative measure in some children. There are small reported series of balloon dilation of congenital mitral stenosis that resulted in adequate relief of valvular obstruction with only modest regurgitation and permitted palliation for sometimes many years prior to the need for surgical intervention.

As noted in Chapter 103 on Aortic Valve Repair, valve reparative procedures are certainly preferable to valve replacement in all children and young adults if a reasonably durable repair can be achieved. In the mitral position, even only modest improvement in stenosis can result in significant palliation. When valve reparative techniques have failed and there is significant pulmonary hypertension or increasing congestive heart failure, valve replacement may be warranted. It is not uncommon to perform multiple valve reparative procedures for mitral regurgitation or stenosis if necessary to avoid valve replacement until an older age when a prosthetic valve can be implanted without increased morbidity and mortality.

The situation of the Shone complex of aortic and mitral valve disease remains difficult. Many patients with this complex anatomy have significant aortic disease as newborns that require balloon dilation. The balloon dilation can then result in only temporary relief of stenosis or can cause significant aortic regurgitation. These patients may then require an autograft aortic valve replacement as a newborn or young infant. Often the nature of the mitral valve disease is not readily apparent until the relief of the distal aortic obstruction is achieved. In situations in which the mitral valve is abnormal in structure, it may be best to consider single-ventricle reconstruction for these patients. Nevertheless, decision-making continues to be difficult, and many patients have an intervention on the aortic valve before consideration is given to the single-ventricle pathway. If aortic regurgitation becomes significant, then it may preclude a single-ventricle reconstruction approach such as the Norwood operation.

There has been a recent interest in attempting to create biventricular repair in patients with certain forms of hypoplastic left heart syndrome. Many of these patients have critical aortic stenosis and endocardial fibroelastosis with secondary effects on the mitral valve with mitral stenosis. The group in Boston has suggested resection of endocardial fibroelastosis and debridement from mitral valvar tissue to encourage left ventricular growth

(*continued*)

EDITOR'S COMMENTS (*continued*)

and growth of the mitral valve. While there have been successes with these approaches, the long-term effects on ventricular function and the relative function of the mitral valve and pulmonary pressures and resistance have not been completely identified. Many of these patients continue to have relative mitral stenosis with some insufficiency and some may eventually require mitral valve replacement at a relatively young age.

As suggested by Dr. Brizard, mitral valve replacement in young children should be avoided if at all possible. While results with supra-annular mitral valve replacement in very small children are suboptimal, success can be achieved with careful implantation of the device. Many of these patients continue to have relative pulmonary hypertension due to lack of compliance of the left atrial chamber above the valve prosthesis, which can limit the functional result. In addition, the need for chronic anticoagulation in growing children makes follow-up difficult.

Mitral valve procedures in children should be considered palliative. In experienced centers such as the Royal Children's Hospital, Melbourne, VIC, the success of mitral valve reparative operations is enviable and a standard to which other pediatric cardiac programs should aspire.

TLS

103 Aortic Valve Repair in Children

Chawki el-Zein, Anastasios Polimenakos, and Michel N. Ilbawi

Surgical management of aortic valve disease in children presents a difficult dilemma. On the one hand, early surgery protects the myocardium from volume and pressure overload, decreases the chance of fibrosis and remodeling, and is consistent with current surgical philosophy of early complete repair of all congenital heart defects. On the other hand, early valve replacement in children is suboptimal because of the lack of an ideal valve substitute that allows growth and does not need anticoagulation or frequent replacement. Autologous pulmonary valve has emerged recently as an attractive aortic valve substitute that fulfills these criteria but concerns persist over the long-term fate of the pulmonary valve in the aortic position.

The dichotomy created by the absence of the ideal valve substitute and the deleterious effects of long-standing ventricular volume and/or pressure overload associated with aortic valve disease has renewed interest in aortic valvuloplasty especially in children. Although several techniques, such as annular reduction, commissural resuspension, and cusp extension were used in the past, aortic valvuloplasty remained an evolving approach rather than a definitive treatment, due in part to incomplete understanding of the functional anatomy and geometry of the aortic valve. Recently, success in atrioventricular valve repair, progress in myocardial protection, refinements in three-dimensional imaging of the aortic valve, and detailed analysis of valve anatomy and function have led to improved results of aortic valve reconstruction.

ANATOMY AND FUNCTION OF THE AORTIC VALVE

The three leaflets of the aortic valve are attached to the aortoventricular junction. The collagenous condensation at the point of attachment of each leaflet has been termed the annulus fibrosis. There is, however, no true "ring" of annular tissue supporting the leaflets in a straight circular plane. The hemodynamic stresses on the leaflets, therefore, are counteracted at several structural levels. The margin of coaptation of a competent valve is more than a finite point of contact. It extends along the whole margin of the leaflet in length and several millimeters in depth. Beneath the apices formed by leaflet attachment, the so-called commissures, there are subcommissural or interleaflet triangles (Fig. 103.1). The wide base of these triangles follows the ventricular contraction pattern and allows optimal retraction of leaflets during systole. The sinotubular bar marks the junction with the ascending aorta. It is thicker than the adjacent sinuses. It is circular with areas of increased collagen. It acts as a suspension post that supports the peripheral attachments (the commissures) of the valve leaflets. The parabolic shape of the leaflets resembles a suspension bridge. Their attachments to the sinotubular bar are several millimeters above the level of coaptation. As these support poles stretch outward by as much as 16% to 44% during early systole, the leaflet edges (the cables) become straighter, aiding in the opening of the valve.

The aortic root is also a complex hemodynamic system. Its component parts change in size and shape during the cardiac cycle. Its distal portion is exposed to the aortic pressure. It expands to allow leaflet retraction. Its base is exposed to ventricular dynamics. It contracts during the peak of systole to decrease the distance between the leaflets and to reduce the stress forces applied to leaflets in early diastole. Moreover, the leaflet–sinus assembly behaves as an independent unit to store the diastolic pressure within. It allows the aortic valve to remain competent even if the interleaflet triangles are partly incised. The instantaneous changes in aortic valve orifice have been shown to precede movement of blood in the ventricle. The transformation of the aortic orifice from a closed position to a triangle and then to a circle without causing flexion deformity of cusp tissue is related to aortic root distensibility and the mechanism of leaflet suspension.

PATHOLOGY AND FUNCTION OF THE ABNORMAL AORTIC VALVE

Aortic Valve Stenosis

The Congenital Bicuspid Aortic Valve

In type I, there is no median raphe at the junction of two cusps. As a result, there are two rather symmetric aortic sinuses and leaflet base attachment. The valve orifice is central. The commissural triangle is rather well developed. The leaflets are suspended at the sinotubular bar and have adequate depth. In type II, which is more prevalent, a median raphe is present. The cusps are asymmetric and the fused leaflet is longer, shallower, and takes up more of the circumference of the valve. In contrast to the normal tricuspid valve, the leaflet edges are excessive and sagging. As a result, there is increased folding and crossing and a compensatory extension of the area of leaflet approximation from their edges (doming). The opening of the valve is eccentric due to discrepancy in leaflet sizes. The orifice also has an elliptical rather than a circular opening. The resultant distortion in blood flow pattern exaggerates turbulence and predisposes to degenerative changes. Frequently, there is commissural fusion that limits the leaflet movement and further exaggerates the eccentricity of valve opening and the decrease in its effective orifice diameter. The narrowed opening, often combined with annular hypoplasia, impairs the ability of the leaflets to escape systolic or diastolic pressure load, further exaggerating the stress on the valve. The subcommissural triangle is severely attenuated. It limits leaflet movement in early systole and the change in orifice configuration necessary for appropriate leaflet coaptation at the end of systole. The leaflet edges are suspended below the sinotubular bar. This, combined with redundant leaflet edges, results in shallow sinuses, decreases coaptation area,

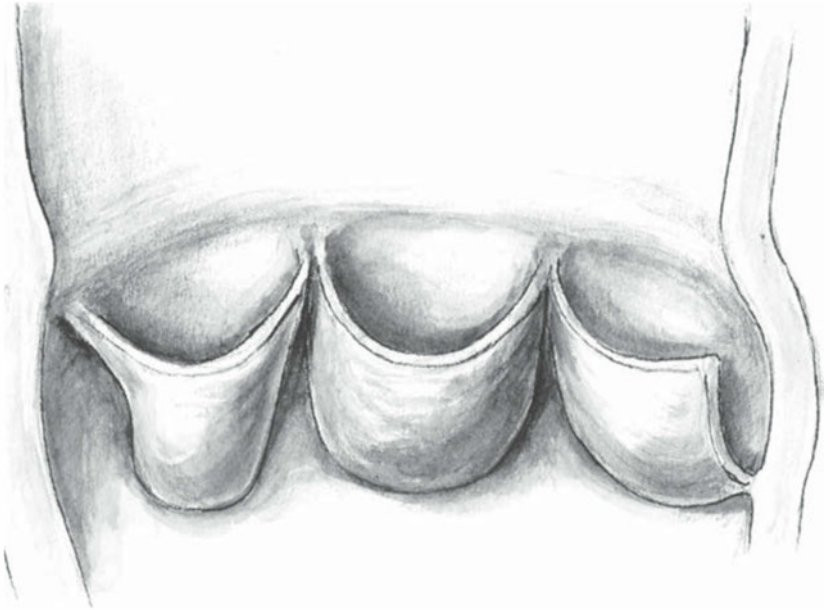


Fig. 103.1. Anatomy of normal aortic valve opened longitudinally. The relationship of the sinotubular bar to commissures is shown. The subcommissural triangle is wide and deep.

and exaggerates leaflet-deforming dynamic forces (Fig 103.2).

The Rheumatic Aortic Valve
The continued inflammatory process causes progressive scarring and thickening of the leaflets and fusion of the commissures. The valve becomes progressively stenotic.

Aortic Valve Regurgitation

There are three types of regurgitant aortic valves. Type I is dilatation of the aortic annulus, sinotubular bar, or ventriculoaortic junction. Type II is leaflet prolapse. Type III is leaflet retraction and scarring. It is the most common pathology of the congenital regurgitant valve (Fig. 103.3).

Regurgitation Associated with Ventricular Septal Defect
There is discontinuity between the aortic media and the crest of the ventricular septum with consequent decrease in the support of the sinus wall and progressive prolapse and deformity of the involved cusp (Type II). The sagging leaflet edge loses coaptation contact with the other two leaflets and central regurgitation ensues. The noncoronary cusp is usually affected with perimembranous ventricular septal defects, whereas the right coronary cusp is involved with the subarterial, more anterior (supracristal) ventricular septal defect.

Regurgitation in Patients with Congenital Valvar Stenosis
The continued trauma to the leaflet edges produced by hemodynamic stress and abnormal flow patterns results in progressive scarring, thickening, deformity, and retraction of the leaflet edges and subsequent lack of coaptation (Type III).

Aortic Regurgitation Secondary to Subaortic Fibromuscular Stenosis
The abnormal blood flow pattern produced by the subaortic stenosis results in progressive deformity of the leaflet. Tethering of the leaflets by the subvalvar fibrous tissue, causing the obstruction, exaggerates the regurgitation (Type III).

Regurgitation in Marfan Syndrome
The pathology is progressive dilation of the aortic root wall due to fragmentation of its elastic support. The dilated sinotubular bar and valve sinuses stretch apart the commissural suspension and leaflet edges. The increase in hemodynamic stress due to changes in the leaflet suspension mechanism combined with enlarged aortoventricular junction leads to poor leaflet coaptation and central regurgitation (Type I).

Postballoon Regurgitation
This condition is usually caused by leaflet(s) tear close to the fused commissure. The leaflet becomes flail and eccentric regurgitation results (Types II and III).

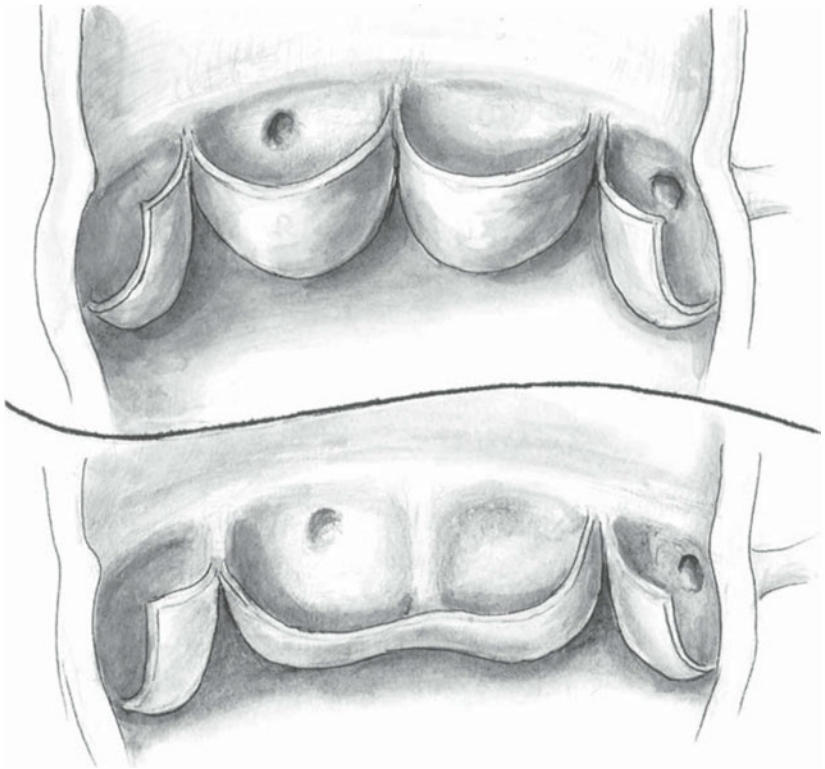


Fig. 103.2. Comparison of normal and bicuspid stenotic valves. The sinuses are shallower and the edges of the leaflets are sagging. The sinotubular bar is superior to the commissures.

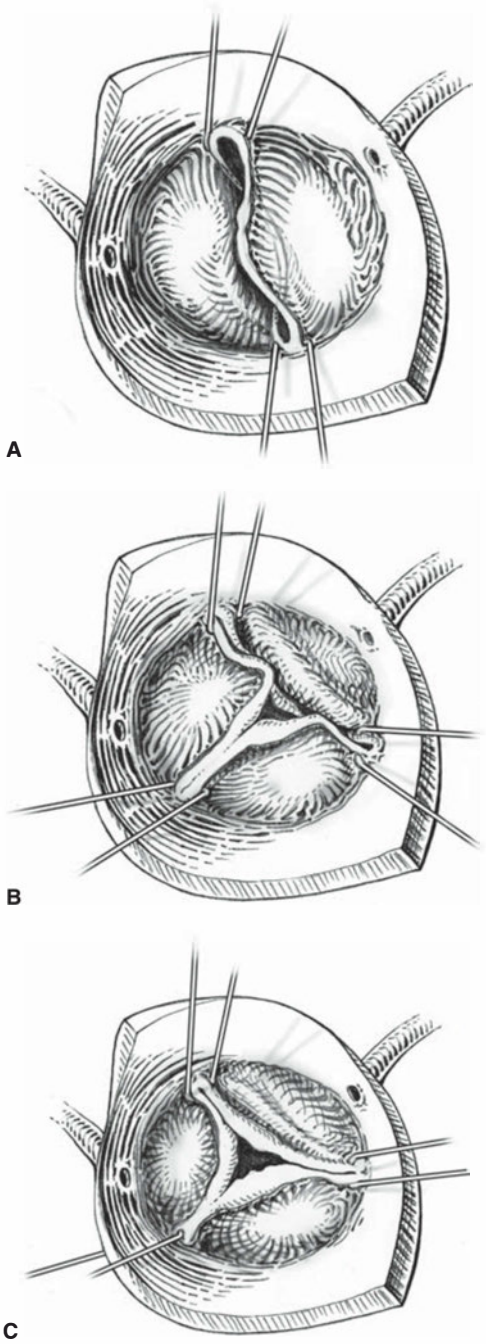


Fig. 103.3. The different pathology types or congenital aortic valve regurgitation. (**A**) Leaflet prolapse. (**B**) Leaflet retraction. (**C**) Annular dilatation.

Regurgitation after Arterial Switch Operation

Regurgitation in these cases is related to disruption of the sinotubular mechanism and undue dilation of the aortic sinuses caused by the implantation of large coronary artery buttons or preoperative pulmonary artery banding. Delayed closure of ventricular septal defect associated with D-transposition of the great arteries also predisposes to long-term neoaortic regurgitation following the switch operation (Type I).

Aortic Regurgitation Secondary to Rheumatic Disease

There is cusp retraction secondary to inflammation and scarring. The hemodynamic sequelae result in progressive annular dilation and worsening of the regurgitation (Types II and III).

TIMING OF SURGICAL INTERVENTION

To achieve optimal short- and long-term results, surgical intervention should be timed appropriately. The decision relies on achieving the goals of valve surgery, which include relief of symptoms, restoration of exercise capacity, improved quality of life, and, most importantly, protection of the myocardium from chronic pressure and/or volume overload. Most of the reported guidelines for timing of valve surgery are based on studies in the adult population and on the premise that valve replacement is the only therapeutic option. These studies use mortality rates as a follow-up endpoint but fail to analyze myocardial performance and reserve several years postoperatively. They utilize as their database several single-center observational studies and very few prospective, randomized trials.

The introduction of and refinement in valvuloplasty techniques have prompted critical evaluation of these older guidelines for the timing of surgical intervention on the diseased aortic valve. The availability of a surgical alternative that avoids valve replacement or anticoagulation has liberalized the older rigid criteria. Although large-scale, long-term data on repaired valves are not available, there is unquestionable evidence that valvuloplasty extends the functional longevity of the native aortic valve in children and may safely delay the need for replacement, thus justifying earlier surgical intervention. Waiting for symptoms to appear or for ejection fraction to decrease prolongs the duration of ventricular pressure and volume overload and may lead to irreversible ventricular dysfunction.

Timing of valvuloplasty involves several two- and three-dimensional echocardiographic and Doppler-derived indices. For isolated aortic valvar stenosis, pressure gradients of 40 to 50 torr associated with progressive left ventricular hypertrophy or impaired exercise tolerance are indications for intervention. Measurement of effective valve orifice and the extent of valve pathology are also helpful in deciding the timing of surgery. In aortic valvar regurgitation, a diameter ratio of regurgitant jet to annulus of $\geq$0.4 and the progressive increase in indexed end-diastolic left ventricular dimensions for two consecutive measurements, if they exceed a *Z*-score of 3, have been found to correlate with early ventricular dysfunction before onset of symptoms, and therefore constitute valid and rather objective indications for intervention. This is especially pertinent as isolated data have shown that patients with significant dilatation of the left ventricular cavity have an increased incidence of recurrent aortic insufficiency even with a successful valvoplasty. However, in specific situations such as balloon-induced aortic regurgitation, there are probable benefits for delaying surgery if the residual pathology is not severe. Remodeling of the leaflets and enlargement of the annulus from increased blood flow during the waiting period may improve the long-term outcome of the valvuloplasty. Other parameters, such as changes in ejection fraction have also been used to time interventions; however, these have not been found to correlate objectively with optimal surgical outcome.

Techniques of Surgical Valvuloplasty

Most of the surgical techniques used for aortic valve repair were devised many years ago or have been used for a long time. Recent improved understanding of the valve pathology and pathophysiology and refinements in cardiopulmonary bypass and myocardial protection in children have made their successful application possible. In addition, several principles have evolved that helped in improving outcome. These include the following: (1) detailed pre- and intraoperative analysis of pathology is essential. This is best achieved by two- and three-dimensional echocardiography. Objective preoperative assessment, however, is rather difficult. Pliability of each cusp is estimated by the apparent change in its area from systole to diastole. Adequacy of cusp tissue for coaptation is estimated by measuring the curvilinear height of the leaflet. Three-dimensional echocardiography helps in measuring leaflet-free edge, the depth of the sinus, and areas of tissue deficiency or prolapse. (2) More than one technique needs to be performed to achieve both competence and relief of obstruction. The different steps in the procedure should be tailored to address the specific pathology types. (3) Reconstructive steps should be preceded by relief of obstruction as completely as possible. All areas of leaflet fusion or stenotic lesions should be relieved first even if that reduces leaflet support. Subsequent reconstructive steps of such leaflets should aim at restoring the normal morphology, function, and support.

(4) Repair of only one leaflet or cusp is inadequate and leads to early failure. (5) Fresh autologous tissues such as pericardium or fascia lata cannot withstand dynamic stress when used for repair and tends to retract and scar with time; therefore, glutaraldehyde fixation is necessary. (6) "Overcorrection" in cases of aortic valve incompetence might be needed, but excessive correction may lead to crowding and distortion of the repaired valve if the root is normal or smaller than normal in diameter. (7) Centralizing blood flow through the valve decreases turbulence and extends the longevity of the repair; therefore, tricuspidization of the valve is advantageous when possible. (8) It is essential to incorporate in the procedure the necessary steps that address the interaction between aortic root dynamics and valve mechanics, namely, maintaining annular and commissural flexibility and movement in order to avoid accelerated stress-induced valve degeneration. Mobilization of the subcommissural triangle and avoiding subtotal excision of the leaflets close to the aortoventricular zone are important to avoid the disruption of the delicate and complex relationship between the root and leaflet. (9) Continued root dilatation whether at the aortoventricular or sinotubular bar junction induces recurrent aortic insufficiency and render the repair ineffective. The integrity of these junctions should be restored whenever they are dilated. (10) The abnormal ascending aorta should be replaced at the time of valvuloplasty especially in patients who demonstrate associated morphologic, histologic, or molecular abnormalities of the aortic wall.

All valvuloplasties are performed through an oblique aortotomy above the sinotubular bar. Antegrade and retrograde blood cardioplegia are used. Placement of commissural sutures aids exposure. Axial traction (perpendicular to the annulus plane) applied to the commissural sutures allows assessment of the valve leaflets and annulus. In addition, a centrally placed suture helps in evaluating the structure, the deficiency, and the redundancy of the different cusps by traction on the suture and pushing the leaflets gently toward the ventricle (Fig. 103.4).

Techniques for Isolated Stenotic Valve with or without Annular Hypoplasia

Unrolling and Thinning of Leaflets

Patients with chronic stenosis, turbulence, and abnormal blood flow patterns have progressive leaflet thickening and distortion.

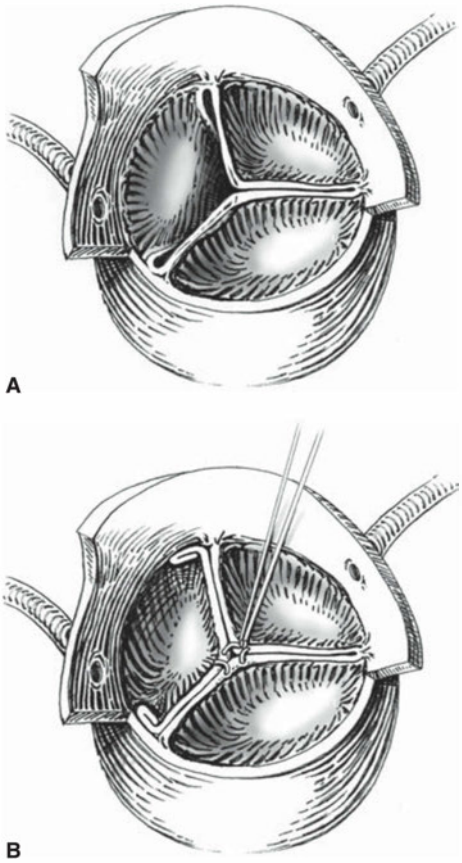


Fig. 103.4. Centrally placed suture helps in outlining the different abnormalities of the leaflets. (**A**) Leaflet prolapse. (**B**) Place of the central suture.

As a result, nodules of fibrous tissue are formed and edges are rolled away from the center, further restricting leaflet mobility. Thinning and unrolling of leaflet edges may improve valve function but has to be combined with other techniques to yield long-lasting benefits.

Simple Commissurotomy

The technique involves incising along the fused edges of the leaflets to allow improved mobility. Traditionally, the incision is extended up to the commissure but stops short of the aortic wall to avoid leaflet detachment and subsequent regurgitation. Although this technique results in larger valve orifice, it fails to address several pathologic features of congenital aortic valve stenosis and therefore has had poor long-term outcome.

Extended Commissurotomy

This approach attempts to improve on the results of simple commissurotomy. The incision along the edge of the fused leaflets is extended into the aortic wall in curvilinear fashion, splitting the inner media at the leaflet junction with the wall. The added extension results in a longer free leaflet edge, mobilizes the subcommissural triangle, and centralizes blood flow across the orifice (Fig. 103.5). However, it decreases leaflet support and may result in long-term progressive regurgitation if it is not combined with leaflet augmentation and suspension.

Techniques for Regurgitant Valves with or without Annular Dilatation

1. *Commissural reduction annuloplasty:* U-shaped pledgeted sutures are placed at one or more commissure(s) to plicate the aortic wall and reduce its total circumference. It is primarily applicable to valves with normal cusp mobility and moderately dilated annulus. Severe annular dilation cannot be repaired using this technique (Fig. 103.6).

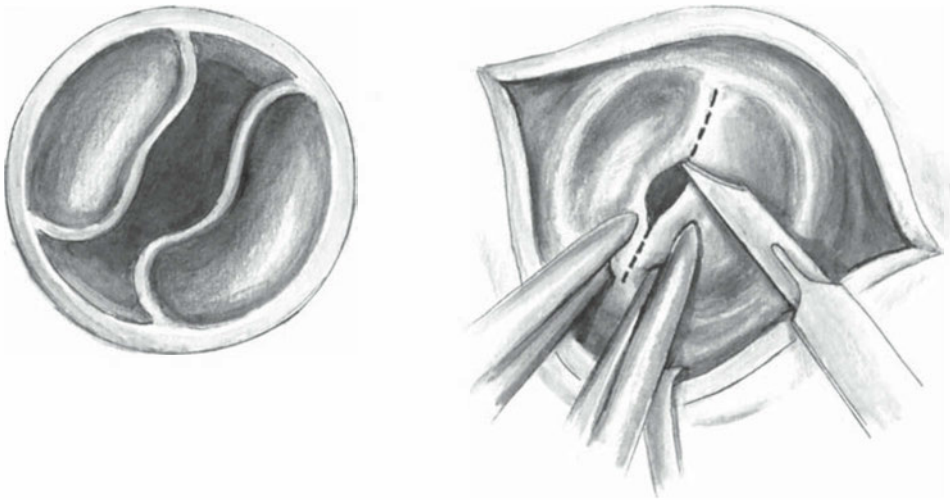


Fig. 103.5. Technique of extended commissuratory. The incision is extended into the aortic wall on both sides to increase the mobility of leaflets.

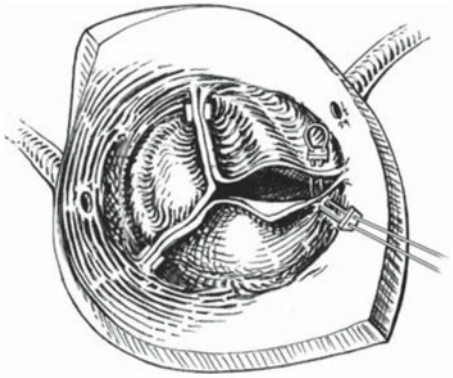


Fig. 103.6. Annular plication: sutures are placed at the base of the commissure and adjacent aortic wall to reduce the diameter and support the leaflet(s).

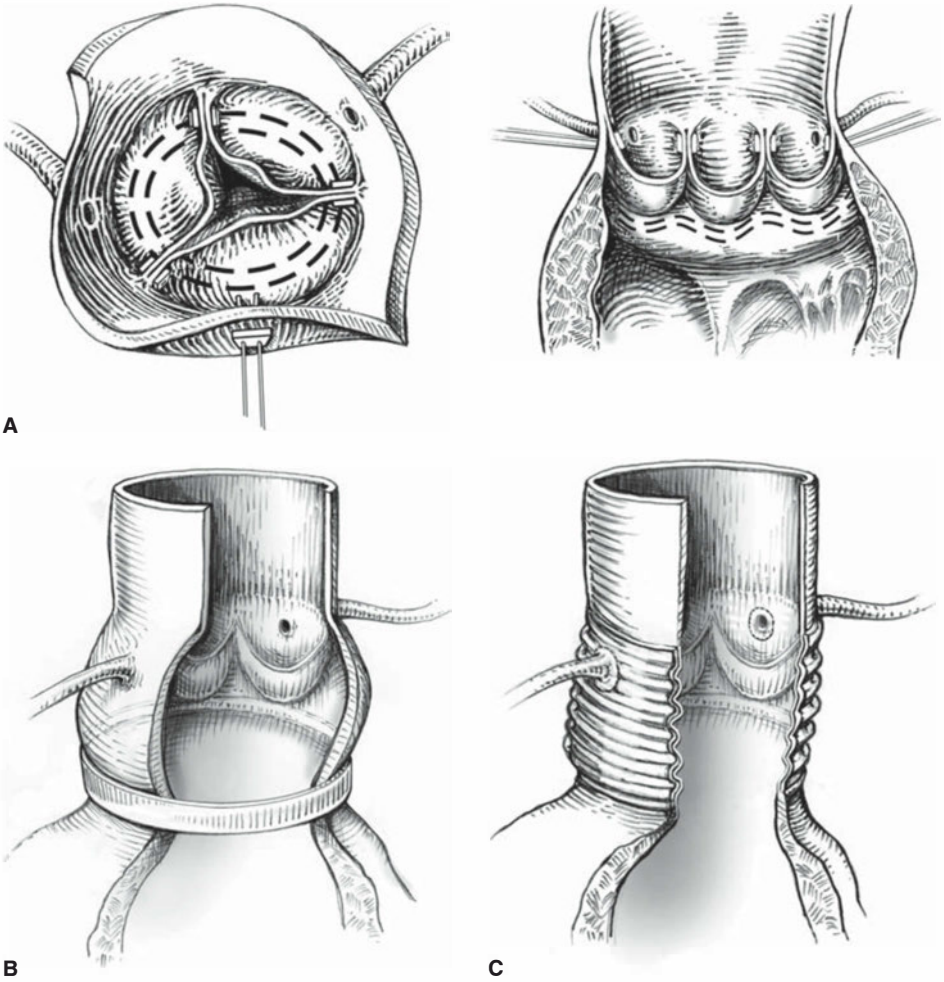


Fig. 103.7. Techniques of reduction annuloplasty: (**A**) A suture is passed in and out at the base of the aortoventricular junction in a circular fashion shown in transverse and longitudinal view. (**B**) Similarly, a wide band is passed around the base. (**C**) A cylindrical support for the dilated root.

2. *Circumferential reduction annuloplasty:* Annular reduction using a circumferential suture around the base of the valve leaflets at the aortoventricular junction can reduce the dilated annulus diameter and effect better leaflet coaptation. Alternatively, the use of external "subvalvar" prosthetic ring or band annuloplasty can also help to reduce root dilatation especially in patients with connective tissue diseases. The technique involves extensive dissection of the root down to the aortoventricular level, and adjusting its diameter over a dilator. Anatomically, however, there might be limitations to placing the circumferential band or suture at one plane, resulting in lack of symmetry of the annular constriction and consequent distortion of the valve. Similarly, placing a cylinder of prosthetic material around the whole valve apparatus reduces aortic root dilatation but could cause crowding and distortion (Fig. 103.7). The use of these different circumferential reduction annuloplasty techniques limits growth potential and results in future stenosis especially in children and young adults.
3. *Patch closure of discrete leaflet perforation:* Isolated perforation of the leaflet due to infective endocarditis or catheter intervention can be easily repaired using a small patch of autologous or bovine pericardium secured in place with multiple sutures. This simple approach is applicable only to cases where the rest of the valve apparatus is essentially normal. Continuous suture should be avoided to prevent purse-string distortion of the leaflet (Fig. 103.8).
4. *Plication of the prolapsing leaflet:* The free margin of the leaflet is plicated and resuspended to the aortic wall, or the prolapsed leaflet is pulled cephalad at both commissures and the redundant edge plicated above the level of both commissures (Fig. 103.9). These techniques are rarely successful when performed as an isolated step. Additional leaflet augmentation with a thin strip of pericardium and resuspension of the reconstructed leaflet at the commissures (see below) are needed to support the repair.



Fig. 103.8. Patch closure of a leaflet perforation.

5. *Partial leaflet resection:* In the presence of restrictive fibrotic or calcified raphe, a triangular resection of the portion of the leaflet involving the raphe is made. Edges are approximated either primarily or using a triangular piece of pericardium (Fig. 103.10).
6. *Leaflet augmentation and resuspension* is the most versatile technique. After thorough mobilization of all aortic valve leaflets (see techniques for reduced mobility), a piece of autologous or bovine pericardium is fashioned (Fig. 103.11A). It is cleared completely from attached fatty tissue. The curvilinear longitudinal dimension GHI should equal the length of the leaflet free edge plus two 0.5-cm extensions on both sides (FA and EJ). The exact dimensions of the pericardial patch could be obtained from echocardiographic studies done on normal aortic valve in which the relationship between the intercommissural distance and the leaflet dimensions (depth and

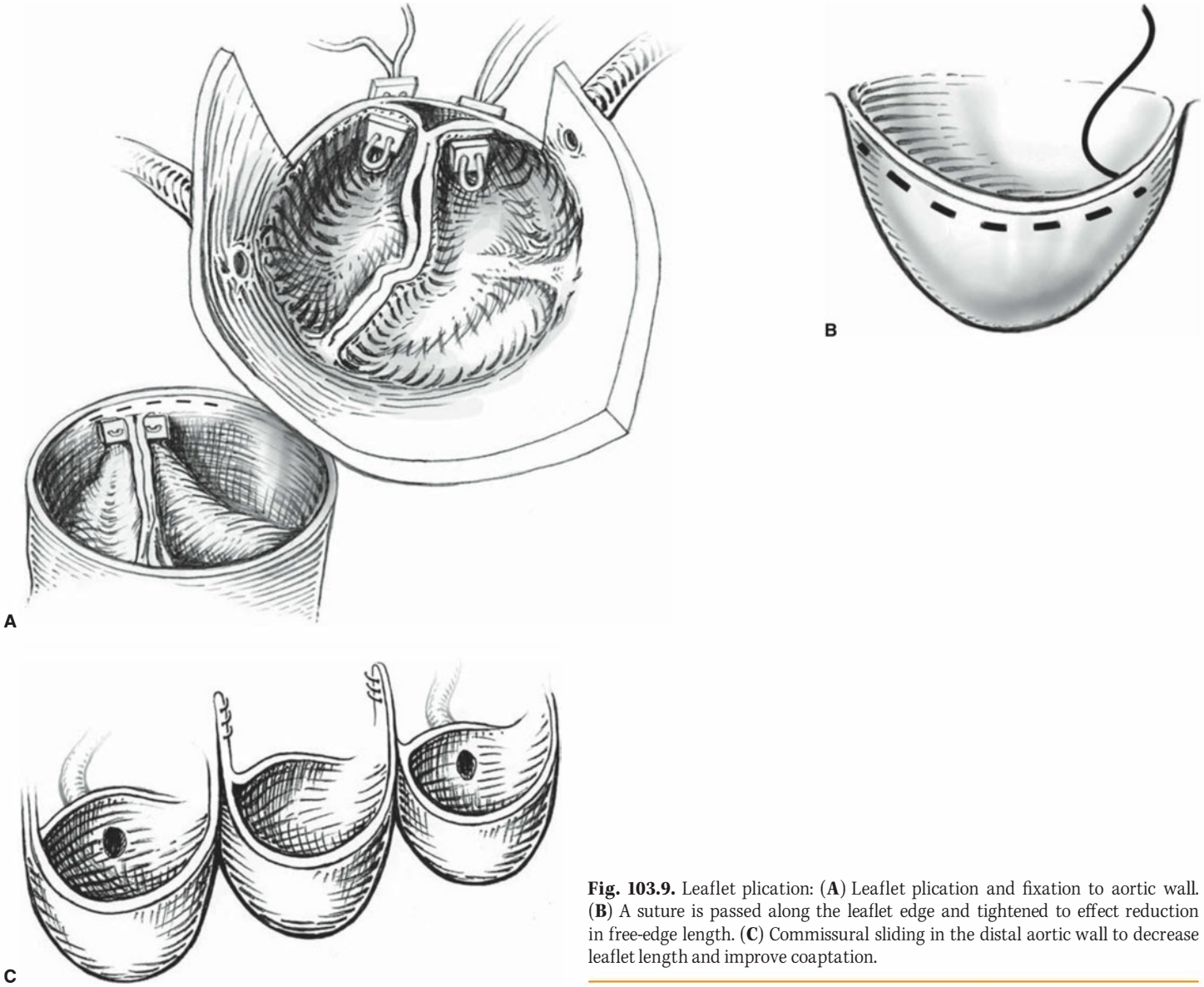


Fig. 103.9. Leaflet plication: (**A**) Leaflet plication and fixation to aortic wall. (**B**) A suture is passed along the leaflet edge and tightened to effect reduction in free-edge length. (**C**) Commissural sliding in the distal aortic wall to decrease leaflet length and improve coaptation.

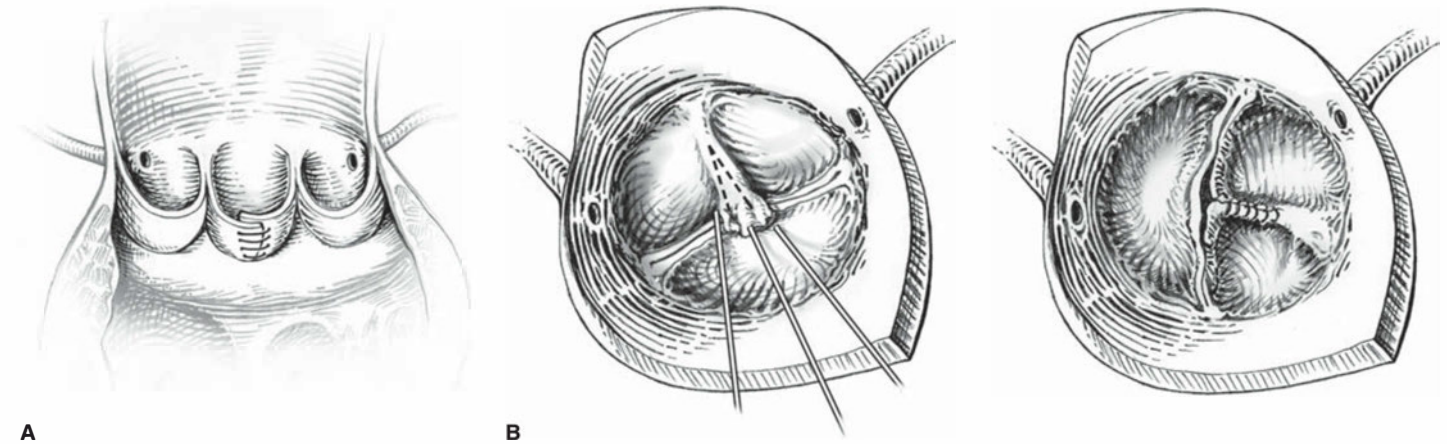


Fig. 103.10. Triangular resection and repair of redundant leaflet. (**A**) The leaflet is reconstructed primarily. (**B**) The raphe is resected and the edges resutured.

length) has been established. The patch is tailored to fit the deficiencies and irregularities of the leaflet edge. The longitudinal dimension (BCD) should be slightly straighter and shorter than the GHI (Fig. 103.11A). The width (CH) should provide enough additional depth to the leaflet so that the reconstructed leaflet free edge is in level with the sinotubular bar at the commissures but deeper (more caudad) at the center (C). The pericardial patch is sutured to the free edge of the leaflet using 5-0/6-0 polypropylene running suture starting at the leaflet center. The patch ends (or wings) are then sutured to the aortic wall with pledgeted suture, resulting in a rounded leaflet edge suspended at the sinotubular bar (Fig. 103.10B). The suspending sutures are placed in such a way as to enhance the sinotubular bar. The step is repeated for at least one additional leaflet. The heights (width) of the augmenting patches should equalize the depth of all sinuses (Fig. 103.11C and 103.11D). This allows effective leaflet enlargement and increases the surface of coaptation in the center of the sinuses. In bicuspid valves with fused but well-developed rudimentary commissure, the commissure is incised all the way to the aortic wall and the valve is converted into a tricuspid valve. The three leaflets are then augmented. An alternative approach to increase sinus depth and improve leaflet coaptation area is basal enlargement of the cusp with an autologous pericardial patch. An incision is made directly at the base of the cusp that needs enlargement. It is extended in both directions toward the commissures. The patch is trimmed to an oval form few millimeters larger than the defect created by the incision and its patch width is determined by the extent of the deficit in the coaptation surface. It is sewn into the leaflet using running and interrupted sutures to avoid purse-string deformation of the cusp. Resuspension of the leaflet edges at the commissure is then performed to allow normal coaptation plane (Fig. 103.12).

Additional Techniques for Aortic Valve Repair

1. *Tricuspidization of quadricuspid valves:* This technique is especially helpful in very young patients with regurgitant truncal valve where one cusp is rudimentary and dysplastic. The edges of the abnormal cusp are sutured to the adjacent leaflets edges resulting in three

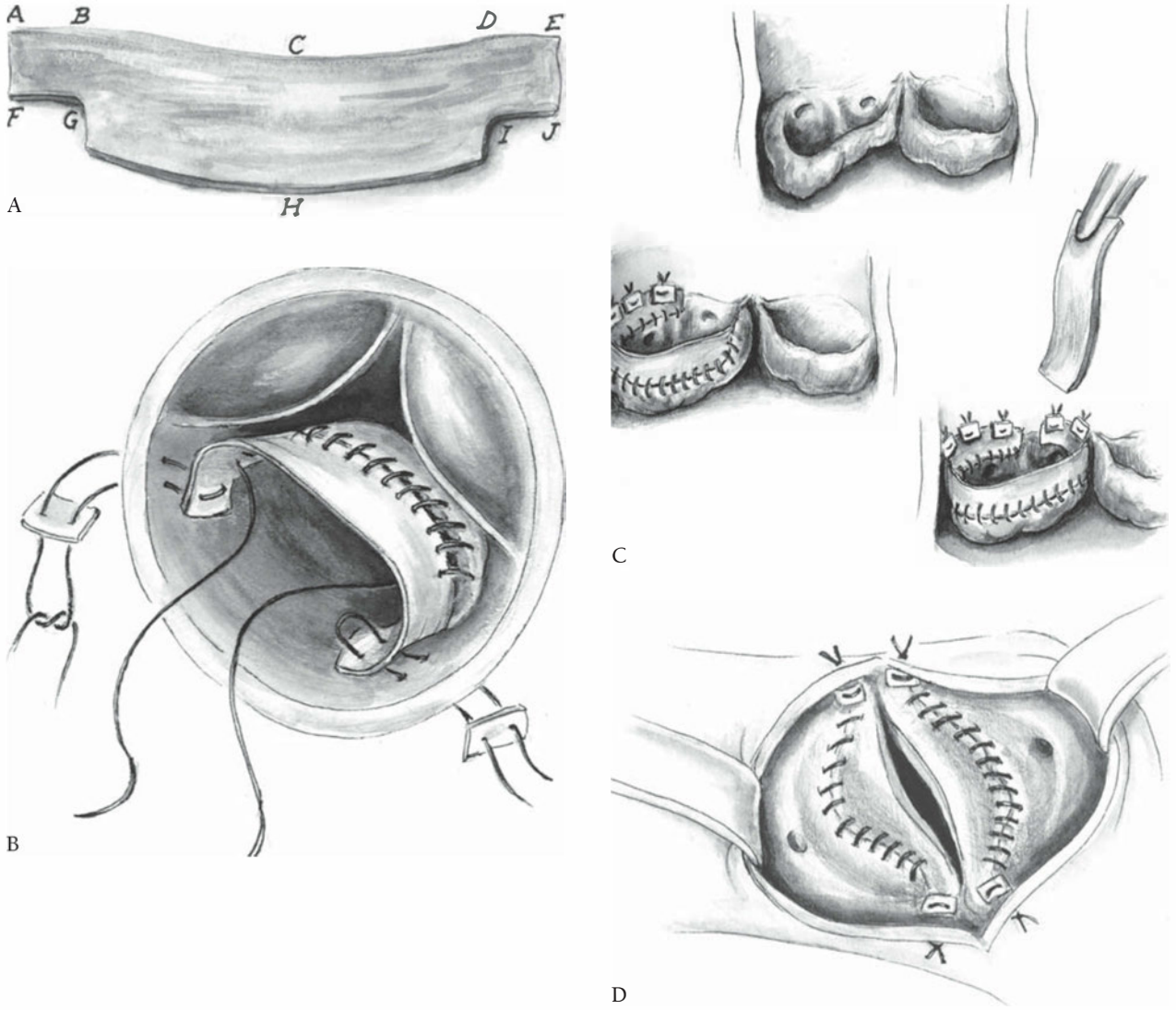


Fig. 103.11. Technique of augmentation valvuloplasty. (**A**) The harvested pericardial patch. (**B**) Augmentation of the leaflet with patch. (**C**) Side view. (**D**) Final result.

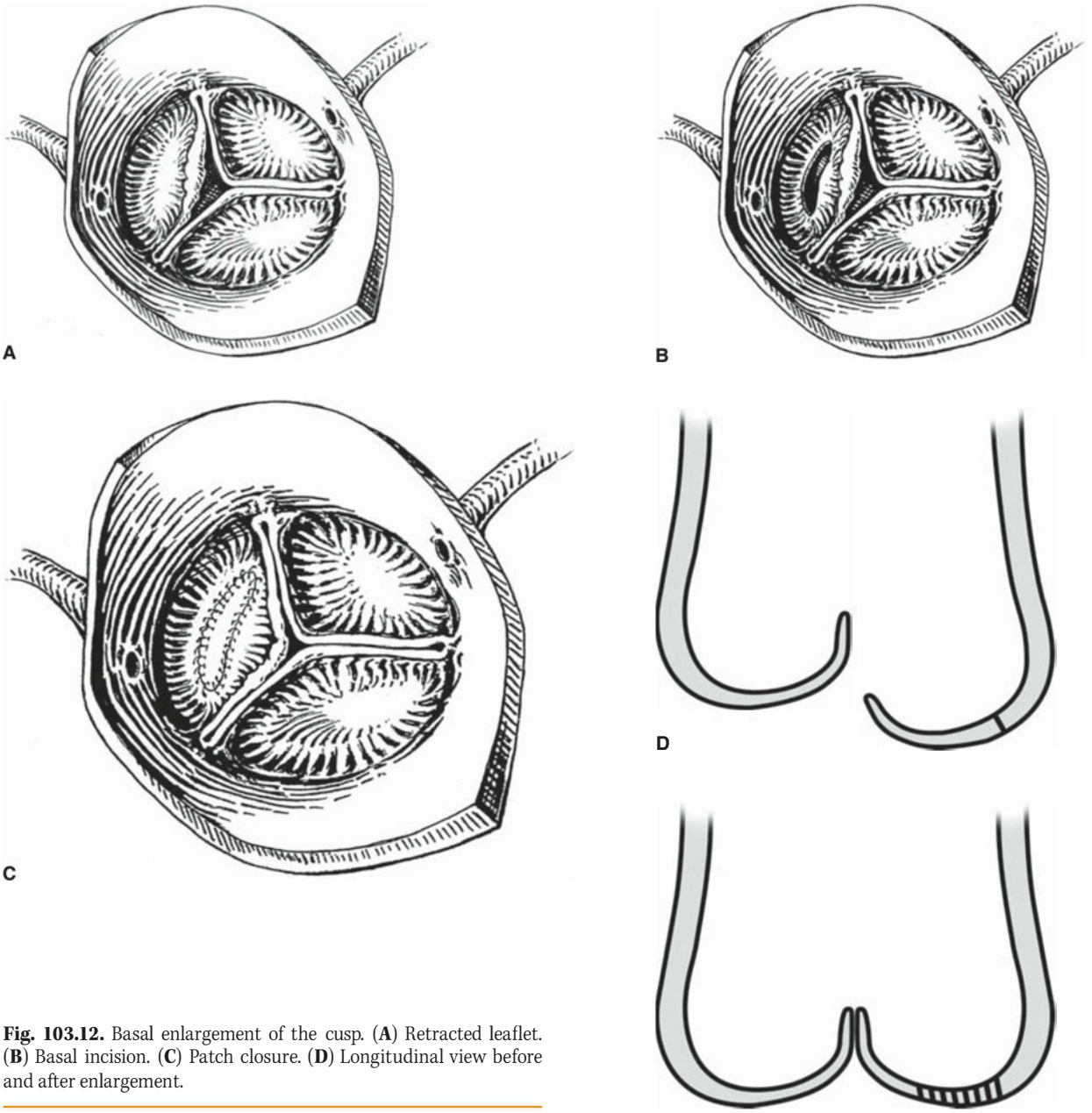


Fig. 103.12. Basal enlargement of the cusp. (**A**) Retracted leaflet. (**B**) Basal incision. (**C**) Patch closure. (**D**) Longitudinal view before and after enlargement.

functional leaflets and by providing support to the prolapsing and rudimentary leaflet, regurgitation is minimized (Fig. 103.13). Another approach is to excise the dysplastic leaflet and its corresponding sinus wall thus effecting a reduction in the circumference of the valve and improving coaptation. An additional subvalvar annuloplasty can be applied to the dilated aortic (truncal) root if further reduction in diameter is needed.

2. *Subtotal leaflet excision and reconstruction:* Excision of all the leaflets of the diseased valve and leaving a narrow strip of cusp tissue at the aortoventricular junction have been used. The leaflets

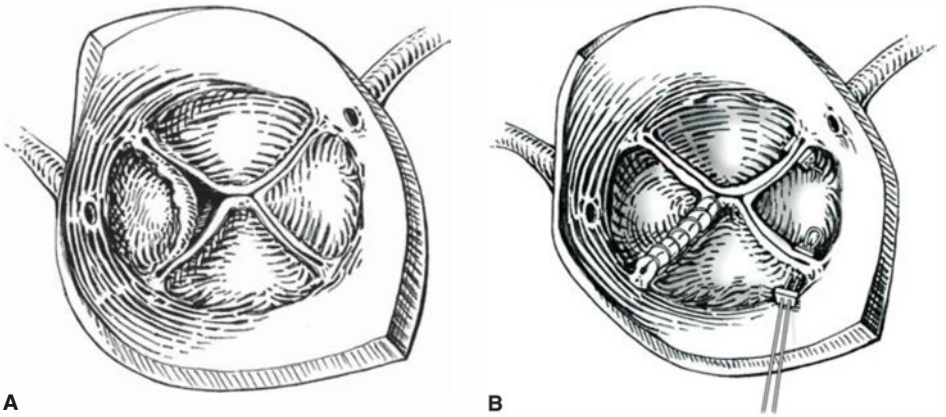


Fig. 103.13. Tricuspidization of aortic valve. (**A**) The rudimentary leaflet. (**B**) Conversion to three leaflets and commissuroplasty.

are then reconstructed by a continuous strip of glutaraldehyde-treated autologous pericardium, fashioned into three cusps and sutured to the leaflet remnants. The newly constructed leaflets are then suspended at the commissural area to prevent prolapse. This approach may impair the dynamic interaction of the aortic valve leaflets with the root and the left ventricle, leading to early failure of the repair.

3. *Bicuspidization of tricuspid valves:* This approach is rarely used. It is an easy surgical alternative in patients with tricuspid aortic valves and a large annulus when one of the three leaflets sags or prolapses, thereby causing significant regurgitation. Its use, however, is limited to patients in whom two of the three cusps are normal.
4. *Leaflet creation:* This technique is used when there is at least one relatively normal leaflet. The rudimentary part of the fused leaflet is partially detached from the annulus and used to reconstruct a normally functioning leaflet. The unguarded area of the annulus thus created is filled with a cusp made of pericardium (Fig. 103.14).

The choice of patch material for valve augmentation has evolved through the years. As mentioned earlier, fresh autologous pericardium does not withstand the hemodynamic stress. Treating the pericardium with a high concentration of glutaraldehyde (2%) resulted in a very stiff patch and predisposed to early calcification and stenosis. The use of low concentration of glutaraldehyde (0.6%) with a short fixation time (3 minutes) results in a more pliable patch material and decreases the incidence of restenosis. Fixing the pericardium on a curved structure such as a malleable retractor shapes the patch like a cusp (Fig. 103.15). Thin bovine pericardium, processed using infrared wave technology, provides a good alternative with minimal calcification or stenosis on short follow-up; however, it has failed to withstand the long-standing hemodynamic stresses that the aortic valve is exposed to. As a result, mid-term degeneration of the patch has resulted in relatively early recurrence of regurgitation. The newly introduced patch material made of decellularized intestinal mucosa (CorMatrix) has been used for reconstruction of the pulmonary and aortic wall. It is pliable, strong, and does not calcify. However, its longevity on the aortic side especially as a valve leaflet has not been objectively evaluated.

TECHNIQUES FOR DILATED AORTIC ROOT

As mentioned above, aortic root dilation is an important aspect of aortic valve disease. Higher freedom from recurrent aortic regurgitation has been observed where the valve repair was combined with root replacement, especially when the dilated aorta has an asymmetric configuration.

RESULTS

The techniques of aortic valvuloplasty are in evolution. Several series have reported results using one or more of the techniques mentioned here. None of these results, however, reflects the present knowledge of the surgical anatomy or the outcome of valve repair when these technical steps are used in combination.

Review of our total experience with these different approaches revealed a significant drop in pressure gradient across the valve, a decrease in aortic regurgitation as judged by grade and by ratio of the regurgitant jet to aortic annulus diameter, and a decrease in indexed left ventricular end-diastolic volumes.

Long-term postoperative follow-up revealed a progressive increase in pressure gradient in 42% of patients, associated with stiffening or calcification of the patch used for leaflet augmentation. When restenosis was analyzed further, it was apparent that patients who had valvar stenosis and annular hypoplasia had the highest incidence of restenosis. This increased incidence might have been due to the crowding of the hypoplastic aortic root when aggressive overcorrection was used early in our experience. Recurrence of regurgitation was not

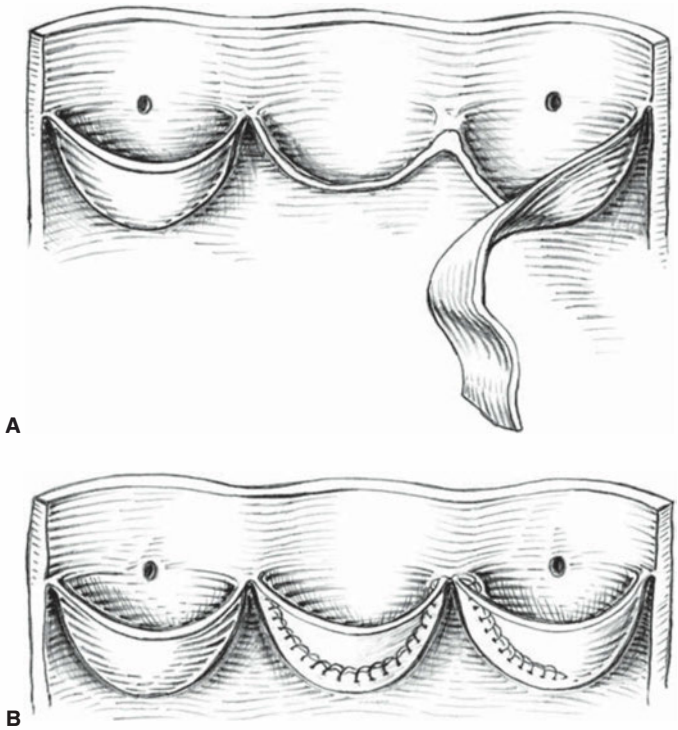


Fig. 103.14. New leaflet reconstruction. (**A**) Removal of the scared and deformed leaflet. (**B**) Reconstructed leaflet. The middle cusp is made of pericardial patch.

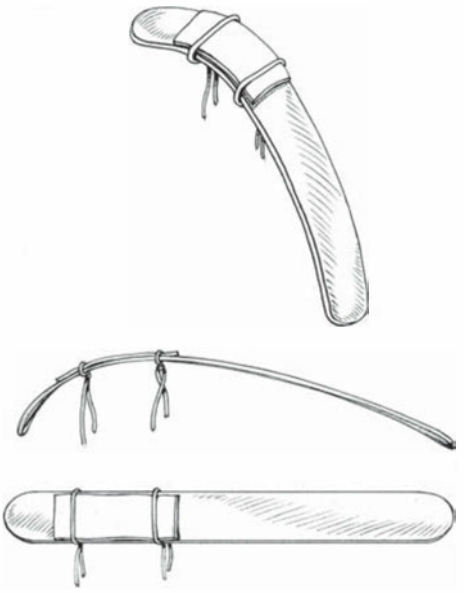


Fig. 103.15. Fixation of the patch on a curved structure allows better configuration of the leaflet edges.

a problem in follow-up, and most patients maintained competent valves. Based on the initial follow-up results, we have adopted a selective approach to aortic valve disease in children. Patients with primary valvar stenosis and associated annular hypoplasia undergo Ross with Konno procedure. On the other hand, patients with primary regurgitation or annular dilatation are best managed early by using the described valvuloplasty techniques.

CONCLUSIONS

Aortic valvuloplasty as currently used has very low operative mortality. It provides an excellent alternative to valve replacement, especially in children and young adults, in whom anticoagulation and need for repeated valve replacements are serious drawbacks. It maintains the patient's own valve and does not preclude other alternatives when deemed necessary. It is probably superior to the Ross procedure in patients who are very young or have significant annular dilation due to regurgitation or other causes. Its use in patients with aortic annular hypoplasia should be limited to avoid recurrent stenosis. The use of patch material not fixed with glutaraldehyde might provide a decrease in the incidence of restenosis.

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EDITOR'S COMMENTS

There has been a resurgence of interest in aortic valve repair based on the pioneering work of Dr. Ilbawi and his associates. As very nicely summarized in this chapter, the complex geometry of the aortic valve requires multiple approaches to achieve a competent repair, and the procedure must be tailored to the individual patient's anatomy and pathophysiology. The use of valve extension techniques as pioneered by Dr. Ilbawi has permitted the creation of trileaflet valves even in patients with bicuspid valves to achieve a more central orifice and improve the long-term outcome.

Valve reparative techniques may offer the option for earlier surgical intervention in patients with significant aortic stenosis and insufficiency. The indications for intervention, however, remain somewhat vague. In most cases, stenosis with a measured peak-to-peak gradient of >40 to 50 mmHg is a relative indication for surgical or balloon intervention, especially when there is associated left ventricular hypertrophy or strain pattern on electrocardiogram. Exercise testing with evidence for ventricular dysfunction is another relative indication. Symptoms are a late finding in aortic stenosis and should not be the indication for surgical intervention. Aortic insufficiency of a severe degree causing diastolic flow reversal in the descending aorta and significant progressive ventricular volume overload are indications for surgical intervention. Documented increase in left ventricular end-diastolic dimension on serial measurements that are greater than three standard deviations above normal despite use of afterload reduction therapy is a relative indication for surgery, as is a decrease in systolic performance on exercise. Exercise intolerance is another relative indication for intervention.

An increasing number of patients are coming to aortic valve repair or replacement after balloon dilation of aortic valve stenosis in infancy. In many cases, these valves can be repaired by the techniques described by Dr. Ilbawi's group. However, many of these patients have relative hypoplasia of the aortic annulus, and the reparative techniques to alleviate the regurgitation may result in varying degrees of stenosis. Therefore, these valves may be best suited to a valve replacement rather than valve repair. Concerns about longevity and aortic root dilation over time of the autograft aortic valve replacement have led to an increasing use of valve reparative techniques as a temporizing measure. We use valve repair whenever possible if we believe a relatively durable (>5 year) good result can be anticipated. If the valve is significantly distorted or if valve reparative techniques will result in relative stenosis, we believe that the Ross operation may be a better choice. Nevertheless, patients with predominant aortic insufficiency can often have a quite durable valve repair, which may allow children to grow and provide more options for valve replacement in the future.

We consider valve repair a palliative procedure that will ultimately require reoperation and valve replacement; however, in small and growing children, female patients, and young adults it may be advisable to perform a valve repair of limited durability to allow the patient to grow into adulthood, when mechanical valve replacement may be a better option. In addition, the increasing interest in development of tissue-engineered valves lends hope that in the future viable valves can be developed that will alleviate many of the concerns with bioprosthetic or mechanical valve replacement.

The tissue used for valve leaflet extension is controversial. As noted, glutaraldehyde-fixed tissue tends to calcify and stiffen over time and may lead to valve degeneration and recurrent regurgitation or stenosis, requiring reoperation. Fresh pericardium tends to shrink and may lead to a relatively unstable early repair. The use of any nonautologous tissue such as homograft material, bovine pericardium, or CorMatrix® has not been examined at long enough follow-up to determine whether these tissues might provide a more reliable alternative. Nevertheless, the use of nonviable tissue for reconstruction is likely to result in degeneration over time, as in tissue valve prostheses.

The complex geometry of the aortic valve and the difficulty in assessing the valve function in the operating room make the use of newer imaging modalities for pre- and postoperative assessment important. Three-dimensional echocardiography is a new modality that may preoperatively identify the mechanism of valve dysfunction and guide the surgical intervention for valve repair. When three-dimensional echocardiography can be done in real time, it may significantly improve our ability to perform accurate valve repairs. Transesophageal 3D transducers are now available for adult size patients. Hopefully, smaller 3D transducers will become available for use in children in the future.

The complications of valve replacement as noted in Chapter 104 have led most pediatric centers to begin more aggressive repair of valves in all positions in the heart in the hope of providing reasonably durable palliation before anticipated later valve replacement.

TLS

104 Aortic Valve Replacement in Children

John W. Brown, Parth Patel, Mark Rodefeld, and Mark W. Turrentine

INTRODUCTION

Left ventricular outflow track obstruction (LVOTO) occurs in 3% to 5% of patients with congenital heart disease. Valvular aortic stenosis (VAS) makes up 50% to 65% of the cases of LVOTO in children. Bicuspid aortic valve (BAV) is the most common type of LVOT pathology and is seen in 1% of human hearts. One-third of BAVs will require surgical intervention during childhood and a second one-third will require replacement during adulthood. The remaining one-third will not require treatment during a normal life expectancy. The aortic valve is the second most common valve to require replacement in childhood superseded only by the pulmonary valve.

Aortic valvular pathology in infants and children poses numerous challenges to the pediatric cardiac surgeon. Without question, valvular repair is the goal of initial intervention because improvement of anatomy and physiology using native tissue allows growth and a potentially better long-term outcome. When reconstruction fails or is not feasible, valve replacement becomes inevitable. The timing of valve replacement and the type of prosthesis remains controversial. Few reports compare the outcomes of the Ross aortic valve replacement (AVR) and other valve substitutes (non-Ross) in children. The goal of this chapter is to address these controversial issues and draw from our institutional experience and review current literature to support our preference for the Ross AVR in children when possible.

Most infants and children with LVOT obstruction due to a BAV in North America will be subjected to an initial attempt at balloon aortic dilatation (BAD). The BAD procedure indiscriminately tears the valve at its weakest point. The stenotic neonatal aortic valve is usually dysplastic and visual identification of commissures can be difficult. There has been little outcome advantage for surgical aortic valvuloplasty (SAV) compared with BAD in neonates. Visualization of the anatomy is not a problem in infants beyond the neonatal period, and in these cases surgical valvuloplasty should be considered as the principal palliative measure.

We recently compared our last 20 years experience with SAV in infants >2 months of age with our last 10-year experience with BAD and found that survival was equivalent. However, SAV gave twice the initial gradient reduction, half the degree of early aortic valve regurgitation, and had 3 to 4 times the palliative durability of BAD. Ten and twenty year freedom from AVR following SAV was 80% and 70%, respectively. Freedom from AVR after BAD was 75% at 10 years and an additional 25% required balloon or surgical reintervention during the same time period.

BAD usually reduces the LVOT gradient but may produce important degrees of regurgitation in up to 20% of neonates and BAD has no advantage in risk of reintervention and mortality in this age group. Other types of LVOT obstruction in children, which involved subvalvar, supravalvar, or multilevel obstructions, are initially referred for surgical treatment. While this report will concentrate on AVR in children, treatment options for patients who have additional areas of LVOTO will be included.

INDICATIONS FOR INTERVENTION IN AORTIC VALVE PATHOLOGY

Infants and children with aortic valve pathology should be offered intervention if they have symptoms of congestive heart failure, angina, or syncope. An asymptomatic child should be offered intervention if the peak LVOT gradient is over 70 mmHg, the mean LVOT gradient is more than 50 mmHg, the aortic valve area is <0.8 cm/m^2 of the body surface area, or if they have more than moderate regurgitation with LV dilation and/or dysfunction.

When surgical palliation fails or is not feasible, valve replacement becomes inevitable. Prosthetic valve selection for infants and children requires several considerations that include: aortic annular size, associated LVOT obstructive lesions, associated connective tissue abnormalities, presence and/or quality of the pulmonary valve, and the ability and experience of the operative surgeon.

Controversy over the prosthetic choice for AVR in children has dropped dramatically in the past 10 to 15 years because of the growth in popularity and excellent results obtained with the Ross AVR in many centers (Table 104.1). Mechanical, xenograft, and allograft valves in most institutions in the current era are reserved for children with connective tissue disorders or patients whose native pulmonary valve, if present, is unsuitable for translocation to the aortic position.

AORTIC VALVE REPLACEMENT USING AN ALLOGRAFT AORTIC VALVE

Fresh antibiotic-sterilized aortic allografts were among the first valves to be used for AVR by Ross and others in the early 1960s. Most were implanted as free-hand valves in the subcoronary position of the recipient's aorta. Their initial function in most instances was satisfactory but within 2 to 6 years the valves failed by calcified stenosis and/or regurgitation.

Aortic allografts were rarely used in North America in the 1970s and early 1980s due to their lack of availability and xenograft and mechanical valves predominated. Cryopreservation techniques became the accepted mode of allograft preservation and storage in the early 1980s and availability of aortic allografts in various sizes for AVR was made possible. In many centers, the aortic root replacement with the allograft aortic root gave more predictable results with less initial regurgitation than did the free-hand sub-coronary

Table 104.1 Ross AVR in Children: Literature Survey

Author, Institution, Publication year	Year of study	*N*	Age (y)	Type of AVR	Early mortality (%)	Late mortality (%)	Embolic event (%)	Bleeding at Ross AVR admission (%)	Endocarditis (%)	Rate of redo AVR (%)	Rate of redo RVOTR (%)	Follow-up (y)	Rate of any reoperation (%)
Piccardo, Marseille, France 2009	1993–2006	55	10 ± 6.2	Ross	2	4	0	4	2	4	5	5.5 ± 3.8	13
Alsoufi, Riyadh, Saudi Arabia, 2009	1990–2006	215	11.4 ± 4.6	Ross	2	0	0	0	1	13	Not Recorded	5.7	23
Lupinetti, Seattle, WA, 2002	1994–2001	78	10.7 ± 6.6	Ross	4	1	1	1	0	1	1		6
Al-Halees, 2002	1990–2000	53	8.0 ± 5	Ross	6	2	0	2	2	2	2	4	4
Hraska, Bratislava, Slovakia, 2004	1997–2003	66	12.9	Ross	0	2	0	2	2	2	2	2.4	6
Hazekamp, Amsterdam, Netherlands, 2005	1994–2003	53	9.15 ± 5.07	Ross	6	6	0	9	0	8	6	5.5	15
Bohm, Stuttgart, Germany, 2006	1995–2004	60	12.6 ± 4.4	Ross	0	3	0	3	0	0	10	3.5 ± 2.25	12
Kalavrouziotis, Liverpool, UK, 2006	1996–2004	35	10.6 ± 5.4	Ross	0	3	0	3	0	0	0	4.1 ± 2.4	0
Brown, Indianapolis, IN, 2012	1970–2012	112	11.4 ± 5.7	Ross	1	4	0	0	4	5	11	7.0 ± 5.4	23
Alsoufi, et al. 2009; Hraska, et al. 2004; Hazekamp, et al. 2005; Bohm, et al. 2006; Kalavrouziotis, et al. 2006; Piccardo, et al. 2009		727	11.0	Total Ross	2.2	2.1	0.1	1.8	1.2	5.9	2.9	—	14.9

AVR, aortic valve replacement; RVOTR, right ventricular outflow tract reoperation.

technique. The cryopreserved allografts functioned well in most older adults for up to 10 years but when used in pediatric patients, especially infants, their durability was often <5 years and most centers abandoned their use. The mechanism of early allograft failure is thought to be immunologic. Reoperation after a failed allograft AVR root can be challenging due to dense calcification of the entire root.

Decellularization of allografts valves were introduced in the early 2000s and this process removed the endothelial cells and most of the other viable cells in the interstitial matrix decreasing allograft immunogenicity without weakening the tissue. Decellularized pulmonary allografts have been used by us and others in substantial numbers and their durability in children and adults is better than standard cryopreserved pulmonary valves. Decellularized aortic allografts have only been used in limited numbers by a few investigators and appear to be more durable than standard cryopreserved aortic allografts but the numbers of patients and length of follow-up is too short to draw meaningful conclusions.

XENOGRAFT AORTIC VALVE REPLACEMENT IN CHILDREN

Stented xenograft valves have had very limited use in the pediatric aortic position due to their bulk and large annulus to effective orifice area ratios. Stentless xenografts have a much better orifice area and hemodynamic efficiency. We have used stentless xenografts for aortic root replacement when the pulmonary autograft is unavailable or unsuitable and when anticoagulation for mechanical valves is impractical as encountered with young females who want to consider having children. However, due to rapid calcification and early dysfunction, xenografts are a poor choice in younger children. Xenograft pericardial valves usually fail by calcific obstruction and porcine valves usually fail by regurgitation. Third- and fourth-generation xenograft valves may prove more durable than prior generation xenografts.

MECHANICAL VALVES IN CHILDREN

Mechanical valves in the aortic position of children can be more durable than allograft and xenograft valves, but they have no growth potential and develop patient prosthesis mismatch (PPM) as the child grows. The smallest mechanical valve manufactured for AVR is 16 to 17 mm, and its effective orifice area is slightly over 1 cm^2. PPM is defined as an effective orifice area of <0.85 cm/m^2 of body surface area. Mechanical valve size <23 mm will likely develop PPM as the child reaches adulthood. Mechanical valves can also suffer from pannus ingrowth that can obstruct the inflow and/or outflow of the prosthesis and lead to early prosthetic dysfunction. The development of left ventricular hypertrophy, secondary to PPM or pannus ingrowth, leads to increased LV mass, increased systolic and diastolic dysfunction, and shortened patient survival.

All mechanical valves at present require Warfarin anticoagulation that leads to bleeding problems in 1% to 2% of patients per year. Bleeding tendency can be a particularly troublesome issue for female patients with monthly menses and females who want to become pregnant. Warfarin is also teratogenic for the fetus. Anticoagulation monitoring and compliance is another important issue with teenage patients who are beyond parental control.

Potential for a thromboembolic event is a constant risk factor in mechanical AVR with and without Warfarin anticoagulation and such events occur in 1% to 4% of patients per year. Central nervous system thromboembolic complications, if not fatal, can be devastating for any patient regardless of age.

Quality of life in children with mechanical aortic valves is less than other valve prosthesis because of the need for monthly monitoring of anticoagulation, fear of thromboemboli and bleeding complications, and inability of the child to participate in active and/or contact sports due to anticoagulation and the attendant bleeding risk.

PROSTHETIC ENDOCARDITIS

Prosthetic endocarditis is higher for xenografts than for mechanical valves, homografts, or autografts. Mechanical and xenograft valve endocarditis frequently leads to septic emboli and the need for early prosthetic replacement. Table 104.2 is a literature survey of outcomes in non-Ross AVR in children and shows a higher early and late mortality, higher postoperative complication rate, and a reoperation rate of nearly 20%.

AUTOGRAFT AORTIC VALVE REPLACEMENT (ROSS) IN CHILDREN

Replacement of the diseased aortic valve by pulmonary autograft, a procedure initially described and performed experimentally by Shumway and Lower in the early 1960s and then performed clinically by Ross in 1967, has provided excellent hemodynamic results, both in children and in adults. In experienced hands, the Ross procedure has low morbidity and mortality (Table 104.1). The Ross AVR was slow to be accepted in North America until Elkins and Stelzer introduced it in the early 1980s. They modified the implantation technique to a full root replacement, which gave the Ross AVR more consistent results. Most American surgeons follow their lead and perform the Ross AVR as a root replacement. The Ross AVR is the only AVR, which can place a child or young adult back on a normal life expectancy curve.

ROSS TECHNIQUE IN CHILDREN

The Ross AVR is most often performed as an autograft root replacement as described previously. All patients have intraoperative transesophageal echocardiography (TEE). Standard techniques of cardiopulmonary bypass were used, with bicaval cannulation, moderate hypothermia to 26°C, and antegrade (when possible) and/or retrograde cold blood potassium cardioplegia. The autograft is harvested using standard techniques and leaving 2 to 3 mm of RV muscle attached to the pulmonary annulus. Only 3 to 5 mm of main pulmonary artery is beyond the pulmonary valve commissures is harvested.

After the aortic root is dismantled and debrided, and the tops of the aortic commissures shortened to achieve a near circular but modified aortic annulus, the coronary arteries are removed as large buttons and mobilized for 5 to 8 mm. After the autograft commissures are marked to enable equal distribution of sutures, pledgeted (3×7 mm) interrupted mattress sutures of monofilament or braided Dacron (Boston Scientific Corp, Natick, MA) are used to encircle the modified aortic annulus and these sutures are placed in mattress fashion through the autograft annulus. The proximal autograft suture line is fixed or reduced with a Dacron strip when the aortic annulus is larger than the autograft annulus and when annular growth is not desired. Annular reduction is indicated when the aortic valve annulus is more than 4 mm larger than the pulmonary valve annulus. Annular

Table 104.2 Non-Ross AVR in Children: Literature Survey

Author, Institution, Publication year	Year of study	*N*	Age (y)	Type of AVR	Early mortality (%)	Late mortality (%)	Embolic event (%)	Postoperative bleeding (%)	Endocarditis (%)	Rate of redo AVR (%)	Rate of redo RVOTR	Follow-up (y)	Rate of any reoperation (%)
Alsoufi, Riyadh, Saudi Arabia, 2009	1986–2006	131	14.0 ± 3.8	M	6	15	3	2	1	6	Not Recorded	8.3	15
Lupinetti, Seattle, WA, 2002	1994–2001	25	10.2 ± 5.5	H + M	0	0	4	0	0	12	0		12
Alexiou, Southampton, UK, 2000	1972–1999	56	11.2	M	5	5	4	0	0	7	0	7.3	9
Lupinetti, Seattle, 1999	1987–1998	50	12.1 ± 4.6	M	10	8	8	0	4	20	NR	5.4	NR
Brown, Indianapolis, IN, 2012	1970–2012	73	12.1 ± 5.8	H + M	7	11	7	1	11	26	8	11.8 ± 9.7	37
Lupinetti, et al. 1999, Alexiou, et al. 2000, Alsoufi, et al. 2009		335	12.6	Total Non-Ross AVR	6.3	10.1	4.8	1.2	3.3	13.1	3.9	—	18.9

AVR, aortic valve replacement; H, homograft/allograft valve replacement; M, mechanical valve replacement; RVOTR, right ventricular outflow tract reoperation.

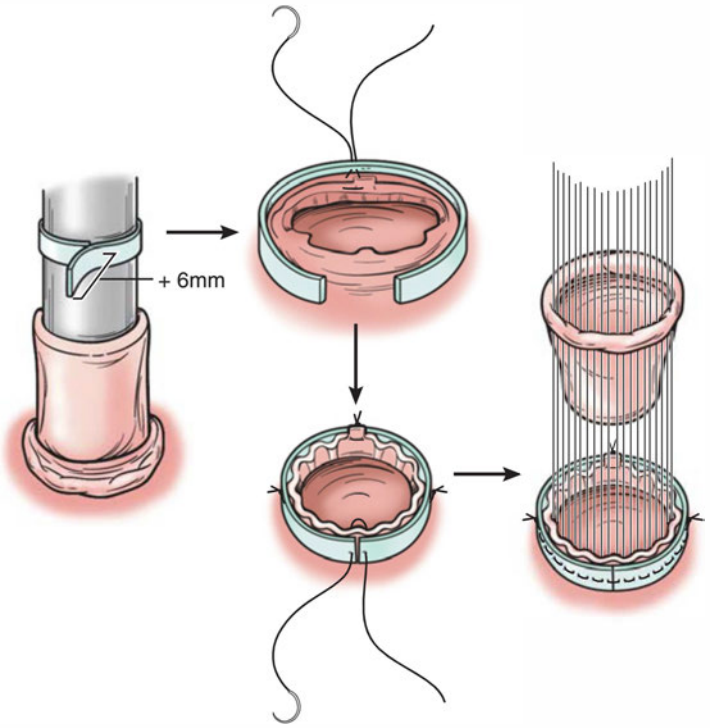


Fig. 104.1. This figure illustrates the annular reduction technique for the Ross aortic valve replacement.

reduction is performed using a 5 mm wide Dacron strip placed circumferentially around the aortic annulus and taking smaller bites of the Dacron, and wider bites of the aortic annulus, thus pleating the aortic annulus 1 to 3 mm per mattress suture along the left and noncoronary portions of the aortic annulus (Fig. 104.1).

The annular reduction is confirmed by placing an appropriately sized Hegar dilator through the reduced aortic annulus.

The autograft is oriented in an anatomic manner keeping the anterior wall anterior and suturing the left-coronary button in the middle of the posterior autograft sinus using 5-0 or 6-0 monofilament suture. The right coronary button is placed in the anterior autograft sinus after careful orientation and before completing the distal aortic anastomosis or left unattached until after the aortic root is allowed to distend.

The right ventricular outflow tract (RVOT) is then reconstructed with an appropriately oversized (6 to 10 mm larger than the autograft) standard cryopreserved pulmonary homograft, a decellularized pulmonary homograft (SynerGraft, CryoLife, Inc., Marietta, GA) or a bovine jugular vein conduit (Contegra, Medtronic, Inc., Minneapolis, MN). The bovine jugular vein conduit is preferred for RVOT reconstruction in children <6 years because it has been more durable at our institution in this younger patient age group.

The distal autograft anastomosis is completed after RVOT reconstruction with a 5-mm Dacron strip incorporated into the distal suture line if growth is not desired. The ascending aorta in older or larger children is replaced to include a hemiarch if it is significantly dilated and replaced with a synthetic Dacron tube graft. The mean ascending aortic graft size is 25 ± 2.6 mm (range, 20 to 30 mm). A drawing of the heart after the operation is shown in Figure 104.2.

A TEE is performed intraoperatively in all the patients after cardiopulmonary bypass weaning in order to evaluate the competence of the Ross and RVOT valves. PI and AI have been minimal in our experience and never greater than mild.

ROSS–KONNO MODIFICATION

The Ross–Konno is reserved for patients with multilevel obstruction. If the aortic annular diameter is 4 mm smaller than the pulmonary annulus, then enlargement of the aortic root is accomplished by the Konno technique. This involves transecting the aortic annulus for 4 to 8 mm at the commissure between the left and right cusp or at a level just below the pulmonary annulus if the aortic valve is bicuspid. The autograft is carefully harvested leaving a 10 to 12 mm anterior lip of subannular free wall muscle for the Konno patch. The Konno patch is kept very short and an aggressive subannular myectomy is performed if subannular obstruction is encountered. The autograft is trimmed and sutured to the widened aortic annulus (root) placing the anterior tip of the retained RV free wall to patch the triangular defect created by transecting the aortic annulus and the proximal ventricular septum. Pledgeted interrupted mattress sutures are used for the Konno extension (Fig. 104.3).

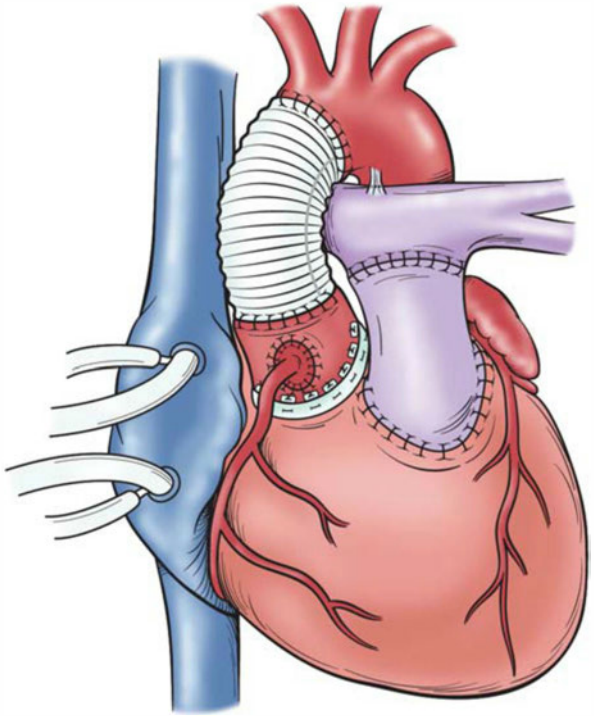


Fig. 104.2. Current Ross technique.

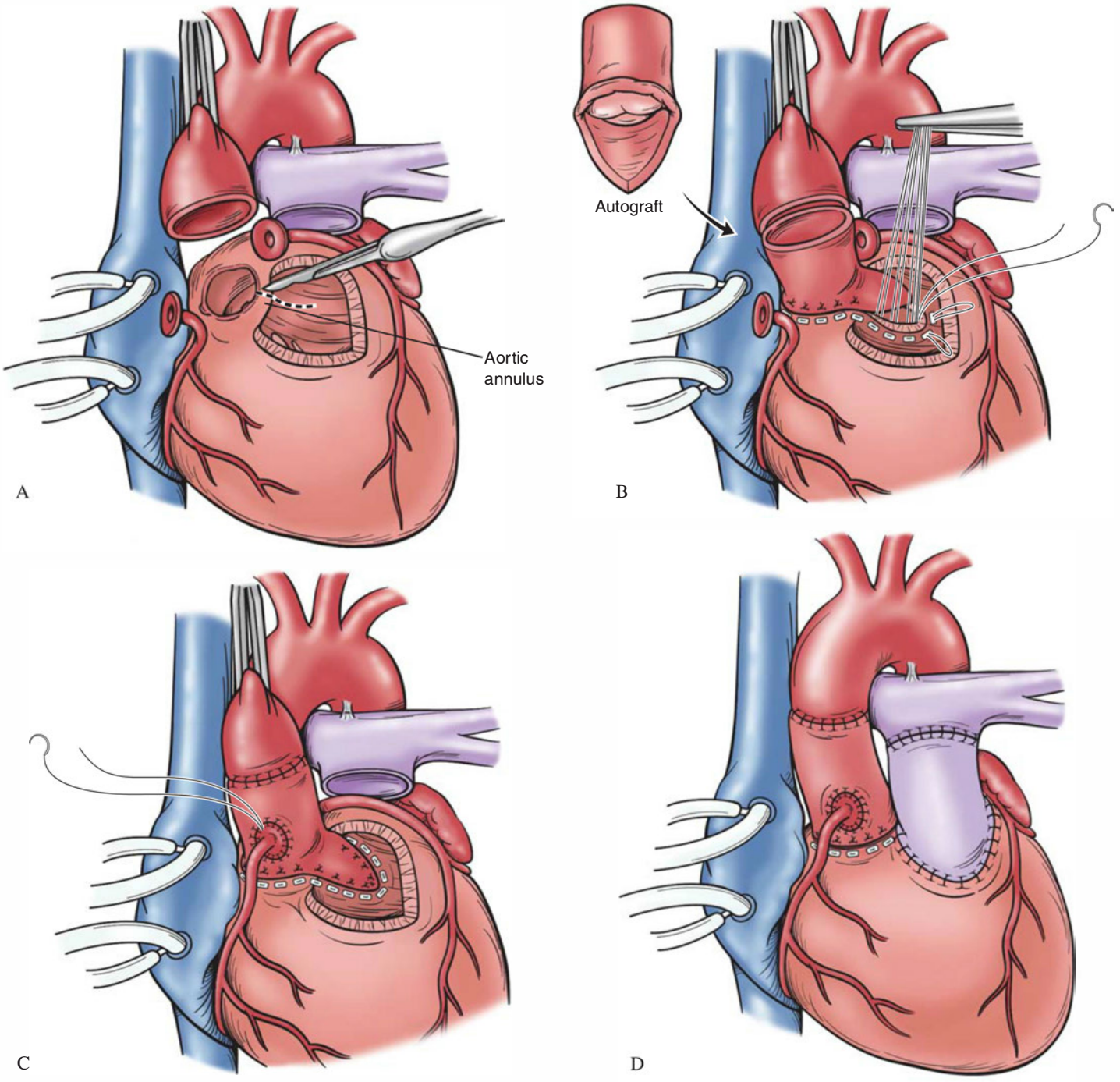


Fig. 104.3. Ross–Konno procedure (**A**) The aortic annulus is enlarged by incising the interventricular septum (dashed line) between the left and right commissures. The length of the incision is frequently limited to just beyond the annulus. A myectomy will also increase orifice area, limiting the extent of ventriculoplasty incision. (**B**) The autograft is sutured to the margins with interrupted pledgeted sutures. A short apron is left on the autograft to fill the ventriculoplasty site. (**C**) Coronary buttons are reimplanted into the autograft. (**D**) Completed reconstruction.

Mavroudis and coauthors have described placing the Konno incision in the middle of the left sinus to reduce the incidence of acquired heart block (Fig. 104.4). We have used this modification on occasion but it requires more extensive mobilization of the left-coronary artery. Additionally, postoperative bleeding in this area if encountered would be difficult to control.

OTHER METHODS OF ANNULAR ENLARGEMENT

If a more limited aortic annular enlargement is required for AVR using any aortic valve substitute, the Nicks or Manouguian techniques can be considered. The Nicks annular incision is performed in the middle of the noncoronary sinus and the Manouguian incision is made at the commissure between the non- and left-coronary cusps and carried down to the mitral annulus.

DISCUSSION

There are several advantages of the pulmonary autograft that benefit both the adults and pediatric patients. These advantages

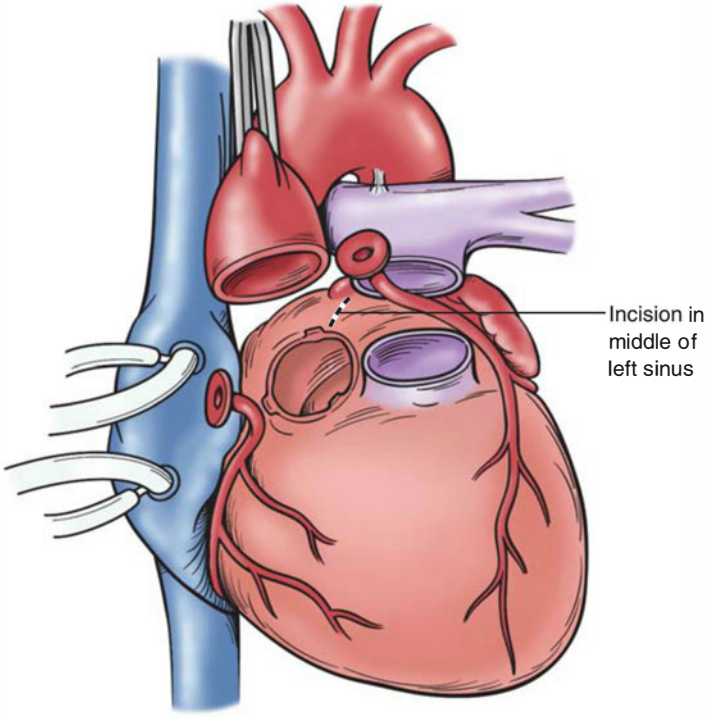


Fig. 104.4. Image of Ross–Konno operation, Mavroudis technique. The arrow points to the dotted line where the incision is made to enlarge the left ventricular outflow track.

include superior hemodynamics and better regression of LV mass, better long-term survival, no risk of thromboemboli, no age or size limitation for infants and children, growth potential for infants and children, lower risk of endocarditis than other prosthetic types, excellent midterm durability of the autograft and allograft when compared with other prosthetic types, acceptable risk of reoperation, and no bleeding issues related to anticoagulation (Tables 104.1 and 104.2). All of these advantages translate to a better quality of life for children because the Ross AVR allows unrestricted physical activity, more normal psychological development due to lack of need for frequent blood draws to check anticoagulation status, and the Ross AVR allows for pregnancy in young women.

Even with all of these advantages, the early experience of the Ross operation was marred by some problems and disadvantages. Disadvantages include the increased technical difficulty of the Ross AVR. The harvesting of the pulmonary valve necessitates putting two valves at risk for a single-valve disease. Additionally, a prior Konno procedure is likely to have damaged the autograft annulus and exclude a Ross AVR. The most significant problems include a higher than expected operative mortality rate in some centers. Late dilation of the autograft root occurred in as many as 20% of patients on long-term follow-up with the root replacement technique; this led to a higher than expected rate of reoperation. Dilatation is specifically shown to occur at the valve annulus, sinuses of Valsalva and sinotubular junction (STJ) of the pulmonary autograft. This increase has been attributed to a variety of causes including secondary to passive dilatation due to exposure to higher blood pressure, mismatch of the pulmonary autograft and the aortic annulus or ascending aorta, an intrinsic abnormality of the pulmonary root possibly associated with congenital BAV disease, or normal somatic growth in the pediatric population.

There is no consensus on the rate and nature of autograft dilatation. Tantengco and colleagues noticed pathologic dilatation according to z values during the first year after the Ross AVR, but without progress thereafter. Pasquali and colleagues demonstrated a significant increase in z values of the annulus, sinus, and STJ over time. Kouchoukos and colleagues observed a consistent increase of the sinus and STJ dimensions over time, but not of the annulus dimensions; however, his measurements were not indexed to the patients' body surface area. Horer and colleagues noticed that the annulus grows proportionally to somatic growth but the curves depicting the z values of the sinus and the STJ increased over time. The rate of autograft dilatation calculated on serial echocardiographic assessments was reported by Takkenberg and colleagues at 0.5 mm/year in an adult population and by Pasquali and colleagues at 2 mm/year in a mixed pediatric and young adult population. Neo-aortic regurgitation and neoaortic root dilatation occur over time in some adolescents and adults; however, the correlation between AR and aortic root dilatation has varied between institutions. Kouchoukos identified dilatation of the STJ as an independent predictor for the progression of AR. Horer and colleagues determined that the aortic root dimensions immediately after the Ross AVR were larger than in normal healthy children but the annulus grows proportionally to the somatic growth of the child. They found in their series that AR develops slowly but significantly and was associated with a dilated STJ. Patients with BAV and dilated ascending aorta or patients with dilated aortic root and primarily aortic insufficiency have been considered the highest risk group for dilation and neoaortic insufficiency in some series. Other studies have shown that no association exists between bicuspid valve disease and histologic changes in explanted pulmonary autografts. When the root dilatation leads to significant autograft valve regurgitation or when the dilatation rapidly progresses into an aneurysm, reoperation is indicated. These problems are uncommon in our experience (Table 104.1).

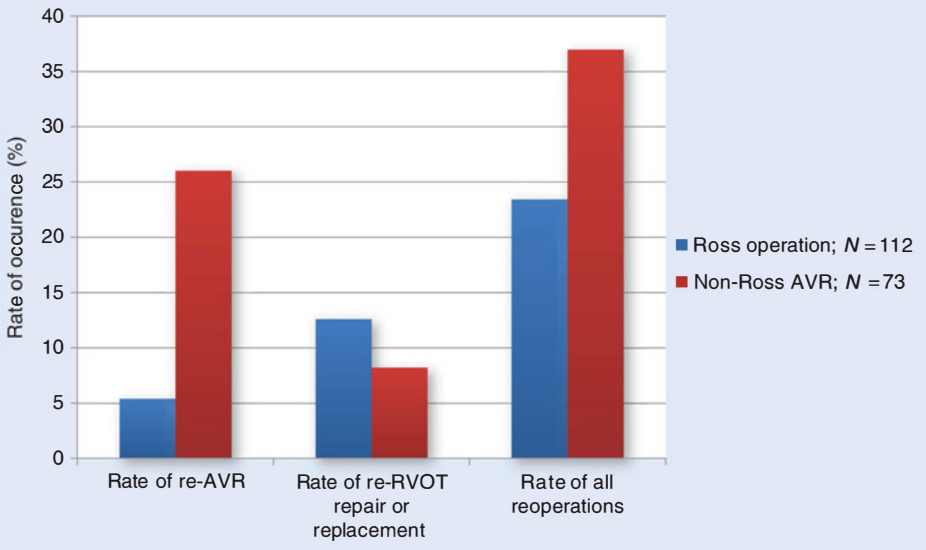
Because of the early experiences with root dilation and/or AR many centers modified their Ross technique in an attempt to prevent autograft dilatation. Our institution currently reinforces the annulus and STJ with Dacron, replaces the ascending aorta if it is significantly dilated ($z > +2$), and aggressively manages perioperative and postoperative hypertension for up to 6 months. We reduce the aortic annulus if it is dilated. The aortic annulus and STJ should be at least 20 mm in diameter before we restrict its growth with a Dacron band or ascending aortic graft. The youngest patient in our pediatric Ross experience who needed aortic annulus reduction was an 8-year-old boy. We reduced his annulus to 20 mm. Elkins and colleagues describe that the only independent predictors for the development of moderate or greater autograft regurgitation were increased age at the time of the operation, autograft regurgitation at completion of the Ross AVR, and increased follow-up time.

Moritz and associates reduced the diameter of the aortic annulus and wrapped the pulmonary autograft with an absorbable mesh in a few patients as an attempt to prevent postoperative AI. Pacifico and colleagues described an operative technique in which the entire pulmonary autograft was wrapped with glutaraldehyde-fixed bovine pericardium to prevent dilation. The Ungerleider technique of placing a Dacron sleeve

around the autograft was recently introduced to address this concern in adults. Other techniques to prevent post-Ross autograft dilation have also been suggested. Excessive autograft length in the root technique may lead to late dilation at the STJ and above, as shown by de Kerchove and coworkers. They conclude that in the root technique the autograft is less likely to dilate if it is as short as possible and when the distal suture line is reinforced with a Dacron strip or extended with a Dacron graft. We adopted these modifications in 2000. These annular and STJ reinforcement techniques may not fully protect against autograft sinus dilation, but some data suggest a benefit from support at and above the STJ . We also advocate aggressive treatment of systemic hypertension during the first 6 months postoperatively.

A comparison of our freedom from reoperation before and after 2000 when we modified our technique and aggressively treated hypertension is shown in Figure 104.5.

Table 104.3 Incidence of re-AVR, re-RVOTR, and all reoperation after the Ross Operation (N = 112; 1993-2012) and the Non-Ross AVR (N = 73; 1969-2011) at Indiana University.



AVR, aortic valve replacement; RVOT, right ventricular outflow tract.

Ross Reoperation

Acquired Ross root dilatation with or without aortic regurgitation was seen before 2000 in our Ross experience in 15% to 20% of our cases. Salvaging the Ross valve using a valve-sparing root replacement technique with reimplantation of the valve has been possible in the majority of our Ross reoperations.

Other treatment options for Ross root dilation and AR include a mechanical valve Bentall procedure, re-replacement of the aortic root with a homograft, replacement of the aortic valve with supported or unsupported aortoplasty, separate replacement of the aortic valve and the ascending aorta, or valve-sparing root replacement.

The rate of pulmonary autograft replacement in our overall adult and pediatric series of 350 patients is 8% and 5% in children. The results in small children <6 years are not as good but an 81% freedom from any reintervention is still acceptable. Tabe 104.3 compares freedom from any reoperation in our Ross versus non-Ross children and shows a superiority of the Ross over non-Ross AVR.

Our good results with RVOT reconstruction in Ross AVR in children are in part due to excising most of the muscle from the proximal end of the allograft, oversizing the allograft by 6 to 10 mm expecting the allograft to shrink 15% to 25%, and treating all patients with low-dose aspirin and ibuprofen to lessen the inflammatory response of the recipient to donor tissue for 6 months postoperatively. With these modifications, we have a 88.5% freedom from replacement of the RVOT in our 112 children and 95% freedom from RVOT reoperation in our 350 combined pediatric and adult series.

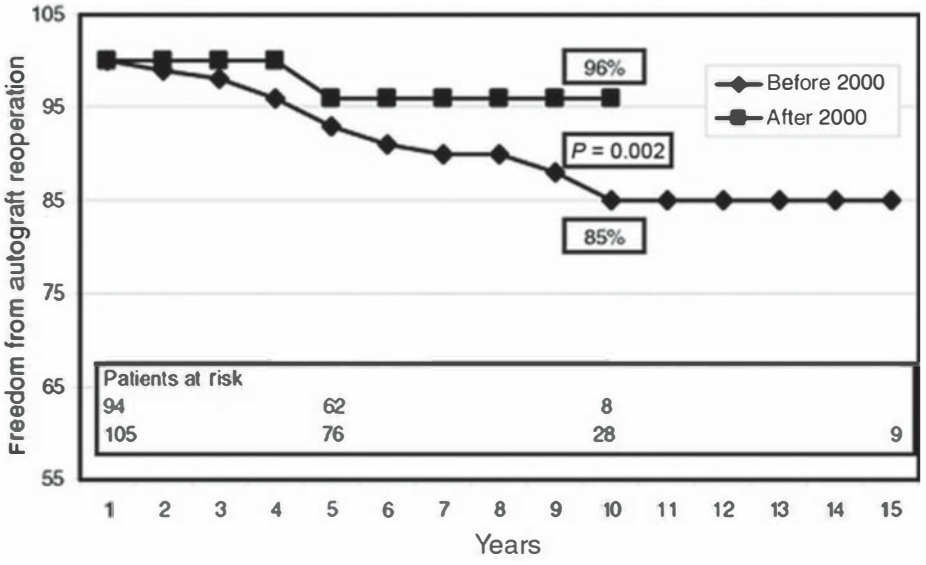


Fig. 104.5. Freedom from reoperation for autograft failure before 2000 (*diamonds*) and after 2000 (*squares*). Modified Ross technique started in 2000.

SUMMARY

AVR in children is challenging. With the Ross procedure, the LVOT gradient is almost always completely eliminated and there is the expected regression of left ventricular mass that should improve the long-term prognosis.

When valve replacement becomes necessary we prefer the Ross AVR as a full root technique as described above.

Ross AVR in infants and children gives superior results in most high volume centers as compared with other prosthetic types (Tables 104.1 and 104.2; Fig. 104.6).

The Ross AVR is a challenging surgical procedure. Many of the early problems seen with the Ross root technique, that is, dilation and regurgitation can be prevented. When dilatation and/or regurgitation are/is encountered, the surgeon has another option of performing a valve-sparing root

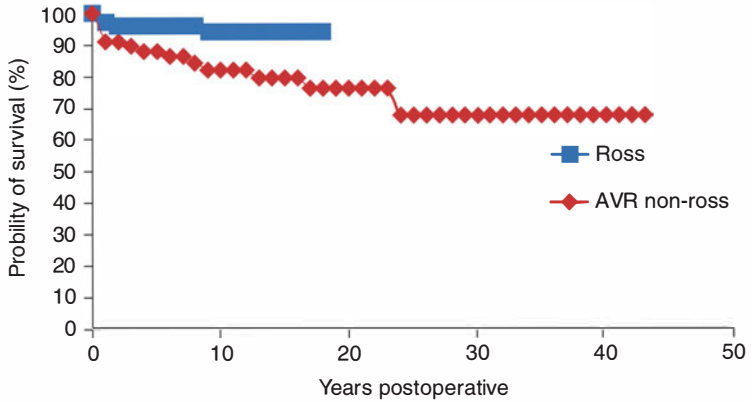


Fig. 104.6. Actuarial survival of IU Ross experience ($N = 112$; 1993-2012) vs. IU non-Ross aortic valve replacement experience ($N = 73$; 1969-2011).

replacement thus avoiding the worrisome complications of other prosthetic valves. The Ross operation has superior survival with less postoperative complications and gives a child a better quality of life. The Ross is the AVR procedure of choice in children requiring valve replacement.

SUGGESTED READINGS

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EDITOR'S COMMENTS

The advent of balloon valvuloplasty for valvar AS and the development of the pulmonary autograft procedure for the replacement of the aortic valve and root (Ross operation) have dramatically changed the approach to children with congenital aortic valve disease. In most centers, surgical intervention by aortic valvotomy has become an unusual operative procedure and balloon valvuloplasty is the primary procedure of choice for aortic valve stenosis in newborns or infants. Dr. Brown and his group have shown an excellent series of surgical valvotomies and valvuloplasties that seem to have results superior to current balloon valvuloplasty results. In spite of this information, most centers will still perform balloon valvuloplasty in preference to surgical intervention because of the less invasive nature of the procedure.

Pulmonary autograft valve replacement has permitted replacement of stenotic or regurgitant aortic valves earlier in life. In the past, patients with significant valvar disease often were managed medically for prolonged periods at the expense of ventricular function, because the options for valve replacement were limited in children and suboptimal. Mechanical valve replacement in children is associated with the need for reoperation as the child outgrows the valve size, and the difficulties of anticoagulating young children are significant, with the added inconvenience of multiple blood drawings for monitoring. Other alternatives for valve replacement are also unappealing, including tissue valves, which degenerate rapidly in children, and homograft valves, which do not have the growth potential of autografts and also have a reasonably high incidence of late degeneration. However, the Ross pulmonary autograft valve replacement as described in this chapter and as extensively performed by Dr. Brown and others in many children with complex forms of left ventricular outflow tract obstruction has proved to be a quite durable and satisfactory procedure. Dr. Brown has shown conclusively that the autograft valves can grow in young children, and the natural endothelial surface and viability of the autograft seems to be ideal for

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EDITOR'S COMMENTS (*continued*)

aortic valve replacement. Thus, the favorable characteristics of the autograft valve replacement may actually permit earlier replacement of the aortic valve rather than repeated valvotomies or other procedures, which may provide only short-term palliation and potentially exacerbate ventricular dysfunction by maintaining chronic volume and pressure loads on the ventricle. The pulmonary homograft valve used to replace the right ventricular outflow tract in the Ross operation may not grow with the child; however, in most cases a much larger pulmonary homograft can be inserted than the size of the autograft valve, which in most children >3 or 4 years of age permits the use of an almost adult-sized valve. Although stenosis of the homograft valve has rarely occurred, in most cases the valves become insufficient with time, which is well-tolerated hemodynamically on the right side of the heart. When the pulmonary homograft used to replace the right ventricular outflow tract after the Ross operation becomes insufficient, it is now possible to consider using stent-mounted transcatheter valve implantation to preserve valve function and extend the life of the pulmonary homograft conduit. The use of these techniques may eliminate some of the concern about late RV outflow tract obstruction after the Ross operation.

On rare occasions, the pulmonary valve is abnormal, and some have suggested not to use bicuspid pulmonary valves for the Ross operation. However, in young children in whom other valve replacements are unappealing or a homograft replacement of the aortic valve would likely be associated with a significant risk of reoperation because of the patient's growth, we use the bicuspid pulmonary autograft, because it may have satisfactory function over the intermediate term. However, other abnormalities of the pulmonary valve, including any pulmonary valvar insufficiency related to leaflet abnormalities such as quadricuspid valves have been associated with autograft regurgitation when used in the aortic position. Therefore, if significant pulmonary valve abnormalities are noted, even in the presence of delicate valve leaflets, we now believe that the autograft should not be used.

Dr. Brown has described nicely the Konno modification of the Ross operation for patients in whom enlargement of the left ventricular outflow tract is necessary. We have also used the technique of excising a portion of right ventricular outflow tract muscle with the autograft as a way of closing the incision in the ventricular septum; however, in patients in whom a larger septal incision is necessary, we have used a Gore-Tex patch to fill in the septal defect and have sewn the autograft to the superior margin of the patch. We have used a slightly different technique for the implantation of the pulmonary autograft than that described by Dr. Brown. While it has been well shown that implantation of the autograft in an inclusion technique inside the aortic root will have less issue with late dilation and insufficiency, the vast majority of children with aortic valve disease have an aortic annulus that is significantly smaller than the pulmonary root and therefore inclusion techniques cannot be successfully utilized. It is not uncommon to need to enlarge the aortic root slightly to accommodate the size of the pulmonary autograft. The incision in the aortic annulus as described by Dr. Brown in the Ross–Konno operation is usually all that is necessary to allow adequate size matching of the autograft and aortic outflow tract. We have tended to use a running technique for autograft implantation in young children and have found that the valve can be inverted into the left ventricular outflow tract to place a circumferential suture line in larger children or simple running suture can be used to implant the annulus in very small children. Reinforcement of the aortic annulus in these small children is not necessary or desirable due to the need to allow for growth and therefore annular reduction techniques and the use of Dacron reinforcement as described by Dr. Brown is not necessary. The need to stabilize the STJ is important; however, this must be done in such a manner as to allow for growth in very young children and therefore a pericardial strip is used if necessary rather than rigid Dacron material. The replacement of the ascending aorta with a graft will eliminate aortic root dilation up to the level of the innominate artery and may also serve the advantage of stabilizing the STJ to prevent late autograft dilation and insufficiency.

The incidence of root dilation requiring reoperation varies largely from one series to the other and Dr. Brown has perhaps the best series with freedom from this complication. The majority of patients who come to reoperation have root dilation that is sufficient to cause the onset of aortic insufficiency and in many of these patients the autograft root can be saved with valve-sparing root replacement, maintaining the benefits of a living valve, and the lack of need for anticoagulation long term. The intermediate term results for valve-sparing root replacement appear to be quite good; however, we have seen occasional patients who have had dehiscence of the valve leaflets from inside the Dacron aortic graft, possibly related to the distortion or abnormal coaptation of often elongated valve leaflets in these patients. This appears to have been a problem primarily in patients who have had valve-sparing root replacement for Marfan's disease but has also been seen in patients after autograft root replacement.

When the pulmonary autograft is much larger than the native aortic root after reconstruction, the pulmonary homograft used to replace the right ventricular outflow tract is relatively large and must swing around the new aortic root, which is larger than the original. Thus, it is important to be sure that there is adequate length of the homograft used for right ventricular outflow reconstruction, and, as the suture line is brought toward the right side of the incision in the right ventricular outflow tract, that it does not become compressed by the autograft aortic root. The homograft has a tendency to be draped across the autograft root. The natural curvature of an aortic homograft may provide a better option for right ventricular outflow reconstruction than a pulmonary homograft in some cases.

Dr. Brown has also described the technique for decreasing the size of the aorta for the distal anastomosis when the ascending aorta is somewhat dilated with the autograft root replacement procedure. We have used a similar technique; however, we have found that the pulsatility of the autograft in the aortic root caused by the elasticity of the pulmonary artery can cause significant bleeding when there is a diameter discrepancy at the distal suture line, and we therefore tailor the distal

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EDITOR'S COMMENTS (*continued*)

aorta, but also reinforce the suture line with a strip of Teflon felt or pericardium to decrease the pulsatility at the suture line and decrease the risk of bleeding.

The greatest long-term concern after Ross pulmonary autograft aortic root replacement is the development of progressive dilation of the autograft and AI. Although there does appear to be an incidence of progressive aortic regurgitation in these patients from dilation of the proximal autograft pulmonary artery wall and distortion of the sinuses, the incidence appears to be relatively low and the progression slow. Nevertheless, reoperations will likely be necessary in some of these patients. However, a valve replacement procedure that can be done in young children with a likely reoperation-free rate at 5 years of 80% to 90% or greater, as has been noted for the autograft, is a significant advance for the management of valvular disease in children.

With longer follow-up of the use of the Ross pulmonary autograft as a root replacement in children and young adults, there has been an increasing incidence of autograft root dilation. The sinuses and aortic wall appear to dilate, and even with annular fixation techniques that preserve valve function, significant aneurysms of the proximal aorta can develop. It is unclear at what size intervention is justified, because there is a distal suture line that potentially could prevent distal dissection as seen in Marfan syndrome; however, there has been an isolated case report of dissection of the autograft wall in association with root dilation. Therefore, most centers expect that when root dilation progresses to 5.5 to 6.0 cm, root replacement should be performed. There is an increasing experience of use of the valve-sparing root replacement in this condition to preserve the autograft valve for as long as possible. In most cases, the autograft valve leaflets remain delicate and can coapt well when the autograft wall is replaced with a Dacron graft. There has also been increasing interest in supporting the autograft wall at the time of implantation in older children and young adults, when growth is not an issue, in the hope of preventing the complication of late aortic root dilation. The effect of a rigid prosthetic stenting of the autograft root on overall late autograft function is not known.

TLS

105 Arrhythmia Surgery and Fontan Conversion

Constantine Mavroudis, Barbara J. Deal, and Carl L. Backer

INTRODUCTION

Arrhythmia surgery in young patients is most commonly performed in the setting of concurrent repair of structural heart disease, and rarely performed in patients with structurally normal hearts with arrhythmias refractory to ablations and medications. As the success of surgical repairs of congenital heart disease has improved survival, the late sequelae of repaired congenital heart disease include late hemodynamic/structural problems, arrhythmias, and congestive heart failure. By adulthood, the development of atrial arrhythmias in the setting of congenital heart disease is associated with a 50% increase in mortality, a twofold increase in stroke or congestive heart failure, and a tripling of the need for cardiac interventions. The incidence of reoperations for complex lesions parallels the incidence of arrhythmias and underscores the complex electromechanical interactions in congenital heart disease. In patients with congenital heart disease, arrhythmia surgery may be performed as therapy for coexisting arrhythmias, or prophylactically to reduce the risk of developing late atrial arrhythmias. Efforts to minimize arrhythmia occurrence can be expected to reduce the morbidity related to arrhythmias and hospitalizations and may alter the risk of sudden death. It has been our practice to integrate anti-arrhythmia therapies into all primary repairs as needed, and to view any reoperation for congenital heart disease as an opportunity to improve both the electrical as well as the hemodynamic status of the patient. A unique subset that we will focus on in this chapter is the patient with a prior atriopulmonary Fontan. These patients undergo a Fontan conversion with arrhythmia surgery.

Arrhythmia surgery in patients with congenital heart disease is challenged by the range of anatomic variants, arrhythmia types, and intramyocardial scar location. Anatomic variants include unusual atrioventricular (AV) as well as ventricle to great vessel connections, juxtaposition of the atrial appendages, anomalous pulmonary and systemic venous return among a myriad of other complex lesions. Arrhythmia types can generally be grouped into specific mechanisms, but location and ablation can be perplexing and demanding as each of the arrhythmias is ablated by different procedures indicated by the arrhythmia substrate. Scar formation is generally induced by previous reparative operations that are required to achieve a separated pulmonary and systemic circulation, but not always. Scar formation leading to intractable arrhythmias may occur owing to intracavitary hemodynamic jet lesions, atrial dilatation, myocardial tumors, and cardiomyopathy.

The purpose of this chapter is to review arrhythmia mechanisms and operative techniques for arrhythmia surgery, with attention to particular challenges associated with congenital heart disease, including resternotomy techniques for safe mediastinal reentry. The historical review is based on our previous publications in this field involving 248 patients between 1987 and 2010 from Children's Memorial Hospital in Chicago and the Cleveland Clinic Children's Hospital (Tables 105.1 to 105.3).

ARRHYTHMIA MECHANISMS AND ARRHYTHMIA SURGERY TECHNIQUES

A simplistic approach to the categorization of arrhythmias is summarized in Table 105.4. The most common types of arrhythmias associated with congenital heart lesions are atrial reentry tachycardia (ART), atrial fibrillation (AF), accessory connections, AV nodal reentry tachycardia (AVNRT), focal or automatic atrial tachycardia, and ventricular tachycardia (VT). Reentry is the most common mechanism for clinical arrhythmias and accounts for more than 60% of late postoperative arrhythmias. A reentrant rhythm requires a circuit, with unidirectional block and slowed conduction allowing the electrical impulse to "reenter" the previously blocked area. Reentry may occur in the atria (atrial flutter or atrial reentry), in the ventricles (postinfarction or scar-related VT), or involve all chambers (reciprocating tachycardia utilizing an accessory connection). Reentrant arrhythmias may be initiated or terminated by pacing, allowing the reproduction of the arrhythmia for elucidation of the mechanism in the catheterization laboratory or operating room. Most reentrant arrhythmias are suitable for catheter or surgical ablation, by interrupting a key component of the reentrant circuit. The majority of reentrant circuits are large, or "macro-reentrant," such as atrial flutter. In typical atrial flutter, the electrical impulse courses up the atrial septum and down the right atrial free wall, with slowed conduction in the isthmus of tissue between the coronary sinus ostium and the tricuspid valve. In some instances, the reentrant circuit is more circumscribed, or "micro-reentrant," and may be amenable to targeted focal ablative techniques in a discrete region. In contrast, automatic arrhythmias are local areas of increased firing and cannot be terminated by pacing or cardioversion; therapy is directed at slowing the rate of firing, or eliminating the specific focus of automaticity with ablation or resection. Examples of automatic rhythms include sinus rhythm, junctional ectopic tachycardia, and accelerated ventricular rhythms. The correct characterization of the arrhythmia circuit is essential to the success of arrhythmia surgery, as the most perfectly executed maze procedure for AF will not eliminate a focal source of atrial automaticity.

Surgical and transcatheter techniques have been highly effective in ablating these arrhythmias in pediatric and adult patients with congenital heart disease and normal anatomic hearts. Difficulty with transcatheter ablation techniques arises with complex anatomic variants, or

Table 105.1 Arrhythmia Surgery and Congenital Heart Disease Repair: Children's Memorial Hospital and the Cleveland Clinic Children's Hospital Experience 1987 to 2010 ($n = 248$)

Mean age	21.3 ± 10.9 y (range: 7 d to 64 y)
Associated congenital heart disease	236 patients
Resternotomy	83%
Functionally univentricular heart	67%
Two ventricles	33%
Symptomatic due to arrhythmia recurrences	100%

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Table 105.2 Arrhythmia Types

	N
Macro-reentrant atrial tachycardia	117
Right-sided	104
Right- and left-sided	13
Atrial fibrillation	86
AV nodal reentry tachycardia	6
Accessory connections (WPW)	19
Focal (automatic) atrial tachycardia	6
Ventricular tachycardia	14

AV, atrioventricular; WPW, Wolff–Parkinson–White syndrome. (Reproduced with permission from Mavroudis C, Deal BJ, Stewart RD. Operative techniques in association with arrhythmia surgery in patients with congenital heart disease. World J Pediatr Congen Heart Surg 2013;4:85-97.)

Table 105.3 Associated Diagnoses

	N
Functionally univentricular heart	164
Tetralogy of Fallot/PA-VSD/DORV	22
Transposition of great arteries (1 ccTGA)	11
VSD (1 multiple)	7
AV canal (5 partial)	7
ASD (1 sinus venosus)	5
PA/PS-IVS	5
Aortic stenosis/atresia	3
Ebstein's anomaly	4
Mitral regurgitation	3
Truncus, Uhl, absent PV, Scimitar	5
Structurally normal heart	12

ASD, atrial septal defect; AV, atrioventricular; ccTGA, congenitally corrected transposition of the great arteries; DORV, double-outlet right ventricle; IVS, interventricular septum; PA, pulmonary atresia; PS, pulmonary stenosis; PV, pulmonary valve; VSD, ventricular septal defect. (Reproduced with permission from Mavroudis C, Deal BJ, Stewart RD. Operative techniques in association with arrhythmia surgery in patients with congenital heart disease. World J Pediatr Congen Heart Surg 2013;4:85-97.)

complexities posed by limitations of venous access or surgical baffles or conduits. Patients with double-outlet right ventricle with subaortic conus can harbor an accessory connection in the subaortic conus between the discontinuous aortic annulus and mitral annulus, a situation that does not exist in normal hearts where the ablative techniques have been standardized. Patients with heterotaxy syndrome with either a functionally univentricular heart or two separate ventricles may have absence of the coronary sinus, presence of a left superior vena cava, separate atrial entry of the hepatic veins, and/or juxtaposition of the atrial appendages, in addition to surgical baffles limiting access to regions of the atria. Patients with congenitally corrected transposition of the great arteries (ccTGA) have a propensity to display multiple accessory connections in association with Ebstein's anomaly of the systemic tricuspid valve. In addition to these anatomic variants, repaired tetralogy patients have a high incidence of ART caused by atrial dilatation and scar formation, as well as VT in the area of the ventricular septal defect closure and right ventriculotomy. Transcatheter ablation in the older Fontan patient is hampered by both the multiplicity of arrhythmia mechanisms and circuits, location of some arrhythmia circuits in the left atrium or under surgical patches over the atrial isthmus, as well as the marked atrial hypertrophy limiting the ability to create transmural conduction block. The contemporary congenital heart surgeon should have a comprehensive understanding of all arrhythmia types and the potential methods of ablation, as well as hurdles of anatomic complexity for arrhythmia surgery if a cardiac operation or reoperation for an anatomic repair/re-repair is required in a patient with arrhythmias.

ARRHYTHMIA-SPECIFIC SURGERY TECHNIQUES

The basic techniques for arrhythmia surgery include dissection, linear or focal ablation (sometimes in tandem with dissection), and maze procedures. Focal or micro-reentrant tachycardias require targeted dissection or ablation of the localized arrhythmogenic focus. Accessory connections are treated with discrete ablation, or endocardial dissection along the AV annulus for multiple or complex-branching connections, as seen with Ebstein's anomaly of the tricuspid valve. maze procedures are designed to eliminate macro-reentrant

Table 105.4 Types of Arrhythmias

Arrhythmia	Examples
Bradycardia	Sinus node dysfunction; atrioventricular block
Tachycardia	
	Supraventricular
	Atrial flutter/reentry
	Atrial fibrillation
	Focal
	Accessory connection (WPW)
	AV nodal reentry or junctional
	Ventricular
	Secondary: inotropes, electrolytes, injury
	Scar-related
	Ion channelopathy mediated
Sudden death	Arrhythmic:
	Bradycardic: asystole, or complete atrioventricular block
	Atrial tachycardia
	Ventricular tachycardia/fibrillation
	Nonarrhythmic: pulmonary embolus, myocardial ischemia, electromechanical dissociation
	Cerebrovascular accident

AV, atrioventricular; WPW, Wolff–Parkinson–White syndrome.

circuits in the atria, while preserving intact AV conduction and atrial transport function, and may be performed using a combination of resection and linear ablative lesions. The right atrial maze procedure is performed for atrial reentry or atrial flutter, and both a right and left atrial maze procedure is performed for AF. The right atrial maze procedure alone is not adequate to eliminate AF, which is primarily of left atrial origin.

MACRO-REENTRANT ATRIAL TACHYCARDIA; ATRIAL REENTRY TACHYCARDIA

As noted previously, typical atrial flutter is a macro-reentrant circuit of the right atrium with an area of slowed conduction between the inferior vena cava (IVC), tricuspid valve, and the coronary sinus, characterized by saw-toothed flutter waves visible on electrocardiogram, and a relatively rapid atrial rate. In repaired congenital heart disease, macro-reentrant atrial tachycardia may occur in either atrium and may involve areas of slow conduction posed by electrical or surgical scar (crista terminalis, atrial septal defect patch) or anatomic orifices (inferior or superior vena cava, atriopulmonary anastomosis, entry of pulmonary veins or anomalies of venous return); in this setting the arrhythmia is labeled "atrial reentrant" or "intra-atrial reentrant" or "scar-mediated atrial reentrant" by various authors. Typical flutter waves are not usually apparent, and atrial rates are slower than typical atrial flutter, resulting in discrete P waves visible on electrocardiogram with variable AV conduction. Ablative lesions are used to interrupt the macro-reentrant circuit in the areas of slowed conduction with anatomic barriers, most commonly at the inferior right atrial isthmus, defined as the area between the coronary sinus, the tricuspid annulus, and the IVC. The ablative lesion, therefore, transforms an area of slow conduction to an area of no conduction, thereby eliminating the circuit. In normal hearts, the therapeutic lesions are noted in Figure 105.1.

Multiple areas of slow conduction may exist in patients with congenital heart disease from prior incisions, patches, or wall stress and the ablative lesions target the bridges between anatomic barriers. Identification of the relevant macro-reentrant circuits and targeting the potential areas of slowed conduction suitable for linear ablations (connecting anatomic barriers) are key to successful ablation. While the anatomic variants are many, the tenets of ablative therapy are constant, namely, that lines of block are established between two or more anatomic barriers. For right ART in the setting of congenital heart disease, the surgeon is concerned with the inferior and superior vena cavae, hepatic venous entry,

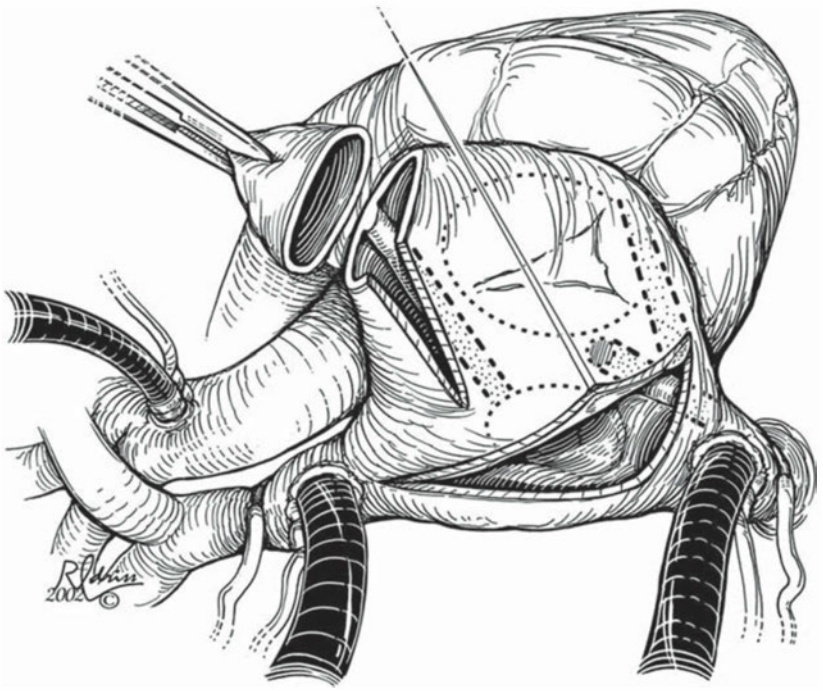
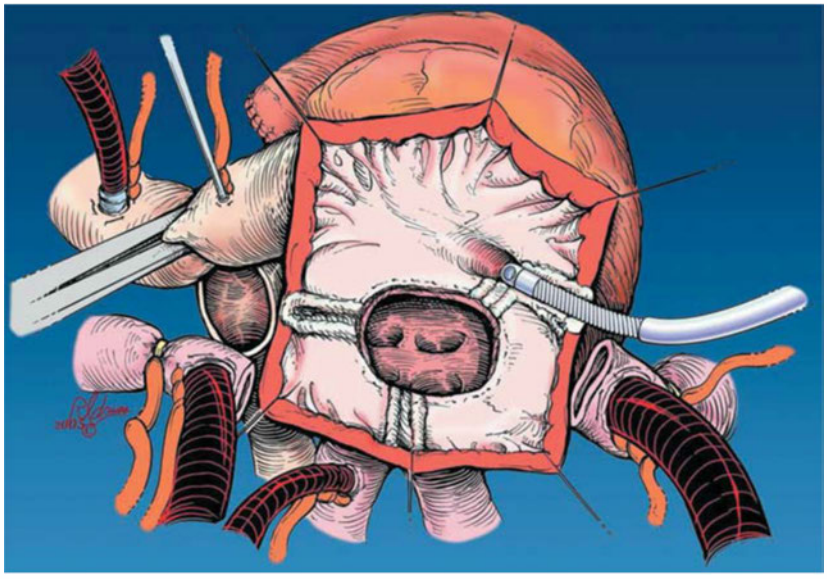
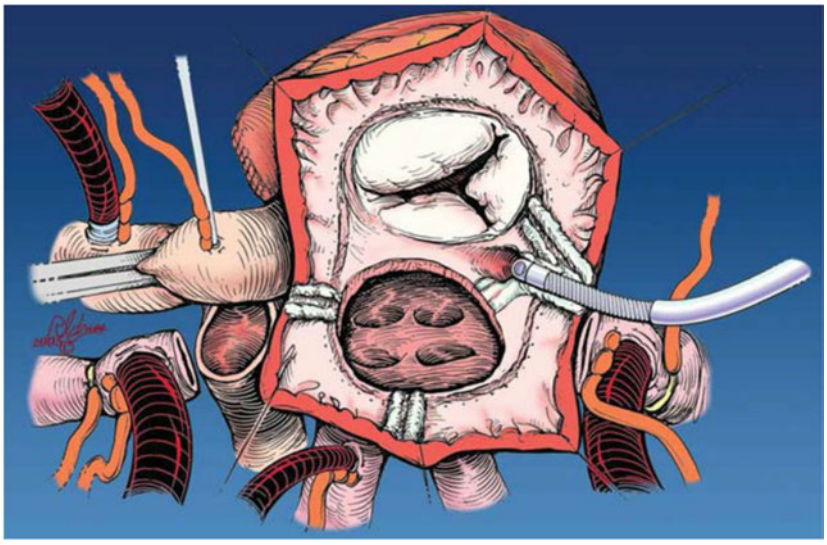


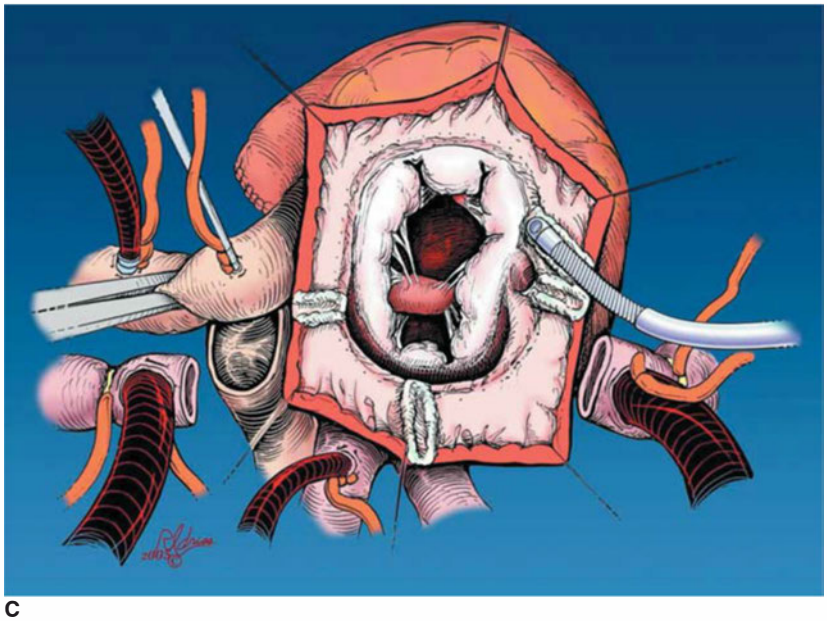
Fig. 105.1. Right-sided maze procedure used in patients with atrial reentry tachycardia and a two-ventricle circulation. Some of the lesions are "cut and sew," others are performed with a cryoablation catheter (−160°C × 1 minute). (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL. Arrhythmia surgery in association with complex congenital heart repairs excluding patients with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2003;6:33-50.)



A



B



C

AV valve, coronary sinus, atrial appendage, and the fossa ovalis or atrial septal patch. When anatomic barriers are absent or anomalous, creative application become necessary by delivering ablative lesions using the guidelines mentioned previously. For example, since there is no tricuspid valve in tricuspid atresia, cryoablation lesions are applied as noted in Figure 105.2A. Figure 105.2B and 105.2C demonstrates the lesions that are required for single right ventricle/mitral atresia and functionally univentricular hearts with unbalanced AV canal, respectively. Figure 105.3 is a complex anatomic diagrammatic representation of the potential anomalies of systemic and pulmonary venous return, juxtaposed atrial appendages and proposed lines of block that are recommended for right- and left-sided maze procedures. The ablative lesions noted in Figure 105.3 are designed to be guidelines that are executed based on the preoperative electrophysiologic study and the type of arrhythmia that has been characterized. As an example, biatrial maze operations are performed if suture lines extend into the left atrium or if there is anomalous left superior vena caval drainage.

FOCAL OR AUTOMATIC ATRIAL TACHYCARDIA

Focal (automatic) atrial tachycardia is characterized by a localized area of electrical impulse generation, which may be caused by discrete micro-reentry or by an automatic focus. Either form of focal tachycardia inhibits the slower rate of normal sinus node firing and results in tachycardia. The electrical impulse from this ectopic focus is conducted and propagated through the atria, often with prolongation of the PR interval, resulting in stimulation of the AV node and ventricles. Surgical treatment of focal (automatic) atrial tachycardia usually requires resection and/or ablation of atrial tissue; more extensive

Fig. 105.2. These are a series of illustrations of the use of cryoablation in patients undergoing Fontan conversion. (**A**) The modified right-sided maze procedure in a patient with tricuspid atresia. (**B**) The modified right-sided maze procedure in a patient with double-outlet right ventricle and mitral atresia. (**C**) The modified right atrial maze procedure in a patient with a functionally univentricular heart (unbalanced atrioventricular septal defect). (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, Johnsrude C, Strasburger J. Total cavopulmonary conversion and maze procedure for patients with failure of the Fontan operation. J Thorac Cardiovasc Surg 2001;122:863-871.)

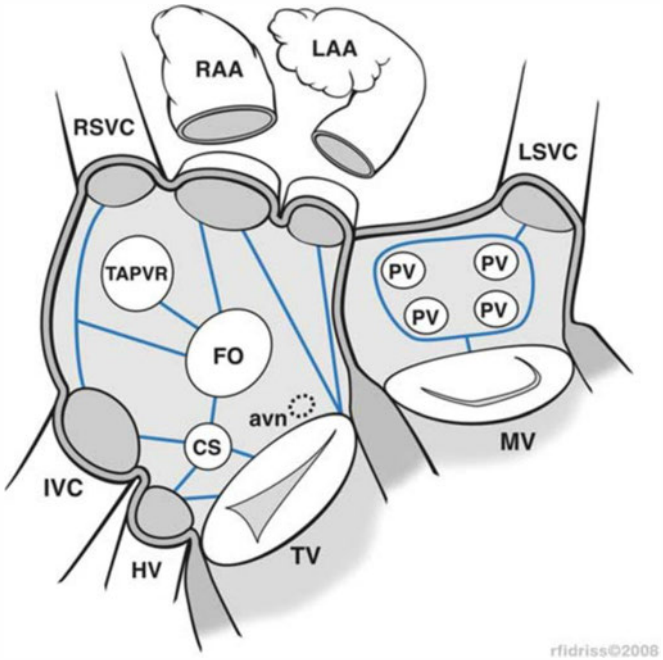


Fig. 105.3. Schematic representation of the possible lines of ablation to treat macro-reentrant atrial tachycardia in the presence of various atrial anomalies associated with complex congenital heart disease. avn, atrioventricular node; CS, coronary sinus; FO, foramen ovale; HV, hepatic vein; IVC, inferior vena cava; LAA, left atrial appendage; LSVC, left superior vena cava; MV, mitral valve; PV, pulmonary valve; RAA, right atrial appendage; RSVC, right superior vena cava; TAPVR, total anomalous pulmonary venous return; TV, tricuspid valve. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL, Tsao S. Arrhythmia surgery in patients with and without congenital heart disease. Ann Thorac Surg 2008;86:857-868.)

areas of focal tachycardia may require electrical anatomic isolation. Favorable outcomes have been achieved with cryoablation and excision of automatic foci. Automatic right atrial foci are often localized to the right atrial appendage and can be resected. Multiple ectopic foci resulting in arrhythmia recurrence stimulated surgeons to use more extensive and creative techniques such as pulmonary vein isolation, left atrial isolation, right atrial isolation, and His bundle ablation with pacemaker insertion in difficult cases.

We have successfully performed atrial appendage resection for automatic atrial tachycardia in two neonates and five older children (Fig. 105.4). One neonate underwent a concomitant Norwood procedure for hypoplastic left heart syndrome and the other, with a normal heart, was spared extracorporeal membrane oxygenation (ECMO) after the arrhythmia site (right atrial appendage) was resected and ablated in the operating room. Of the five older children, three underwent concomitant atrial ($n = 2$) and ventricular ($n = 1$) septal defect repairs with successful focal cryoablation and the other two had normal hearts with depressed ventricular function due to the arrhythmia. Atrial tachycardia can cause "idiopathic" dilated cardiomyopathy; appropriate and aggressive arrhythmia treatment may be curative, and allow ventricular function to recover. One teenaged patient in our series presented with cardiogenic shock due to atrial tachycardia. The patient underwent emergent median sternotomy for the placement of a mechanical assist device. However, a focal right atrial tachycardia originating in the right atrial appendage was identified and resected, resulting in sinus rhythm obviating the need for the ventricular assist device. Subsequently, ventricular function recovered over a period of weeks. In the current era, automatic atrial tachycardia is usually treated by transcatheter ablation. Rare refractory cases, however, require an individualized treatment plan for accurate diagnosis and ablation.

ACCESSORY CONNECTION-MEDIATED TACHYCARDIA

Accessory connections may "manifest" on electrocardiogram during sinus rhythm resulting in the appearance of a delta wave, or Wolff–Parkinson–White (WPW) syndrome in this setting there is fusion of conduction from atria to ventricles over both the AV node and the accessory connection. More commonly accessory connections are "concealed," and conduct only in the retrograde direction from ventricle to atrium; a delta wave is not present during sinus rhythm. The accessory connection provides the substrate for a macro-reentrant circuit including the atria, AV node, ventricular myocardium, accessory connection, and return to the atria. Thus, the existence of an accessory connection may contribute to several forms of tachycardia, most commonly orthodromic reciprocating tachycardia, but also antidromic reciprocating tachycardia, and AF with rapid antidromic conduction via a manifest accessory connection. During orthodromic reciprocating

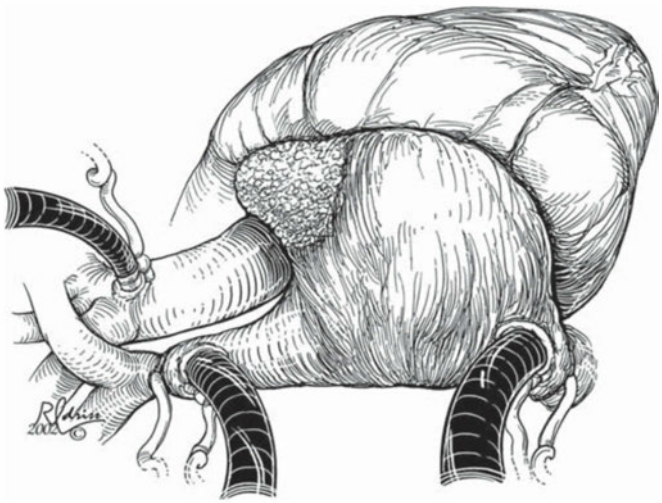


Fig. 105.4. Gross anatomic findings of dysplastic atrial tissue confined to the right atrial appendage. This was a focus of automatic atrial tachycardia. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL. Arrhythmia surgery in association with complex congenital heart repairs excluding patients with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2003;6:33-50.)

tachycardia, conduction proceeds normally from the right atrium through the AV node to the ventricle, and then via the accessory connection to transmit the electrical wavefront from the ventricle back to the atrium. Interruption of this macro-reentrant circuit at either the AV node (adenosine) or at the accessory connection (procainamide, ablation) interrupts the circuit and terminates tachycardia. Antidromic, as the word indicates is a circuit moving in the opposite direction and is associated with unidirectional block in the AV node, with conduction proceeding antegrade via a manifest accessory connection from atrium to the ventricle; if the impulse reenters via the AV node in the retrograde direction, antidromic reciprocating tachycardia ensues. Because the manifest accessory connection may be capable of very rapid antegrade conduction (without the inherent electrical delay of the AV node), the potential exists for triggering ventricular fibrillation and is the cause of sudden death in the setting of WPW.

In the normal heart, accessory connections are most frequently left-sided, usually left laterally or posteriorly. In congenital heart disease, accessory connections may be associated with pathology of the corresponding AV valve. Thus, Ebstein's anomaly has the highest association of accessory connections related to the tricuspid valve. In the setting of ccTGA, the accessory connections are associated with the left-sided, tricuspid valve. Currently, accessory connections are optimally treated by transcatheter ablation, with acute success rates of 70% to 98% depending on accessory connection location. Excellent results have been achieved using this approach, and transcatheter ablation of manifest accessory connections is recommended prior to planned operative therapy for anatomic correction/palliation. In the setting of Ebstein's anomaly, due to the frequent association of concealed right-sided accessory connections, preoperative electrophysiology study and ablation as necessary is recommended. When an operative procedure is planned, the team must take into consideration all variables that attend the eventual repair.

The transcatheter ablation procedure can precede the anatomic repair. Alternatively, anatomic repair and the ablation procedure may be performed together in the operating room. The conditions influencing operative repair include but are not limited to right-to-left shunt, number and location of accessory connections, previously failed attempts at catheter ablation, projected cross-clamp time for the planned procedures, experience of the operating team using ablative procedures for accessory connections, and the degree of ventricular dysfunction. A staged and collaborative approach of preoperative mapping and localization of accessory connections, with transcatheter ablation as possible, and identification of anatomic considerations such as an epicardial accessory connection location or multiple accessory connections guiding operative arrhythmia surgery is optimal.

ATRIOVENTRICULAR NODAL REENTRY TACHYCARDIA

AV nodal reentry occurs when there is *functional* dissociation of conduction to the AV node between "fast" and "slow" conducting tissue in the approaches to the compact AV node, or His bundle. Normal AV nodal conduction occurs over the anterior located fast "pathway," with a normal PR interval. Should block occur in this tissue, conduction via the slow tissue, usually located more inferiorly and posteriorly, occurs with a longer PR interval. Reentry may then occur via the fast pathway, allowing *typical AV nodal reentry* to sustain (antegrade "slow," retrograde "fast" pathways). This arrhythmia is characterized by a narrow QRS with the retrograde (fast) P wave buried within the QRS; frequently appearing as an rSr' pattern in lead V1 on electrocardiogram. *Atypical* forms of AV nodal reentry occur less commonly, including antegrade fast and retrograde slow circuits, as well as intermediate variants. Initial attempts at catheter ablation of AVNRT targeted the region of the fast pathway just proximal to the compact AV node and carried the risk of developing AV block. Presently, the usual catheter ablation approach targets the slow AV nodal pathway, in the region anterior to the coronary sinus ostium and carries success rates of over 95%. AVNRT occurs in association with certain forms of congenital heart disease, involving either anatomic variation of AV nodal tissue (twin AV nodes in heterotaxy syndrome), or surgical manipulation in the region of the approaches to the AV node (Mustard or Senning repairs of transposition of the great arteries). In patients with prior atrial baffle repairs, successful ablation targets atrial tissue on the pulmonary venous side of the baffle, necessitating a retrograde approach to the pulmonary venous chamber, or transvenous baffle perforation.

At the present time, AVNRT is treated by either the cryoablation or radiofrequency technique. Operatively, a linear lesion is placed from the posterior inferior rim of the coronary sinus ostium to the IVC. In the presence of a right-sided AV valve, a linear lesion from the valve to the posterior ostium of the coronary sinus is delivered. The operative approach in a beating nonworking heart using discrete cryolesions around the coronary sinus is no longer used. Results of catheter modification of the AV node slow pathway in the region of the coronary sinus ostium indicate that it is not desirable to attain prolongation of the PR interval, as previously performed. In patients with prior Mustard or Senning procedures undergoing reoperation, we have elected to perform ablation of AVNRT directly intraoperatively and avoid the retrograde catheter approach to the pulmonary venous atrium.

ATRIAL FIBRILLATION

AF is predominantly a left atrial arrhythmia characterized by very rapid chaotic atrial activity with variable AV conduction. On ECG, consistent P waves are replaced by rapid oscillations or fibrillatory waves that vary in amplitude, shape, and timing. AF is frequently associated with left-sided valvar pathology and carries a risk of atrial thrombosis formation and stroke, in addition to decreased cardiac output in patients dependent on atrial contributions to cardiac output, such as patients with a functionally univentricular heart. The mechanism of AF may be due to multiple micro-reentrant atrial wavelets, focal triggers from sleeves of cardiac muscular tissue often within pulmonary veins, or localized reentrant circuits with fibrillatory conduction. Sympathetic and parasympathetic innervation from autonomic ganglia located on the epicardial surface of the posterior left atrium may contribute to perpetuation of fibrillation. In addition to a trigger, an anatomic substrate is needed for the maintenance of AF, and atrial remodeling occurs rapidly with the onset of AF. Despite characterization as multiple reentrant wavelets, AF does not terminate with atrial overdrive pacing.

In the setting of congenital heart disease, AF is associated with unoperated atrial septal defects in older patients, mitral valve regurgitation or stenosis, hypertrophic cardiomyopathy, and in patients at risk for recurrent atrial arrhythmias, such as atrial repairs of transposition of the great arteries, Ebstein's anomaly, or the Fontan procedure. The risk of developing AF late after closure of atrial septal defects increases with advancing age, such that as many as 60% of patients over 40 years of age at the time of intervention will develop AF. For patients undergoing surgery for ASD closure, preoperative atrial arrhythmias or age >40 years should prompt

consideration for concurrent AF surgery. Surgical intervention solely for hemodynamic problems will not eliminate AF, and arrhythmia surgery is recommended concurrently for at-risk patients undergoing structural surgery. Right atrial arrhythmia surgery is not an effective treatment for AF. The surgical treatment of AF was introduced in 1987 by Cox, Boineau, and Schuessler, and involved a "cut and sew" approach designed to prevent perpetuation of reentrant circuits, while preserving AV conduction and atrial transport function via a "maze." There have been many subsequent modifications of the maze lesion sets in an effort to minimize the extent of surgery, including epicardial approaches and thoracotomies, or a hybrid approach; freedom from recurrent AF is improved in patients undergoing the more extensive full lesion sets. The goals of the full lesion set are to isolate the pulmonary venous atrium, and to connect the encircling pulmonary venous lesion to the mitral annulus and the base of the resected left atrial appendage, in addition to a dome lesion at the roof of the left atrium, either to the anterior mitral annulus or to the right atrial appendage. The ability to use linear or specially designed ablation catheters has largely eliminated the need for the lengthy "cut and sew" approach. Different energy sources are used, including cryoablation, radiofrequency, bipolar radiofrequency, ultrasound, and laser. In adults with unoperated atrial septal defects, atrial flutter and fibrillation occur in 14% to 22% of patients. Surgical ablation for AF prevents AF recurrence in over 90% of adult patients, often in association with mitral valve repair, atrial septal defect closure, or coronary artery bypass surgery. We have incorporated successfully the Cox–maze III procedure into repair of complex congenital heart disease, including patients with functionally univentricular hearts undergoing Fontan conversion (Fig. 105.5), atrial septal defect closure, and mitral valve repair in patients as young as 4 years of age. We have had no late recurrence of AF in our patient population undergoing the maze III lesions, although organized ART has recurred in up to 17% of patients, similar to the incidence reported from other centers. The addition of lesions to the mitral annulus aligned with a corresponding epicardial lesion, and dome lesions is performed in an effort to reduce the risk of late tachycardia recurrence.

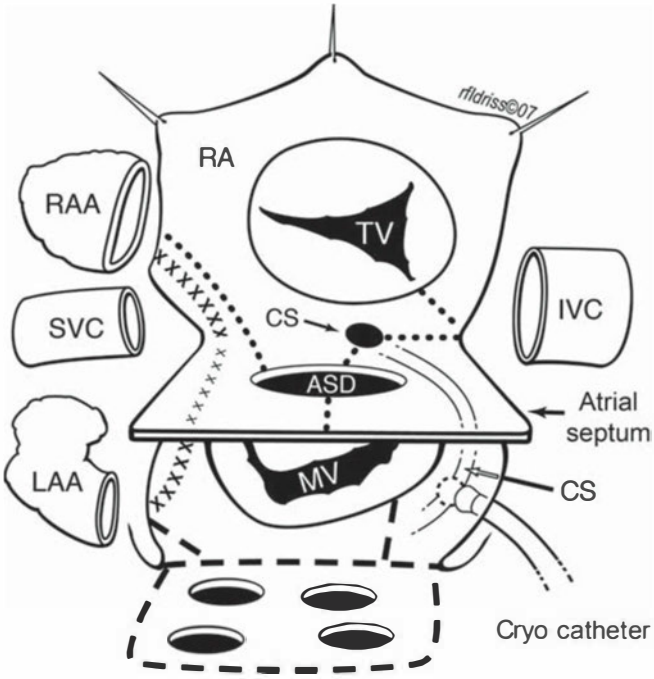


Fig. 105.5. Diagrammatic representation of the modified right-sided maze procedure for right atrial reentry tachycardia (*dotted lines*) and the left atrial Cox–maze procedure (*dashed lines*) for atrial fibrillation and left atrial reentry tachycardia. The (*xx*) represents a line of block that encompasses both atria and not just one. ASD, atrial septal defect; CS, coronary sinus; IVC, inferior vena cava; LAA, left atrial appendage; MV, mitral valve; RA, right atrium; RAA, right atrial appendage; SVC, superior vena cava; TV, tricuspid valve. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL, et al. 111 Fontan conversions with arrhythmia surgery: surgical lessons and outcomes. Ann Thorac Surg 2007;84:1457-1466.)

VENTRICULAR TACHYCARDIA

VT is defined as three or more consecutive ventricular beats at rates >150 bpm in older patients, with *sustained VT* defined as lasting >30 seconds or requiring intervention prior to that time due to hemodynamic compromise. Among pediatric patients, the most common setting for VT is following repair of lesions requiring ventricular surgery, particularly tetralogy of Fallot and double-outlet right ventricle. Late VT is being increasingly recognized in older patients with atrial repairs for transposition of the great arteries, Fontan procedure for functionally univentricular heart, and Ebstein's anomaly. Nonsustained VT in these patients may be a marker for declining ventricular function, and successful reoperations for residual hemodynamic problems may decrease the frequency of ventricular arrhythmias. However, patients with sustained VT prior to surgery are at increased risk for recurrences, and intervention for the arrhythmia is recommended. VT is associated with cardiac tumors, most notably fibromas, as well as rhabdomyomas, myxomas, or hemangiomas. Intractable VT of infancy is associated with histiocytoid cardiomyopathy and ion channel disorders. Patients with VT refractory to medications and transvenous catheter ablation techniques as well as patients undergoing concomitant repair of structural heart disease are considered for intraoperative ablation of VT.

As noted earlier, incessant life-threatening VT in infancy is associated with ventricular hamartomas or histiocytosis. Before the availability of intravenous amiodarone, such infants underwent surgical resection of the left ventricular endomyocardium with some success. In older patients, usually adolescents, idiopathic VT in the ostensibly normal heart typically arises from either the right ventricular outflow tract (left bundle branch block, normal to rightward QRS axis morphology) or the septal surface of the left ventricle (right bundle branch block, left axis morphology). Both forms of idiopathic VT are amenable to catheter ablation techniques, with success rates of 70% to 80%. Rarely, VT may originate from the epicardial surface of either the left or right ventricle and is amenable to epicardial arrhythmia surgery with success rates over 70%. Detailed preoperative and intraoperative mapping of the arrhythmia focus is essential to the successful performance of such procedures.

STRUCTURAL HEART DISEASE

Patients with postoperative tetralogy of Fallot or double-outlet right ventricle are at risk for late postoperative sustained VT, occurring in 5% to 8% of cases. Moreover, the risk of late sudden death is 2% to 6%. Risk factors for VT include older age at initial repair, residual right ventricular hypertension, right ventricular outflow tract patch or aneurysm, significant pulmonary regurgitation, prolonged QRS duration over 180 ms on resting electrocardiogram, abnormal signal-averaged electrocardiogram, and longer duration of follow-up. At least 15% of patients with repaired tetralogy of Fallot undergo reoperation for residual defects or late pulmonary valve regurgitation. Ambulatory electrocardiographic monitoring and exercise testing are excellent diagnostic tools to demonstrate sustained VT as well as significant atrial and ventricular arrhythmias. Electrophysiologic studies are indicated for patients with symptoms of palpitations, syncope or cardiac arrest, sustained wide-QRS tachycardia, or QRS prolongation >180 ms. Sustained VT may be ablated preoperatively; should catheter ablation fail, direct endocardial resection and cryoablation may be performed intraoperatively. We and others have repaired residual hemodynamic abnormalities in the adult patient with tetralogy of Fallot concomitantly with resection and cryoablation of the tachycardia circuit. This oftentimes eliminates the need for antiarrhythmic drugs or an implanted defibrillator. Preoperative and intraoperative mapping of tachycardia will demonstrate the common sites of arrhythmia origin that are related to the ventriculotomy, outflow tract patch, and perimeter of the ventricular septal defect. Postoperative ventricular stimulation is indicated to determine efficacy. Defibrillator implantation is performed if sustained VT remains inducible. Successful surgical treatment of VT in this setting ranges from 50% to 70%. Reasons for decreased success for surgical therapy of VT following repair of congenital heart disease may be due to inadequate mapping, perhaps related to deep intramural origin with late activation of the endocardial surface, as well as the limited ability to safely resect significant amounts of ventricular myocardium.

In rare cases, patients with congenital heart disease may have sustained VT before primary surgical repair. Under these circumstances, preoperative electrophysiologic mapping can successfully localize the arrhythmia source, which may be associated with a hemodynamic jet lesion. We have successfully resected such lesions in two young patients undergoing VSD closure. Large white plaque lesions noted opposite to the VSD jet were resected and cryoablated. VT associated with dilated nonischemic cardiomyopathy may be approached surgically with a decrease in frequency of recurrences, as reported in older patients; most patients undergoing VT surgery also receive implanted defibrillators.

VT may be secondary to ion channelopathies including long QT syndrome, and catecholaminergic VT associated with disorders of ryanodine receptors or calsequestrin. Severe ventricular arrhythmia storms may present in infancy, and such patients have an increased incidence of "double hits" for gene disorders. The frequency of electrical storms can be substantially reduced by adjunctive therapy using left cervical sympathectomy. Surgical sympathetic denervation is guided by video-assisted thoracotomy (VATS) techniques, which identify the thoracic ganglion allowing partial resection. We have successfully used bedside anesthetic nerve block of the cervical ganglion prior to proceeding with surgical sympathectomy in such an infant.

OPERATIVE TECHNIQUES SPECIFIC TO CONGENITAL HEART DISEASE

Techniques of Resternotomy and Cannulation

Eighty-three percent of our patient population required resternotomy. Since our unwanted cavitary entry rate is low, we describe our techniques for resternotomy. While femoral cannulation has long been employed for emergency or prophylactic cardiopulmonary bypass in difficult cases, recent publications have offered axillary cannulation, especially for cases involving aortic ascending aneurysms or sternal adherence of cardiac chambers in which there is a high probability of myocardial injury during resternotomy. The incision and dissection of the axillary artery in preparation for arterial cannulation are shown in Figure 105.6A. The dissection is performed

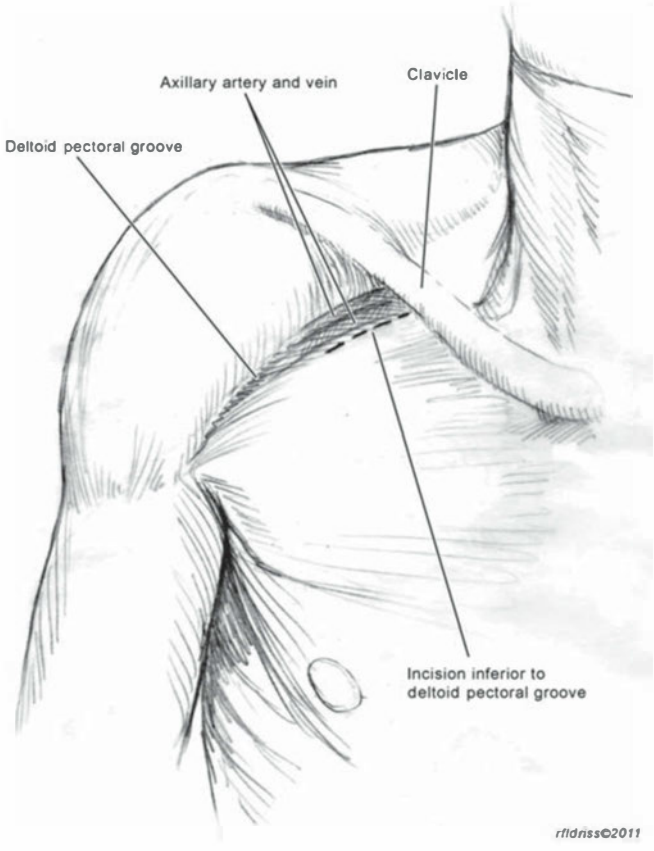
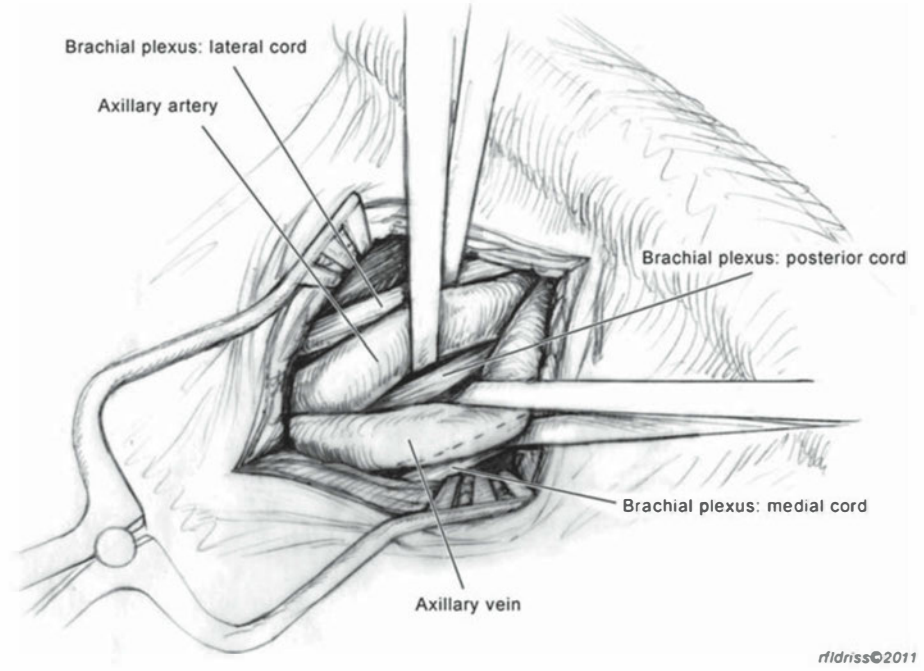
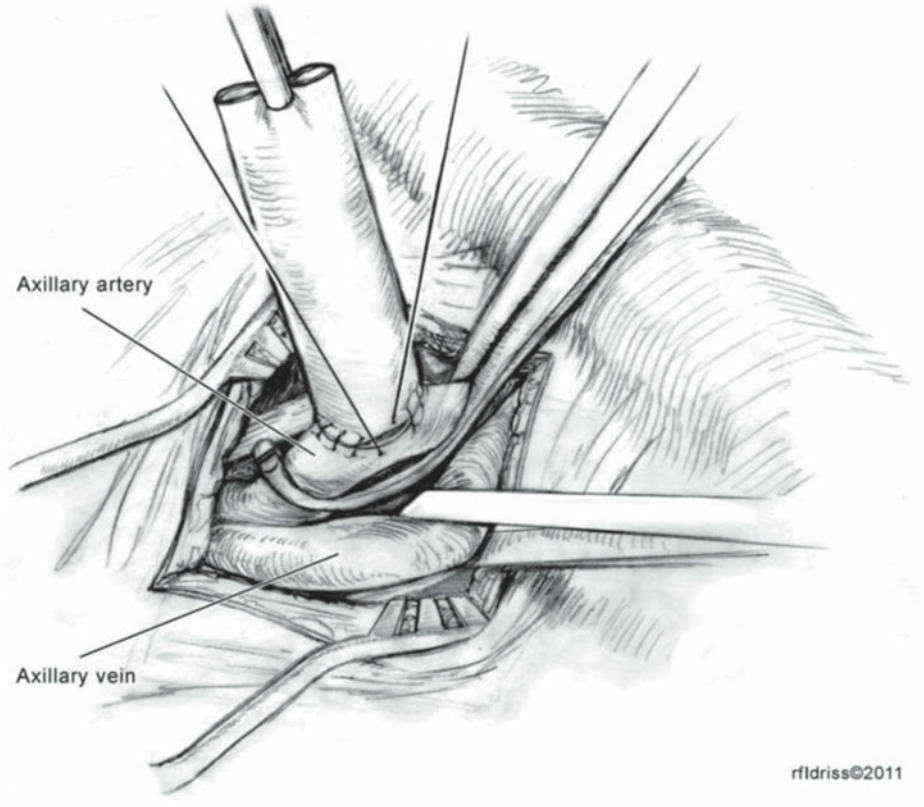


Fig. 105.6. (**A**) Surface anatomy for incision and dissection of axillary artery in preparation for axillary arterial cannulation. (**B**) The axillary artery is dissected from the brachial plexus cords. (**C**) A polytetrafluoroethylene graft is anastomosed end-to-side to the axillary artery. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL, Stewart RD. Operative techniques in association with arrhythmia surgery in patients with congenital heart disease. World J Pediatr Congen Heart Surg 2013;4:85-97.)



B



C

Fig. 105.6. (*Continued*)

in the deltopectoral groove (Fig. 105.6B). The axillary artery and the brachial plexus cords are carefully dissected; proximal and distal arterial control are easily achieved. After systemic anticoagulation (heparin), a polytetrafluoroethylene (PTFE) graft of appropriate size is anastomosed end-to-side to the axillary artery (Fig. 105.6C). An arterial cannula, usually a femoral cannula, is connected to the graft in preparation for cardiopulmonary bypass. This method allows continuous cardiopulmonary bypass without direct ascending aortic recannulation. Some surgeons prefer this technique when the aortic arch requires repair or replacement since the cannulation site is distant from the operative exposure. Furthermore, regional cerebral perfusion can be performed using this technique.

There are many options for effective and safe resternotomy. Some surgeons employ self-retaining internal mammary artery retractors that lift the sternum away from the mediastinal structures, thereby facilitating the dissection (Fig. 105.7A). The lower sternal exposure is initiated by scissors dissection where a plane can be developed (Fig. 105.7B). The heart can usually be identified by its contractile motion and the sternum by its firm osseous nature. The technical challenge is to identify the appropriate space between them. Once this is determined, the surgeon may elect to use blunt and electrocautery dissection alternately by creating fenestrated adhesive tissue bands. These can then be lysed by electrocautery all the time directly visualizing the beating heart and the extent of the dissection. A superiorly advancing "delta" dissection plane is created and resternotomy can proceed using scissors, electrocautery, and a standard sternal saw as necessary for a safe mediastinal reentry. Precautionary measures are instituted and include presternotomy defibrillation pads, preference for scissors dissection in the event that electrocautery causes premature ventricular contractions or an episode of ventricular fibrillation, and preference for scissors dissection for aortic adherence to the underside of the sternum. After safe sternotomy, standard aortobicaval cannulation techniques are performed.

Fontan Conversion

A unique subset of patients are those patients with a functionally univentricular heart who have had a prior atriopulmonary Fontan procedure. These patients can develop right atrial dilatation, atrial tachycardia, and AF. This results in hemodynamic compromise and progressive heart failure symptoms. Dr. Hillel Laks reported an experience with three patients with atriopulmonary connection who developed late arrhythmias and atrial thrombus. These three patients were treated with surgical conversion of the atriopulmonary connection to a *lateral tunnel*-type Fontan. Our series at Children's Memorial Hospital began in 1994. We have now performed 136 Fontan conversions. Our primary contribution has been the addition of arrhythmia surgery, consisting of either a right-sided

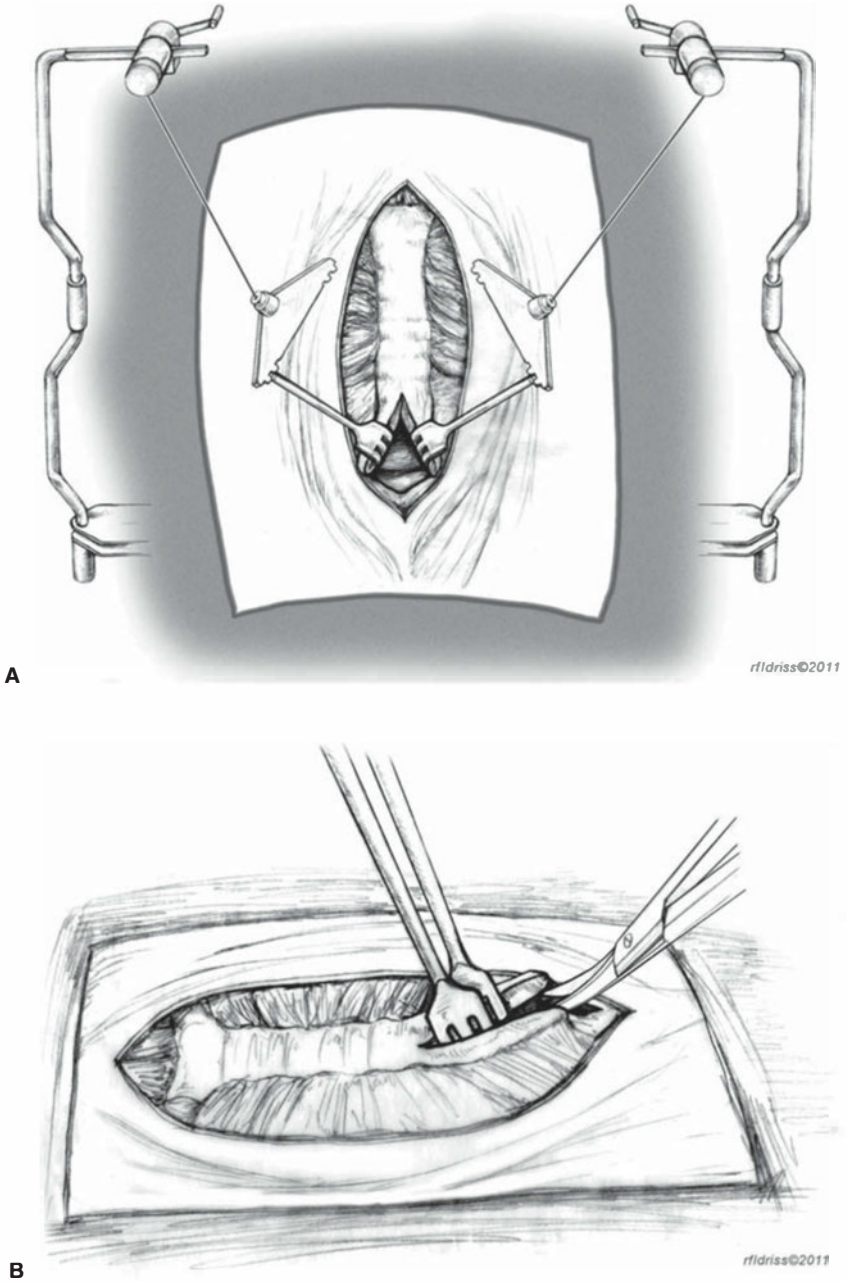


Fig. 105.7. Sternal reentry: (**A**) Using self-retaining internal mammary artery retractors to lift the sternum from the mediastinal structure. (**B**) Substernal scissors dissection. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL, Stewart RD. Operative techniques in association with arrhythmia surgery in patients with congenital heart disease. World J Pediatr Congen Heart Surg 2013;4:85-97.)

maze or biatrial maze. We have also used an *extracardiac* Fontan rather than the lateral tunnel type of connection. The majority of these patients had a diagnosis of either tricuspid atresia or double-inlet left ventricle.

The primary steps of the Fontan conversion procedure are as follows:

1. After placing the patient on cardiopulmonary bypass (Fig. 105.8), the IVC is transected from its entrance into the right atrium and a No. 24 mm PTFE tube graft is sutured to the IVC.
2. Cardioplegia is administered and the majority of the enlarged right atrium is excised.
3. An atrial septal defect is created for the purpose of draining the coronary sinus blood to the left atrium.
4. Right maze or right and left-sided maze are performed with cryoablation lesions as previously illustrated in Figures 105.2, 105.3, and 105.5.
5. The right atrial incision is closed and the cross-clamp is released.
6. Bidirectional superior cavopulmonary anastomosis is created.
7. PTFE tube graft is beveled and anastomosed to the undersurface of the pulmonary artery.
8. Epicardial dual chamber atrial antitachycardia pacemaker is placed.

The completed Fontan conversion procedure is illustrated in Figure 105.9.

From 1994 to 2011, 136 patients have undergone the Fontan conversion with arrhythmia surgery. The mean age at the time of operation was 23 years. Operative mortality was 2 of 136 (1.5%). The mean age at the original Fontan procedure was 7 years. The mean interval from original Fontan procedure to Fontan conversion was 16 years. At 10 years following Fontan conversion, 85% of patients are alive and the freedom from arrhythmia recurrence is also 85%. An analysis of the first 111 patients demonstrated the following risk factors for death or transplant: protein-losing enteropathy, CPB time >4 hours, preexisting moderate-to-severe AV valve insufficiency, and right or ambiguous ventricle.

There are anatomic variations of the Fontan operation that have proven to be a challenge for Fontan conversion and arrhythmia treatment. The following sections describe the conditions that may be encountered that will require special surgical management for an effective arrhythmia ablation procedure. Figure 105.8 shows the cannulation for Fontan conversion. Occasionally, a right atrial clot, which is associated with low cardiac output, can be found in Fontan conversion patients. The conduct of the dissection becomes complicated since oftentimes cardiopulmonary bypass will be required before the caval veins are cannulated. Figure 105.10 shows venous catheter placement away from the atrial clot, which can be identified by epicardial or transesophageal echocardiography, for temporary cardiopulmonary bypass. This maneuver avoids the possibility of clot embolization of the venous catheter resulting in ineffective venous drainage.

Takedown of Right Atrial-to-Right Ventricular Bjork-Fontan Modifications

The Bjork–Fontan modification either with or without a valve was designed to take advantage of a small but contractile right ventricle. The natural history of this operation, however, resulted in right atrial enlargement and atrial arrhythmias. Oftentimes, the small right ventricle has grown

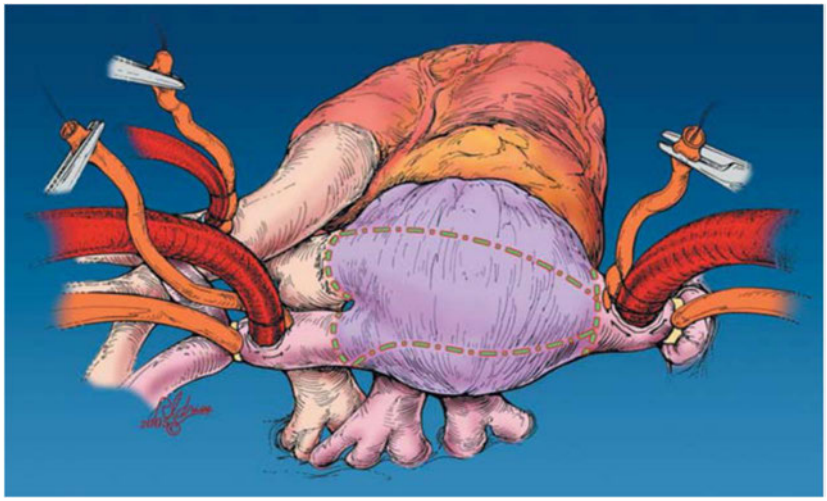


Fig. 105.8. Technique of cannulation for Fontan conversion. Note direct caval venous cannulation. Projected right atrial resection is shown (*dotted lines*). (Reproduced with permission from Backer CL, Deal BJ, Mavroudis C, Franklin WH, Stewart RD. Conversion of the failed Fontan circulation. Cardiol Young 2006;16(Suppl 1):85-91.)

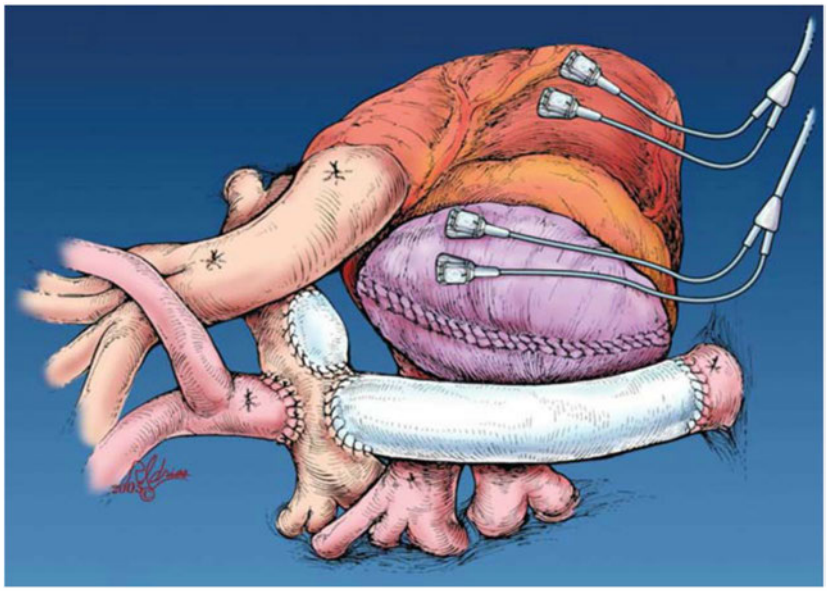


Fig. 105.9. Completion of total cavopulmonary connection with extracardiac polytetrafluoroethylene tube graft, bidirectional superior cavopulmonary anastomosis, and epicardial bipolar atrial and ventricular pacemaker leads. (Reproduced with permission from Mavroudis C, Deal BJ, Backer CL, Fontan conversion/arrhythmia surgery. In Kaiser LR, Kron IL, Spray TL, eds. Mastery of Cardio-Thoracic Surgery, 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2007:1035-1041.)

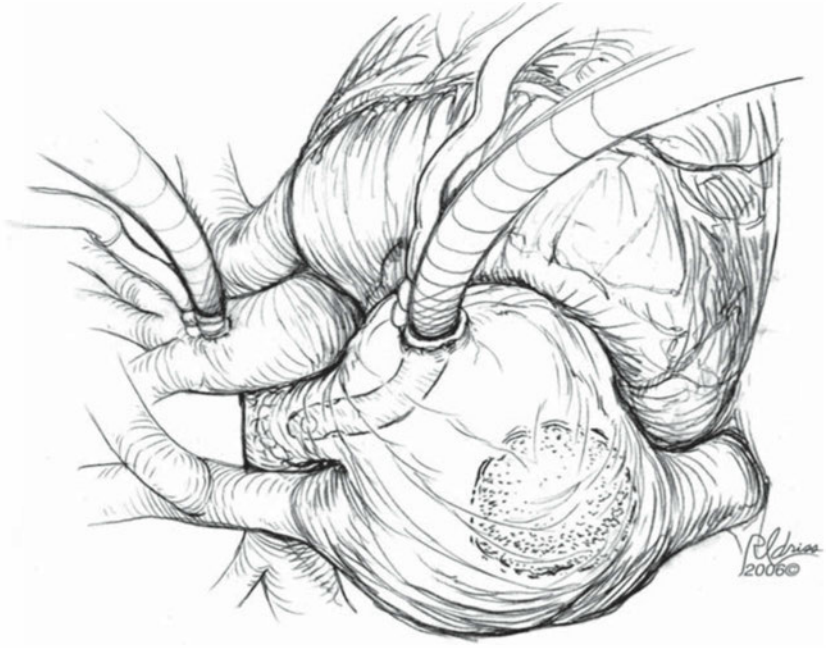


Fig. 105.10. Venous catheter placed away from the clot in the inferior portion of the right atrium. After establishing uniatrial cardiopulmonary bypass (CPB) safely the patient can be converted to bicaval CPB. (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, et al. Evolving anatomic and electrophysiologic considerations associated with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2007;10:136-145.)

during the period of somatic growth. During surgical conversion of a Bjork–Fontan modification to a total cavopulmonary artery connection, the dilemma arises as to the management of the right ventricle.

Disconnecting the main pulmonary artery and leaving the right ventricle without an egress of blood flow has the potential for right ventricular dilatation and leftward interventricular septal deviation caused by accumulation of the besian drainage. Therefore, the surgical considerations en route to total cavopulmonary artery connections are (1) takedown of the Bjork–Fontan anastomosis and (2) management of the right ventricle.

Patients without a bioprosthetic valve interposition graft may have a communication formed by a posterior reversed right atrial flap and an anterior prosthetic patch, which forms the right atrial to right ventricular connection. Surgical takedown involves careful dissection of the right AV grove which facilitates right atrial appendage amputation, right atrial wall reduction, epicardial pacemaker lead placement, and adequate right ventricular wall patch closure. Care is taken to avoid injury to the right coronary artery (Figure 105.11A). We have preserved the right ventricular to main pulmonary artery connection allowing egress of Thebesian blood flow into the pulmonary arteries. Right ventricular pressure is low under these circumstances because of decreased right ventricular preload and does not affect the nonpulsatile blood flow established by the TCPC extracardiac connection. In most cases, the pulmonary valve is competent. Figure 105.11B shows a right ventricular patch and right atrial closure in association with the extracardiac TCPC.

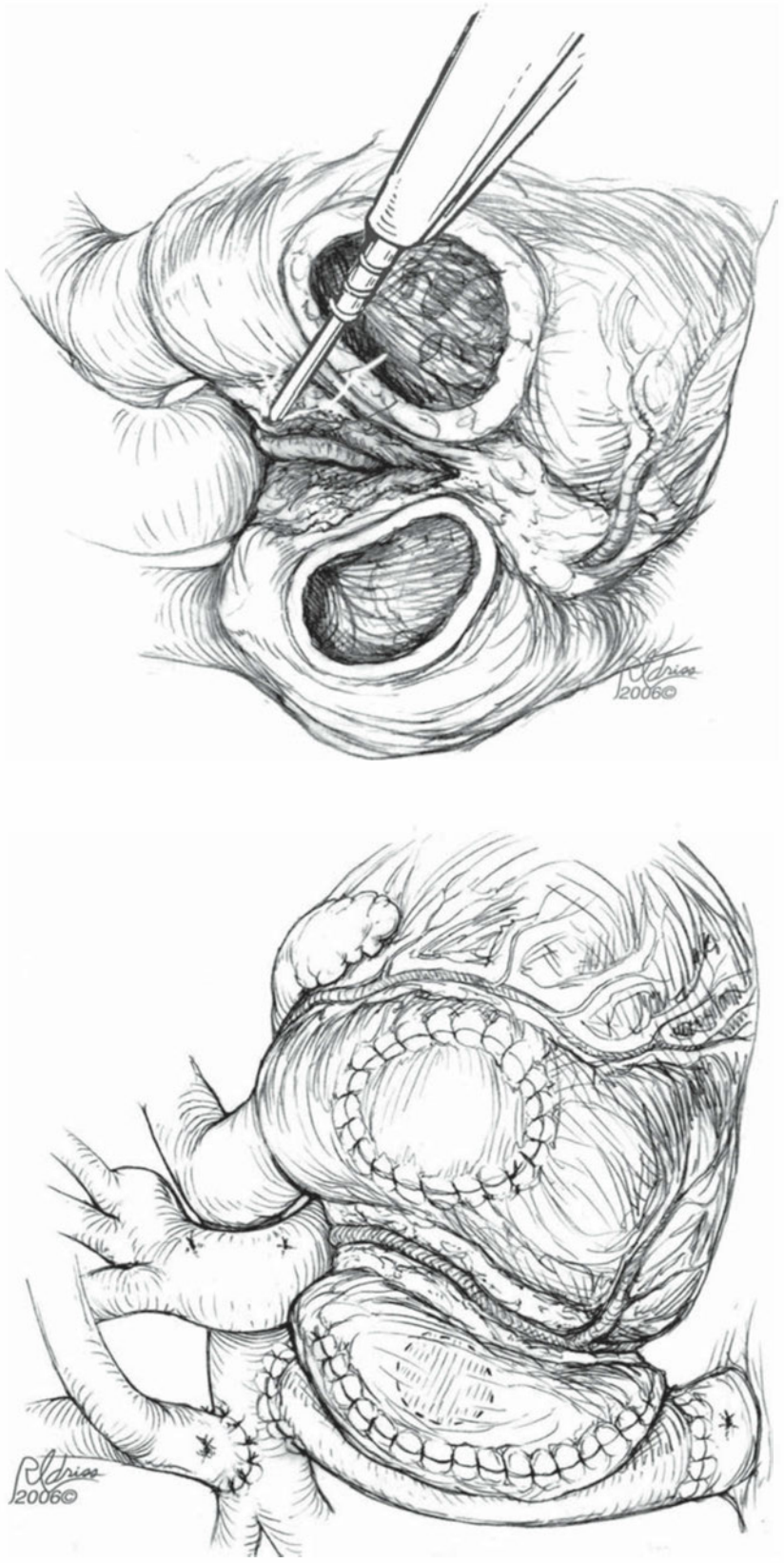


Fig. 105.11. (**A**) Electrocautery dissection at the atrioventricular groove commencing at the base of the aorta with extension to the right ventricular free wall. (**B**) The completed extracardiac polytetrafluoroethylene tube graft from the inferior vena cava to the pulmonary artery is shown. Right atrial wall reduction and closure is noted by the long atrial suture line. Right ventricular to main pulmonary artery continuity is maintained by a right ventricular patch. (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, et al. Evolving anatomic and electrophysiologic considerations associated with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2007;10:136-145.)

RIGHT VENTRICULAR HYPERTENSION AND TRICUSPID REGURGITATION AFTER ATRIOPULMONARY FONTAN FOR PULMONARY ATRESIA AND INTACT VENTRICULAR SEPTUM

Patients with pulmonary atresia and intact ventricular septum can present with a small right ventricle necessitating an eventual Fontan operation. In the early experience with this operation, the small tricuspid valve was intentionally made incompetent. Following atriopulmonary repair, the right ventricle enlarges over time, with marked hypertrophy and suprasystemic right ventricular pressure since there is no forward flow. Consequently, significant tricuspid

regurgitation results in marked right atrial dilatation and hypertension. If an extracardiac TCPC is performed without addressing the regurgitant tricuspid valve, increased common atrial pressure can cause compromise of the Fontan circulation. After performing the modified right-sided maze procedure, we isolated the tricuspid valve with a fenestrated (8 mm) Gore-Tex patch, which restricts blood flow into the right ventricle, and allows blood egress from the ventricle into the common atrium (Fig. 105.12). By unloading the right ventricular volume, RV pressure is significantly reduced without impacting the common atrial pressure.

Takedown of Atrioventricular Valve Isolation Patch for Right-Sided maze Procedure

After the initial successes with the Fontan procedure for tricuspid atresia, it was adapted to more complex anatomy such as double-inlet ventricles, criss-cross hearts, and straddling AV valves. The smaller right-sided AV valve may have been isolated using a patch or the atrial baffle. Patching of this valve accomplished separation of the circulations by establishing an atriopulmonary connection together with an atrial septal defect closure. The relationship of the patch to the valve annulus and the coronary sinus has important anatomic and electrophysiologic considerations. The patch may have partitioned the coronary sinus on the ventricular side to avoid complete heart block. The anatomic right atrial isthmus, therefore, is partitioned to the pulmonary venous atrium. This is problematic since in the presence of right atrial macro-reentry tachycardia, the right-sided maze cannot be safely performed because the traditional landmarks, namely the coronary sinus and the tricuspid valve, are not exposed (Fig. 105.13A). The patch over the right-sided valve therefore must be removed to allow cryoablation according to the tenets of adequate landmark identification (Fig. 105.13B). This aforementioned partitioning and restriction of access to the right atrial isthmus accounts for failure of a transcatheter ablation and will contribute to failure of right atrial maze surgery without resection of the prosthetic patch material. If the tricuspid valve is competent by bulb syringe testing, one can elect to allow the valve to function normally without valvar isolation. In the setting of valve incompetency, the valve can be again isolated using Gore-Tex patch anchored to the tricuspid valve leaflets near the annulus (Fig. 105.13C).

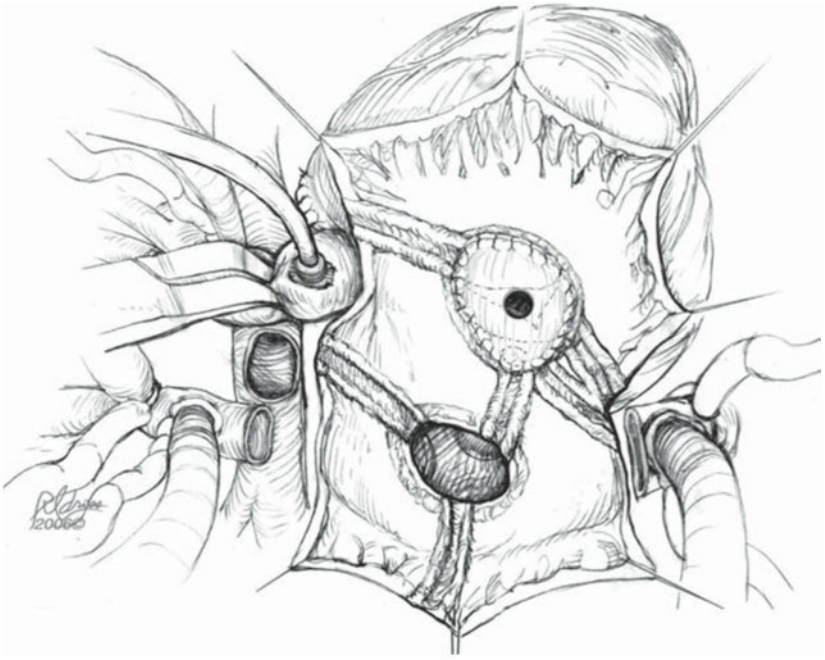
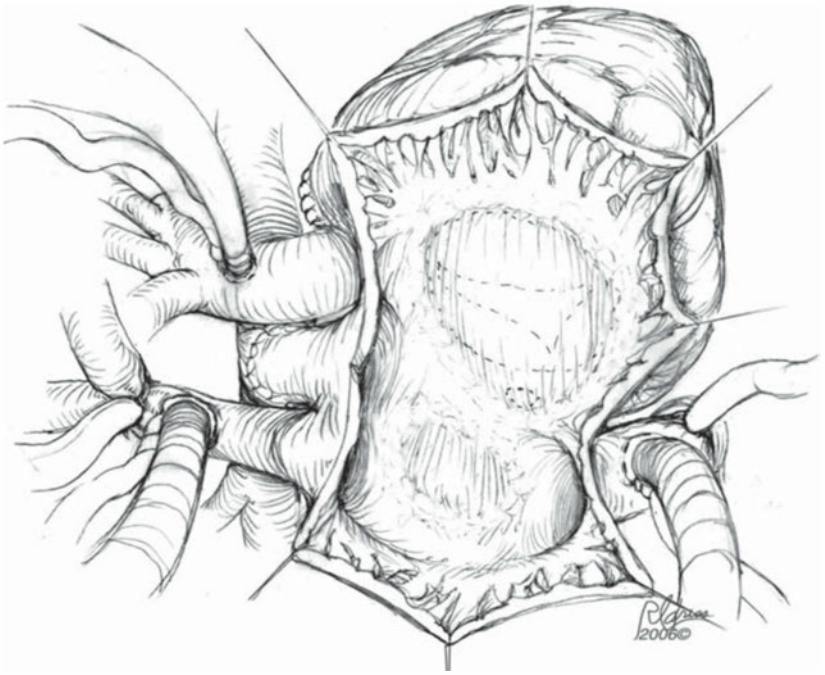
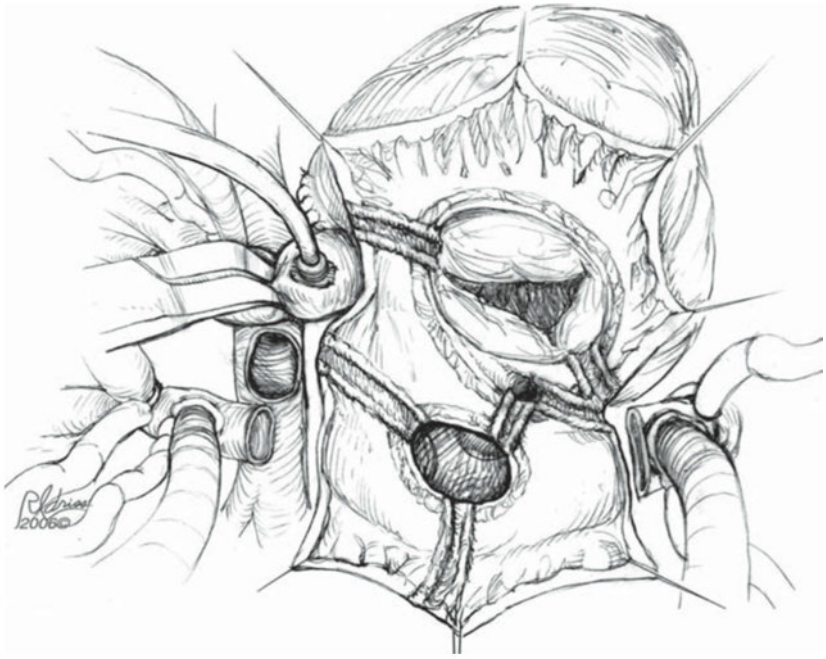


Fig. 105.12. Cryoablation lines noted for the modified right-sided maze procedure as well as the fenestrated tricuspid valve isolation patch. The idea of fenestration is to limit the inflow to the right ventricle in diastole, which will decrease the developed pressure of the right ventricle in systole. (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, et al. Evolving anatomic and electrophysiologic considerations associated with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2007;10:136-145.)

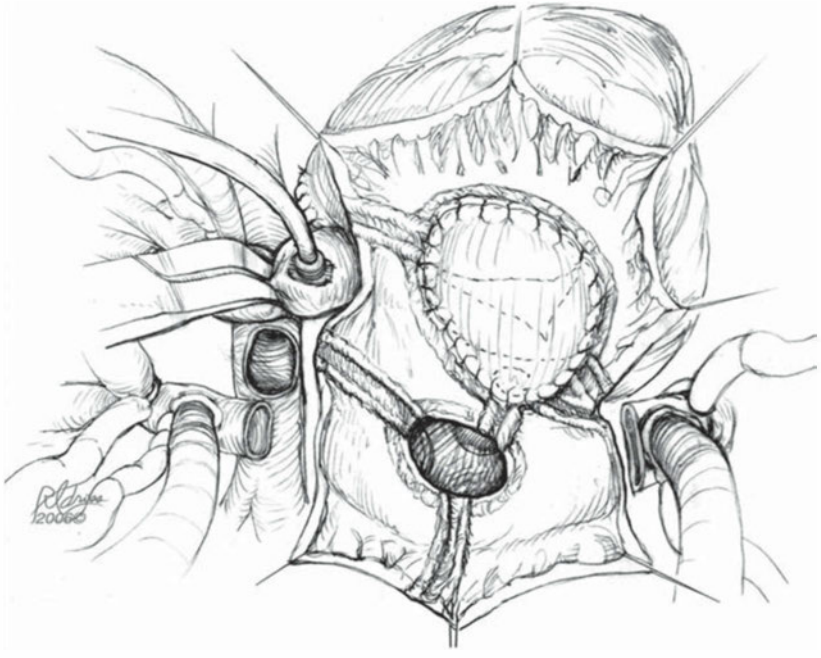


A

Fig. 105.13. (**A**) Right atrial view of a patient with double-inlet ventricle who had an atriopulmonary Fontan with tricuspid valve isolation and atrial septal defect closure. (**B**) Right atrial view of isolation patch removal and atrial septal defect creation. The cryoablation lesions are shown after proper identification of the tricuspid valve annulus and coronary sinus. (**C**) Right atrial view of polytetrafluoroethylene patch tricuspid valve isolation after cryoablation. (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, et al. Evolving anatomic and electrophysiologic considerations associated with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2007;10:136-145.)



B



C

Fig. 105.13. (*Continued*)

SURGICAL TRANSLOCATION OF ATRIAL SEPTAL ALIGNMENT

Some surgeons revised the atriopulmonary connection in an effort to create a wider atriopulmonary anastomosis by aligning the posterior pulmonary artery directly with a revised translocated atrial outflow. Anastomosing the confluence of pulmonary arteries to the dome of the right and left atrium was performed by resecting the superior atrial septum in a subset of patients. We have named this reconstruction the "dropped atrial septum technique." Separation of the circulations was accomplished by a synthetic septal patch that in effect dropped the atrial septum into the left atrial dome, thereby aligning the pulmonary artery with the newly formed systemic atrium (Fig.105.14). Important arrhythmia considerations include the potential for left ART based on the suture lines that were placed in the dome of the left atrium. Successful arrhythmia surgery will require left atrial suture line resection and biatrial maze procedure. Prevention of pulmonary vein stenosis may require atrial reconstruction with a Gore-Tex patch in the dome of the left atrium.

CONCLUSION

Arrhythmia surgery requires a collaborative approach between the surgical team and electrophysiologists, with careful preoperative evaluation to correctly characterize the arrhythmia substrate. Prior knowledge of specific surgical variations, familiarity with the principles of arrhythmia mechanisms, and understanding the tenets of surgical ablation are used to design and guide the appropriate therapy. These techniques have been particularly useful for the patients requiring Fontan conversion. Evolution in mapping systems, improved energy delivery technology, and prospective clinical studies can be expected to simplify the application of arrhythmia surgery to routine surgical interventions.

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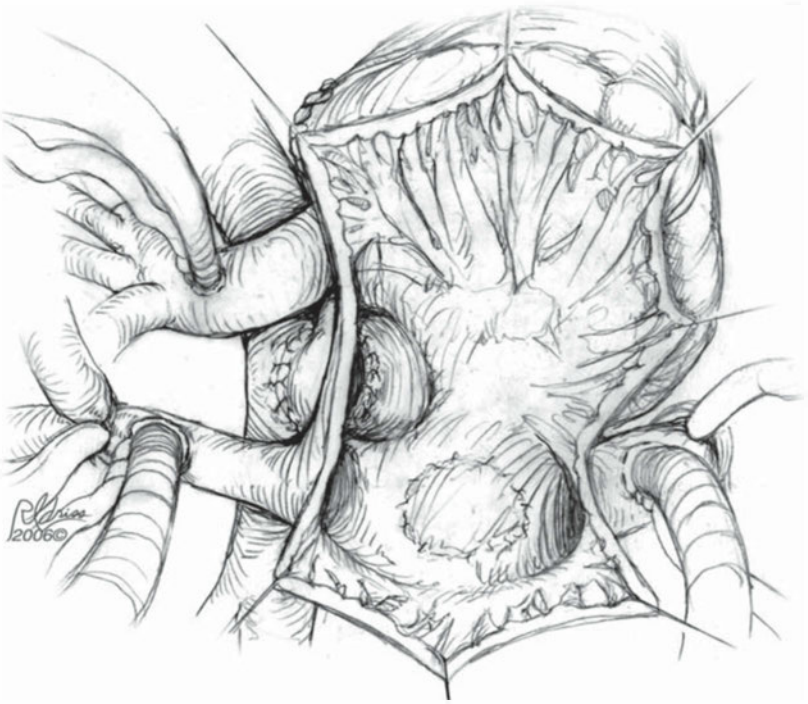


Fig. 105.14. Right atrial view of atriopulmonary Fontan performed by moving the atrial septum posteriorly, which realigned the outflow of the right atrium to conform to the more posterior main pulmonary artery. This "dropped atrial septum" required a suture line in the left atrium, which caused scarring and an area of slow conduction leading to atrial reentry tachycardia. In addition, when this anastomosis was taken down, a significant moiety was created which required attention lest pulmonary venous obstruction be created. (Reproduced with permission from Mavroudis C, Backer CL, Deal BJ, et al. Evolving anatomic and electrophysiologic considerations associated with Fontan conversion. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2007;10:136-145.)

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EDITOR'S COMMENTS

In this chapter Dr. Mavroudis and his associates have given an excellent outline of issues regarding arrhythmia surgery in patients with congenital heart disease. In the majority of cases of accessory AV pathways or AV nodal reentrant tachycardias, surgical intervention is not required, as most of these patients may be dealt with using ablation techniques in the cardiac catheterization laboratory. It is, however, imperative for surgeons to understand the anatomy related to these arrhythmias in certain cases where access is limited and surgical intervention may still be required on occasion. It should be noted that most of the ablation techniques being used today rely on earlier studies by surgeons who initially ablated these bypass tracts surgically and identified the basic anatomy and location, which could then subsequently be dealt with interventional transcatheter ablation techniques.

Dr. Mavroudis and his associates have clearly demonstrated that patients with the Fontan circulation who have inefficient venous pathways with areas of stasis and associated arrhythmias and ventricular dysfunction can be effectively treated with Fontan conversion and arrhythmia surgery with a marked improvement in exercise performance and clinical status. These results confirm the concept that the Fontan circulation should be made as hemodynamically efficient as possible, which lends support to the total cavopulmonary connection and extracardiac conduit Fontan modifications that are now the most popular techniques for Fontan completion. The single-ventricle Fontan physiology creates a situation of relatively low cardiac output, and therefore, anything that can be done to maximize the cardiac output and maintain ventricular performance should result in improved long-term outcomes.

It is unknown whether the newest modifications to the Fontan operation (such as the extracardiac conduit), which have the potential of decreasing the number of atrial suture lines, will in fact decrease the risk of late arrhythmias. The follow-up for the extracardiac conduit modification is not yet long enough on average to determine whether the incidence of late arrhythmias will be significantly decreased with this technique. There are encouraging early reports of a lower incidence of arrhythmias at 5 to 8 years of follow-up; however, because the majority of atrial arrhythmias do not occur until 10 years or later after the Fontan operation, the overall effect is still unknown. There may well be anatomic substrates for arrhythmias in the single-ventricle population that will make patients at risk for late arrhythmias regardless of the type of Fontan connection performed. In many cases, the need for atrial septectomy as a first-stage operation in association with other reconstructive procedures means that many patients will have atrial suture lines regardless

(*continued*)

EDITOR'S COMMENTS (*continued*)

of the type of completion Fontan. Many patients late after the Fontan operation will have relative diastolic dysfunction of the single ventricle. Diastolic dysfunction has been associated with atrial arrhythmias in normal hearts as patients age and this same phenomenon may lead to the incidence of arrhythmias, which has been seen even in patients with the extra cardiac conduit Fontan that is not associated with elevated atrial pressures. Thus, the technique of Fontan reconstruction may be less important in terms of late development of arrhythmias than the underlying substrate of the ventricular performance.

The majority of Fontan revision procedures that have been performed in most centers have converted an old-style atriopulmonary Fontan connection with very dilated atrial chambers and thick atrial walls to the more hemodynamic extracardiac Fontan. There will probably be an increasing number of lateral tunnel Fontan operations that develop enough dilation of the atrial pathway to require conversion in the future, although the incidence of this phenomenon is not yet clearly determined. It is likely, however, that an increasing number of patients will be referred for arrhythmia modification and conversion as the very large population of children and young adults with the Fontan circulation age. The majority of those Fontan patients who have had the procedure for hypoplastic left heart syndrome are now just entering their preteenage years. These patients may represent an increasing challenge for pediatric cardiac surgeons as they develop potential complications of Fontan physiology as they age into adulthood. Surprisingly, even after many years of follow-up, only a relatively small percentage of Fontan patients have required cardiac transplantation, as nicely demonstrated by Dr. Mavroudis and his colleagues. The primary goal with staged surgical reconstruction in single-ventricle physiology is to maintain ventricular function as much as possible to delay the need for cardiac replacement.

The increasing need for surgical arrhythmia ablation in adults with congenital heart disease has made it imperative that congenital heart surgeons learn these techniques. Our own lesser experience with ablative surgery for arrhythmias and Fontan conversion mirrors Dr. Mavroudis' results. Despite the complexity of the operations, which are often quite prolonged and difficult, the results have been gratifying. Most patients achieve a significant improvement in exercise performance and control of arrhythmias, which dramatically improves their quality of life.

TLS

106 ECMO and Ventricular Assist Devices

Andrew J. Lodge and Robert D. B. Jaquiss

INTRODUCTION

Mechanical circulatory support (MCS) in children is used for both profound respiratory and circulatory failure. Occasionally, these conditions coexist in the same patient. Extracorporeal membrane oxygenation (ECMO) has been relatively commonly used in neonates since the initial successful use in the mid-1970s. Indications include persistent pulmonary hypertension related to meconium aspiration, sepsis, idiopathic causes, and congenital diaphragmatic hernia. As results with this population improved, the use of ECMO was expanded to a variety of causes of respiratory and circulatory failure in other infants and children.

End-stage heart failure is a growing problem in the pediatric population in patients with primary cardiomyopathies and the increasing number who are surviving with repaired or palliated congenital heart defects. These patients are conventionally managed with a combination of oral and intravenous medications. When these fail, MCS is indicated. Particularly in small patients, ECMO has been historically used for this purpose. In adults, ventricular assist devices (VADs) have become the standard of care. There are a variety of devices that are approved by the U.S. Food and Drug Administration (FDA) for both short- and long-term support of these patients. Over the past several years, improvements in technology have now made VADs available to infants and children. These devices may be indicated as a bridge to recovery of the native heart function, or, more commonly in children, as a bridge to transplant. At the current time, there are devices in clinical use or being developed that will further improve the care and outcomes of these patients. This chapter will focus on the use of ECMO and VADs in children.

RESPIRATORY AND HEART FAILURE IN PEDIATRIC PATIENTS

Profound respiratory failure may occur in infants in children due to a variety of etiologies including those mentioned above as well as other infectious or inflammatory causes. In a minority of patients, conventional medical therapy including inhaled nitric oxide and alternative ventilation strategies may be inadequate to sustain adequate gas exchange. Severe respiratory failure in the neonate may precipitate pulmonary hypertension, shock, and circulatory collapse.

In contrast to adults, heart failure is relatively uncommon in pediatric patients. As in adults, the ultimate therapy for pediatric patients with end-stage heart failure is cardiac transplantation. Worldwide, 1,500 to 2,200 children are listed for heart transplant and each year 347 to 386 are transplanted. Congenital heart disease now surpasses primary cardiomyopathy as the most common indication for heart transplant in children. Since there are a growing number of children with successfully repaired or palliated congenital heart defects, one could anticipate that the number of listed patients and their waitlist times will increase. An important subset of this population is children with a diagnosis of hypoplastic left heart syndrome or related lesions. Results of conventional staged palliative surgery for these patients are improving, leading to a larger number of children with this diagnosis surviving. A number of these patients will experience failure of the single-ventricle circulation at various stages of palliation, including after the Fontan procedure. As a result, larger numbers of these patients will be evaluated for transplant. Compared with adults, the waitlist times for children are, on average, longer. Waitlist mortality is also higher in pediatric patients compared with adults with a threefold increase in the death rate on the waitlist that is essentially unchanged over the last decade. If medical therapy for advanced heart failure fails, some children will progress to the need for MCS. In the most recent era, the percentage of children supported with a VAD at the time of transplant was 13%, which has progressively increased over time.

RATIONALE AND INDICATIONS FOR THE USE OF EXTRACORPOREAL LIFE SUPPORT

Severe respiratory failure may become manifest by inadequate gas exchange despite maximal medical therapy, or excessive and toxic ventilator support being required to maintain such gas exchange. In such cases, ECMO is indicated to sustain life and allow time for lung recovery. Most recently, ECMO has been used successfully to bridge children with end-stage lung disease to lung transplantation.

End-stage heart failure may become manifest as refractory or unmanageable symptoms on conventional medications or, in babies, by poor growth, difficulty feeding, and failure to thrive. When more advanced, function of other organs such as the kidneys may become impaired. If these conditions do not improve with intravenous inotropes, MCS is indicated. In some children, end-stage heart failure is treated in part with tracheal intubation, sedation, and mechanical ventilation. This may reduce metabolic demand and oxygen consumption sufficiently to make the heart failure symptoms more manageable. However, remaining intubated and confined to bed has many risks. In adults, the use of VADs has allowed patients to improve from a metabolic, end-organ function, and nutritional standpoint. They can be extubated, become ambulatory, and even be discharged home as they await transplant. In many cases, this therapy has allowed patients who otherwise would have died to survive to transplant, and in others it has allowed them to become better transplant candidates. These same benefits can now be realized in children.

MCS may be indicated to treat postcardiotomy failure, acute cardiomyopathy, chronic cardiomyopathy, or failed corrective or palliative surgery for congenital heart disease. The indication, as well as

the intention to use MCS as a bridge to recovery, a bridge to transplant, or permanent (destination) therapy, has implications for the type of support chosen, the technical details of implementation, and the expected results and risks. Importantly, whenever the use of ECMO or VAD support is contemplated, careful thought should go into the eventual outcome. If a reversible cause of the underlying problem is unlikely, or if the patient is not a candidate for heart or lung transplant, MCS may be contraindicated.

EXTRACORPOREAL MEMBRANE OXYGENATION

ECMO systems include a blood pump, an artificial lung (oxygenator), tubing, and monitoring equipment (Fig. 106.1). ECMO support is generally established by inserting relatively large-bore arterial and venous cannulae in peripheral vessels. ECMO may be venovenous (VV), in which case drainage and return of blood are both from the venous side of the circulation, or venoarterial (VA), in which case blood is withdrawn from the venous side and returned to the arterial circulation. VV ECMO is indicated for purely respiratory support and may be sufficient if cardiac function is relatively preserved and the patient is not in profound shock or on excessive inotropic or vasopressor support. In most cases, a specially designed double-lumen cannula, similar to a dialysis catheter, is used and placed through the right internal jugular vein to the right atrium (Fig. 106.2). An alternative is drainage from one cannula in the femoral vein and return via a different cannula in the internal jugular vein.

In infants and smaller children, the right common carotid artery and internal jugular vein have traditionally been used as these vessels are usually easily accessible and amenable to rapid exposure. These are typically accessed by a transverse skin incision on the right side of the neck approximately one to two fingerbreadths above the clavicle depending on the size of the child. The subcutaneous tissue and platysma are incised with the cautery. The medial border of the sternocleidomastoid muscle is mobilized. The right common carotid artery and internal jugular vein are isolated. Meticulous hemostasis is maintained since the patient will be anticoagulated. At this point, we administer 50 U/kg of intravenous heparin and allow 2 to 3 minutes for circulation. If VA ECMO is used, the carotid artery is cannulated first. Care is taken to avoid the vagus nerve, which is adjacent to the artery. A silk ligature is used to ligate the more cephalad portion of the exposed vessel. The ligature is clipped to the drape such that the vessel is placed on some traction. A loose ligature is placed around the artery proximally, and a fine vascular or bulldog clamp is used to temporarily occlude the vessel. Using a No. 11 blade, a transverse arteriotomy is made to open the artery. The appropriate-sized cannula, usually 8F or 10F for a neonate or small infant, is inserted to a depth of 2 to 3 cm and secured by tying the proximal ligature around the artery and cannula. We tie the ligature over a small piece of vessel loop to prevent damage to the vessel wall at decannulation. The more distal ligature is then tied around the cannula for security. The right internal jugular vein is cannulated in similar manner. Occasionally, a branch such as the facial vein requires ligation and division. The venous cannula, which has more side holes, is usually larger than the arterial by one or two sizes and is inserted to a greater depth such that the tip is well into the right atrium (Fig. 106.3). Both are then secured to the skin with two additional sutures each. The incision is then closed around the cannulae either with a simple running monofilament suture or with a layer of absorbable suture to reapproximate the platysma layer followed by the skin closure. Ideally, a small skin bridge

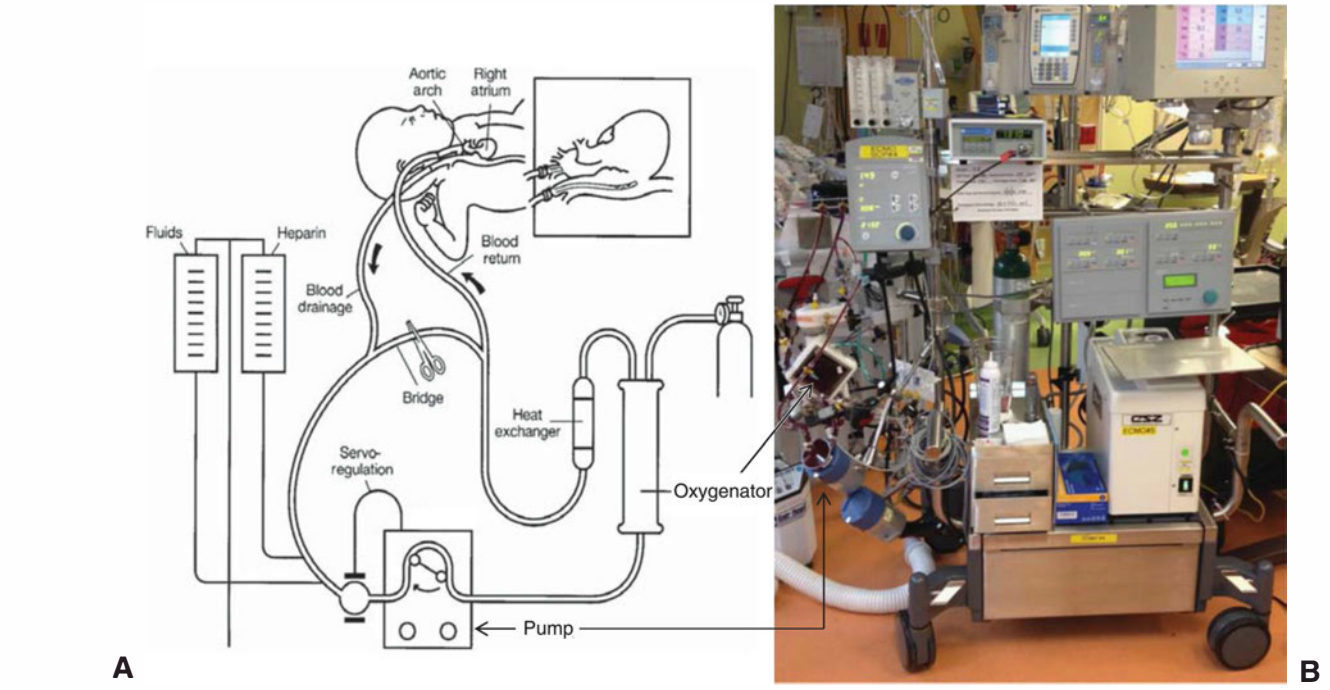


Fig. 106.1. (**A**) Schematic diagram of an extracorporeal membrane oxygenation (ECMO) circuit is shown, demonstrating the various components. The inset shows the position of the cannulae inserted via the cervical vessels. (**B**) A photograph of an ECMO circuit in clinical use. (http://www.nichd.nih.gov/publications/pubs/images/Efig1p26.gif.)

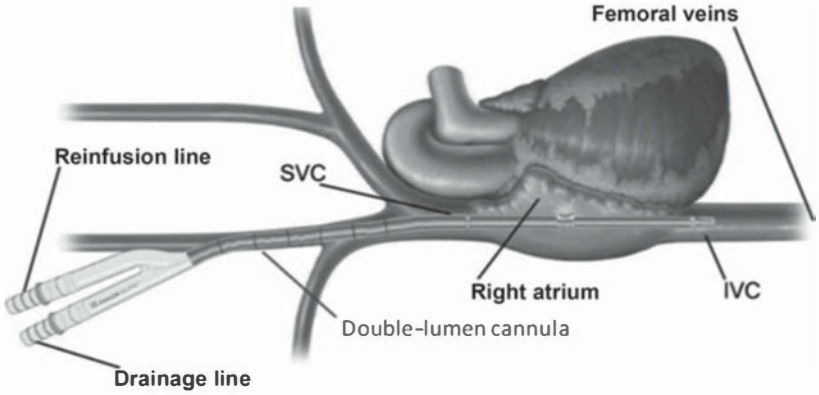


Fig. 106.2. A double-lumen extracorporeal membrane oxygenation (ECMO) cannula and its proper positioning from a right internal jugular vein approach. IVC, inferior vena cava; SVC, superior vena cava. (Avalon Laboratories, LLC, Rancho Dominguez, CA.)

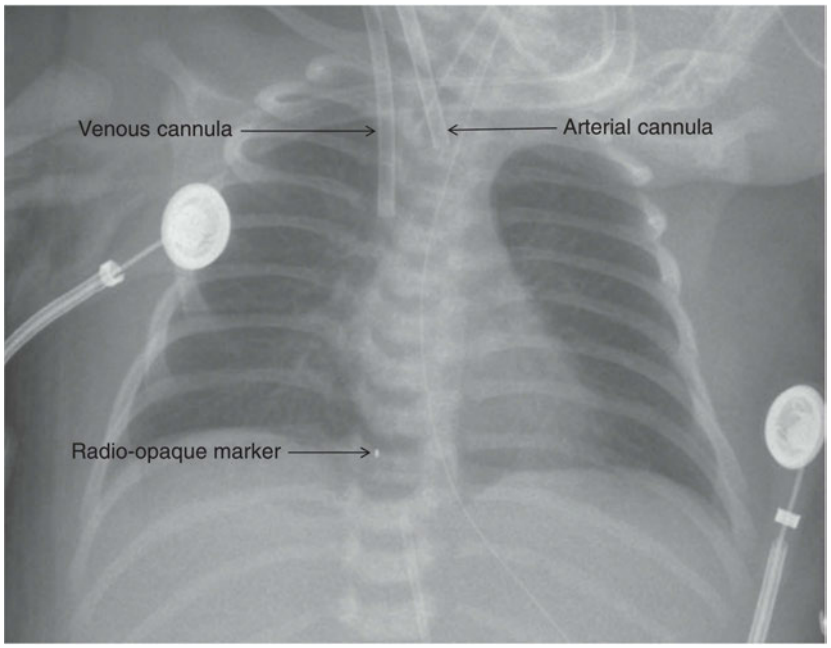


Fig. 106.3. Chest X-ray demonstrating appropriately positioned cannulae for venoarterial (VA) extracorporeal membrane oxygenation (ECMO). There is a radio-opaque marker at the tip of the venous cannula to mark its position.

is left between the arterial and venous cannulae.

Depending on the center and clinical circumstances, when the child is weaned from ECMO support and the cannulae are removed, the vessels may remain permanently ligated or be repaired. It is unclear whether one approach is superior to the other. If the vessels are repaired, the ligatures must be removed carefully while maintaining proximal and distal control with a combination of vessel loops or fine vascular clamps. A small amount of back-bleeding is allowed to assess vessel patency and expel any debris or thrombus. The vessels are then repaired with fine, typically 7-0, monofilament sutures. Either a running or interrupted technique can be used.

In older children, there may be reluctance to ligate the carotid artery, although this has been done successfully in several centers. Other options for cannulation include the femoral, iliac, and axillary vessels. Like the cervical vessels, the femoral artery and vein are relatively superficial and amenable to reasonably rapid exposure by cutdown. Alternatively, if the child is large enough, with currently available cannulae, these vessels can be accessed percutaneously. The lower extremities are more susceptible to ischemic complications related to cannulation as collateral circulation is not as robust as in the brain. This can be treated with the placement of a more distal arterial catheter, which is connected to the arterial limb of the ECMO circuit. In small children, it may be difficult to get cannulae of adequate size to achieve full flow in the femoral vessels. In these cases, the external iliac vessels may be used. These vessels are accessed by performing an extraperitoneal dissection via an incision above the inguinal ligament. It is generally not safe to access these vessels percutaneously. If the axillary vessels are used, which is unusual, they can be accessed via an infraclavicular incision. The brachial plexus is at risk of injury from this exposure. Most commonly, this incision is used for arterial access only. A vascular graft is usually sewn end-to-side to the artery and the cannula is placed in the graft rather than cannulating the artery directly. The femoral, iliac, and axillary vessels are not permanently ligated after discontinuing ECMO.

PREPARATION FOR VENTRICULAR ASSIST DEVICE IMPLANTATION

The placement of any VAD requires meticulous preparation. The appropriate device should be selected based on the patient size and indication as discussed above. The patient should be free from any active infection. Neurologic status should be carefully assessed prior to surgery as an acute intracranial ischemic or hemorrhagic process represents a contraindication. The presence of any intracardiac shunting should be excluded by echocardiogram. If a shunt such as a PFO is present, it should be closed at the time of VAD implantation. The patient should be rendered as metabolically stable as possible, although not infrequently conditions such as metabolic acidosis and renal insufficiency represent part of the clinical syndrome that makes MCS necessary and will improve after successful VAD placement. Any preexisting coagulopathy should be reversed to the extent possible prior to surgery.

TEMPORARY VENTRICULAR ASSIST DEVICES

Although ECMO can be used for cardiovascular support and has some advantages such as the potential for rapid institution (which is useful in the setting of cardiopulmonary resuscitation) almost anywhere in the hospital and its combination of

cardiac and respiratory support, these are balanced against many negative features that make it suboptimal as a durable form of circulatory support. These include the relatively unstable cannulation, which has traditionally required the patient to be fairly immobilized, nonambulatory, and generally confined to a mechanical ventilation, the relatively large artificial blood contacting surface that results in hemolysis and platelet destruction, ongoing transfusion requirements and coagulopathy, and potentially inadequate decompression of the left ventricle leading to ongoing left atrial hypertension, pulmonary edema, and in the worst cases, pulmonary hemorrhage. In addition, inadequate unloading can impair any potential recovery of the compromised ventricle. In these situations, additional steps must be taken to decompress the left side of the circulation. This can be accomplished in the cardiac catheterization lab with atrial septostomy or septoplasty, or by the surgical placement of a left atrial or ventricular vent that is connected to the venous limb of the ECMO circuit. All of these factors make ECMO suitable for short-term support, but very suboptimal for longer term use.

Better temporary support options may include centrifugal pump-based VAD systems. These systems, which employ a centrifugal blood pump, tubing, and cannulae similar to those used for cardiopulmonary bypass (CPB), are designed for short-term use, can be customized to provide single or biventricular support, can be adapted to fit a variety of different sized patients, and are relatively inexpensive. Temporary cannulation is employed, which is not typically stable enough to allow much patient movement. Frequently, these types of systems are placed with only temporary sternal closure in the postcardiotomy support setting, which limits the ability to mobilize the patient. Until recently, pediatric-specific pumps were not available, leading to potential problems with lower pump speeds predisposing to thrombosis and thromboembolism in this group. Recently, the Levotronix® Corporation (Waltham, MA) developed a smaller version of their magnetically levitated centrifugal blood pump, the CentriMag®, called the PediMag® that obtained 510k clearance from the FDA in 2009 for short-term use as a pediatric blood pump (Fig. 106.4). This pump has a priming volume of only 14 ml and 1/4 inch connections, and a maximum flow of 1.5 l/min making it potentially useful as a temporary VAD for infants and small children. The PediMag® has been successfully used as a bridge to transplant in an infant. Its use may increase as further experience with it is gained.



Fig. 106.4. A CentriMag® console and pump and PediMag® pump are shown. The PediMag® is similar in design to the CentriMag and uses the same console but is smaller and has ¼ inch connections.

Cannulation for LVAD support is usually accomplished with direct aortic cannulation for outflow and cannulation of either the left atrium or the left ventricular (LV) apex for inflow. Apical cannulation provides more complete ventricular decompression, whereas atrial cannulation is more easily accomplished but may risk ventricular thrombus formation due to stagnant flow. RVAD support is generally accomplished by cannulation of the right atrium for inflow and the pulmonary artery for outflow. All cannulae should be secured with two concentric purse-string sutures, which are securely tied to the cannulae themselves to maximize hemostasis and immobilization. Cannulae may be selected that can be tunneled through the body wall like a chest tube to permit chest closure during the period of support. When these devices are used for postcardiotomy support with anticipated ventricular recovery, weaning at the bedside can be accomplished reasonably easily, and the temporary nature of the cannula placement facilitates device removal if sufficient recovery occurs.

PARACORPOREAL DEVICES

The most commonly used VADs in children today are so-called first-generation paracorporeal devices. These are displacement pumps that function much like the native ventricle with inflow and outflow valves and a pumping chamber that is pneumatically driven. These devices can be used for single or biventricular support, but employ cannulation that is stable and allows for patient extubation, mobilization, and rehabilitation. In older children, adult devices such as the Thoratec paracorporeal VAD (PVAD; Thoratec Corporation, Pleasanton, CA) have been used. Unfortunately, the blood pump and cannulae for the device are large, which leads to potential problems with thromboembolism, excessive stroke volume, low pump flow rates, and difficulty with cannula positioning. From a practical standpoint, these systems cannot be used in children under 20 kg, and even in that size range, the use of the pump is problematic.

The most important current manufacturer involved in pediatric VAD development is Berlin Heart GmbH (Berlin, Germany). In 1989, they modified an adult device in order to allow VAD placement in small children (Fig. 106.5). The Berlin Heart EXCOR® Pediatric VAD is produced in a variety of sizes, with an applicable size range from 3 kg to adult size. For approximately a decade, the use of the device was confined to Western Europe. After the first usage of the device in the United States was reported in 2005, its use spread to multiple centers in North America, all under so-called compassionate use protocols. The positive experience at implanting centers led to the initiation in 2007 of a large multicenter trial, constructed to potentially permit formal FDA approval of the device. This trial has now been completed and has resulted in the recent approval of the device in the United States.

Implantation of all the paracorporeal devices is similar. For LVAD support, apical cannulation for device inflow is preferred as discussed above. Using normothermic or mildly hypothermic CPB, the pericardium is opened and the apex of the heart is identified. A skin exit site is selected for the inflow cannula in the left upper quadrant below the costal margin on a line corresponding to the location of the apex. A disc of skin slightly smaller than the diameter of



Fig. 106.5. A 7-year-old patient with a Berlin Heart EXCOR® pediatric left ventricular assist device (LVAD). The pump is connected to the heart by transcutaneous cannulae (inflow more lateral, outflow more medial) and to the system controller via a pneumatic driveline (connection just visible at far left).

the cannula is excised, and an extraperitoneal tunnel is created into the pericardium. Incision of the pericardium out toward the apex and sometimes partial takedown of the anterior diaphragmatic attachments facilitates this maneuver. Care must be taken to maintain hemostasis. The tunnel can be maintained by passing a chest tube or umbilical tape through it. The apex of the heart is mobilized and elevated into the field by placing sponges in the pericardium behind the heart. We prefer CO_2 insufflation of the operative field to assist with deairing of the heart. Particularly in the reoperative setting, it is important to establish hemostasis in the pericardial space prior to positioning of the apical cannula as this space will be inaccessible once the device is positioned. Depending on surgeon preference, an LV vent may or may not be used. Apical cannula placement can be accomplished with the heart beating, or with fibrillatory arrest. Once the heart is positioned, a core of apical LV muscle is removed at the intended site of cannula placement. In general, this should be at or just lateral to the apical dimple with enough of a margin between the ventriculotomy and left anterior descending coronary artery to permit suture placement. The ventricle should be inspected for thrombus, which should be removed if present. Any trabeculae that could interfere with inflow are excised. If an LV vent is not present, a cardiotomy suction catheter can be placed through the ventriculotomy. A series of 8 to 12 pledget-supported horizontal mattress sutures is then placed around the ventriculotomy. Relatively wide bites are taken through the ventricular muscle into the ventricle. If preferred, the suture may then be brought out through the epicardium near the edge of the ventriculotomy (Fig. 106.6). Once all of the sutures have been placed, they are brought out through the flange on the inflow cannula. The sutures should be evenly spaced but should not have gaps between them. If the inflow cannula has a bevel, it should be positioned such that the bevel faces the septum. The cannula is placed into the LV apex until the flange is flush with the epicardium. The cannula should fit snuggly into the ventriculotomy to optimize hemostasis. The sutures are tied. Depending on surgeon preference, a surgical sealant may be applied to the suture line but if used it is important to exclude this from the blood that is returned to the cardiotomy reservoir. It is our preference to then place a polytetrafluoroethylene (PTFE) membrane with a hole cut in the middle over the apical cannula such that the membrane covers the apex to prevent severe adherence to the pericardium and chest wall. We have found this helpful when explanting the device. The apical cannula is then passed through the previously created tunnel. Care must be taken to maintain its orientation and support the connection to the ventricle such that it is not disrupted. After passage through the tunnel, the apical cannula can be vented to the cardiotomy reservoir. The flange of the cannula should be flush with the diaphragm.

Attention is then turned to the placement of the outflow cannula. An exit site for this is selected which is typically more medial, often just to the right of the midline. The tunnel is created as described above. The cannula may be tunneled prior to connection to the aorta. This may be preferable in the case of the somewhat rigid Berlin Heart cannulae to avoid tension on the aortic connection with tunneling. A site for the outflow cannula placement on the aorta is selected. This is typically anterior or slightly offset rightward on the greater curve of the ascending aorta. It must be above the sinotubular junction but should also leave room distally for later aortic cannulation and transection for eventual heart transplant. The smaller Berlin Heart outflow cannulae have a tip beyond the sewing ring that is inserted into the lumen of the aorta when the sewing ring is approximated to the adventitia of the aorta. An aortotomy is made after the

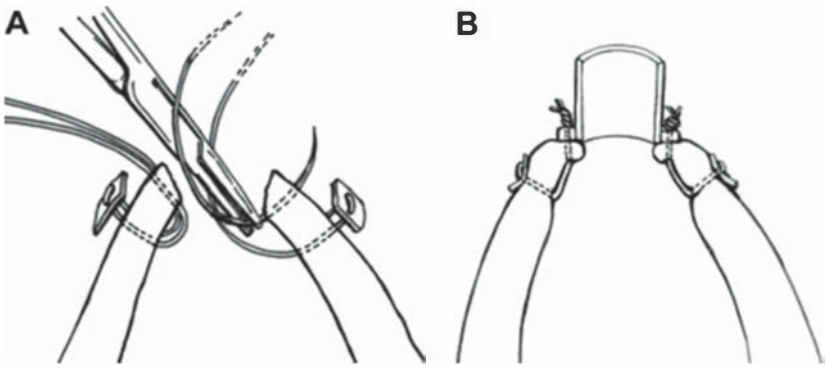


Fig. 106.6. Diagram showing technique for suture placement around the apical ventriculotomy used for implantation of the apical cannula. (**A**) Suture placement. (**B**) Sutures are tied.

application of a partial occlusion clamp to the aorta to isolate the selected site. If the aorta is difficult, cross-clamping with cardioplegic arrest can be performed, but this is avoided if possible, especially in the case of LVAD support alone as ischemia to the right ventricle is undesirable. A longitudinal incision is made in the aorta sufficient to match the size of the outflow cannula. In smaller children, it may be possible to secure the cannula with two concentric purse-string sutures, the ends of which are brought up through the sewing ring of the cannula. These are placed prior to the application of the partial clamp. The aortotomy is then made within the purse-strings after the clamp is applied and the cannula tip is introduced into the aorta as the purse-string sutures are tightened. Care must be taken to make the purse-strings large enough that the aortotomy can be made with an adequate margin of tissue around it. The sutures are secured and the cannula is deaired. In larger patients or if this technique is not selected, or in the case of a larger cannula, a series of interrupted polypropylene mattress sutures with or without pledgets is placed around the aortotomy and brought up through the sewing ring. The cannula is then lowered down to the aorta, the sutures are tied, and the cannula is deaired and clamped. Hemostasis is confirmed at the anastomosis. Some surgeons may prefer a running suture technique, but this is technically more difficult (Fig. 106.7). The Thoratec PVAD has an outflow cannula with a flexible vascular graft designed to be sewn to the aorta. In this case, the graft must be properly sized prior to anastomosis to the aorta, anticipating some lengthening when it is pressurized. A running suture is more suitable to this type of cannula.

Preparations are then made for connection of the pump. The pump should be previously primed on the back table by the perfusionists or nursing staff. The pump is connected to the cannulae after appropriate trimming, if necessary, of the cannulae to prevent kinking. Great care is taken to avoid the introduction of air during this process. Attention is then turned to deairing of the heart and pump. The Berlin Heart pump has a deairing nipple, which is accessed prior to connection of the pump when the pump is fully primed. In pumps with outflow cannulae that have vascular graft material, deairing can be facilitated with the placement of a needle in the graft. If left ventricular venting has been ongoing, it is discontinued at this time. The patient is placed in the Trendelenburg position and the lungs are ventilated. The heart is gently agitated as the heart is slowly filled. Transesophageal echocardiography is helpful in monitoring the amount of air in the heart. Once the pump has been fully deaired, the VAD is activated with single strokes from the controller or hand pump, depending on the device. The pump is observed for air. If air is not present the patient is weaned from CPB, then the VAD pumping is initiated at a conservative rate. Activating the VAD prior to weaning from CPB can lead to the entrainment of air in the heart. If LVAD support only is used, adequate support of native right ventricular function must be provided. We have found that a combination of epinephrine, milrinone, vasopressin, and inhaled nitric oxide to be useful. The pump, echocardiogram, and patient hemodynamics are observed and the VAD pumping rate is adjusted as necessary. Typically, the pump rate, positive (ejection) pressure, and negative (filling) pressure can be adjusted. The pump ejection pressure will typically need to be at least 80 to 100 mmHg higher than the patient's systolic blood pressure. Only enough negative pressure to achieve adequate pump filling is used. Excessive pressures can contribute to hemolysis.

If biventricular support is required, another pump can be used as a right ventricular assist device (RVAD). The RVAD is typically placed with inflow from the right atrium and outflow to the main pulmonary artery. The right ventricle is more problematic to cannulate due to its geometry and highly trabeculated nature. Occasionally, if it is thought that RVAD support will only be required for a short time, a temporary centrifugal pump may be employed as described above. The use of such a pump facilitates RVAD removal and may simplify management. If longer term RVAD support is required, then a second Berlin Heart or similar pump is indicated. The outflow cannula is secured to the main pulmonary artery beyond the pulmonary valve in a manner similar to that described above for the aorta. The Berlin Heart atrial cannula is inserted through the free wall of the right atrium and secured either with purse-string sutures as described above for the aorta, or mattress pledget-supported sutures as described for LV apical cannulation or a combination of these techniques. In general, the RVAD flow should not exceed the LVAD flow. It is generally slightly lower to avoid pulmonary congestion. Importantly, if biventricular support is required, care must be taken to maintain adequate

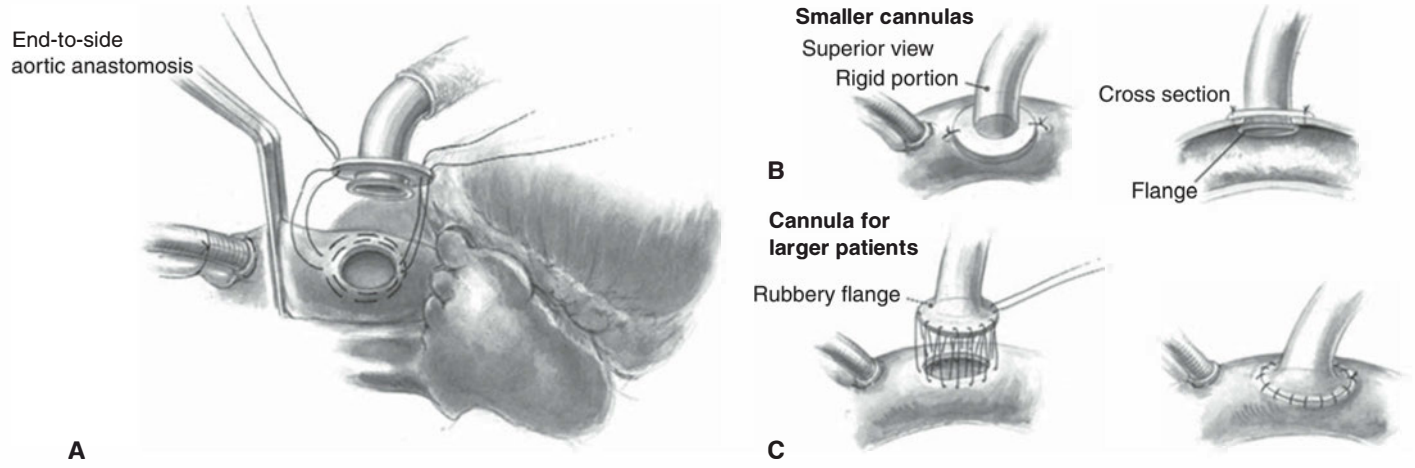


Fig. 106.7. Technique for the connection of the Berlin Heart outflow cannula to the aorta. (**A**) In this depiction, aortic cross-clamping and the two concentric purse-string suture technique is used. (**B**) The final position of the cannula. (**C**) Technique for anastomosis of the larger cannula that does not have the intraluminal extension, in this case, using a running suture.

skin bridges between all cannula exit sites to avoid necrosis and infection.

Bleeding may be an issue at the conclusion of these cases due to the degree of illness and preoperative coagulopathy in these patients. Aggressive early management of this coagulopathy helps to establish adequate hemostasis. Sometimes packing of the chest with temporary chest closure is useful. Adequate drainage to avoid pericardial tamponade, including a posterior pericardial drain, is useful. We have routinely tacked a PTFE membrane to the pericardial edges to prevent adhesions to the sternum and facilitate redo sternotomy.

IMPLANTABLE DEVICES

Further device development has led to the emergence of implantable VADs. These pumps are placed within the body and are connected to an external controller via a transcutaneous electrical driveline. The current versions are the second-generation axial flow pumps and the third-generation centrifugal pumps. Both types produce continuous rather than pulsatile flow. These devices are designed as LVADs so are generally not capable of biventricular support. Some reports of the use of these adult devices in children have appeared with mixed results. These include the VentrAssist (Ventracor Limited, Chatswood, NSW, Australia), a centrifugal LVAD (which is no longer available), and the Thoratec HeartMate® II, an axial flow device (Fig. 106.8). Because of the size of the implantable hardware, these devices can only be used in children with a body surface area of at least 1.2 to 1.4 m^2.

Most recently another adult device, the HVAD (HeartWare, Inc., Miami Lakes, FL), that has relatively recently been approved for use as a bridge to transplant in adults in the United States, has been successfully used in children. This device is an implantable centrifugal pump LVAD (Fig. 106.9). It is the smallest of the adult-implantable devices studied so far, and therefore may be more ideally suited for children, including those smaller than mentioned above. A German group has reported the successful use of this device to bridge children to transplant. It has also been used several times in the United States for this purpose, including a 10-year-old boy with dilated cardiomyopathy in our institution.

Although implantable devices are now commonly used in adults, since the pediatric market is substantially smaller and the process of developing such a device and bringing it to market is lengthy, cumbersome, and expensive, until recently there has been relatively little motivation for the development of such devices specifically for children. Several companies have now engaged in this challenge, however. An important example is Micromed Cardiovascular, Inc. (Houston, TX). This company produced the first VAD, and the only implantable device, approved for use in children in the United States. This device, the HeartAssist 5® (formerly called the DeBakeyVAD Child), received FDA approval as a humanitarian device exemption in early 2004. The approval was granted for patients between 0.7 and 1.5 m^2, between ages 5 and 16. The device is an axial flow pump similar to the HeartMate® II and designed for LVAD usage only. After the initial U.S. implant in 2004, the device has not gained widespread acceptance predominantly because of thrombotic concerns, and very few additional reports of its usage have appeared in the literature.

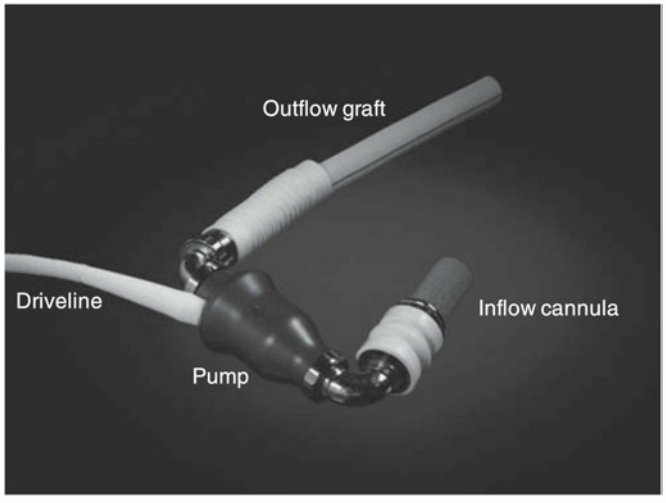


Fig. 106.8. Photograph of a HeartMate® II left ventricular assist device. The components of the device are labeled.



Fig. 106.9. The HeartWare HVAD pump. Its small size facilitates placement in the pericardium rather than a preperitoneal pocket that is used for other implantable devices.

Implantation of all of these LVADs is similar. The primary differences from paracorporeal device placement are that the pumps reside within the body and the electrical driveline is tunneled transcutaneously. For all but the HVAD, a preperitoneal pocket is created for the pump by dividing the anterior attachments of the left hemidiaphragm from the chest wall and dissecting posterior to the abdominal fascia. This can be done before initiating CPB. The driveline is tunneled from a preselected exit site in the right upper quadrant to this space. The electrical connection for the driveline is typically larger in diameter than the driveline itself, so we generally narrow the exit site with sutures after the driveline is passed and hemostasis is confirmed. For each device, there is an apical connector that is secured to the LV apex in much the same manner as described above for the paracorporeal VAD. The devices typically come with a coring tool that removes a round core of apical muscle. For the HVAD, the apical sewing ring is secured prior to coring. The pump is primed on the back table, and then connected to the apical attachment according to the manufacturers' guidelines. The outflow graft is then connected to the aorta and to the pump. These outflow grafts comprise a vascular graft connected to an adapter that connects to the pump. An end-to-side anastomosis is made with a running polypropylene suture, again using a partial occlusion clamp just to the right of anterior on the proximal ascending aorta. The graft will stretch once pressurized so

it must be trimmed to the appropriate length prior to sewing the anastomosis. The proximal portion of the graft is protected with a semi-rigid outer sheath to prevent kinking. Deairing of the pump is facilitated by placing a needle in the outflow graft and then partially occluding the graft distal to the needle. The patient is weaned from CPB as described above with gradual increase of the pump speed.

PEDIATRIC SPECIFIC ISSUES

Despite the early stages of development of pediatric VADs, and, in fact, the failure of some to make it through the early clinical use phase successfully, it is still clear that these devices represent an improvement in support compared with ECMO. This observation is borne out in a study by Jeewa et al. in which these two forms of support were compared. Patients supported with VADs had improved waitlist mortality and hospital survival.

Perhaps the most vexing problem in the field of MCS, shared in all generations of devices, and patients of all ages and sizes, is the issue of arterial thromboembolism. This is especially true with events involving the central nervous system. Although such events may be reversible or only result in minor disability, in other circumstances the events may be devastating or even fatal. Despite multiple advances in VAD design, the problem persists to such an extent that all devices that might be used in children require intense anti-thrombotic therapy. This typically includes both anticoagulation with heparin or warfarin and anti-platelet medications. As an example, a protocol has been developed for use with the Berlin Heart and is employed by essentially all implant centers in some form. The protocol involves the introduction of unfractionated heparin within the first day after implantation, in the absence of ongoing bleeding. This is later converted to either low molecular weight heparin or a vitamin K antagonist and combined with anti-platelet medications, most commonly aspirin and dipyridamole. Despite the use of such strategies, strokes have been described in most published experienced, with rates typically in the range of 25% to 40%. Bleeding is also an issue and the transfusion requirement consequent in major bleeding is especially problematic for pretransplant VAD patients in that exposure to foreign HLA antigens increases the likelihood of the development of anti-HLA antibodies.

The most important current issue in pediatric VAD practice is the lack of an approved device with an acceptably low-risk profile and the versatility to provide durable support for the entire range of size and anatomic complexity. Although the Berlin Heart device has been used successfully in a wide range of patient sizes, there remain concerns about its risk profile and patients generally remain confined to the hospital after implantation.

Unique to pediatric VAD candidates is the issue of anatomic complexity related to congenital structural heart disease. This may be manifested in abnormalities of cardiac situs, in the case of dextrocardia or dextroversion of the cardiac apex. In other cases, the systemic ventricle may be a morphologic right ventricle, as in the case of a patient who has previously undergone an atrial switch operation for transposition of the great arteries, and this may require an alteration in VAD cannula shape or orientation (or both). Perhaps the most difficult patients are those with univentricular hearts in whom the power source for pulmonary blood flow is either the failing ventricle, or systemic venous pressure (either a superior cavopulmonary connection or total cavopulmonary connection). Conventional VAD configurations do not account for the variations in pulmonary blood flow in children with single-ventricle hearts, and consequently the experience with VAD support in these patients is very limited. A recent literature survey found only 10 single-ventricle patients implanted with VADs. Of the 10 patients reported, 6 went on to receive cardiac transplantation.

FUTURE DIRECTIONS

Initially, the expansion of the use of the Berlin Heart may result in slightly less favorable results as inexperienced centers learn to use the device. In the longer term, the overall outcomes should be improved, in part by reducing waiting list mortality. It may be speculated that VAD-supported patients might even become better transplant candidates with the opportunity for improved nutrition and rehabilitation. This phenomenon has been observed in adults. Furthermore, the urgency to accept a marginally suitable heart for a rapidly declining recipient may vanish, allowing optimization of patient–recipient matching.

Also on the near horizon is the initiation of the second phase of the National Heart Lung and Blood Institute (NHLBI)-sponsored trial, termed the Pumps for Kids, Infants, and Neonates (PumpKIN) Trial. For this trial, the NHLBI has awarded four contracts totaling over $23 million to begin preclinical testing of pediatric circulatory support devices. At time of this writing, a Data and Clinical Coordinating Center (DCCC) is being identified for the trial. After the DCCC is identified, it is anticipated that clinical trials of the two VAD devices in the trial will begin, possibly by 2013. The devices to be evaluated in the PumpKIN trial include two ECMO devices that will not be discussed further, and one VAD (Fig. 106.10) the Jarvik 2000 [Jarvik Heart, Inc., New York]). The VAD is an implantable axial flow device, designed

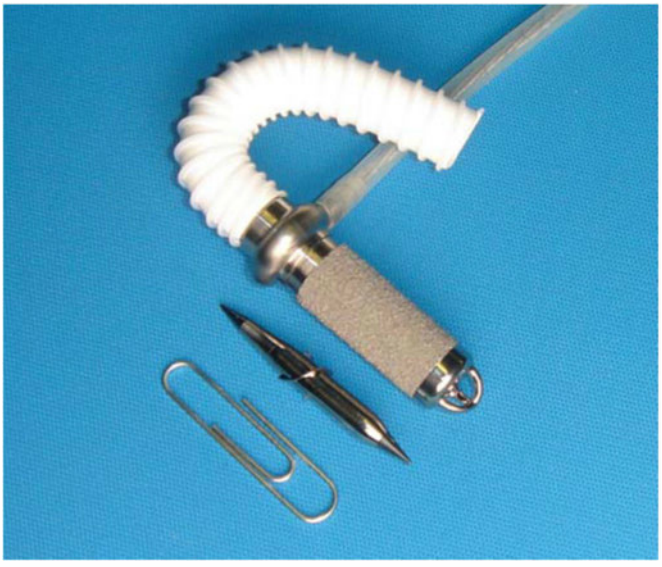


Fig. 106.10. The infant Jarvik pump. As can be seen in the photograph, this device is substantially smaller than the other implantable devices discussed, allowing for potential implantation even in small infants. As with other implantable pumps, there is a transcutaneous electrical driveline.

for LV apical and aortic cannulation to allow support of the left ventricle, which follows the trend in adult VAD practice, whereby the initial pulsatile devices have now been supplanted by continuous flow pumps. Flow rates from the device would permit complete systemic ventricular support for children in the 3 to 20 kg range. However, it is capable of biventricular support. Furthermore, the degree of anticoagulation required for the device is as yet undefined, as is the attendant stroke risk. Because the Berlin Heart has only been recently approved, and because the PumpKIN device study design has not yet been finalized, study sample size for device evaluation and comparison and the effect of the approval of the Berlin Heart device on the pace of enrollment of subjects in the VAD arms of the PumpKIN trial are undefined. In the Berlin Heart IDE trial, a single-arm study in 15 centers required 3 years to enroll a total of 24 patients in the size range of patients who will be enrolled in the PumpKIN trial. It seems very likely that the PumpKIN trial will require at least as long.

Beyond the Berlin Heart and PumpKIN devices, as durable VADs become better, the concept of destination therapy in children or adolescents is on the horizon. In children who have heart failure but are so highly sensitized as to not be candidates for transplantation, such a concept may be worth considering. Another population that comes to mind is that with a failing Fontan circulation. In other subjects, if the morbidity attendant to durable MCS can be reduced, an informed patient may elect to persist with MCS in preference to the well-known transplant related morbidity such as immunosuppression related infection and malignancy, graft vasculopathy, hypertension, diabetes, and chronic renal failure. At the 2010 meeting of the International Society for Heart and Lung Transplantation, Dr. O. H. Frazier proposed a new paradigm whereby heart transplantation is reserved for the elderly and younger people are supported by cardiac pumps. This concept, somewhere between bridge to transplant and destination therapy may be termed "bridge to postponement." In fact in Western European adults, there were more VAD implants in 2010 than heart transplants, and it is anticipated that a similar relationship of MCS to heart transplants will pertain in the United States in the near future.

In the longer term future, several other issues may significantly alter the practice of pediatric MCS. For example, in other areas of medicine, novel oral anticoagulant medications appear likely to provide more favorable risk–benefit profile than the traditionally used warfarin. Such agents may prove useful in MCS patients. Regardless of the specific advances in MCS that the future holds, it is clear that we are now much closer to our goal of durable mechanical support for children of all ages with a minimum of associated complications.

SUMMARY

Although pediatric MCS is an underdeveloped field, substantial progress has been made in recent years. For the first time, there is an approved VAD for pediatric patients of all ages and sizes in the United States. In the immediate future, a trial of a new implantable pediatric device will begin. With all of the activity in the field, and as more centers gain experience, VADs in pediatric patients will become more routine, expanding both their indications and applicability to increasing numbers of children with end-stage heart failure.

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EDITOR'S COMMENTS

This chapter summarizes the current state-of-the-art use of ECMO and assist device support in children with congenital heart disease. While ECMO has been the mainstay of support for both heart and lungs in children with ventricular failure or failed cardiac repair, the limitations of cannulation and the need for intense anti-coagulation with this therapy have limited its use to short-term support. It is difficult to maintain a patient on ECMO support for more than a few weeks as a bridge to cardiac transplantation. Thus, ECMO is rarely used as a sole bridge to transplantation therapy since the waiting times are now so long that it is unlikely that the patient can be supported solely with ECMO as a bridge to transplant. Early in the use of ECMO for postoperative cardiac support and as a bridge to transplantation, the waiting times were short enough for young children such that ECMO could be used as the sole bridge; however, waiting times have continuingly increased for cardiac transplantation even in very small infants and children.

As noted in this chapter, there have been increasing reports of ECMO being used as a bridge to lung transplant in children. However, again the problems with this approach are that waiting times are so long for lung transplantation that ECMO is not a very effective bridge. In patients with purely respiratory issues, venovenous ECMO can be used as a bridge to transplant since it can be maintained for a longer period of time than venoarterial ECMO and patients can in fact even be ambulatory or even extubated with venovenous ECMO support for respiratory causes. Nevertheless, the relative lack of mobility and the instability of cannulation for venovenous ECMO for respiratory support continue to limit this technique and in most cases we would not consider using ECMO as a bridge to pulmonary transplantation. Newer devices are being developed that can assist in oxygenation and ventricular support such as the Novalung Oxygenator that can be placed between the ventricle and atrium in such a way as to decompress the right ventricle in patients with severe pulmonary hypertension yet oxygenate the blood, but use the ventricle as the cardiac pump.

When ECMO is utilized it is generally preferable to repair the neck vessels at the time of decannulation. While long-term patency of the carotid artery has been demonstrated, the patency of the jugular vein is rarely accomplished long term, but repair of the vessels may at least permit recannulation if the patient deteriorates after ECMO decannulation via the same vessels. We have routinely repaired the ECMO vessels at the time of decannulations since the early 1980s and have had minimal complications of vascular disruption, hemorrhage, or pseudoaneurysm formation.

In the majority of circumstances today, ECMO is used only as a short-term bridge to cardiac recovery in patients following cardiac surgery or for resuscitation for cardiac events acutely in the postoperative period. When ECMO is used with little chance of cardiac recovery, the patient is now bridged with ECMO to implantation of a more permanent ventricular assist device such as the Excor Berlin Heart. If a patient does not recover cardiac function within 5 to 7 days of initiation of ECMO support, then bridging to more permanent support is indicated, generally as a bridge to cardiac transplantation. Some cases of recovery of ventricular function after implantation of ventricular assist devices have been observed, such that the devices can ultimately be removed after return of cardiac function. As noted by the authors of this chapter, implantation of assist devices permits patients to have better recovery of overall organ function and can result in mobilization with both pulmonary and systemic rehabilitation that can decrease the risk at the time of later cardiac transplantation. As has been noted in the adult experience, ventricular assist devices are now commonly being used in pediatric patients as a bridge to cardiac transplantation such that at least 25% of patients

(*continued*)

EDITOR'S COMMENTS (*continued*)

undergoing transplants now have had a previously implanted assist device. I expect that the percentage will increase progressively as it has in the adult world such that the vast majority of patients will have assist devices as a bridge to cardiac transplant in the future as waiting times continue to lengthen.

The pediatric ventricular assist device market is somewhat limited and there have been few devices developed for our pediatric patient population. It is encouraging that device manufacturers are now looking to the pediatric market and developing newer axial-type flow devices. The PumpKIN trial that is currently being developed will help assess new devices for our patients for the future. Hopefully, these devices will become easier to implant and will have lower risks of thromboembolism and hemorrhage, which are currently the major problems with assist device use in children. As noted in the chapter, anti-coagulation is required for all of these devices and a risk of postoperative hemorrhage is significant even late after device implantation. In addition, thromboembolism is a recurring problem and the majority of patients will have some thromboembolic event with a device implanted for over 2 to 3 months. It is becoming important now for pediatric cardiac centers to have a well-established protocol for therapy of thromboembolism of patients on devices with very acute intervention and thrombolytic or thrombectomy catheter-based therapies for embolic strokes, which can improve the overall neurologic outcome of these patients.

The total artificial heart has now been developed for implantation in older children and adults and may have a particular applicability in patients after the Fontan operation. As noted by the authors, the patients with the Fontan circulation represent a particularly difficult subgroup for assist device implantation. It is unclear whether support of the right-sided circulation with pushing of blood through the lungs, or support of the left-sided circulation with pulling the blood through the lungs, is the most appropriate approach in any individual patient. Great interest is being shown now in developing assist devices for the venous side of the circulation in the Fontan situation, and implantable devices for optimizing pulmonary blood flow may appear in the near future. Until that time, the total artificial heart may be the most appropriate option as a bridge to transplant.

TLS

107 Introduction and Overview of Pacemaker/ICD Therapy in Children

Peter Aziz, J. William Gaynor, and V. Ramesh Iyer

INTRODUCTION TO PACEMAKER THERAPY

HRS/American College of Cardiology Recommendations/Guidelines for Pacemaker Implantation

The clinical indications for pacemaker implantation in children, adolescents, and young adults with congenital heart disease (CHD) are summarized below.

Class I

1. Advanced second- or third-degree atrioventricular block (AVB) associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output.
2. Sinus node dysfunction (SND) with correlation of symptoms during age-inappropriate bradycardia.
3. Postoperative advanced second- or third-degree heart block that is not expected to resolve or that persists >7 days after cardiac surgery.
4. Congenital complete AVB with a wide ventricular escape rhythm, complex ventricular ectopy, or ventricular dysfunction.
5. Congenital complete AVB in an infant with an average rate of 55 bpm or less or a rate of 70 bpm or less in a patient with CHD.

Class IIa

1. CHD and sinus bradycardia for the prevention of recurrent episodic intra-atrial reentrant tachycardia (IART).
2. Congenital complete AVB beyond infancy with an average heart rate of <50 bpm, abrupt pauses in ventricular rate that are more than two to three times that of the sinus cycle length, or in patients with symptomatic bradycardia.
3. Sinus bradycardia in patients with complex CHD with a resting ventricular rate of <40 bpm or pauses of more than 3 seconds.
4. CHD and impaired hemodynamics due to sinus bradycardia or loss of atrioventricular (AV) synchrony.
5. Unexplained syncope in a patient with prior congenital heart surgery complicated by transient complete heart block with residual fascicular block.

Class IIb

1. Transient postoperative complete AVB that reverts to sinus rhythm with residual bifascicular block.
2. Congenital complete AVB in a patient with an acceptable rate, narrow QRS escape, and normal ventricular function.
3. Asymptomatic sinus bradycardia after biventricular repair of CHD with a resting heart rate <40 bpm or ventricular pauses of more than 3 seconds.

Class III

1. Not indicated for transient postoperative complete AVB.
2. Not indicated for asymptomatic bifascicular block after CHD surgery in the absence of prior transient AVB.
3. Not indicated for asymptomatic second-degree AVB type I.
4. Not indicated for asymptomatic sinus bradycardia with pauses <3 seconds and an average heart rate >40 bpm.

Common Clinical Indications for Pacemaker Implantation

Complete Congenital Heart Block

Complete congenital heart block (CCHB) is the most common indication for permanent pacemaker in the pediatric population in the absence of CHD. Approximately 20% to 30% of paced pediatric patients received implants for this indication. Though maternal exposure to lupus antibodies is a common etiology for CCHB, most cases are idiopathic. The importance of confirming lupus exposure is highlighted by the fact that some infants exposed to lupus antibody are also more likely to develop dilated cardiomyopathy as a result.

L-transposition, heterotaxy, and common AV valve defects are congenital lesions commonly associated with complete heart block. The most common association of CHB in the newborn is heterotaxy with left atrial isomerism. In these patients with significant fetal compromise (hydrops, poor ventricular function), outcomes remain poor. Isolated CCHB in the absence of CHD, however, carries an excellent prognosis.

Postoperative Heart Block

Before the advent of pacemakers, surgical complete heart block (SCHB) following repair of CHD carried a dismal prognosis with a 50% mortality rate. Currently, pacemaker implantation is recommended for persistent SCHB for at least 7 days, with no expected resolution. The study by Weindling et al. showed 97% recovery of AV node conduction by 10 days suggesting an extended observation period before making the decision of pacemaker implantation. Although the majority of patients with resolution of SCHB demonstrate intact AV conduction in follow-up, a small subset of patients develop recurrence of AV node disease several years after surgical intervention. As such, pacemaker implantation is a class II indication in patients with unexplained syncope and residual bifascicular block following resolution of SCHB as these patients may be at risk for recurrence or higher grade heart block. Unfortunately, no definitive predictors of recurrence have been established in patients with normal electrocardiograms (ECGs; after resolution of immediate postoperative heart block) and these patients require close follow-up attention postoperatively.

Epicardial vs. Transvenous Considerations

Epicardial pacing systems (Fig. 107.1) are generally utilized in younger patients (<5 years or so), patients with single ventricle physiology and intracardiac shunts. In a multicenter study by Khairy and colleagues comparing transvenous versus epicardial pacing systems, the authors found a greater than twofold increase in thromboembolic events with transvenous devices in patients with significant left-to-right or right-to-left shunts. The advantage of transvenous systems is that transvenous leads tend to have lower pacing thresholds, higher lead impedances, and longer battery longevity in comparison to the epicardial leads. Of course, epicardial systems also obviate the need for a sternotomy or thoracotomy. The risks of venous occlusion and lead compromise are significantly increased in infants with transvenous pacemakers because of smaller patient and vessel lumen size. The major disadvantage of epicardial systems in young children is the higher risk of lead fracture due to mechanical stress. More importantly, maintaining long-term vessel integrity is imperative for infants requiring life-long pacing and, therefore, an epicardial system is ideal at a young age until adequate vascular growth is achieved and transvenous leads are an option.

Transvenous leads in the pediatric patient pose a unique challenge. Adhesions between the leads and endocardial or vascular tissue and patient growth can cause shear and stress tension on the lead and may compromise lead function. To circumvent this issue of growth, many implanters leave an extra loop of lead (such as right atrial loops) to minimize tension (Fig. 107.2). However, looping may create the presence of more surface lead area in the heart and promote further adhesions, thus complicating future lead extraction if needed.

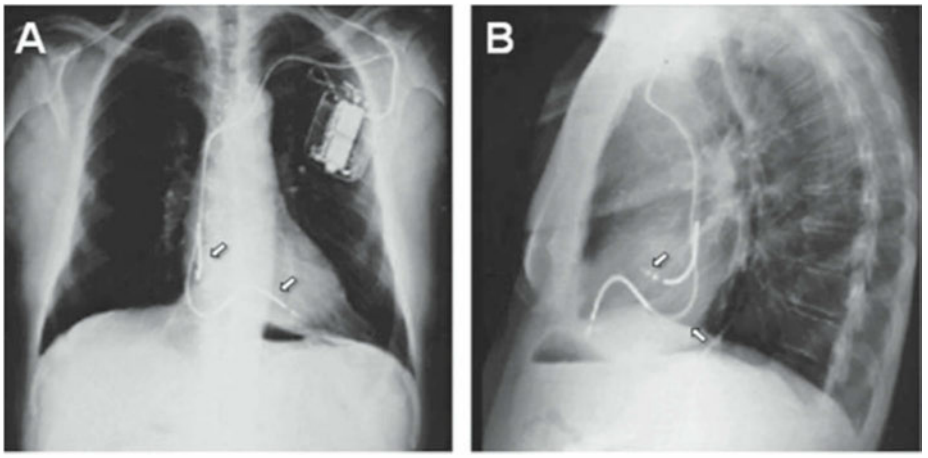


Fig. 107.2. X-rays showing transvenous atrial and ventricular lead (*arrows pointing*) with extra loop for growth in anteroposterior (AP) (**A**) and lateral (**B**) views.

Though epicardial systems are preferred in infants, the decision regarding implantation type in young children is more difficult. The risk of thromobembolism in young patients must be balanced with the risk of compromise to lead integrity. In a study of 63 children, echocardiography and venography were routinely performed to assess the risk of thrombus formation. Twenty-one percent were found to have moderate or severe thrombosis at follow-up. Patients found to have thrombosis were younger (4.5 vs. 8.2 years) and had larger lead diameter to body surface area ratio suggesting that children <5 to 6 years of age should have epicardial systems implanted. Dual chamber (two lead) systems were also found to be prothrombotic (because of two leads occupying a larger vessel diameter) and should be avoided in the younger patient population.

Epicardial systems (Fig. 107.1) are preferred in all patients with CHD and intracardiac shunting, regardless of age, because of increased risk of systemic thromboembolism. Unipolar leads (Fig. 107.1) have a single electrode with the pacemaker generator acting as the anode. Steroid-eluting leads provide lower (better) pacing thresholds and therefore preserve battery longevity. Bipolar leads (suture on) offer better sensing

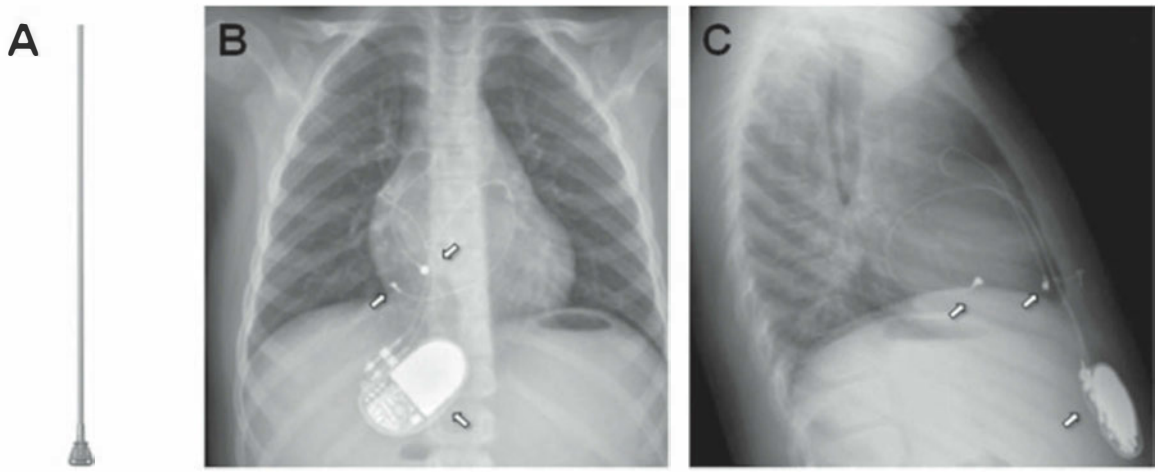


Fig. 107.1. Illustration of an epicardial unipolar suture on lead (**A**), X-rays of epicardial unipolar atrial and ventricular lead and abdominal generator (*arrows pointing*) in an anteroposterior (**B**) and lateral view (**C**).

thresholds as compared with unipolar leads since they avoid far-field cardiac or skeletal muscle sensing. Ideally, bipolar leads (Fig. 107.3) should be placed on the myocardium free of scar to optimize sensing and pacing thresholds. Bipolar screw-in leads are also available for use in older patients with thicker myocardium (Fig. 107.4). In patients with multiple prior sternotomies, the left thoracotomy approach enables the surgeon to access more posterior structures such as left atrium and ventricle and avoid scar tissue (from sternotomies), thus facilitating dissection.

Subpectoral vs. Subcutaneous Considerations

Subpectoral devices are preferred in children as they offer the most optimal cosmetic result. However, the implant may be more technically challenging (more submuscular dissection) and need an experienced implanter. Muscular protection of the proximal lead, header, and generator improve longevity as well. Since subpectoral implants are anteriorly and posteriorly surrounded by muscular tissue, bipolar sensing and pacing are preferred to minimize muscular interference and muscular stimulation, respectively.

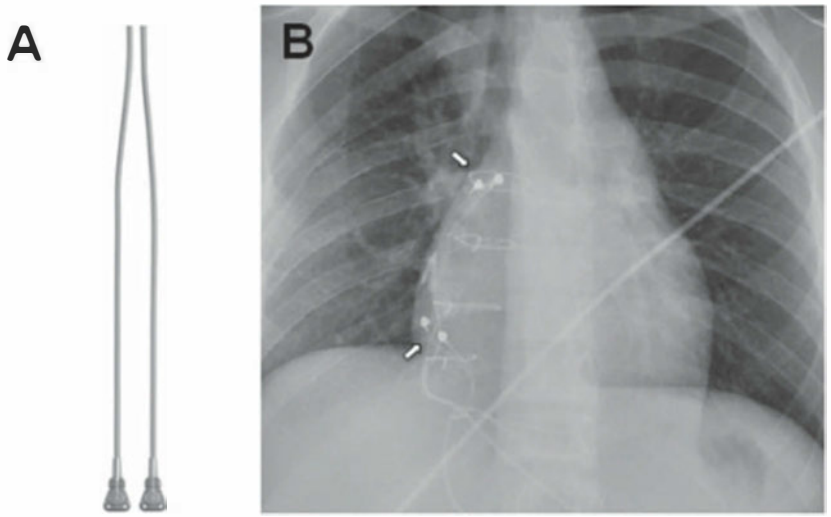


Fig. 107.3. Illustration of an epicardial bipolar suture on lead (**A**), X-rays of epicardial bipolar atrial and ventricular lead (*arrows pointing*) in an anteroposterior view (**B**).

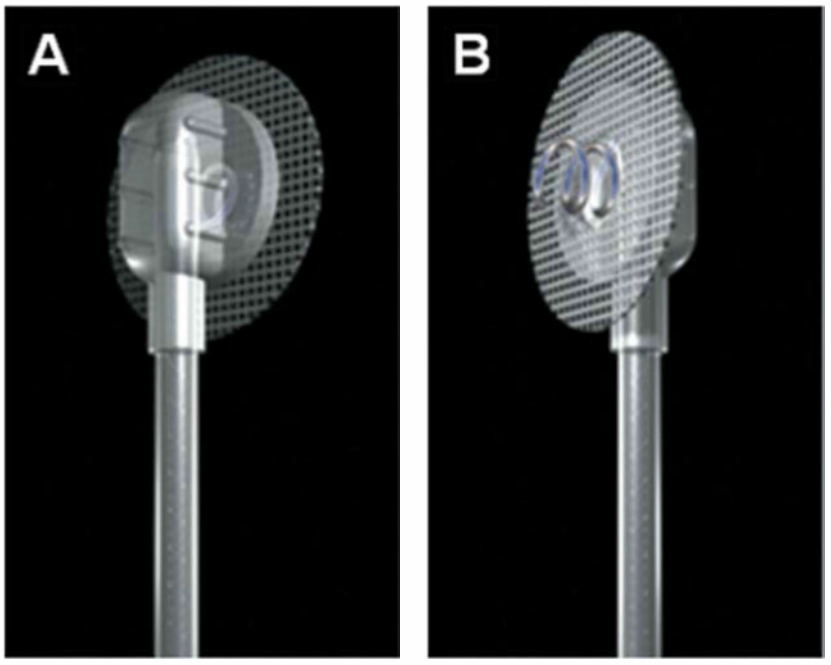


Fig. 107.4. Picture of screw in bipolar epicardial lead in two views.

Pacing in Operated Congenital Heart Disease

The indications for pacemaker implantation in patients with and without CHD are important to distinguish. In patients with normal cardiac anatomy, bradycardia, even advanced atrioventricular heart block, can be well tolerated from a symptomatic standpoint and is not an indication alone for pacemaker implantation. However, patients with operated CHD may require closer evaluation for residual hemodynamic issues, which may influence pacemaker implantation decision-making. One such example is an adolescent with single ventricle physiology with AV valvar insufficiency who has SND with bradycardia and junctional rhythm. Such a subject may be a candidate for pacing (atrial pacing) to improve his symptoms and hemodynamics. Consideration must be given to residual hemodynamic issues such as AV valve or semilunar valve insufficiency and chamber enlargement in addition to the presence of sinus or AV node dysfunction.

Pacing for Arrhythmias

Intra-atrial Reentrant Tachyarrhythmias

Pacemaker implantation is recommended for patients with CHD and bradycardia-induced IART. The purpose of pacing in these individuals is to prevent premature atrial complexes and short–long–short extrastimuli sequences, which have been implicated as tachycardia triggers. Anti-tachycardia pacing is a useful programming tool available in select devices, which has been shown to terminate atrial reentrant tachycardia in more than 50% of instances in a multicenter trial of pediatric patients with CHD. The use of anti-tachycardia pacing is limited since it is not effective in terminating atrial fibrillation and may actually induce atrial fibrillation in susceptible patients.

Long QT Syndrome

Life-threatening arrhythmias such as polymorphic ventricular tachycardia (VT) (Torsade de pointes, TdP) can be triggered by bradycardia or pauses in some forms of long QT syndrome (LQTS). Cardiac pacing has been employed to prevent pause-dependent TdP and has been shown to be effective in preventing ventricular arrhythmias and sudden death. Physiologic pacing has also been shown to shorten the QT_c interval. Pacing has also improved the clinical

outcome in LQTS patients with 2:1 AVB at birth, long to be considered extremely high risk for sudden cardiac death (SCD).

Biventricular Pacing in Pediatrics

The objective of cardiac resynchronization therapy (CRT) is to coordinate right and left ventricular function in patients with clinical heart failure, particularly those with a left bundle branch block. Numerous studies in adult patients have demonstrated the benefits of CRT in those with dilated cardiomyopathy and electromechanical dyssynchrony. These controlled and randomized studies have shown an improvement in New York Heart Association (NYHA) class, exercise stress testing parameters, and mortality. Unfortunately, there are no such pediatric studies that have shown a similar benefit, but there are small series and case reports that have shown some promise of CRT in children with and without CHD. Currently, cardiac resynchronization is a class I indication in adult patients with an ejection fraction of ≤35%, QRS duration of ≥120 ms, and a NYHA class of III or IV.

The efficacy of CRT in the pediatric population, particularly those patients with CHD and right ventricular (RV) dysfunction, is not well established. As occurs in most studies in patients with CHD, the degree of heterogeneity in this population precludes large studies and conclusive statements. Moreover, traditional measures of improvement (QRS duration, tissue Doppler imaging (TDI), ventricular function) are complicated by underlying congenital cardiac lesions and establishing benefit in these patients is exceedingly difficult. In a small study, CRT was shown to increase arterial blood pressure and decrease QRS duration in the acute postoperative setting in patients with CHD. One prospective study by Jeewa et al. showed no acute benefit to CRT (cardiac index, blood pressure, TDI) in the congenital population. Long-term results in the largest retrospective study to date showed improved NYHA class and ejection fraction in 87% of patients undergoing CRT. Unfortunately, no conclusive study on CRT exists in pediatrics and most studies show conflicting results. It remains to be determined whether CRT will play a role in the management of patients with CHD, particularly those with RV dysfunction and right bundle branch block.

Surgical Considerations

Implantation of pediatric pacemakers is best performed by a team (cardiologists, surgeons, and anesthesiologists) skilled in the management of children. Particular appreciation for anatomical considerations is often the key in patients with CHD as anatomical variations often dictate the surgical approach. Ideally, general anesthesia should be performed by a pediatric cardiac anesthesiologist with special attention to effects of anesthesia on the hemodynamics of the cardiac lesion.

Careful screening for patients deemed pacemaker dependent (defined as an unreliable underlying escape rhythm) is a crucial part of surgical preparation. Anesthesia commonly suppresses conduction and may exacerbate brady arrhythmias. Pacemaker-dependent patients undergoing generator changes must also have an appropriate mechanism for maintaining hemodynamic stability. Isuprel administration or a temporary transvenous pacing lead via the femoral vein are useful adjuncts in preparation for pacemaker implantation in patients with unreliable escape rhythms.

Patients with CHD frequently have variations in venous anatomy, thereby complicating the transvenous approach. As such, venography of the subclavian, innominate, and superior vena cava can document patency and provide a "road map" for venopuncture. Clear elucidation of intracardiac anatomy and the presence or absence of shunts is also imperative. The presence of an unrecognized patent foramen ovale may lead to inadvertent placement of the ventricular lead in the left ventricle (Fig. 107.5). This could be avoided by careful fluoroscopic imaging of the leads using different angles to confirm anterior lead placement. On the other hand, the ventricular lead placement in a patient with transposition of the great arteries (in the pulmonary ventricle-anatomically left ventricle) will be posterior fluoroscopically compared with a patient with normal segmental anatomy (Fig. 107.6). Demonstration of patent systemic venous return in patients with atrial switch operations (Mustard, Senning) is imperative as occlusion prevents transvenous access to endocardial tissue. Narrowing, stenosis, or interatrial leaks of the atrial baffle needs to be recognized by angiography and addressed (surgically, by stent or atrial septal defect device placement) before transvenous leads are placed (Figs. 107.6 and 107.7). Cardiac angiography in this patient population is also indicated to assess for baffle leaks that are a risk for paradoxical emboli and stroke. Baffle leaks need to be closed (either by device closure or surgical approach) before transvenous leads are inserted. The use of transvenous approach for atrial and ventricular pacing has been described in patients with single ventricle physiology and Fontan palliation. However, this poses technical challenge of finding viable atrial tissue capable of capture (in Fontan baffle with prosthetic material) and also obviates the need for chronic anticoagulation and its associated complications.

LEAD EXTRACTION

Summarized below are the indications for lead extraction as it pertains to the pediatric patient:

Class I

1. Sepsis related to device infection.
2. Life-threatening arrhythmia or immediate physical threat related to retained lead fragments.
3. Clinically significant thromboembolic event caused by lead fragment.
4. Lead that interferes with subsequent implantation.

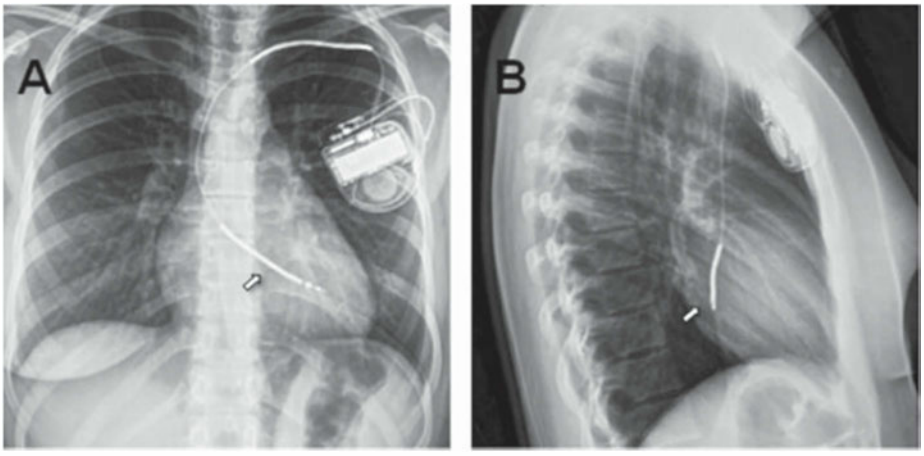


Fig. 107.5. X-rays in an AP (**A**) and lateral (**B**) view showing inadvertent placement of transvenous ventricular lead (*arrows*) across a patent foramen ovale into the left ventricle.

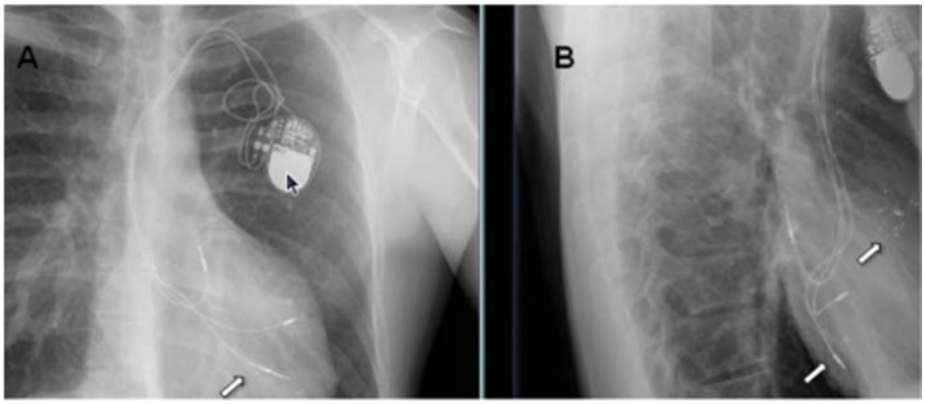


Fig. 107.6. X-ray in AP (**A**) and lateral (**B**) views showing transvenous atrial and ventricular lead placement in a patient with TGA after Mustard operation. Note the ventricular lead is in the left ventricle (pulmonary ventricle) (*arrows*). Also there is an atrial septal defect device in place for closure of baffle leak.

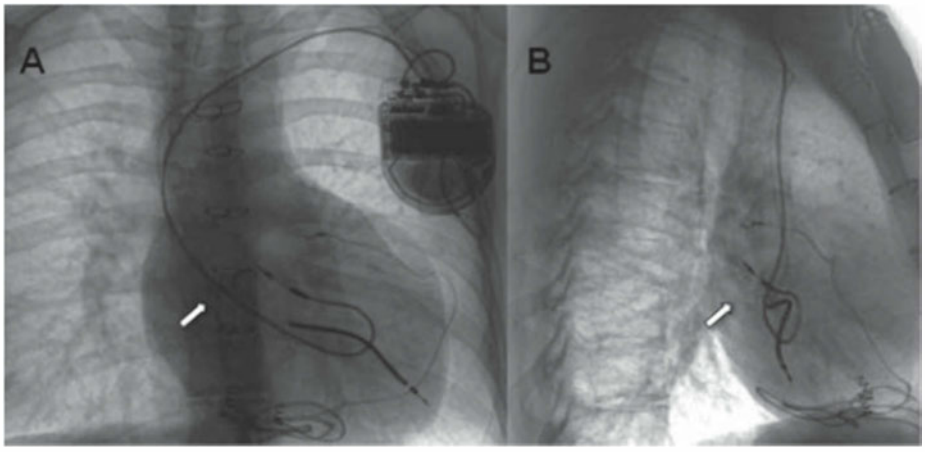


Fig. 107.7. X-ray in (AP) (**A**) and lateral (**B**) views illustrating a stent in Mustard baffle (placed for stenosis) (*arrows*) and transvenous atrial and ventricular pacing leads across the stent in a patient with transposition of great arteries (TGA).

Class II

1. Localized pocket infection.
2. Pocket site that causes significant discomfort.
3. Lead that poses potential threat due to lead design or failure.
4. Nonfunctional lead in a pediatric patient.

Class III

1. Risk of removal outweighs benefit.
2. Lead that can be reused at time of generator change.

The most common class I indication for lead extraction is pocket/device infection (Fig. 107.8). Successful treatment of pacemaker-related infection requires complete removal of all prosthetic (pacemaker and lead) material. In a study from the Cleveland Clinic of 123 patients undergoing hardware removal for infection, four patients experienced recurrence of infection and all four patients had retained device remnants, both transvenous and epicardial. Complete extraction was achieved in 118 patients, all of whom had no infection recurrence. Lead malfunction in the pediatric population is the most common indication for lead extraction. Lead insulation disruption and inner pacing or sensing coil fracture (Fig. 107.9) are common causes of lead malfunction, particularly in pediatrics as trauma and patient growth can compromise lead integrity. Traditionally, epicardial leads are abandoned (after new lead/s placement) without the need for extraction.

Thrombus formation around the lead is the major impediment to extraction. Fibrous tissue then forms over the lead in the course of days to months. Typical sites of fibrous accumulation (on the lead) include the venous entry site, the anastomosis of the superior vena cava and innominate vein, and the distal lead tip. In younger patients with older leads, calcification may further strengthen the fibrous matrix, thereby complicating extraction.

Several tools have been devised to facilitate lead extraction. In the past, direct traction was the only available method. Not surprisingly, direct traction often led to complete lead fracture with retained fragments requiring surgical removal. The advent of locking stylets allows for traction to be placed on the distal lead tip, increasing the likelihood of complete extraction. Telescoping sheaths and laser sheaths provide counterpressure and countertraction, both disrupting fibrous tissue adhesion to vascular structures thus facilitating lead removal.

Life-threatening complications of lead extraction include myocardial avulsion, vascular tear, pneumothorax, pulmonary embolism, arteriovenous fistula, and death. Minor complications are pericardial effusion, hematoma, venous thrombosis, and arrhythmia. Recent reported complication rates are between 0.4% and 1.0% with lead extraction success rates of >95%. The most important factors predicting extraction failure are increased implant duration, physician inexperience, younger patient age, and ventricular versus atrial leads. The risk of failed extraction, based on this database, doubles for every 3 years of implant duration. Operators who have performed fewer than 20 extractions had lower success rates. As described earlier, the pediatric population is more likely to develop calcified fibrous matrices lending to lower success rates in younger patients. Because of the increased risks, such complicated procedures should be limited to experienced operators with all available resources including surgical operating room backup for emergency surgical intervention in the event of any of the above-mentioned life-threatening complications.

Pacemaker Follow-up Challenges

The objective of pacemaker follow-up is to optimize device function, prolong battery longevity, and identify possible lead/generator issues. At implant, threshold margins (both pacing and sensing) are programmed with 2 to 3× safety margin. This is because myocardial edema and scaring may transiently affect threshold values in a fresh lead implant. Lead maturation occurs over the course of 6 weeks to 3 months at which time thresholds remain stable (chronic thresholds) and reprogramming to optimize battery longevity is indicated.

Pacemaker status evaluation can be performed in a variety of settings. Conventionally, device checks are performed in the office by an electrophysiologist or a cardiologist with pacemaker programming

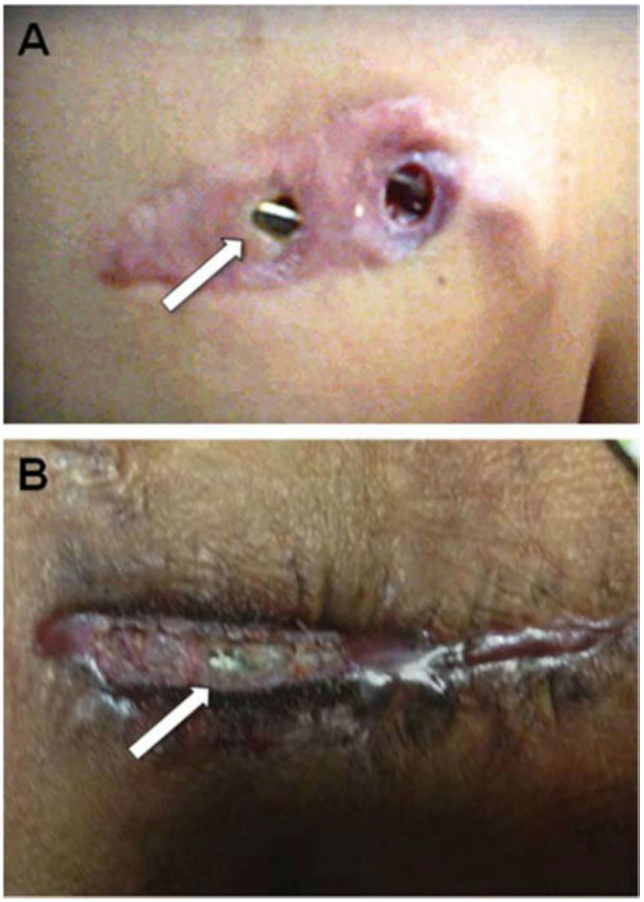


Fig. 107.8. Pictures showing pacemaker pocket infection (**A**) and device extrusion (*arrows*) (**B**).

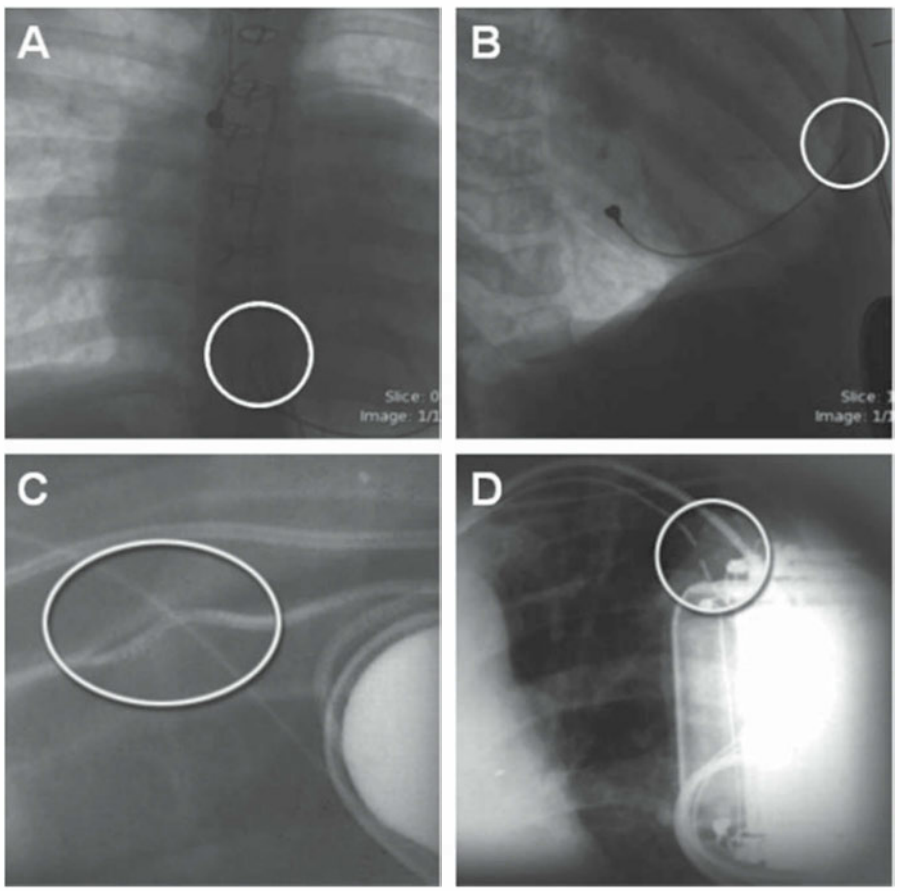


Fig. 107.9. X-rays in AP and lateral views showing epicardial (**A** and **B**) and transvenous pacing lead fracture (*circles*) (**C** and **D**).

expertise. With the advent of remote monitoring, transtelephonic checks can provide information regarding sensing/pacing thresholds, stored electrograms in the device for arrhythmia troubleshooting, and battery longevity. However, formal device interrogations by a technician or an electrophysiologist are required on a yearly basis, and more often for those patients with CHD or ongoing pacemaker issues.

Generator Replacement

The commonest indication for generator replacement is battery depletion. Several factors influence battery longevity in pacemakers. Current drain for pacing is dependent on pacing rate, percent pacing, programmed voltage, programmed pulse width, and lead impedance. Ohm's law ($V = IR$, V = voltage, I = current, R = resistance) dictates that the current is inversely proportional to impedance at any given voltage. Therefore, leads with higher impedances (such as transvenous bipolar leads) tend have lower current drain (for a similar voltage) and longer battery longevity. Moreover, contemporary pacemakers have several features obligating current including sensing measurements, rate response sensors, and algorithm implementation.

Most devices are equipped with an algorithm that will estimate battery life. Devices that reach elective replacement indicator (ERI) will typically revert to an industry-dictated mode that is recognizable to the clinician. For example, Medtronic devices will pace VVI at 65 bpm if ERI is reached. Depending on variables mentioned, generators will have only months of battery life at ERI and generator replacement planning during that time frame is required. Patients with CHD who are reliant on AV synchrony will become quite symptomatic in an asynchronous pacing mode (e.g., VVI). As such, many clinicians will opt to replace the generator just prior to ERI in such selected patients. Of course, a thorough investigation of lead competence should be performed prior to generator change in all patients.

Pacemaker Recalls/Device Malfunction
Pacemaker malfunction is rare, occurring in approximately 0.4% of all implanted pacemakers translating into 1.3 malfunctions per 1,000 person-years. Unfortunately, device and lead recalls carry an enormous financial and psychological burden. Regular monitoring of lead and device performance by various device companies and subsequent FDA reporting has paved the way for physicians to closely monitor some of the "recalled" leads and devices. As technology continues to improve, device malfunctions will hopefully be limited to nonclinically significant events.

Improvement in magnetic resonance imaging (MRI) and pacemaker device technologies has seen the introduction of "MRI safe" pacemakers in the last couple of years. However, all risks of MRI exposure have not been completely understood and the use of MRI in patients with pacemakers or implantable cardioverter defibrillators (ICDs) should be carefully considered after discussing the risks and benefits of the diagnostic testing with the patient and treating physician.

Special Challenging Situations

Older Fontan
Bradyarrhythmias and tachyarrhythmias cause significant morbidity in patients with Fontan physiology. Incidence of SND and atrial flutter increases with time after the Fontan operation. Loss of AV synchrony in Fontan patients can lead to significant hemodynamic compromise. As such, 6% to 10% of patients with Fontan physiology require pacing 5 years postoperatively.

Venous anatomy in Fontan patients precludes direct access to atrial tissue limiting the ability to implant transvenous devices, particularly in patients with an extracardiac conduit. Though reports of transvenous implantation in patients with lateral tunnel Fontans have been reported, the risk of thromboembolism mandates coumadin therapy. For these reasons, surgical placement of epicardial pacing leads is the preferred option. Adhesions and scaring can prolong dissection times and limit the amount of viable tissue to obtain adequate pacing and sensing thresholds. Extensive dissection has been theorized to increase the duration and longevity of postoperative pleural effusions (and thus prolonging hospital stay) though a comparison of Fontan versus non-Fontan patients has shown no difference in length of chest tube drainage, particularly since the advent of the Fontan fenestration.

Difficult Vascular Access
Patients with complex CHD, multiple cardiac catheterizations, or a history of venous thrombosis who are being considered for transvenous pacemaker placement should undergo vascular studies as part of the preimplant evaluation. Ultrasonography and/or venography are crucial in demonstrating feasibility of access, certainly prior to pocket formation. In patients with occlusion of the left innominate vein, right-sided access is possible and safe. Sometimes, right-sided access is preferred in patients who are left-handed as well. If vascular access precludes transvenous placement, the epicardial approach is indicated.

Newborn with Congenital Heart Block
Permanent pacing in patients with congenital CHB is quite common with 53% to 63% of patients requiring pacemaker implantation, 33% within the first 9 days of life. Among patients paced, the mortality rate in the first year of life is 17%, mostly related to dilated cardiomyopathy. Fetal hydrops is associated with a particularly poor outcome with a mortality exceeding 80%. Though anecdotal, no clinical trials have proven the efficacy of these measures.

The surgical approach in this patient population is often determined by limitations in patient size and myocardial access. Atrial tissue in a neonate tends to be friable and single chamber pacing with ventricular leads is often preferred as the initial permanent pacing system. Also, the atrial rates in newborns are significantly faster (120 to 160 bpm) and AV synchronous pacing at faster rates may be detrimental to achieving optimal cardiac output. As mentioned, asynchronous pacing may be poorly tolerated depending on baseline hemodynamics and may not be appropriate for all patients, particularly those with significant CHD.

Premature Infants with/without Congenital Heart Disease
Prematurity, low birth weight, low ventricular rate (<55 beats/min), significant structural heart disease, evidence for ventricular dysfunction or associated cardiomyopathy and presence of hydrops fetalis all predict poor outcome in neonates with complete heart block. Antenatal treatment has been pursued in these high-risk patients and consists of (1) inotropic medications to increase fetal heart rate, (2) plasmaphoresis and steroids to counteract an inflammatory component, (3) digoxin and/or lasix to treat ventricular dysfunction, and (4) fetal pacing. Unfortunately, no clinical trials are available to demonstrate the efficacy of these methods. In fetuses that are profoundly affected with evidence of hydrops, early delivery and pacing have been performed.

In the premature infant, technical challenges in combination with relative hemodynamic instability complicate permanent pacemaker implantation. Reports of pacemaker implantation in premature infants >2 kg exist. However, the importance of early stabilization in these patients may trump the need for permanent pacing. One approach is to place temporary pacing wires immediately after birth for stabilization, thus circumventing the possibility of low cardiac output and acidosis in the neonatal period. This approach also allows for achieving adequate growth, thus facilitating permanent epicardial pacing at a later date.

The premature patient with significant CHD continues to portend a dismal outcome with nearly uniform mortality. Unfortunately, no approach has been shown to demonstrate a more favorable result in this patient group. Certainly, fetal counseling about possible poor outcome is imperative for the family with an in utero diagnosis.

INTRODUCTION TO ICD THERAPY

HRS/American College of Cardiology Recommendations/Guidelines for ICD Implantation

The clinical indications for ICD implantation in children, adolescents, and young adults with CHD are summarized below.

Class I

1. Survivors of cardiac arrest (CA) after exclusion of reversible causes.
2. Symptomatic sustained VT in association with CHD.

Class IIa

1. Recurrent syncope of undetermined origin in the presence of ventricular dysfunction or ventricular arrhythmias at electrophysiology study (EPS).

Class IIb

1. Recurrent syncope associated with complex CHD and advanced ventricular dysfunction with no other identifiable cause.

Class III

1. Not indicated in patients with a life-expectancy of <1 year.
2. Not indicated in patients with incessant VT of VF.
3. Not indicated in patients with significant psychiatric illness that may be aggravated by a device.

4. Not indicated in patients with syncope of undetermined cause with no inducible VT and no structural heart disease.

The indications for ICD implantation in young patients is mostly based on adult data and has evolved substantially in the last 15 years. Unfortunately, pediatric patients experience a high rate of mortality compared with adults following resuscitation after a sudden CA. Fewer than 1% of all ICDs are implanted in the pediatric patient, thereby limiting our ability to assess the efficacy in this vulnerable population. Generally, the risk of sudden death is likely smaller in a pediatric patient than in an adult with the same disease process. As such, guidelines based on adult literature may not be extrapolated to the pediatric patient.

Appropriate patient selection is imperative. One must balance cumulative life-time risk of a presenting condition with the cumulative life-time risk of having an ICD before making the decision. Typical indications for ICDs in the pediatric patient include CHD with significant arrhythmias and risk of sudden death, cardiomyopathies, and inherited arrhythmia syndromes. Antiarrhythmic therapy is variably effective with significant toxicity in some. Patient compliance, particularly in the teenage population, offers yet another obstacle to medical management. Though the frequency of device implantation has increased in recent years, technological advances have not translated into freedom from inappropriate shocks and device/lead malfunction. Therefore, the decision to implant a device must accompany a candid conversation of risks and benefits with the patient and family. As will be discussed, the specific clinical situation and diagnosis are the crux of the decision-making process.

Common ICD Clinical Situations

Primary prevention therapy is intended to prevent SCD in patients susceptible based on risk factors. Secondary prevention therapy is intended to prevent SCD in patients with a previous aborted cardiac event.

Long QT Syndrome

LQTS is a cardiac channelopathy associated with syncope and SCD as a result of ventricular arrhythmias, particularly TdP. The management of LQTS is aimed at preventing significant arrhythmia and SCD. Current therapy consists of beta-blockers, ventricular pacing (in some situations) (as discussed in the previous section) and left cardiac sympathetic denervation. ICD therapy is recommended for patients with recurrent syncope despite medical therapy, sustained or nonsustained ventricular arrhythmias indicating a risk of sudden death or an aborted SCD event. Current guidelines state that primary prevention ICD therapy may be considered in patients with a strong family history of SCD, though this is controversial. In a study by Kimbrough et al., sudden death in the family does not increase the risk of lethal events in other family members. Certainly, in these scenarios, emotional considerations complicate the conversation and decision-making process.

Genotype considerations have received a great deal of attention in the literature and offer critical insight into the decision-making process. Though not formally part of the HRS/AHA guidelines for device implantation, these associations cannot be ignored as the risk of SCD is not the same among different genotype classifications. Fortunately, the most common genotype (LQT1) confers the least risk of SCD. In a study evaluating 187 LQT1 patients treated with beta-blockers, the incidence of CA was 1.1%. In this same study, the incidence of CA among 120 LQT2 patients was 6.6%, none of which experienced sudden death. Therefore, approximately 99% and 93% of patients with LQT1 and LQT2, respectively, are free from significant cardiac events on beta-blocker therapy. These results contrast with that of 28 treated LQT3 patients of which 14% experienced CA in the same study. In these higher risk LQT3 patients, beta-blockers clearly do not offer the same protection against SCD and the threshold to implant an ICD should be presumably lower.

Arrhythmogenic Right Ventricular Cardiomyopathy

Arrhythmogenic RV cardiomyopathy/dysplasia (ARVC/D) is characterized by fibrofatty replacement of ventricular myocardium leading to ventricular arrhythmias. In younger patients, SCD may be the presenting symptom. As such, ICD implantation has become an emerging treatment modality for ARVC. Historically, the decision to implant an ICD was largely driven by merely meeting diagnostic criteria. In a study by Corrado et al., nearly 50% of patients with an ICD required appropriate ICD intervention despite antiarrhythmic drug therapy over a mean follow-up period of 3.3 years. Fatal arrhythmias (ventricular fibrillation) were experienced by 24% of patients. This study concluded that a history of either CA or VT with hemodynamic compromise, younger age, and LV involvement are all independent risk factors for fatal arrhythmias. Induction of VT during an EPS was thought to be of limited value in this study though a more recent study states that a positive EPS is a strong predictor of lethality.

According to the AHA/HRS guidelines, patients with prior CA, syncope due to VT, extensive RV disease, LV involvement or presentation with polymorphic VT and apical RV aneurysm are at higher risk of SCD. The guidelines also state that the decision to implant an ICD should ultimately be based on "experience, judgment and available data" until more information becomes available.

Brugada Syndrome

The Brugada syndrome has the characteristic ECG findings of right precordial ST elevation and is associated with a high incidence of sudden death. Typically, events occur during sleep or with fevers and are caused by a primary cardiac, sodium channelopathy. Like other channelopathies, therapy is aimed at preventing SCD. Unfortunately, ICDs are the only intervention shown to prevent SCD; therefore, risk stratification serves an important role. Much like LQTS, a family history of sudden death does not predict a higher risk for events in other family members. Patients with a spontaneous Brugada-type pattern and a history of syncope are at a sixfold increase risk of SCD.

As with ARVC, the role of EPS in risk stratification remains controversial. With a low positive predictive value of 23%, but a high negative predictive value over 3 years of 93%, some argue its utility in identifying low-risk patients. Supraventricular tachycardia exists in 20% of Brugada patients and the role of EPS is mainly to eliminate the causes of supraventricular tachycardia (atrial fibrillation, AVNRT, WPW) prior to ICD placement as SVT will increase the likelihood of an inappropriate shock. Though gene testing can identify approximately 50% of Brugada patients, specific mutations in the *SCN5A* gene have not been useful in identifying patients at risk for SCD.

Based on a consensus statement, ICD implantation is recommended for patients with a Brugada pattern on ECG (whether spontaneous or induced) who present with an aborted sudden cardiac event. ICD is also recommended for patients with symptoms such as syncope, seizure, or nocturnal agonal respiration after exclusion of noncardiac causes. Management of asymptomatic patients with positive ECG findings and a family history of Brugada

is controversial. Some studies have shown that the use of EPS may be helpful in risk stratification. Asymptomatic patients with a negative family history and an induced ECG pattern should be followed closely.

Catecholaminergic Polymorphic Ventricular Tachycardia

Catecholaminergic polymorphic VT (CPVT) is characterized by ventricular arrhythmias in the setting of physical or emotional stress. Patients with CPVT have normal resting ECGs, unlike patients with LQTS. Symptoms typically develop in childhood during exertional activities. The ryanodine receptor (RyR2) is responsible for approximately half of all the autosomal dominant cases of CPVT. The autosomal recessive type is associated with mutations in the calsequestrin gene (CASQ2). Beta-blocker therapy is the accepted mainstay of therapy and has been shown to reduce SCD. A minority of the patients continue to have VT and/or syncope on adequate beta-blocker treatment and are candidates for ICD therapy. Fortunately, CPVT is a rare disease and as such, risk stratification does not currently exist.

Epicardial vs. Transvenous Considerations

Compared with standard pacemaker leads, ICD coils tend to be thicker thus increasing the risk for vascular damage and thrombus formation, particularly in smaller children. Also, in order to provide adequate therapy charge, ICD generators are much larger than the standard pacemaker generator. Therefore, the age limitations for ICDs tend to be more conservative than that of pacemakers. As with pacemakers, infraclavicular implantation (whether subcutaneous (SQ) or subpectoral) is possible in the older patient (usually >7 years of age) undergoing transvenous placement. In the younger child, abdominal implantation is preferred with an epicardial lead system. Intracardiac shunts are a contraindication to transvenous lead implantation.

The technical challenges in ICD implantation are not only the result of lead and device size but also due largely to limitations in establishing an appropriate shocking vector. Several configurations have been explored with the goal of providing an adequate shocking threshold while limiting invasiveness and procedural complication. Conventionally, epicardial patches have been employed in the past for pediatric patients. Unfortunately, this technique requires a thoracotomy or sternotomy and restrictive pericarditis (secondary to the patch) is a known complication of epicardial patch implantation. As such, alternative defibrillator configurations have been developed, particularly the SQ array. Other configurations employ transvenous leads placed on the epicardium (Fig. 107.10) with an active can and/or a SQ array. Though no major differences in configuration type are described in the literature, establishing an adequate shocking vector continues to be challenging.

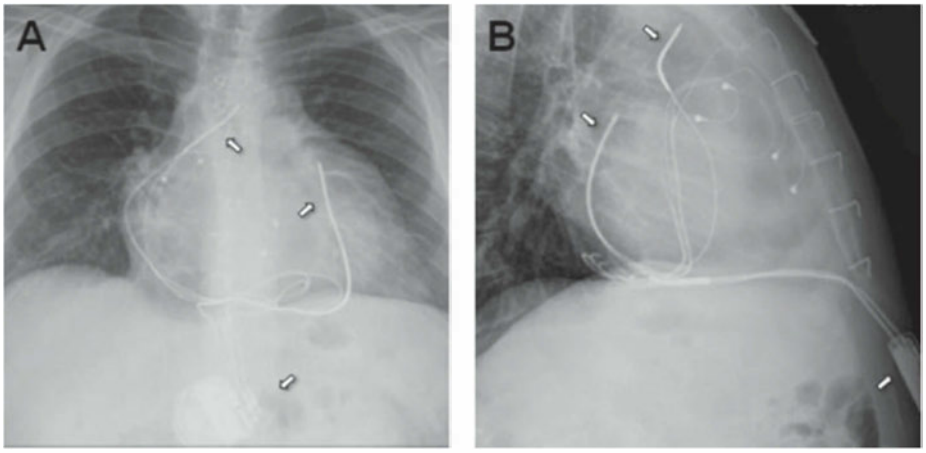


Fig. 107.10. Epicardial ICD placement in a patient with Fontan surgical correction. The arrows in panel (**A**) show an anteroposterior depiction of the epicardial electrode placement around the heart and panel (**B**) shows a lateral depiction of the same with an abdominal generator.

SQ sensing and defibrillation leads are being developed, though their use in pediatric patients is likely not imminent. These devices do not require intracardiac or intravascular hardware. Unfortunately, "leadless" ICDs (or SQ ICDs) do not allow for pacing. More importantly, sensing would be compromised, which could increase the risk of inappropriate shocks, particularly during sinus tachycardia or supraventricular tachycardia that are very common in the pediatric population. However, this technology appears to have promise for the pediatric population.

ICDs in Patients with and without Congenital Heart Disease

Due to the heterogeneity of CHD in the pediatric patient, there is a lack of large pediatric studies for broad statements regarding ICD indications, complications and benefit are not available in the literature. As such, risk stratification continues to evolve. The following is a brief summary of ICD indications and challenges in patients with CHD according to specific lesions.

Tetralogy of Fallot

The risk of SCD in tetralogy of Fallot (TOF) has an incidence of 1.2% to 3% per decade. Though other mechanisms of SCD have been proposed in this patient group, including rapidly conducting IART, sustained VT is thought to be the final common pathway to SCD in most instances. Though predictors of VT and SCD have received much attention over the last 30 years, no agreed upon risk stratification methodology has emerged. A history of ventricular ectopy, inducible VT at EPS and wide QRS duration (>180 ms) have prognostic value in determining at-risk patients. Certainly, a patient with an aborted CA or with sustained VT will benefit from ICD implantation. Patients with symptoms (palpitations, dizziness, syncope) should undergo a thorough clinical evaluation including an EPS to determine their arrhythmia substrate. A positive ventricular stimulation study has been shown to be predictive of clinical VT and warrants primary prevention ICD implantation. The clinical approach to the asymptomatic patient with nonsustained VT on surveillance ambulatory monitoring remains variable and controversial.

D-Transposition of the Great Arteries

Complete transposition of the great arteries with atrial switch operation is among the highest risk congenital lesions for SCD. The incidence of SCD in this patient population is more than three times that of TOF and is the most common cause of late mortality. Also unlike tetralogy, D-TGA atrial switch patients are at a higher risk for atrial tachyarrhythmias. In a multicenter study by Khairy and colleagues, SVT was found to coexist or precede VT during intracardiac ICD tracings prior to defibrillation. Therefore, rapidly conducting atrial tachycardia, and VT, are both proposed mechanisms for SCD in this high-risk patient population. A positive ventricular stimulation study was

not predictive of appropriate ICD shocks in this study suggesting that EPS for risk stratification has a low yield. Additionally, secondary prevention ICD implantation for RV dysfunction, nonsustained VT, and syncope are not currently supported in the literature.

Early reports of the arterial switch operation are promising though much remains to be seen regarding the risk of SCD in this group. Long-term results are emerging and are equally promising thus far. In a study by Tobler and colleagues, no arrhythmia deaths were reported and one patient had VT out of 65 patients followed over 20 years. As this population reaches adulthood, risk stratification for ICD implantation will be elucidated.

Hypertrophic Cardiomyopathy

Known risk factors for SCD in hypertrophic cardiomyopathy (HCM) are as follows: (1) previous CA, (2) sustained VT, (3) nonsustained VT, (4) family history of SCD, (5) syncope, (6) septal thickness >30 mm, and (7) abnormal blood pressure response to exercise. In these high-risk patients, primary prevention ICD interventions were common and frequently restored sinus rhythm. It is worth mentioning that severity of disease and symptoms such as chest pain, dyspnea, and exercise intolerance do not correlate with SCD risk and should not be used as justification for ICD implantation.

Left Ventricular Noncompaction

Left ventricular noncompaction is a congenital disease caused by abnormal embryogenesis of the endocardium and epicardium. Complex ventricular arrhythmias occur in approximately 40% of pediatric patients with LV noncompaction and sudden death is the most common etiology for mortality. Though data do not currently exist to risk stratify these patients, primary prevention ICD implantation has been performed and is a "reasonable clinical strategy" according to guidelines.

Technical Considerations

Similar to pacemaker implantation, meticulous attention to anatomical detail is essential to successful ICD implantation. Systemic venous anomalies are important to elucidate as part of procedural preparation. In the case of a persistent left superior vena cava (LSVC) draining to the coronary sinus (with an absent right SVC), positioning the lead toward the tricuspid valve from the coronary sinus may require use of long sheath or atrial looping to direct the lead toward the ventricle. Venous occlusion, particularly in patients with prior leads and CHD, is also important to consider. In patients with suspected occlusion, venography to ensure vein patency prior to pocket creation is recommended.

Though epicardial ICD implantation is preferred in the pediatric patient after a Fontan repair, endocardial ventricular lead placement has been described via fenestrations or atriotomies. Little exists in the literature describing placement and efficacy of ICD units in patients with Fontan physiology.

The atrial switch operation presents a unique subset of challenges in ICD implantation. Endovascular stent placement to circumvent baffle obstruction may be required for endocardial access. Caution must be taken when manipulating the ICD lead in the morphologic left ventricle, which tends to be thinner and less trabeculated and then the right ventricle. Locating viable atrial tissue for adequate sensing and SVT discrimination can be difficult due to extensive scarring that occurs during the atrial switch operation. Viable regions within the atrium are the left atrial appendage, left anterior atrial roof or SVC/RA junction. Left atrial appendage pacing can cause phrenic stimulation and therefore may not be practical in some patients (more of a consideration in dual chamber devices).

As with pacemaker implantation, intracardiac shunts are a contraindication to transvenous placement though trivial left-to-right shunts are likely not high risk for thromboembolic events. Otherwise, shunt closure or epicardial device placement is warranted.

Often most challenging is establishing an adequate defibrillation vector. This issue will be discussed below.

Long-Term Complications

Acute complications of device implantation are similar to that of pacemakers and include infection, bleeding, lead placement issues including dislodgement (Fig. 107.11), and vascular damage. In a study from the pediatric ICD registry of 443 patients, 64 acute (<30 days) complications occurred in 55 patients, the most common of which were lead dislodgement and inability to defibrillate at an acceptable defibrillation threshold testing (DFT). Chronic complications in this study included conductor/insulator breech and changes in electrical properties and occurred in 116 patients.

Issues related to growth, development, and trauma are unique among the pediatric population. ICD malfunctions related to these issues can be quite debilitating and potentially fatal. Inappropriate shocks have become the most burdensome complication of ICD implantation in pediatrics and, unfortunately, occurs much more commonly than in the adult population. Of 409 patients with ICDs, 87 (21%) received an inappropriate shock with a mean of six shocks per patient compared with appropriate shocks in 105 patients (26%). Common etiologies for inappropriate shocks in the study group included lead failure, inability to distinguish sinus tachycardia or SVT, and/or oversensing of T wave. Clinical interventions such as ablation of SVT substrates, adjusted sensing to eliminate T wave oversensing, and initiating anti-arrhythmic medications can reduce

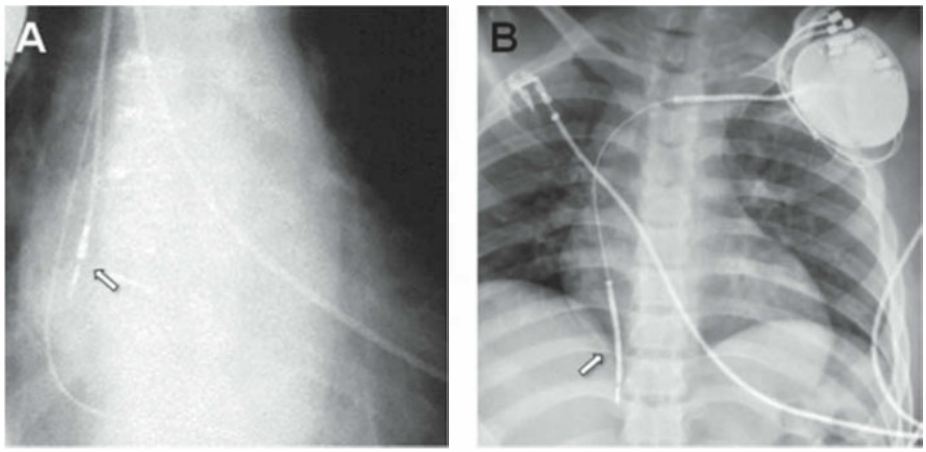


Fig. 107.11. Examples of atrial (**A**) and ventricular lead (**B**) dislodgement seen on X-ray—AP views.

the rate of inappropriate shocks after the etiology for the event is established. Behavioral disorders, anxiety, device dependence, and social withdrawal have been reported in patients with ICDs. Appropriate counseling before and after device implantation with the assistance of psychiatric professionals is warranted in some cases.

Fortunately, the mortality rates in pediatric patients with ICDs are much lower than adults. With a mean follow-up of 7.5 years, arrhythmia-related mortality occurred in only 1% of patients in the pediatric ICD registry. Moreover, the high rate of appropriate shocks justifies implantation in selected at-risk patients as described in previous sections.

Defibrillation Threshold Testing

The objective of DFT is to determine the lowest amount of energy required to convert VF into sinus rhythm as a surrogate for establishing device reliability and margin of safety. DFT was once an obligatory component of ICD implantation and considered standard of care in the early implant years. Recently, this practice has come into question as adult data from the SCD-HeFT (SCD in heart failure trial) has shown no significant benefit in terms of survival in a select group of patients receiving ICD therapy as primary prevention for SCD. In this study, shock outputs were programmed at 30 Joules regardless of DFT values. No differences were found in success of first shock regardless of DFT results. The authors are quick to note that these results should not be extrapolated to other patient groups, particularly those that are higher risk for SCD. The safety and clinical utility of DFT are also debated. Viskin and colleagues thoroughly describe the shortcomings of DFT. Specifically, DFT employs induction and detection of VF, which may not be the clinically relevant arrhythmia in the ambulatory setting. Furthermore, there is no guarantee that external defibrillation as a "bail out" for ICD refractory VF will be successful and death has been reported as a result.

Though the low-risk adult population may not benefit from DFT, most pediatric electrophysiologists perform DFT at implant. Contraindications to DFT are important to recognize. The pediatric relevant contraindications are as follows: (1) intracardiac thrombus, (2) severe aortic stenosis, (3) inadequate sedation, and (4) hemodynamic instability. In those high-risk patients, particularly patients with HCM, restrictive cardiomyopathy or severe dilated cardiomyopathy, surgical support, and ECMO backup are crucial in preventing catastrophic outcomes during DFT.

Routine, follow-up DFT has also been employed in the past but not proven to be useful. With the concerns mentioned in device longevity (such as increasing charge times) in the pediatric population, changes in DFT over time may be clinically significant and require attention. In the largest pediatric study assessing utility of follow-up DFT, the authors concluded that routine testing is not clinically useful though should be considered if clinical changes occur. Currently, no guidelines regarding DFT in pediatrics exist.

ICD Follow-up, Recalls, and Device Malfunction

ICD Follow-up

Due to the high rate of device complications in children, routine follow-up serves the purpose of optimizing device function and ensuring patient safety. Most centers perform outpatient device interrogation every 6 months. These visits are focused on assessing patient symptoms (palpitations, dizziness, syncope, device discharges) and evaluating lead parameters, battery longevity, and battery life (charge time). Depending on the device parameters, intracardiac electrograms may be available to review for high rate episodes. If sinus tachycardia is noted close to the defibrillation zone, adjustments should be made accordingly to avoid inappropriate shocks. All device discharges are also available for interrogation allowing the clinician to determine the rhythm inciting the discharge event.

Frequently, lead fractures can be identified prior to inappropriate shocks highlighting the utility of routine interrogations. A sudden change in lead impedance, accompanied by "lead noise," should prompt further investigation and possible lead extraction and replacement. Low battery life or long charge times suggest the need for generator replacement.

Advanced transtelephonic systems are available in most current devices allowing remote assessment of device parameters. The reports provide pacing and voltage impedance, arrhythmia detection and therapy logs, pacing and sensing thresholds, and programmable alerts (e.g., lead noise). Most clinicians perform remote monitoring approximately every 3 months, or sooner depending on device status.

Device Recalls and Device Malfunction

The implications of device and lead malfunction are severe, particularly in our patient population. The decision to explant an ICD system must be carefully discussed. Factors affecting this decision include presumed risk of an adverse event related to the malfunction and the presumed risk of ICD revision. Certainly, patients that are at high risk for SCD with a high probability of clinically significant consequences from device malfunction should undergo revision.

In a study performed by members of the Pediatric and Congenital Electrophysiology Society, 58 of 233 (25%) ICD patients were identified as having device advisory or recalls in a 5-year period. Thirteen of these patients underwent explantation with a mean time from implant of 3.1 years. None of these patients were found to have defective devices upon investigation by the manufacturer, which highlights the importance of risk assessment in these scenarios.

From the initial commercial availability in 2004 to the lead recall in 2007, 268,000 Sprint Fidelis leads were implanted in patients worldwide. Manufacture concerns regarding high lead fracture rate were publicized and clinicians were left with a difficult decision, particularly the pediatric electrophysiologist as younger patients had a much higher incidence of lead failure. The failure rate for this lead was 2.8% per year, unlike other leads with a failure rate of approximately 0.43% per year. Though deaths were not reported as a result of Fidelis lead failure, inappropriate shocks were extremely common. As mentioned, inappropriate shocks have a pervasive psychological impact.

Special Challenges in ICD Implants

Difficult Vascular Access

Ultrasonography and/or venography are crucial in demonstrating the feasibility of access in those patients considered to be at high risk for venous occlusion. Patients with complex CHD, multiple cardiac catheterizations, or a history of venous thrombosis who are being considered for ICD placement should undergo vascular studies prior to pocket formation. As with pacemaker patients, the epicardial approach is preferred if vascular access precludes transvenous placement.

High Defibrillation Threshold Testings

DFT is influenced by many factors including electrolyte balance, sympathetic tone, and antiarrhythmic drugs. A 10-Joule safety

margin has been widely accepted as standard practice. High DFTs occur in 5% to 6% of patients and are associated with LV dilation and medication use (see below). Presumably, these patients are not adequately protected and are at higher risk of SCD. Several management options are available and all aim to provide the highest discharge output through the most efficient shocking vector.

Prior to exchanging or adding hardware, reversible causes of high DFTs should be investigated. For instance, air is a poor electrical conductor and pneumothoraces must be excluded as a culprit. Medications particularly mexiletine, carvedilol, sildenafil, and amiodarone have all been shown to increase DFT. Perhaps the least invasive approach to circumvent a high DFT situation is to replace the generator with a higher output device. Higher output devices tend to be larger and may not be an option in a smaller patient. An SQ array can be introduced to enhance the shocking vector as well. Some have advocated using additional coils (e.g., SVC coil). Addition of sotalol has been shown to lower DFTs and is a reasonable option in selected patients with a normal baseline QT_c interval.

Single Ventricle Patients

Risk assessment in patients with single ventricle physiology is yet to be clearly defined. Khairy et al. evaluated 261 Fontan patients of which 76 died with a substantial portion experiencing SCD (7 patients, 9.2%). Due to small numbers, no predictors of SCD were proposed in this study. Heart failure from severe ventricular dysfunction predicts a high rate of mortality and is likely not ameliorated by ICD therapy. Typically, patients with severe cardiac dysfunction are better served by cardiac transplantation. Certainly, the risk of anesthesia and surgical ICD placement must be considered and discussed with the family after other interventions are thoroughly evaluated.

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EDITOR'S COMMENTS

The authors have nicely summarized the indications for pacemaker and resynchronization and ICD therapy in children. A particularly difficult group of patients are those with heterotaxy syndrome with hydrops and complete heart block with fetal compromise. It is interesting that despite aggressive pacemaker implantation at birth, these patients do not seem to tolerate pacing well with poor cardiac output and a high mortality rate despite what would appear to be good AV sequential pacing or reestablishment of AV synchrony with a higher ventricular rate. In some cases, the patients may be best served by ventricular pacing without atrial synchronization. The mechanism by which the increased heart rates do not result in improvement in clinical outcome and cardiac output in these patients has not been well elucidated, and they continue to be an extraordinarily high-risk population with poor results.

Postoperative complete AV block is often transient as noted in the chapter. It is preferable to wait 7 to 10 days after initial cardiac surgery with complete AV block prior to committing the patient to a permanently implanted pacemaker system. The significant majority of patients will return to normal sinus rhythm in this timeframe. However, as noted, since AV block can reoccur even after the recurrence of sinus rhythm in some patients, the patient who has a poor escape rate and relatively dense AV block who then recovers sinus node function late at 7 to 10 days after initial surgery may be best served in some cases by placement of a back up VVI generator to protect them from possible late recurrence of lack of AV conduction.

An additional issue with pacemaker implantation in postoperative complete heart block is the fact that although pacemakers are generally implanted after 7 to 10 days of lack of sinus rhythm, after placement of a pacing system some patients will revert back to sinus rhythm over the next several months to years. Thus, it is imperative to follow these patients and consider whether or not they continue to need active pacing systems over time. In some cases, if sinus rhythm can be demonstrated and is stable on Holter monitoring or long-term follow up through the pacing device, it may be possible to explant the device with an assurance that sinus rhythm will be maintained.

As noted by the authors, the relative efficacy of resynchronization pacing therapy in children with congenital heart disease is not well established. There are isolated reports of significant improvement in exercise performance after resynchronization therapy in some patients, including those with single ventricle physiology. Large series, however, have not been accumulated and therefore it is difficult to know when to recommend this type of therapy in the pediatric patient population. In addition, the variations in venous anatomy and ventricular morphology make transvenous implantation of resynchronization leads more difficult in these patients and epicardial leads may be necessary. Deciding on location for epicardial placement of pacing leads can be difficult and previous surgery can make exposure quite challenging.

The indications for placement of ICD devices in the pediatric population are well summarized by the chapter. It is important to emphasize as the authors have done in this chapter that ICD placement carries a significant morbidity due to the psychological impact of inappropriate counter shocks not to mention the psychological issues of the potential for counter shocking at any time. It would seem appropriate that very stringent indications be followed for placement of these devices in the pediatric population due to the high incidence of complications, the relatively short durability of the leads and pacing systems, and the relatively large sizes of the devices which have cosmetic implications in small children.

TLS

108 Conjoined Twins and Ectopia Cordis

Joseph J. Amato

Birth of conjoined twins (twins whose bodies are connected at some level) is indeed very rare and estimated by various authors as 1 in 50,000 to 100,000 births. A majority (40% to 60%) are stillborn and 35% do not or cannot survive after birth. They are associated with a high incidence of maternal polyhydramnios, prematurity, and low birth weight. They are always the same sex and approximately 70% are female. Isomerism is common: twins are born with two right sides, two left sides, in mirror images of normal anatomy, or a mixture of the two with shared organs.

HISTORY

There are numerous historical accounts of the existence of conjoined twins. The earliest recorded history was found in ceramics of ancient Peru in 300 AD.

History then records two Arabian brothers who were joined with ones leg to the others head. They were separated with a sword in 750 AD.

The most famous report described "Siamese Twins" which gave a new popular synonym that is often substituted for "Conjoined Twins." These were the historical twins named Eng and Chang who were born in Siam in 1811 (now Thailand but really they were Chinese) sharing a common liver and joined at the abdominal area. King Rama ordered them to death because he believed that they represented a sign of an evil omen. Later, this decree was withdrawn. Interestingly, they joined the circus, settled in America, married two sisters, had 22 children, became successful businessmen and ranchers, and settled in Wilkes County, NC. In January 1874, Eng woke up to find Chang dead; thus, both died at the age of 63. The social element of this subject of marriage to two separate sisters, a "bed for four," and after a severe argument between the sisters the creation of two separate "homesteads," created a fascinating account. Conjoined twins are not limited to any racial or ethnic group.

Also famous were the Biddenden Maids born together in 1100 and who died together in 1134.

There appeared to be an epidemic of conjoined twinning in South Africa and in Sweden; this cluster of births has, however, been disputed.

There are also published accounts of conjoined twins occurring in cattle, rabbits, cats, hamsters, pigs, rats, and mouse.

MECHANISM OF CONJOINED TWINNING

There are two theories of the mechanism of conjoined twinning (which are incompletely understood and are contradictory). One theory is called "fission" in which there is an incomplete and asymmetrical partial splitting of a single embryonic fertilized egg. The second theory is called "fusion" in which the fertilized egg was destined to become completely separate twins but then stem cells search for like stem cells on the other twin (due to their close proximity) then fuse together. One author states that the "fission" theory is more widely acceptable; however, now the "fusion" theory is stated to look most probable. If the splitting or rejoining of the fertilized egg occurs after 12 days with either theory, the result will be a conjoined twin.

ETIOLOGY OF CONJOINED TWINS

Conjoined twins are monozygotic (one egg) and are monoaminiotic and monochorionic. The one amnion is the inner layer that holds the embryo and later the fetus within the amniotic fluid. The chorion is the inner membrane that contains the embryo and part of the placenta. It is known that twinning may tend to be prevalent in some families.

While the etiology of either fission or fusion may be disputed, various teratogenic agents may influence the incidence of conjoined twins. These may include:

(a) The use of gonadotropins, which are fertility drugs for women and hormonal imbalance in men.
(b) Experimental studies in animals demonstrate that general and local anesthetics, acetaldehyde (a metabolite of ethanol), and some tranquilizers may induce conjoint twinning.
(c) Another cause has been shown to be lack of calcium and magnesium salts.
(d) Seven cases (two published) have been reported in which conjoined twinning occurred with the use of clomiphene for induction of ovulation.
(e) Maternal exposure to valproic acid has been reported when used as an anticonvulsant or with the use of valerian, a common mood-stabilizing agent.
(f) Fungistatic agents as griseofulvin (used as a fungicide) also has been associated with conjoined twins.
(g) Agent Orange and other herbicides. There has been a marked increase in conjoined twins in Vietnam due to Agent Orange and other herbicides.

CLASSIFICATION OF CONJOINED TWINS

A general nomenclature has been established for all types of conjoined twins. Twins are classified according to the common area of the body onto which their bodies are joined and the ending "pagus" from the ancient Greek language is added which means "fixed."

Spencer provides a classification of eight different types of conjoined twins: parapagus (26%), thoracopagus (25%), omphalopagus (17%), cephalopagus (11%), ischiopagus (9%), pygopagus (6%), craniopagus (5%), and rachipagus (2%). However, this classification is difficult to follow for the cardiothoracic surgeon because thoracopagus and omphalopagus often coexist. Several alternative classifications are more useful for cardiothoracic surgeons. In one classification, the types in alphabetical order are described as anterior (front), posterior (back), lateral (side), and ventral (area of abdomen).

This classification specifies the types of twins, which either have two hearts, one

shared heart, and a possible involvement of the heart in these infants. When indicated there is involvement of the heart in most cases and possible involvement in some cases.

I. Ventral union (referring to the front of the body, faces, four ears, and two bodies)
   (a) *Cephalopagus:* Twins joined at the upper part of the body with one head. They are joined by one face with two bodies. There is one face with four ears. These types are extremely rare and do not survive (Fig. 108.1).
   (b) *Cephalothoracopagus (cephalothoracopagus syncephalus, epholothoracopagus, craniothoracopagus):* Joined at head and thorax. There are two faces, one brain, one head, usually share one heart and fused gastrointestinal tracts (Fig. 108.2).
   (c) *Thoracopagus:* Joined at upper thorax to umbilicus. Shared heart in 90% and 75% have a joined heart which cannot be separated, four arms, four legs, and two pelvises (Fig. 108.3).
   (d) *Omphalopagus:* Joined in the area of the umbilicus, sometimes thorax, however, not affecting the heart. Two pelvises, four arms, and four legs (Fig. 108.4).
   (e) *Thoraco-omphalopagus:* A combination of thoracopagus and omphalopagus. Joined at upper thorax to the umbilicus and usually share a heart. Ninety percent have a common pericardium and 75% have a shared heart. They have four arms and legs. Xiphoomphalopagus look similar but their hearts are normal (Fig. 108.5).
   (f) *Xiphopagus (a subset of omphalopagus):* Joined at the xiphoid from lower sternum (gladiolus) to the umbilicus (Fig. 108.6).
   (g) *Ischiopagus:* Twins joined at the level of the umbilicus end to end with the spine. There are four arms with various combinations of legs. Of the above types, there may be only two legs present (ischiopagus dipus), three legs present (ischiopagus tripus), or four legs present (ischiopagus tetrapus/quadripus).
   (h) *Omphalo-ischiopagus:* A combination of omphalopagus and ischiopagus are similar to ischiopagus twins facing each other. There can be cases of thoraco-omphaloischiopagus and xipho-omphaloischiopagus twins who are joined facing each other but are joined at the chest.

II. Lateral union (referring to twins joined side by side with shared umbilicus, abdomen, and pelvis. These rarely involve the heart.
   (a) *Parapagus:* Twins joined at the pelvis with one symphysis pubis and one or two sacrums (Fig. 108.7).
   (b) *Dithoracic parapagus:* Twins joined at the abdomen and pelvis but not the thorax (Fig. 108.8).
   (c) *Dicephalic parapagus (dicephalus):* Twins joined at the abdomen, pelvis, and thorax. All of the above may have two arms (dibrachius), three arms (tribrachius), and four arms (tetrabrachius).

III. Dorsal union (referring to twins joined at the dorsal aspect (at the back). In these twins, there is no involvement of the thorax and abdomen.
   (a) *Craniopagus:* Twins joined at the skull except for the face.
   (b) *Pygopagus (pyopagus or illeopagus):* Twins joined at the sacrococcygeal, perineal, and occasionally the sacrum.
   (c) *Rachipagus:* Twins fused dorsally above the sacrum and the lumbar spine. This type is very rare. The above classification constitutes one of many and excludes many other rare types. Two of these types are:
      - Parasitic twins (asymmetrical, unequal) in which if one dies or



Fig. 108.1. Cephalopagus classification.



Fig. 108.2. Cephalothoracopagus classification.

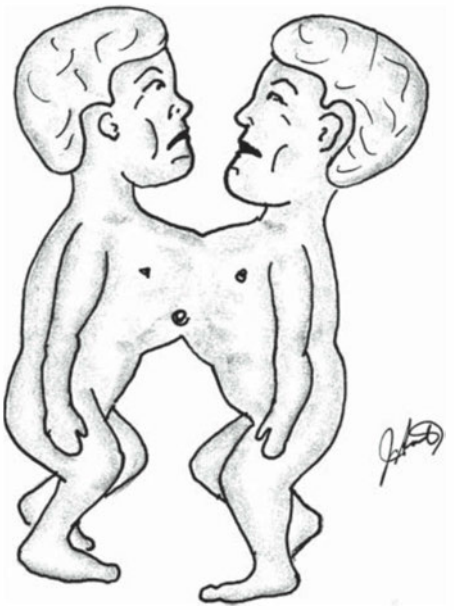


Fig. 108.3. Thoracopagus classification.

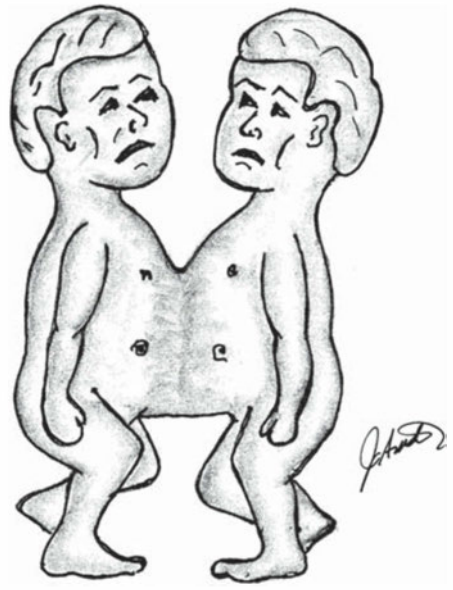


Fig. 108.4. Omphalopagus classification.

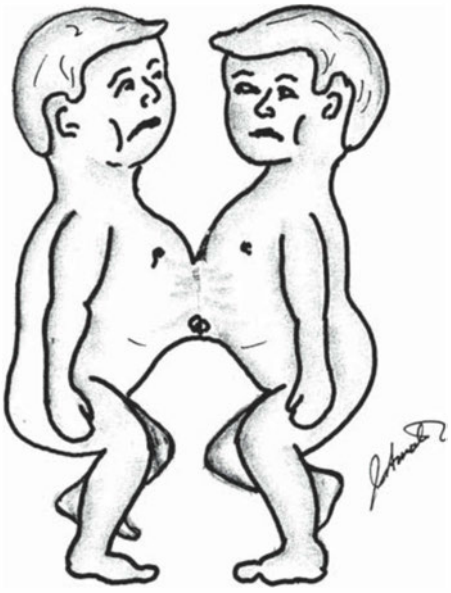


Fig. 108.5. Thoraco-omphalopagus classification.

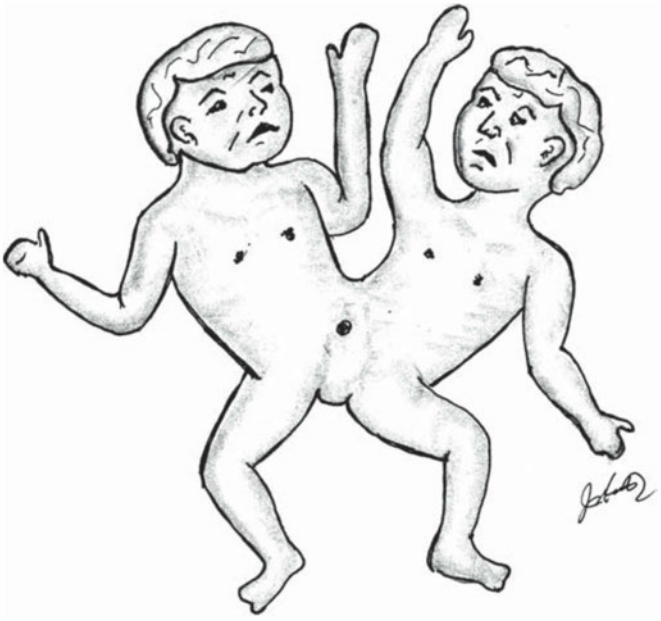


Fig. 108.7. Parapagus classification.

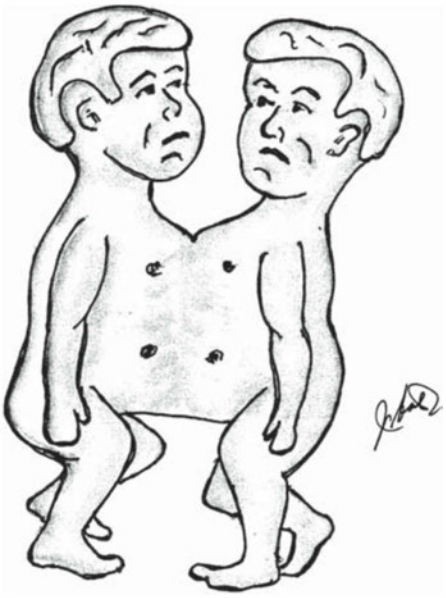


Fig. 108.6. Xiphopagus classification.

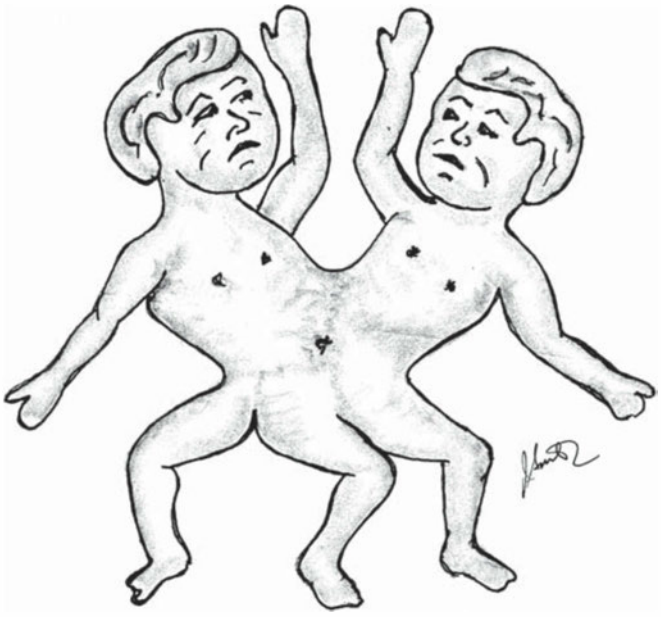


Fig. 108.8. Dithoracic parapagus classification.

receives inadequate nutrition while in the uterus then one embryo maintains dominant development at the expense of the other.

- A cardiac twin is an incomplete twin without a heart that grows separately and receives nourishment from its normal twin.

A second simpler classification listing the frequency of conjoint patterns is as follows:

(1) Thoraco-omphalopagus (twins joined at the chest, abdomen, or both)—74%.
(2) Thoracopagus or xiphopagus (joined at the chest)—40%.
(3) Omphalopagus (joined at the abdomen)—34%.
(4) Pyopagus (joined at the buttocks)—18%.
(5) Ischiopagus (joined at the ischium)—6%.
(6) Craniopagus (joined at the head)—2%.

CARDIAC INVOLVEMENT IN CONJOINED TWINS

For the purpose of this chapter, while other types of twins are possible, our concern is twins with involvement of the cardiovascular structures. These twins are all joined primarily in the thoracic area. There may be either a single heart or partial duplications of cardiac and vascular elements. There can also be two hearts with extensive sharing of the vascular elements. The sternum is either partially or totally absent in all cases, the pericardium in 90% and the hearts fused in 75%.

The other organs that can be involved include the unusual saddle-shaped liver, a joined common duct (while the gall bladders and hepatic ducts might be separate), and hepatic veins either separate or shared. There can be persistent left superior vena cavae, which drain into separate atria. Also obviously, there is a great variation of gastrointestinal tracts. Obviously, those

with two hearts, each with one heart in a thoracic cavity, have a better chance of survival.

One of the most difficult tasks is to present a classification of the morphology and anatomical arrangement of the heart or hearts in these twins. A simple general classification has been presented by Ambar:

(1) Group A: Separate hearts, separate pericardium.
(2) Group B: Separate hearts, common pericardium.
(3) Group C: Fused atria, separate ventricles.
(4) Group D: Atrial and ventricular fusion.

With the above cardiac abnormalities, one must consider the other associated various organ systems, with either multiple or incomplete formation of the liver, intestines, and urinary systems.

The most common atrial malformations include a single atrium with atrial septal defects. The most common ventricular malformations are a single ventricle and large ventricular defects. Combined pulmonary arteries and aorta have not been reported. Of the others, any combinations of basic defects have been described: D-transposition, interrupted aortic arch, and pulmonary stenosis and atresia.

Successful separation of thoracopagus twins in cases of conjoined heart is difficult. There has been only a single report of successful separation of a conjoined heart and in this instance it was conjoined atria. In a series of 13 surgeries on conjoined twins in The Children's Hospital of Philadelphia, PA none of the patients with complex fused hearts survived even after sacrificing one of the twins. Cases of thoracopagus conjoined twins with normal separate hearts, common pericardium, anomalous systemic venous drainage, fused atria, and atrial septal defects have been successfully separated.

DIAGNOSTIC PROCEDURES IN CONJOINT TWINS

There are two phases of diagnosing the presence of conjoined twins: early in the prenatal period and then following delivery. Evaluation of the extent of pathological patterns of both the cardiovascular system and other involved organs that can potentially affect the question of infant separation and survival of both twins is necessary. For the fetuses, prenatal ultrasound has been used as early as 1960 and with more modern techniques, the use of transvaginal ultrasound may give the diagnosis as early as 7 to 14 weeks into the pregnancy.

Prenatal magnetic resonance imaging has better established the intricate shared multiple organs of the fetus. Magnetic resonance imaging has the advantage of being capable in twins to determine the systems of blood flow and the production of three-dimensional and multislice imaging, which can clarify the venous and atrial anatomy.

Early imagery allows proper counseling with regard to prognosis, management, and if indicated, termination of pregnancy. One interesting case diagnosed with a fetal echocardiogram was the condition never reported prior to the year 2000 of a single heart in conjoined twins diagnosed by the pumping of blood though the cord from the one infants single heart to create circulation in the noncardiac infant. The delivery of the two and the cutting of the cord would cause both infants to die. Surgery was performed by a caesarian section. Separation of the infants caused immediate death of the infant without the heart. At separation, the heart did not fit into the chest and protruded in a similar manner to an ectopia cordis. A rib cage was created from the chest wall from the twins tissues. The second infant was alive at 14 months at the time of this report.

ANESTHETIC MANAGEMENT OF CONJOINED TWINS

Perioperative management includes monitoring, airway and fluid management, ventilator support, and anesthetic medications. There are usually two teams of anesthesiologists. There are obviously difficulties in the positioning of the twins; body alignment is of utmost importance because of the ability to intubate because of the various anatomic positions and single or multiple airways, and only the use of airway masks if intubation is impossible. The management of the potential dead twin upon separation is too variable to explain. In a rare case in which one twin had a rudimentary heart and there was severe cardiac dysfunction in the possible normal twin, a method of intubation (an EXIT procedure [ex utero intrapartum treatment]) is performed immediately after the extraction of the fetus from the womb and separation of the dead twin. Prior to cutting of the umbilical cord, the surviving twin is either intubated or tracheostomy is performed.

In many of the twins, there is also marked difference in the twin's circulatory systems, which present with different arterial blood pressures and atrial oxygen saturations.

SURGICAL SEPARATION

The surgical separation of conjoined twins is a delicate and risky procedure, requiring extreme precision and care. Decision to separate twins is a serious one. In 1689, Konig performed the first recorded separation of twins joined at the umbilicus with a ligature. There have been approximately 250 successful operations in which either one or both twins have survived. Of 47 pairs of surgically separated thoracopagus (omphagothoracopagus) twins with completely separate hearts only 42 survived (70%), and in 5 patients with only atrial connections, only 1 patient survived. In nine patients with both atrial and ventricular connections, there were no survivals. The site of the anatomical pericardial and/or cardiac conjoined areas is the most important consideration of a successful outcome.

Once separation is successfully achieved, tight closure of the pericardium and large abdominal and thoracic defects should be avoided. Silastic sheeting or the author's preference of a thin polytetrafluoroethylene (PTFE) membrane should be used. Skin coverage of the separated areas is necessary. Skin flaps, as in ectopia cordis, can be used. In one case, the chest wall of the nonsurviving twin was used. Other surgeons have used a resin called "Cranioplastic" (commonly used by neurosurgeons, to create a "resin plate"). In other cases, siliconized rubber sheets, acrylic materials, and another resin mesh made of polypropylene mesh have been utilized. In the authors surgical management of ectopia cordis, a Gore-Tex® material (PTFE by Gore) was used to stabilize the chest, which was then covered by skin. This can be accomplished by raising skin flaps or use of the expired twins' redundant skin, or use of tissue expanders if separation can be delayed for a few months.

ADDITIONAL CONSIDERATIONS FOR CONJOINED TWINS

Surgical separation of conjoined twins that results in the death of one of the twins raises complex legal, moral, ethical, religious, and financial issues.

Conjoined twins raise the ethical issue of "Is it justified to sacrifice one life to save another." Ethical issues arise for each of the twins in which the following apply:

(a) There is a law of conduct, namely, "Thou shall not kill."
(b) Natural law demands us not to harm anyone's health, liberty, or possessions.
(c) The medical staff understands that life is sacred where every life is equally valuable. The problem lies in analyzing each of the twin's different anatomies and chances of survival.

A suggested approach is as follows: (a) the parents meet the team that is to care for the children. (b) Confidentiality is maintained. (c) Parents speak with their religious advisors. (d) The public relations department maintains a level of guarding the interests of the children and family. (e) Legal advisors consult with the Law Courts and protect the surgeons from accusation of wrongful acts, which might lead to prosecution.

CONGENITAL WALL DEFECTS (ECTOPIA CORDIS)

Ectopia cordis is classified within the broad spectrum of congenital chest wall deformities, which include (1) pectus excavatum; (2) pectus carinatum; (3) Poland's syndrome; (4) sternal cleft defects that include ectopia cordis; and (5) other miscellaneous conditions. Thus, reviewing ectopia cordis is necessary to understand sternal cleft defects and separate ectopia cordis from the other isolated sternal cleft anomalies. The embryological failure of the fusion of the sternal bars within this category create a complete or partial separation of the sternum.

The three categories of chest wall defects include:

Isolated sternal clefts: These occur within the upper sternum and there is normal skin coverage; the heart is within the sternum; the pericardium is intact; the diaphragm is normal; and omphaloceles do not occur. These patients seldom have intrinsic congenital heart disease.

Ectopia cordis: The various forms of ectopia cordis are often associated with other midline defects, which include intrinsic cardiac defects, complete or partial absence of pericardium, omphalocele, and ventral diaphragmatic hernia.

DEFINITION OF ECTOPIA CORDIS

Dr. Anderson emphasizes that the term ectopia, which is derived from the Greek word "ektopos," classically means "away from a place" which could mean the displacement of the heart somewhere within the thorax, but this classical term is now commonly used to refer to the heart either partially or completely outside of the thorax.

Descriptions of ectopia cordis also vary from a complete absence of the sternum to a completely intact sternum in which the heart is located in the cervical area or dropped below the diaphragm.

The term ectopia cordis was first used and independently reported in 1706 by Haller and Martinez. Numerous cases were subsequently reported. Each of these cases had various descriptions of the ectopic heart. In Barardo, references to cases beginning with Martinez in 1706 to 1884 are listed with many giving to the location and position of the heart. Ravitch described a more extensive history of early cases.

The number of these cases increased to the point that many initial classifications were either discarded or added on to. These changes in classification occurred as an attempt to include all forms of ectopia cordis. Even today there is disagreement concerning the multiple classifications.

INCIDENCE

The incidence reported by Abbott in 1936 was 0.8% of all cardiac malformations, which is about 0.004% to 0.005% of the entire population.

However, in 1988, this very rare effect was reported to occur with an estimated prevalence of 0.079/10,000 births (5.5 to 7.9 million live births) and may occur more frequently in females.

The most recent statistics quoted in 1995 and 1996 are 5.5 to 7.9 per 1 million live births.

In evaluating this figure, one must account for cases remaining unreported. One must also account for the varying definitions of ectopia cordis and for incidence figures that include varying numbers of partial ectopia cordis. The purpose of this article is to discuss the *true* or *thoracic* ectopia cordis and its surgical correction.

Helen Taussig stated "little wonder that such a severe anomaly, which must completely alter the forces of stress and strain, is associated with some gross and bizarre cardiac anomaly. It is interesting, however, that this entity is not so rare among the lower animals, especially birds and it has also been described in cattle.

APPLICATION AND USE OF A PROCRUSTEAN CLASSIFICATION

The general classification of the anatomical types has now been well established.

The problem lies in the definition of true "ectopia cordis." Anderson describes "Ectopia Cordis" as a "Procrustean" term. Procrustes was a mythological Greek God who offered hospitality to passing strangers, who were invited in for a pleasant meal and a night's rest in his very special bed. Unknown to the guest was that he has to fit exactly into the bed, stretching him on the rack if he was too short for the bed and chopping off his legs if he was too long. The difficulty is that many definitions include sternal clefts, Pentalogy of Cantrell, and other lower midline sternal defects in the general classifications of "ectopia cordis" in addition to the much more rare and difficult group with true exteriorization of the heart.

Garson writes: "About two hundred cases of Ectopia Cordis have now been reported, though many have had only a part of the heart exposed, and others have been simply cases of bifid sternum, the heart remaining within the thorax, albeit just beneath the skin. The latter cases should not be considered as a Ectopia Cordis." Morales and his associates in 2000 reported four successful cases, including one of ectopia cordis and three of Cantrell's Pentalogy. The first case described as a "thoracic ectopia cordis" is shown in the figure provided as a partial form of this entity with a small amount of ventricle and diverticulum exposed. The other three were described as "Cantrell's Pentalogy," in which the definition includes "the extrathoracic heart is covered by body wall, having a covering of either membrane or skin, therefore adding to the confusing description of ectopia cordis."

In a 1996 article on ectopia cordis (in which all patients were listed as "Ectopia Cordis"), the authors state that of the 15 patients reported, there were only 5 survivors. They explain that Shamberger in 1990 presented these same patients; however, he did not discuss the details of the clinical course, cardiovascular anatomy, hemodynamics, and cardiac operations of these 15 patients in this earlier report.

However, in reviewing this earlier report, which was cited by the previous authors, Shamberger states, "We have reviewed our experience with sternal defects. Sixteen patients with sternal defects were identified. Of the five patients with thoracic ectopia cordis with *a completely naked heart*, all were fatal. Of the eight with a form of thoracoabdominal ectopia cordis with *a covered heart*, five of these patients survived. The three patients with *a cleft sternum* all survived.

A more complete description of "ectopia cordis" should allow the subgroup analysis of sternal defects including:

(1) Anatomical position of the heart.
(2) Partial or complete forms of the ectopic heart.
(3) The heart covered or uncovered by pericardium and/or skin.
(4) The time of intervention (minutes, hours, days) after the infants birth.
(5) Associated intracardiac or vascular anomalies.

(6) Achieved surgical result (exterior or interior placement of the heart within the chest wall).
(7) Time of stages of repair.
(8) Subsequent surgical interventions.
(9) Current status of the patient.

PREFACE ON THE HISTORICAL ASPECT OF ECTOPIA CORDIS

Descriptive History of Ectopia Cordis

The earliest recorded report of a child with ectopia cordis was that of Neil Stensen in 1671. The name "Stensen" was translated by many as "Nicholas Steno" (1638 to 1686).

Stenson was a famous Danish physician and anatomist who first described an "embryo monster" in an abbreviated translation by Willius. Stensen and he observed the following anatomy: "the sternum was split and the heart, liver, with most of the intestine and right kidney have passed through the split in the sternum and thus were uncovered." The anatomy of the heart clearly described what was to be a form of "Tetralogy of Fallot."

EMBRYOLOGY IN THE DEVELOPMENT OF THE HEART AND CHEST WALL

When the embryo reaches its third week of life, it can no longer rely on nutrient and gas exchanges by diffusion. The cardiovascular system begins its formation within the developing embryo and the primary germ layers of ectoderm, mesoderm, and endoderm begin to form. The cellular sponge-like layer of mesoderm splits into splanchnic (visceral) and somatic (parietal) sheets (Fig. 108.9) (cross section of heart). The coelum divides into spaces for the heart and lungs. The heart, which is forming at the same time as the surrounding sternal element, originates from the visceral (splanchnic) layer including the diaphragm, pericardium, and epimyocardium. This early formation of the heart originates in a very cephalad position within the cervical region but reaches its position by flexing ventrically at about 2 to 3 weeks of development. The sternum arises from the parietal (somatic) layer as a pair of mesenchymal bars at about 6 weeks of gestation (Fig. 108.10). These paired mesenchymal bars, which have no connection with the lateral forming ribs, become fused at about 9 or 10 weeks of age. Above these two bars (cranially), another mass of mesoderm tissue in the midline, the future (primordial) manubrium, also called the presternum, is separated by other mesenchymal tissue on each side that eventually becomes the articulating junctions between the manubrium and the clavicle (The red arrow in this figure demonstrates the exterioization of the heart). As the sternal bars fuse together, they separate into three sternebrae (segments): the manubrium, the body, and the xiphoid. Ossification occurs later. Concurrently, the muscle formation of the diaphragm and pericardial cavity occurs by closure of the transverse septum (Fig. 108.11).

Conditions in which failure of the two sternal islands to come together without the failure of the heart to be properly placed are known as a sternal fissure, bifid sternum, sternal cleft, or sternal "fusion defects." These usually occur without other intrinsic cardiac anomalies.

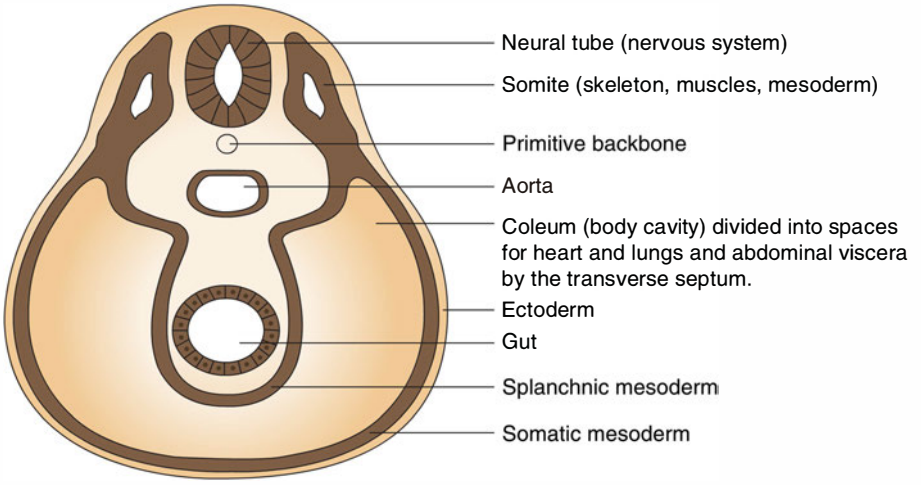


Fig. 108.9. Cross section of the heart.

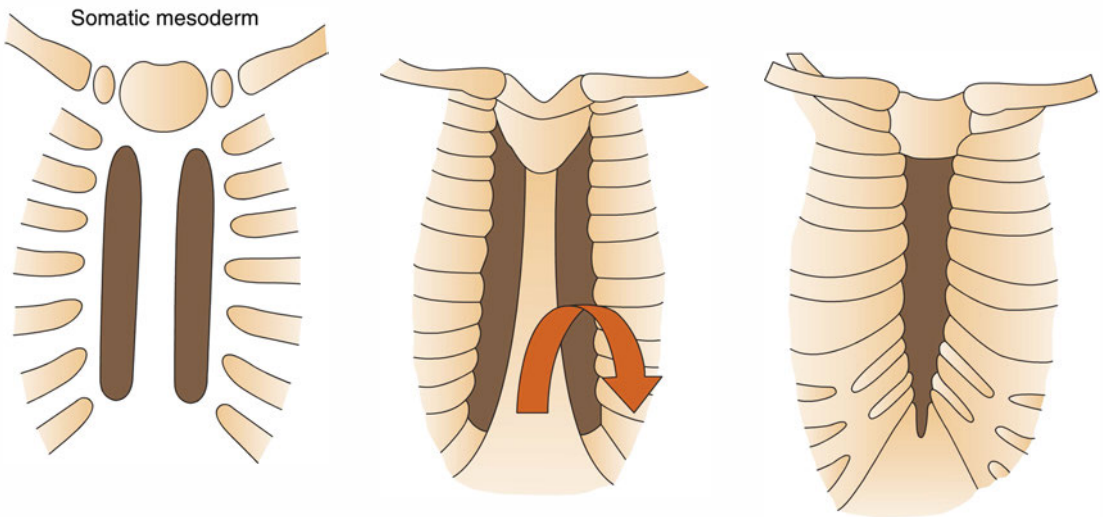


Fig. 108.10. Development of the sternum.

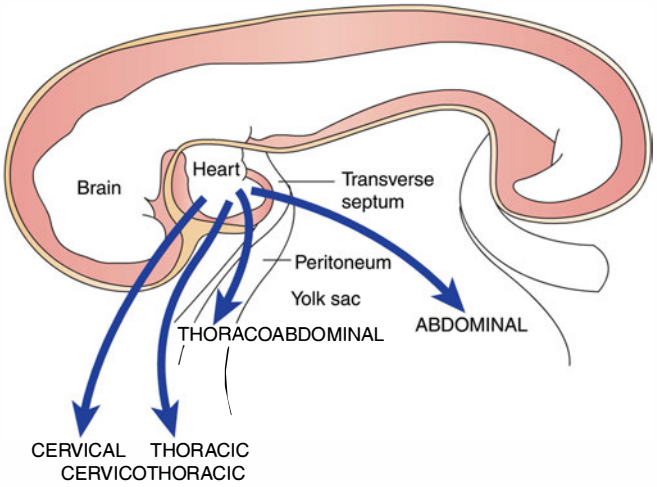


Fig. 108.11. Muscle formation of the diaphragm.

The conditions in which the heart is externally placed have been classified as the "cervical type." These occur when the heart becomes trapped above the sternal closure.

In true ectopia cordis, the failure appears to be secondary to the presence of the heart inserted primarily into this abnormal position. The other forms that will be discussed are the cervical, cervicothoracic, thoracic, thoracoabdominal, and abdominal types (Fig. 108.12).

It is extremely useful in these classifications to describe the outward (external) displacement of the heart as partial or total.

The Pentalogy of Cantrell is a thoracoabdominal type of ectopia within the classifications of the ectopic heart. Cantrell in 1958 described the syndrome, which consists of a malpositioned heart extruding partially or totally from the thorax with (1) defect in the midline supraumbilical abdominal wall (omphalocele); (2) defect in the lower sternum (sternal cleft, bifid sternum); (3) anterior diaphragmatic deficiency (diaphragmatic hernia); (4) defect of pericardium (communication with peritoneal cavity); and (5) intracardiac defects (most common are tetralogy of Fallot, diverticulum of the left ventricle, and ventricular septal defect). Many other cardiac defects have been reported.

These five conditions may not always be present in the complete form, thus should disqualify it as a true ectopia cordis. The heart may also have no intracardiac defects.

PATHOGENESIS OF ECTOPIA CORDIS

The etiology of ectopia cordis in which there is disruption of the midline fusion is unknown. Various theories have been proposed:

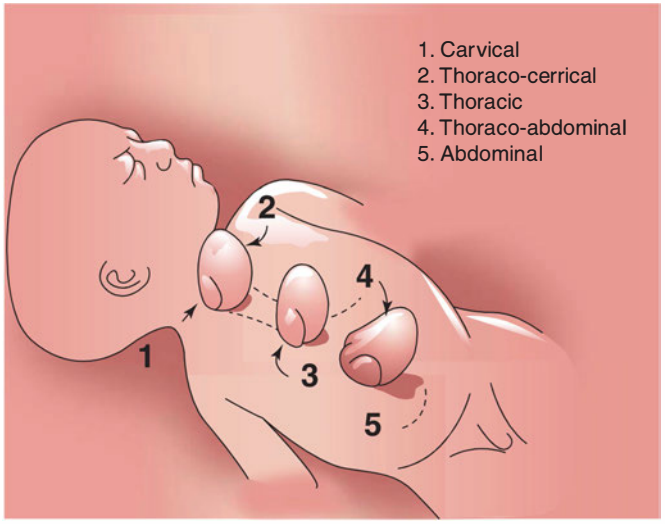


Fig. 108.12. Types of ectopia cordis.

(A) Changes within the sequence of the gestational process of the formation of the heart and thorax include the following:
  (1) Premature rupture of the fetal membrane that surrounds all other membranes.
  (2) Premature rupture of the membrane that enlarges and lies next to the umbilicus.
  (3) Abnormal formation of the coelum.

(B) Ventral body wall defects—an abnormal closing of the ventral body wall over various organs:
  (1) Upon the heart—ectopia cordis.
  (2) Upon the abdominal wall—gastroschisis in which the infant's intestines extrude from the body through a defect on one side of the umbilical cord.
  (3) Bladder exstrophy.

(C) Genetic karyotype abnormalities include trisomy 18 (Edwards syndrome), Turner's syndrome, testicular disorders, craniofacial disorders, and others.

(D) Teratogenic agents include the following:
  (1) Studies on chick embryos demonstrated that administration of hydrocortisone will cause a high incidence of ectopia cordis. Hydrocortisone injected into chicks on the fourth embryonic day caused a 84.8% incidence of ectopia cordis.
  (2) Antitumor agents such as platinum thiamine blue.
  (3) Maternal hormonal therapy.
  (4) Aminopropionitriles that are used for the removal of scar tissue and also as antirheumatic agents used in veterinary medicine.

(E) Environmental factors include the following:
  (1) Gas emissions reported in South Asia from three-wheel autorickshaws in which compressed natural gas caused production of CO_2, CO, NO, and other nonmethane volatile organic compounds. Chick embryos treated with these gases produced four different types of ectopia cordis.
  (2) Genetic karyotypes—genetic testing of the patient and parents for chromosomal variations to determine the possible causative factors should be encouraged.
  (3) Some suggest that other environmental factors such as general air pollution, smoking, alcohol, and drugs could react with genetic patterns.

CLASSIFICATION

The most complete classification, given in 1962, includes five types, which are discussed in the following sections.

Cervical (Ectopia Cordis Cervicalis)

This extremely rare lesion occurs when in the unfolding process, the heart, which initiates in the cervical area, fails to descend from the embryologic submandibular region, and the sternal bars come together leaving the heart behind. Two infants have been described to live only hours after being born. Van Praagh in 1983 dismissed this lesion as occurring only in stillborn.

Cervicothoracic (Ectopia Cordis Cervicothoracolis)

The heart is partially in the neck, with an upper sternal cleft defect. This form is extremely rare; however, Leca reported 18 cases of this type, some of whom had intact sternum.

Thoracic (Ectopia Cordis)

This may be described as the heart protruding from the defective sternum partially or completely. Although the heart completely exposed can be considered true thoracic ectopia cordis, it is difficult to know the exact incidence of this defect. It has been described as occurring in 36%, 58%, and 62% of reported cases.

Thoracoabdominal (Ectopia Cordis Thoracoabdominalis, Ectopia Cordis Ventralis, L'ectopia-Abdominale, Cantrell's Pentalogy, Cantrell's Pentad)

Cywes in 1967 believed this type not to be a true ectopia cordis. The first successful repair of "a thoracoabdominal type" was by Wieting in 1912.

It has been described as varying from an open "incomplete expression" to a severe expression in which the complete form may include at least four of the five defects described by Cantrell. In many cases, only a small portion of the heart is not covered by skin, and even Cantrell stated that the heart appears to be extrathoracic. Even though the heart appears to be extrathoracic, it is truly adjacent to other thoracic elements. One can question whether the presence of an omphalocele places the ectopic heart within the classification of thoracoabdominal rather than thoracic. Hornberger, in 1996, reported 13 cases of ectopia cordis, of which four were described as the thoracic type. Yet, three of the four had an omphalocele. We would classify these three as an "incomplete" expression of the thoracoabdominal type.

Abdominal (Ectopia Abdominalis)

In this form, the heart lies totally within the abdomen with a diaphragmatic defect. In 1809, Cullerier reported a case in which the heart was displaced along with its pericardium into the abdomen through an anterior diaphragmatic displacement into the cavity of an absent left kidney. This defect relates to the incomplete formation of the transverse septum. Although from 11% to 13% of ectopia cordis cases are reported as abdominal thoracic ectopia cordis, Van Praagh insists that the heart was not within the abdominal cavity but displaced downward in communication with the thorax and recommends that this form be deleted from the classification of ectopia cordis.

The diagnosis, if not made by fetal echocardiography, is extremely evident on inspection of the infant at birth. It is essential to determine any intracardiac defects of the infant by sterile careful epicardial ultrasound if not previously detected by prenatal ultrasound. Cardiac catheterization is not necessary for surgical correction. Correction or palliation of any major intracardiac defects is imperative prior to enclosing the heart in the body cavity or skin. Genetic karyotyping maybe performed by chorionic villus sampling for abnormal chromosomal complements; however, this has not always correlated with the diagnosis of ectopia cordis. Finally, prior to surgery, chest and abdominal radiographs should be performed to judge the volume of these cavities. Once the diagnosis of ectopia cordis has been confirmed prenatally, it is recommended if possible that a cesarean section delivery be performed.

SURGICAL CORRECTION

Successful correction must include the following:

(1) Prenatal diagnosis and delivery by cesarean section.
(2) Immediate coverage of the heart with the patient's own skin.
(3) Palliation and/or repair of any congenital defects at presentation.
(4) Placement of the heart within the thoracic cavity.
(5) Delayed repair of the thoracic cage defect.

Prenatal Diagnosis and Delivery by Cesarean Section

Vaginal delivery might prolong compression of the heart or rupture of the atrial diverticulum or omphalocele and can cause damage to the herniated abdominal viscera, if present.

Immediate Coverage of the Naked Heart with the Patient's Own Skin

As previously stated, some partial forms are adequately covered with skin. In these patients, there is adequate time to attend to the remaining factors listed below. If not, then the heart should be covered with sterile dressing after applying Betadine or other antibacterial agents to the skin.

If mobilization of the skin is not possible, rotational flaps have been used with split-thickness grafts in the absent lateral portion. Hochberg, in 1993, used Gore-Tex temporarily with tissue expanders bilaterally to stretch the tissue and then within 48 hours raised bilateral myocutaneous flaps. Seven months later, the Gore-Tex was removed (we would recommend it to be left in). The native tissue was reapproximated in the midline and skin grafts applied to the lateral chest and abdominal walls. Tissue expanders were also used in 1978 to stretch the skin prior to covering this defect. Other alloplastic materials, such as methyl methacrylate, and Marlex composite struts were used in the sternal defect and then covered with bilateral muscle flaps. Silastic membranes have also been used.

Repair of Intracardiac Defects

Only a few cases of complete intracardiac repair have been reported in the literature. These were in cases of partial ectopia cordis. A double-outlet right ventricle was repaired with a Dacron patch and the child's heart returned to the thorax. However, the child died of sepsis on the 12th postoperative day.

Hornberger in 1996 reported four infants with intracardiac defects. Two thoracoabdominal types of these had severe-to-moderate pulmonary hypoplasia. Both infants died within 48 hours. A third infant with a thoracic ectopia cordis and a tetralogy of Fallot had several attempts to return the heart into the left pleural space and cover the heart with a skin flap and the large omphalocele with a silastic membrane and Teflon mesh, but died within 48 hours. The fourth infant with thoracoabdominal ectopia cordis had a double-outlet right ventricle, ventricular septal defect, and pulmonary atresia and received a Blalock–Taussig shunt. The omphalocele and skin defect were repaired. This infant died at 5 weeks of life with candidal sepsis and pericarditis.

This author's second case of total thoracic ectopia cordis presented days after

delivery at another hospital. This child had a double-outlet right ventricle and pulmonary atresia. A single-stage repair was performed and the heart was successfully returned into the thoracic area. One month later, the child died of overwhelming sepsis. Litwin in 1998 operated on a similar child who died of sepsis several months later. Repair of intracardiac defects has been mentioned in the repair of thoracoabdominal ectopia cordis, but few of these were described in detail.

Placing the Heart within the Thoracic Cavity

There have been numerous methods described for enlarging the thoracic cavity to admit the heart. Division and dropping of the anterior diaphragm to attach it inferiorly to the lower part of the defect has been done to create a larger space. The author has done this successfully in three cases. Incision of the costal cartilages and division of some of the lower ribs were done in all three of the author's cases. This maneuver will not cause anterior bulging but rather lateral expansion of the chest wall.

Aytac in 1975 reported performing a left lower lobectomy to increase the intrathoracic space. When the heart became brachycardic, he then performed a left pneumonectomy. The patient lived 20 hours after surgery but then the heart arrested. At autopsy the heart showed a hypoplastic left ventricle, transposition, and anomalous coronary arteries. Also suggested to provide more space was resection of the left lobe of the liver and lengthening of the inferior vena cava to put the heart back into the chest, but no successes were reported.

Repairing and Closure of the Thoracic Cage Defect (Delayed)

These maneuvers are usually performed in cases either with staged repair of the ectopic defect or in cases where the protrusion of the heart is not pronounced. A partial ectopia cordis was repaired and reported by Sabiston in 1957, where the costal cartilages were divided in an oblique manner and allowed to slide to the midline and stabilize the anterior thoracic wall.

Nine months after the initial repair of an ectopic heart by the use of large skin graft flaps, Dobell reported the use of autogenous bone segments from alternating ribs to repair the sternum. With the authors' three cases of total ectopia cordis in which the heart was successfully brought into the thoracic cage without difficulty, a flask-shaped piece of Gore-Tex was sewn to the edges of the widely open sternum. This was followed by skin closure above the Gore-Tex. The second case did not survive due to the tissue swelling from a severe infection, caused by the delay of repair from the time of the infant's birth.

Finally, if it is not possible to create an enclosure of the heart within the chest cavity, it is advised that the skin be mobilized to cover the heart that is extruding from the chest wall and to use an artificial device or a curved external shield fashioned to protect the heart immediately below the skin. It is, however, important to understand that even in the author's single-stage repair with the chest appearing normal, a protective vest should be used to protect the heart until full growth is achieved. At that time, the use of ribs taken alternatively from both sides of the chest can be inserted to protect the heart.

THORACIC ECTOPIA CORDIS

If described well, this category represents the true form of "ectopia cordis." Again the reader is cautioned to read the description of the anatomy with the understanding of partial or total protrusion of the heart. Morello, in a literature review, reported 239 cases of ectopia cordis, and states that most of these cases were of the abdominal and thoracoabdominal types, in which the majority of these cases were "Cantrell's syndrome." Morello then states that only 91 infants had true ectopia cordis but did not distinguish whether these "true" thoracic cases were partial or total ectopia cases.

Many of these were reported as stillborn, whereas others were premature and many survived only a few hours. Only one of the children with major intracardiac defects survived. However, this child was classified as a true ectopia cordis and grouped with three others as thoracic types of ectopia cordis. Three of the four children had an omphalocele, which the authors would have classified as thoracoabdominal cases. It is assumed that there are a number of cases of ectopia cordis that have not been reported in the referred literature.

THORACOABDOMINAL ECTOPIA CORDIS WITH THE INCLUSION OF CANTRELL'S SYNDROME

This condition can be divided into complete and incomplete forms. Partial exposure of the heart and technical advances as previously described render this type more easily repaired and survival more frequent than in true ectopia cordis. Shamberger lists eight methods of repair of thoracoabdominal ectopia cordis in 27 patients. All these patients were repaired with variations similar to the repairs described for true ectopia cordis. Some required secondary epithelialization of the omphalocele or repair of the abdominal wall defect.

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EDITOR'S COMMENTS

Very few textbooks of congenital heart surgery include chapters on ectopia cordis and conjoined twins. Both of these defects are complex and rarely have successful surgical outcome. As noted by Dr. Amato, the classification of ectopia cordis is quite confusing in the literature. I believe that the majority of what has been classified as ectopia cordis actually involves chest wall defects and abdominal defects, which could largely be considered variations of sternal cleft and omphalocele. Pentalogy of Cantrell is usually a distal sternal cleft, which has associated cardiac anomalies and ventricular diverticulae and is usually associated with a superior omphalocele. These defects can generally be repaired with early placement of more rigid covering over the heart and then eventual closure of the sternum or creation of an anterior sternal plate with rib grafts or extensions. These defects have a relatively higher success rate with complete repair and are quite different from true ectopia cordis, which I consider to be a heart that is exposed anterior to the sternum and without a skin covering. These defects have a very high risk for surgical repair primarily because of sepsis and the difficulty in relocating the heart truly into the mediastinal space. Most of these defects should have initial repair immediately to try to prevent infection with relocation as best as possible with diaphragm repositioning with some type of skin coverage and cardiac protection.

Sternal clefts are often considered in the category of ectopia cordis, but these defects are generally easily repaired and do not have the same implication for morbidity and mortality as true ectopia cordis. The incidence of these conditions is extremely low and, therefore, few surgeons have extensive experience with operative repair. Nevertheless, with the techniques described in this chapter some successes can be expected, especially in the more minor defects. It is important to coordinate repair of lower sternal and abdominal defects with general pediatric surgeons so that the attempt to reposition the abdominal contents in the abdomen does not cause additional compression of the diaphragm, which can limit the amount of room in the chest for relocation of the heart. It may be necessary to relocate the heart initially with a silo for the abdominal organs and then gradual closure of the abdominal defect to allow the heart to reposition. Care must be taken when repositioning the heart in the chest to avoid compression of other structures. We have seen an instance where right ventricular outflow tract obstruction has developed from abnormal positioning of the heart in the mediastinum and also limitation of flow through a left superior vena cava to the coronary sinus in another patient.

Conjoined twins are an extremely difficult subgroup of patients to deal with. The vast majority of thoracopagus conjoined twins have hearts that cannot be separated without demise of one or the other twin and often the twin that might be most likely to survive from an abdominal organ standpoint has the most limited cardiac structure. Very rare cases of separation of twins joined at the heart have been reported and most have been unsuccessful. We have seen situations where hearts are joined by a small bridge of tissue at the atrium, which has permitted separation even though the patients had a common pericardium. In this situation, both hearts were separate but there was a bridge of tissue at the base of the atrium connecting the two. Interestingly, the patients had both identical heart rates due to the tissue, which acted as a bypass tract from one atrium to the other. When medications were given to one twin, the other twin's heart rate responded in a similar manner. As soon as the twins were separated surgically, the heart rates became independent.

In the majority of cases of separation of conjoined twins at the thorax and abdomen, chest reconstruction and respiratory insufficiency are the primary cause of morbidity and mortality. These patients often require tissue expanders prior to separation to allow skin coverage of the chest but the lack of significant portions of the rib cage and sternum make respiratory function inefficient. Creation of more rigid thoracic shells may be necessary in some cases to allow for adequate ventilation and many patients will require tracheotomy and long-term ventilation during extensive chest wall reconstruction.

TLS

Index

Page numbers followed by *t* indicate table; those followed by *f* indicate figure.

A

Abdominal phase, of transhiatal esophagectomy, 158–160
Aberrant right subclavian artery, left aortic arch with, 804
Abnormal ostium remodeling, for anomalous coronary artery course, between aorta/pulmonary artery, 1064*f*
Absent pulmonary valve, tetralogy of Fallot with, 730
Accessory left atrioventricular valvar orifice, AVSDs and, 903
Accessory nerve of Kuntz, intraoperative picture, 329*f*
Accessory valve tissue, pediatric mitral valve repair and, 1105
Accountability, 400
Acetaminophen, 338
Achalasia
  definition of, 199
  diagnosis of, 199–200
  etiology of, 199
  incidence of, 199
  non-surgical treatment options for, 200–202
  surgical technique for, 202–207
  symptoms of, 199
  treatment for, 200–202
  vigorous, 199, 202*f*
Acid–base management, neurologic injury prevention and, 393
ACOSOG. *See* American College of Surgeons Oncology Group (ACOSOG)
Acquired mitral valve disease, pediatric mitral valve repair and, 1106
Activated clotting time (ACT), for lung implantation, 236
Acute ischemic mitral regurgitation
  clinical presentation of, 549–550
  diagnosis of, 549–550
  incidence of, 548–549
  myocardial infarction complications and, 548–551
  natural history of, 550
  operative technique for, 551
  operative timing for, 550–551
  pathogenesis of, 548–549
  preoperative therapy for, 550–551
  survival for, 551
Acute pulmonary embolus, 670–675
  anatomic considerations for, 672
  background on, 670
  diagnosis of, 670–671
  operative procedure for, 672–674
  postoperative considerations for, 674–675
  surgery indications for, 671–672
Acute traumatic aortic transection, 611–619
  background on, 611
  diagnosis of, 611–613
  incidence of, 611
  medical therapy for, 613
  natural history of, 611
  surgical management of, 614–617
Acute type A aortic dissection, 565–567
  aortic valve resuspension and, 566*f*
  distal false lumen obliteration and, 567*f*
  surgical technique for, 567–570
Acute type B aortic dissection, 571–572
  intramural hematomas and, retrograde extensions of, 572
  malperfusion syndrome and, 571*f*
  pseudo-coarctation and, 571*f*
Aerodigestive anatomy, 2
AESOP. *See* Automated Endoscopic System for Optimal Positioning (AESOP)
AF. *See* Atrial fibrillation (AF)
Air removal, from heart, in CPB, 366–367. *See also* De-airing, left ventricle
Airway complications, with heart–lung transplantation, 636
Airway reconstruction, carinal resection and, 106
American College of Chest Physicians (ACCP), 16, 24, 120
American College of Surgeons Oncology Group (ACOSOG), 40, 307
Amplatzer repair, of secundum-type atrial septal defects, 820–821
Anastomosis, pericardial fat wrapped around, 73*f*
Anastomotic technique, interrupted, 73*f*
Anesthesia
  for bronchoplastic procedures, 71
  for carinal resection, 100–101
  for OPCAB, 508
  for robotic mitral valve surgery, 421
  for thoracic/thoracoabdominal aneurysms, 577
Aneurysmal dilation, of superior caval system, 783
Annuloaortic ectasia, 556–562
  background on, 556
Annulus enlargement, pediatric, operative technique for, 466*f*
Anomalous coronary artery course
  between aorta/pulmonary artery, 1062–1065, 1063*f*–1065*f*
    abnormal ostium remodeling for, 1064*f*
Anomalous innominate artery, with tracheal compression, 804–806
  anatomy of, 804
  clinical presentation of, 804
  diagnosis of, 805
  surgical indications for, 805–806
  surgical technique for, 805
Anomalous pulmonary venous return, 1041–1052
7F Ansel catheter, 486
Antegrade cannulas, for cardioplegia, 373
Antegrade pressure, of cardioplegia, 374
Antegrade/retrograde cardioplegic perfusion, delivery/monitoring of, 372–374
Antegrade/retrograde perfusion, for myocardial protection, 370–371
Anterior cervicothoracic approach, 37–38, 38*f*
Anterior leaflet commissuroplasty, for mitral valve repair, 413*f*
Anterior leaflet pathology, 413–414
Anterior leaflet prolapse, 425–427, 424*f*–427*f*
Anterior left aortic arch, division of, 806*f*
Anterior mediastinotomy, 19–20, 24*f*
Anterior mitral leaflet, triangular resection of, 416*f*
Anterior retrosternal diaphragmatic hernia of Morgagni, 262–263
  repair of, 265*f*
Anterior scalene muscle
  insertion of, superior sulcus tumor resection and, 319*f*
  off first rib, 247*f*–248*f*
Anterior segmental branch, of pulmonary artery, 55*f*
Anterior thoracotomy, 35, 35*f*
  patient position for, 35
Anterolateral thoracotomy
  pericardiectomy via, 294*f*
Anticoagulation management protocol, 358*t*
Antireflux procedures, 147–156
  complications of, 154
  patient preparation for, 148
  postoperative care for, 154
  results of, 154–155
  surgical principles of, 147–148

Aorta
cardiac magnetic resonance imaging of, 726–730
Aorta-to-right ventricle shunt, PA/IS with, definitive surgical treatment for, 974
Aortic annulus
enlargement, operative technique for, 466*f*
small, 464–467
Aortic arch
aneurysms, 599–609
cerebral protection and, 600–601
diagnosis of, 599
elephant trunk technique for, 602–604, 603*f*
hemi-arch technique for, 601–602, 602*f*
indications for, 599–600
reverse elephant trunk technique for, 604, 608
surgical techniques for, 605–609, 606*f*
total arch technique for, 602, 602*f*
anomalies
classification of, 797–810
embryology of, 797
historical aspects of, 797
incidence of, 797–810
anterior left, division of, 806*f*
double
anatomy of, 797, 805*f*
surgical technique for, 801–803
embryonic, outcome of, 798*t*
interrupted, 861–869
normal development of, 797, 798*f*
obstructions, congenital heart disease and, 753–754
right, with retroesophageal ligament, 801, 803
Aortic cannula/obturator, wire-wound pediatric, 363*f*
Aortic coarctation, 849–858
end-to-end anastomosis and, 853–854
extended end-to-end anastomosis and, 853–854
hypoplastic left ventricle and, 858
interposition graft and, 854
juxtaductal, 849, 850*f*–852*f*
patch aortoplasty and, 852–853
recurrent, 858
special situations for, 857–858
subclavian flap repair for, 851–852, 851*f*
surgical approaches to, 850–851
surgical complications to, 856–857
surgical results for, 857
VSDs and, 857
Aortic cross-clamping
extracorporeal circulation and, 579
pharmacologic manipulation and, 579
Aortic cusp prolapse, repair of, 595
Aortic dissection, 563–573
background on, 563
chronic dissections and, 573
classification of, 563–564, 564*f*
clinical presentation of, 564–565
diagnostic evaluation for, 564–565
follow-up on, 573
incidence of, 563
Aortic flow velocity, 462–464
Aortic graft selection
thoracic/thoracoabdominal aneurysms and, 581
Aortico-left ventricular tunnel, 886–891
clinical presentation of, 886
diagnosis of, 886
morphology of, 886, 888–890, 889*f*
surgical indications for, 889
surgical technique for, 889, 890*f*
Aortic regurgitation
asymptomatic, 463
diagnosis of, 458
Aortic reimplantation, for coronary artery anomalies, 1058, 1059*f*
Aortic root
aneurysm, 556, 557*f*
dilation, 591, 593, 886, 948, 993, 1126
remodeling of, 556–557, 557*f*
replacement of, 377, 465, 560, 561*f*
sinuses, congenital anomalies of, 887
surgical complications of, 561–562
Aortic stenosis (AS), 458
asymptomatic patient of, 462–463
congenital heart disease, 750–752
diagnosis of, 458
low-gradient, 463
LVOTO and, 871–872
natural history of treated, 458
pathophysiology of, 479
symptoms of, 458
Aortic valve
disease, 575
hypoplasia/atresia, interrupted aortic arch with, 864–865
reimplantation of, 556
repair
acute type A aortic dissection and, 556, 562
pediatric, 1118
resuspension, acute type A aortic dissection and, 566*f*
stenosis, pediatric aortic valve repair and, 1118–1119
surgical anatomy of, 458
Aortic valve replacement (AVR), 458–468
operative technique for, 458–461, 466*f*–468*f*
with patient internal mammary artery, 468
pediatric, operative technique for, 1129–1136
Ross procedure for, 471–478
surgical outcomes for, 461
Aortic valve-sparing operations, 556–559, 594–596
Aortopulmonary region, bidigital palpation of, cervical mediastinotomy and, 25*f*
Aortopulmonary shunts
anatomy of, 764, 765*f*
for congenital heart disease, 764–771
Aortopulmonary window, 11, 838–847
anatomy of, 838
approach through, 842*f*
cannulation for repair of, 826*f*, 844*f*
catheter-based approaches for, 843
clinical presentation of, 838–839
diagnostic considerations of, 838–839
embryology of, 838
external appearance of, 840*f*, 844*f*
interrupted aortic arch repair of, 842–843, 845*f*
postoperative care for, 843
preoperative management of, 839–840
repair of, 840, 841*f*
simple, operative technique for, 840–842
surgical complications of, 843
surgical indications for, 820–821
surgical results of, 843, 846–847
Apical-anterior arterial trunk, apical venous branch and, 44*f*
Apical blebs, wedge resection of, 121*f*
Apical resection, for subpleural bleb, of recurrent primary spontaneous pneumothoraces, 117*f*
Apical subpleural blebs, 116
Apical venous branch, apical-anterior arterial trunk and, 44*f*
Arrhythmia. *See also specific* arrhythmia
after pneumonectomy, 66
substrate, 1176
surgery, 1141–1154
background on, 1141
results of, 1154
Arrhythmias, cardiac, 276, 331, 683
Arterial repair, TGA and, 939
Arteriopexy, for innominate artery compression, 810*f*
Ascending aortic aneurysms, 575, 590–597
aortic root anatomy, 591–592
aortic valve and root components, terminology for, 576*f*
aortic valve-sparing operations
aortic cusp prolapse, repair of, 595
aortic root remodeling, 594
aortic root replacement, 595
aortic valve, reimplantation of, 594–595
completion of proximal ascending aortic anastomosis, 595*f*
isolated ascending aorta replacement, 595
open distal anastomosis technique, 595–596
epidemiology and pathophysiology, 591
etiology, 591
imaging, 590–591
indications for intervention, 592–593
operative preparation and initial exposure
bicaval cannulation technique, 594*f*
cardiopulmonary bypass, 593–594
neurologic protection, 593–594
outcomes, 596
presentation, 592
26F Ascendra I device sheath, 487
ASDs. *See* Atrial septal defects
Aspirin, 393, 454, 483, 508–509, 512, 515, 538–539, 973, 1027, 1033, 1036, 1164
Asymptomatic aortic regurgitation, 463
Asymptomatic aortic stenosis, 462–463
risks without operation, 462
unmasking symptoms of, 462
Atherosclerotic heart disease, 493

Atrial fibrillation (AF), 479, 638–646
alternatives to surgical ablation for, 638–641
AV nodal ablation for, 638
current clinical algorithm for, 651
future directions for, 651
maze procedure for, 638, 639*f*, 641, 645
pulmonary vein isolation for, 650
surgical treatment of, 622–630
Atrial lesions, alternate energy sources for, 640*f*
Atrial repair, TGA and, 940
Atrial septal defects (ASDs), 447, 451, 535, 643, 693–694, 695*f*, 700, 706*f*, 816–826
background on, 816
closure recommendations for, 816–818
diagnosis of, 816
echocardiogram of, 817*f*
patch fenestration, deficiency of, 904
skin incision landmarks for, 818*f*
surgical technique for, 818–826
Atrial septectomy, for congenital heart disease, 775, 776*t*, 940, 1030, 1035, 1039
Atrial septum, 451, 667, 693–694, 706–707, 748, 757, 758*f*
Atrioventricular alignments, 692, 692*f*–693*f*
Atrioventricular canal defects. *See* Atrioventricular septal defects (AVSDs)
Atrioventricular canal-type defects, 830
Atrioventricular nodal ablation, for AF, 638
Atrioventricular septal defects (AVSDs), 892–907
accessory left atrioventricular valvar orifice and, 903
anatomic considerations for, 892–893
attaining left atrioventricular valve competence and, 898–901
controversial issues with, 907
diagnostic considerations for, 893–895
eccentric pericardiotomy for, 895, 895*f*
operative technique for, 895–901, 896*f*–902*f*
partial, surgical management of, 904
postoperative care for, 905–907
surgical complications for, 905–907
surgical indications for, 895–901
with tetralogy of Fallot, 902, 1001
unbalanced, 902–903
unusual variants of, management of, 902–904
Atrioventricular valve stenosis, or insufficiency, hemodynamic assessment of, 748
Atrioventricular valve tissue, deficiency of, 903–904
AVR. *See* Aortic valve replacement
AVSDs. *See* Atrioventricular septal defects
Axillary thoracotomy, 34–35
advantages of, 34
incisions, 29*f*
positioning for, 34
Azygos continuation, to right SVC, 783–784
Azygous vein, 63, 193

B

BAC. *See* Bronchioloalveolar carcinoma
Balloon angioplasty
congenital heart disease and, 753–756
for native coarctation, 857–858
Balloon valvuloplasty (BAV), 483*f*, 484–485
for congenital heart disease, 749–750, 751*f*, 752
Belsey fundoplication, 155*f*
with Collis gastroplasty, suturing sequence, 351*f*
Berlin Heart EXCOR® pediatric left ventricular assist devices (LVAD), 1161*f*
Bernoulli theorem, modified, ultrasound physics/applications and, 701
Bicuspid aortic valves (BAVs), 591
Bidigital palpation, of aortopulmonary region, cervical mediastinotomy and, 25*f*
Bilateral lung transplants (BLT), 231, 235–236
Bilateral sequential lung transplantation, 1089
Bilateral superior caval veins
anatomy of, 780
drainage to systemic venous atrium, 780–781
Bilateral thoracosternotomy, 36–37, 1088
Bilobectomy, 49–52
sleeve resection, 73
Blalock-Taussig shunt
for congenital heart disease, 764–765, 765*f*–766*f*, 765*t*
takedown of, with double ligation, 766*f*
Blalock-Taussig shunts, modified, for congenital heart disease, 765–769, 767*f*–768*f*, 767*t*
Bleeding
from CABG, 500
mediastinal biopsy and, 20
as post-transplant complications for, pediatric heart transplantation, 1075
BLT. *See* Bilateral lung transplants
Body surface area (BSA), 366, 475*f*
Botulinum toxin (Botox), 328
BPF. *See* Postpneumonectomy bronchopleural fistula
Brachial plexus dissection
for superior sulcus tumor resection, 321–322, 324
Brachytherapy, for esophageal cancer, 219
Branch pulmonary artery stenosis, congenital heart disease and, 754–755
Bronchial anastomosis, lung transplantation and, 237*f*
Bronchioloalveolar carcinoma (BAC), 79
Bronchoplastic procedures, 71–77
anesthesia for, 71
history of, 71
justification for, 71
operations, 71–75
outcome for, 76–77
postoperative care, 75–76
preoperative assessment for, 71
preparation for, 71
principles of, 71
right-sided resections and, 71–73
Bronchopleural fistula, pneumonectomy and, 67–68
Bronchoscopy, 4–5, 8–10
flexible, 8–9
patient positioning for, 10*f*
procedure for, 9
Bronchus
division, 49*f*–50*f*
in lung transplantation, 236*f*
BSA. *See* Body surface area
Bullous disease, 116–121
classification of, 121*t*

C

CAB. *See* Coronary artery bypass
CABG. *See* Coronary artery bypass grafting
Cannula
antegrade, 362
retrograde, 362
tip, non-deep palpation of, 374*f*
Cannulation, 404–405. *See also* Reoperative cardiac surgery
adult preparation for, 360*f*
aorta incision for, 361*f*
for CPB, 358–362
double, 361*f*–362*f*
final aorta position for, 359*f*
infant preparation for, 360*f*
single venous, 363*f*
Cantrell's syndrome, inclusion of, 1190
Carcinoid heart disease, tricuspid valve and, 447
Cardiac displacement, for OPCAB, 510–511, 510*f*
Cardiac magnetic resonance imaging, 716–744
anatomical uses, 726–733
anatomy, 719*f*
of aorta, 726–730, 728*f*
approach to, 723–726
complex spatial relationships and, 731–732, 731*f*
extracardiac conduits/baffles and, 730–731
of general morphology, 733
great artery anatomy, preoperative/postoperative, 726–730
of miscellaneous diseases, 733
myocardial perfusion with, 735
myocardial tissue/blood tagging with, 735
perfusion/viability/coronary imaging with, 720
physiology/function, 720*f*
for physiology/function, 733–735
of pulmonary arteries, 730, 730*f*
of pulmonary veins, 733
of systemic veins, 732
tissue/blood-tagging imaging with, 724*f*
types of, 724*f*–728*f*
venous connections and, preoperative/postoperative, 732–733
Cardiac malpositions, 698

Cardiac neoplasms
benign, 665*t*
malignant primary, 665*t*
metastatic, 665*t,* 668
primary malignant, 667–668
Cardiac/noncardiac pathologic conditions, associated with VSDs, 831
Cardiac pheochromocytomas, 667
Cardiac repair/lung transplantation, for pulmonary atresia, pediatric, 1091*f*
Cardiac stress test, 60
Cardiac Surgery Consultants Committee (CSCC), 379, 383
Cardiac surgery databases
background on, 379
construction of, 380–382
current adult, 382–385
data collection for, 383
historical considerations on, 379–380
Cardiac surgery, repeat, 375–376
Cardiac tamponade, diagnosis of, 289–290
Cardiac tumors, 664–669
classification of, 664
epidemiology of, 664
historical background on, 664
myxomas and, 664–666, 665*f*
Cardiology Patient Database, of CHP, 780, 782–783
Cardioplegia
antegrade cannulas for, 362, 373
antegrade pressure of, 374
cold blood, 370
cold multidose blood, solution contents for, 372*t*
components, 370
for coronary grafting/valve replacement, 373*f*
coronary sinus injury and, 374
delivery devices for, 373*f*
modified cold blood maintenance solution, 372*t*
monitoring devices for, 372–374, 373*f*
monitoring pressure of, 374
multidose, 372*t*
retrograde cannulas for, 362, 373
warm blood, 370
Cardioplegic perfusion
antegrade/retrograde, 370–371
delivery of, 375*t*
intermittent/continuous, 371
Cardioplegic solutions, 374–375
Cardiopulmonary bypass circuit, 364–365, 364*f,* 606*f*
example of, 364*f*
Cardiopulmonary bypass (CPB), 356–368, 404–405, 593–594, 680, 896*f,* 1010, 1067. *See also* Reoperative cardiac surgery
air removal from heart in, 366–367
alternate arterial cannulation for, 360
CABG using, 493–503
cannulation, 358–362, 358*f*–363*f*
completed cannulation, 368*f*
decompressing left heart in, 365–367
draping and, 357
hematocrit management for, for neurologic injury prevention, 393
incisions for, 357–358
indications for, 362–363
institution of, 365–367
ischemic mitral regurgitation and, 434
lung transplantation and, 236
patient blood volume in, 365*t*
patient positioning for, 357
plan development for, 356
preparation for, 357–363
priming volume per tube length/diameter in, 365*t*
for pulmonary stenosis, open pulmonary valvotomy using, 963–964
required tubing diameter, for different patient weights, 365*t*
skin preparation for, 357
weaning from, 375, 475, 878
Carina, bronchoscopic view of, 3*f*
Carinal resection, 100–106
airway reconstruction and, 104–105
anesthesia for, 100–101
intraoperative evaluation for, 101–102
introduction to, 100
operative technique for, 101–105
patient evaluation for, 100
postoperative care for, 104–105
release maneuvers for, 102, 102*f*
surgical approaches to, 101–102
Carpentier-McCarthy-Adams IMR etlogix ring, for ischemic mitral regurgitation, 435*f,* 436
Carpentier's functional classification, of ischemic mitral regurgitation, 432*f*
Catamenial pneumothorax, 118, 121, 272
Catheter placement, with polyethylene terephthalate cuffs, 281*f*
Cavopulmonary shunts
bidirectional and, 1010–1011, 1010*f*–1011*f*
early postoperative complications for, 997
hemi-Fontan operation and, 989–998
hemi-Fontan procedure and, 1011–1014, 1012*f*–1013*f*
history of, 1006–1007
indications for, 1007–1008
operative technique for, 1008–1014, 1008*f*–1013*f*
preoperative assessment for, 1007–1008
risk factors for, 1007, 1011, 1014–1015
unidirectional, 1008–1009, 1008*f*–1009*f*
CBF. *See* Cerebral blood flow
ccTGA. *See* Congenitally corrected transposition of great arteries
Celiac/mesenteric/renal arteries, reimplantation of, 581–582, 582*f*
CentriMag® console and pump, 1160*f*. *See also* ECMO and ventricular assist devices
Cephalopagus, 1183, 1183*f*
Cephalothoracopagus, 1183, 1183*f*
Cerebral blood flow (CBF), 390, 735, 802, 1010
Cerebral perfusion maintenance, neurologic injury prevention and, 391
Cerebral protection
arch aneurysms and, 600–601
techniques, 390–394
Cervical dissection, 18*f*
in mediastinoscopy, 18*f*
Cervical incision, 33, 34*f,* 96*f,* 163, 165, 195*f*
Cervical mediastinotomy, bidigital palpation of, aortopulmonary region and, 25*f*
Cervical phase, of transhiatal esophagectomy, 160–161
CF. *See* Cystic fibrosis
Chemotherapy, for esophageal cancer, 170
Chest lobe resections, en bloc removal, 323*f*
Chest radiographs (CXR), for pneumothorax, 118–119
Chest tubes
for empyema, 313, 315–316
Chest wall closure, superior sulcus tumor resection and, 325
Chest wall resections, 253–258
en bloc lateral, 254*f*
en bloc removal, 323*f*
indications for, 253
operative planning for, 253
pedicle muscle flaps and, for reconstruction, 257*f*
preoperative evaluation for, 253
prosthesis inset for, 256*f*
sternal resections and, 257, 258*f*
superior sulcus tumor resection and, 320–321, 321*f,* 324–325
synthetic material choices for, 255–256
tissue transfer options for, 256–257, 256*t*
CHF. *See* Congestive heart failure
Children's Hospital of Pittsburgh (CHP), 780
Cardiology Patient Database of, 780, 782–783
Chordal replacement, technique of, 421
Chordal shortening, technique of, 427, 1110*f*
CHP. *See* Children's Hospital of Pittsburgh (CHP)
Chronic obstructive pulmonary disorders (COPD), 231
Chronic postoperative pain syndromes, 330
Chronic pulmonary thromboembolism, 676–685
Chronic thromboembolic pulmonary hypertension (CTEPH), 676–685
alternative treatments for, 679
anesthetic considerations for, 680
clinical presentation of, 677
diagnostic modalities for, 677–678, 678*f*
incidence of, 676
indications for, 679
operative preparation for, 680
operative principles for, 679–680
operative techniques for, 680–683, 681*f*–684*f*
pathology of, 677
pathophysiology of, 677
postoperative care for, 683–684
pulmonary thromboendarterectomy for, 679
reperfusion response management and, 684
surgical complications of, 683–684
Chyle, composition of, 276*t*

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Chylothorax
definition of, 274
diagnosis of, 275
etiology of, 275*t*
internal shunt system for, 279*f*
management of, 274–287
octreotide for, 276, 277*t*
pleuroperitoneal shunt for, 278
pneumonectomy and, 67
somatostatin for, 276, 277*t*, 287
thoracoscopic management of, 278*f*
thoracotomy for, 277, 277*f*
Cine magnetic resonance imaging uses, 734
Circular gastric staple line creation, 345*f*
Citrate phosphate dextrose (CPD), 370
Clamshell. *See* Bilateral thoracosternotomy
Cleft mitral valve, pediatric mitral valve repair and, 1105, 1109
Clopidogrel, 483
Closed tricuspid valvotomy, for neonates, 967–968
Coarctation. *See* Aortic coarctation
Cold blood cardioplegia, for myocardial protection, 3370, 372*t*, 375, 562
Cold multidose blood cardioplegia, solution contents for, 372*t*
Collis gastroplasty, 152, 153*f*, 348, 348*f*
Colon interposition, 214–215
mobilized left, 214
preparation for, 214
procedure for, 214
Compensatory hyperhidrosis (CH), 331
Complete vascular rings
anatomy of, 797
clinical presentation of, 797–800
diagnosis of, 800–801
surgical indications for, 801
surgical technique for, 601–803, 804*f*–809*f*
Complexity theory, 387
Complex spatial relationships, cardiac magnetic resonance imaging and, 731–732, 731*f*
Computed tomography (CT), 61, 119, 200, 314, 493, 494, 575, 1075–1076
Conal septal defect
repair of, 834
Conduits/shunts, congenital heart disease and, 756, 756*f*
Cone resection, with electrocautery, 83*f*
Congenital heart disease
anatomy of, 688–698
aortic arch obstructions and, 753–754
aortopulmonary shunts for, 764–771
atrial arrhythmias in, 643
atrial septectomy for, 775, 776*t*
balloon angioplasty and, 753–756
balloon valvuloplasty for, 749–750, 751*f*
Blalock-Taussig shunt for, 756*f*, 764–769, 765*f*–768*f*
branch pulmonary artery stenosis and, 754–755, 755*f*
chamber/valve/vessel abnormalities and, 692–698
classification of, 688–698
accuracy in, 689
aims/principles of, 688–689
atrioventricular alignments and, 692, 692*f*–693*f*
cardiac segment and, 689–692
intersegmental alignments and, 689, 692–698, 692*f*–697*f*
organization of, 688
precision in, 689
quantification, 689
segmental approach to, 689
segmental notation and, 690–692
ventricular loop and, 689–690, 690*f*
ventriculoarterial alignments and, 689, 692
visceroatrial situs and, 690, 691*f*
conduits/shunts, 756, 756*f*
device closures and, 757–758
echocardiographic evaluation of, 700–714
embolizations and, 756–757
endovascular stenting and, 753–756
imaging children with, 702–706
abdomen view of, 702
atria/ventricles, 687–688
atrioventricular connection, 705–706
situs and, 702–705
venous connections and, 712
intraoperative transesophageal echocardiography/postoperative imaging and, 713–714
miscellaneous palliative procedures for, 775
modified Blalock–Taussig shunts for, 756*f*, 765–769, 767*f*–768*f*, 767*t*
palliative operations for, 764–776
Potts shunt for, 770–771, 771*f*–772*f*, 771*t*
pulmonary artery band for, 759*f*, 771–775, 773*t*
pulmonic valve stenosis/atresia and, 749–750
retrievals and, 758–759
septostomy procedures for, 748
syntax for diagnosis, 698
systemic pulmonary veins and, 755–756
transcatheter therapy for, 746–760
hemodynamic assessment and, 746–760
Waterston/Cooley shunt for, 769–770, 769*f*–770*f*, 769*t*
Congenital wall defects
ectopia cordis, 1186
isolated sternal clefts, 1186
Congenitally corrected transposition of great arteries (ccTGA), 708, 709*f*, 954–960, 1077–1078
anatomic repair for, 957–960
anatomy of, 954
background on, 954
classical repair of, 959
diagnosis for, 954–955
embryology of, 954
palliative operations, 955
subpulmonary obstruction repair for, 956–957
surgical results of, 959–960
surgical treatment for, 955–957, 956*f*–957*f*
ventricular septal defects closure for, 955–956
Congenital mitral valves, pediatric mitral valve repair and, 1112–1113
Conjoined twins
"Siamese twins," 1182
abdominal, 1189
anesthetic management, 1185
Cantrell's syndrome, inclusion of, 1190
cardiac involvement, 1184–1185
cephalopagus, 1183, 1183*f*
cephalothoracopagus, 1183, 1183*f*
cervical, 1189
cervicothoracic, 1189
classification of, 1182–1184, 1188–1189
congenital wall defects, 1186
considerations for, 1185–1186
craniopagus, 1183
diagnostic procedures, 1185
dicephalic parapagus, 1183
dithoracic parapagus, 1183
ectopia cordis, 1186
etiology of, 1182
heart and chest wall development, embryology in, 1187–1188
cross section of, 1187*f*
diaphragm muscle formation, 1188*f*
sternum development, 1187*f*
history of, 1182
incidence, 1186
ischiopagus, 1183, 1184*f*
mechanism of, 1182
omphalo-ischiopagus, 1183, 1184*f*
omphalopagus, 1183, 1183*f*
parapagus, 1183
procrustean classification, 1186–1187
pygopagus, 1183
rachipagus, 1183
surgical correction
heart, placing within thoracic cavity, 1190
heart, immediate coverage of, 1189
intracardiac defects, 1189–1190
prenatal diagnosis and cesarean section, 1189
thoracic cage defect, repairing and closure, 1190
surgical separation, 1185
thoracic ectopia cordis, 1190
thoraco-omphalopagus, 1183, 1184*f*
thoracoabdominal ectopia cordis, 1190
thoracopagus, 1183, 1183*f*
xiphopagus, 1183, 1184*f*
Conotruncal anomalies, 697–698, 697*f*, 709–710
Conotruncal defects, 710, 829–830
Conoventricular defects, 829, 830*f*
repair of, 832–834, 832*f*–834*f*
transventricular closure of, 834*f*
Conscious sedation, 8, 10
Continuous positive airway pressure (CPAP), 1108
Contrast effect, 398*f*
COPD. *See* Chronic obstructive pulmonary disorders

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Coronary artery, 431, 433, 459*f*, 473*f*–474*f*, 735, 1065–1070
fistula
definition of, 1065
results for, 1067
surgery indications for, 1066
surgical techniques for, 1066–1067, 1067*f*, 1068*f*
Coronary artery anomalies
CABG for, 1058
left internal mammary artery grafting and, 1058
left subclavian-to-left coronary artery anastomosis and, 1058
in children, 1055–1070
origin from pulmonary artery, 1055–1062, 1056*f*–1062*f*
results of, 1061–1062
Coronary artery bypass (CAB), 390–395, 500*f*, 507–513, 642
intraoperative management, 508
neurologic injury prevention after, 390–395
procedure, 392
surgical techniques for, 515–517
Coronary artery bypass grafting (CABG), 356, 367–368, 390, 507
bleeding from, 500
complications from, 503
for coronary artery anomalies, 1058
evolution of, 493
future considerations and, 503
infection from, 503
neurologic complications, 503
outcomes from, 501–502
renal dysfunction and, 498
using CPB, 493–503
Coronary grafting/valve replacement, cardioplegia for, 373*f*
Coronary sinus (CoS), 373–374, 440, 693, 706–707, 785, 824–825
anomalies of, 785
atrial septal defect, surgical repair of, 824–825, 824*f*–825*f*
injury, cardioplegia and, 374
Coronary stabilization, for OPCAB, 511
Coronary targets, for OPCAB, 510–511, 510*f*
CoS. *See* Coronary sinus
Cox Maze III modifications, 639*f*–640*f*
Cox Maze procedure
mitral valve surgery and, 642
results of, 644–645
surgical technique of, 1101
CPAP. *See* Continuous positive airway pressure
CPB. *See* Cardiopulmonary bypass
CPD. *See* Citrate phosphate dextrose
Craniopagus, 1183
Crawford classification, of thoracic/thoracoabdominal aneurysms, 575, 576*f*
Crural sutures placement, 348
CSCC. *See* Cardiac Surgery Consultants Committee
CT. *See* Computed tomography
CTEPH. *See* Chronic thromboembolic pulmonary hypertension
Current transcatheter heart valve technology
Edwards SAPIEN and SAPIEN XT, 481–482
Medtronic CoreValve ReValving System, 482
CXR. *See* Chest radiographs
Cyclobenzaprine, 338
Cystic fibrosis (CF), 121, 231, 1085

D

Damus-Kaye-Stansel repair of DORV, with subpulmonary ventricular septal defect, 929, 931*f*
Databases. *See* Society of Thoracic Surgeons (STS) database; Veterans Affairs database
David technique, 594–595
DCT. *See* Double-clamp technique
De-airing, left ventricle, 436
Debakey forceps, 167, 168*f*
Decompressing left heart, in CPB, 365–366
Deep vein thrombosis (DVT), 670, 676
Degenerative mitral disease, 421–422
DES. *See* Diffuse esophageal spasm
Diaphragm, 260–273
abdominal view of, 263*f*, 265*f*
anatomy of, 260–261, 261*f*–262*f*
embryological development of, 260–261
eventration of, 273
incisions in, 262, 264*f*
phrenic nerve and, 260
plication of, 269*f*, 272*f*
PTFE patch for, 263
repair, 263, 265*f*, 269*f*
suturing techniques for, 269*f*
thoracic view of, incisions in, 261*f*
trauma to, 267–268
tumors of, 272
Diaphragmatic flap, 272–273
harvesting technique for, 273*f*
Diaphragmatic pacing, 272–273
Diaphragmatic paralysis, 273
Diaphragmatic surface, transdiaphragmatic fenestration and, 118*f*
Dicephalic parapagus, 1183
Diffuse esophageal spasm (DES), 207–208
definition of, 207–208
diagnosis of, 208
etiology of, 208
treatment of, 208
Diffuse subaortic stenosis, 878
Dilators, flexible, 7*f*
Discrete subaortic stenosis, 876, 878
Distal false lumen obliteration, acute type A aortic dissection and, 567*f*
Dithoracic parapagus, 1183
Diverticula. *See specific* diverticula
Domino-donor procedures, with heart–lung transplantation, 636
Donor heart procurement, for heart transplantation, 622–623
Donor lung retrieval, 231–234
Doppler principle, ultrasound physics/applications and, 700–701
Dor fundoplication in laparoscopic esophagomyotomy, 203*f*
Double aortic arch, 713, 724, 727, 797–798
anatomy, 797
surgical technique for, 801–803
Double cannulation, final position for, 362*f*
Double-clamp technique (DCT), 392
Double-outlet left ventricle (DOLV), 916–918
anatomy of, 934
complications of, 934–935
definition of, 933–934
postoperative considerations of, 934–935
surgical technique for, 934
Double-outlet right ventricle (DORV), 710–711
anatomic repair of, with subpulmonary ventricular septal defect, 910–912, 911*f*
classification of, 924
complications of, 915–916
Damus-Kaye-Stansel repair, with subpulmonary ventricular septal defect, 929, 931*f*
definition of, 924
doubly committed ventricular septal defect with, 924, 930
noncommitted ventricular septal defect with, 930–932
postoperative considerations of, 932–933
subaortic ventricular septal defect and, 924–926, 926*f*
with subaortic ventricular septal defect with pulmonary stenosis, 924, 926–927, 927*f*
subpulmonary ventricular septal defect and, 924, 927–929, 928*f*
surgical techniques for, 924–925
tunnel repair of, 932*f*
Double venous cannulation, 361*f*
Doubly committed ventricular septal defect, with DORV, 924, 930
Duke Lung Transplant Program, 231, 232*f*, 238
Duke University immunosuppression protocol, for lung transplantation, 239*t*
Duke University infection prophylaxis for lung transplantation, 240*t*
Dumbbell tumor. *See* Posterior mediastinal neurogenic dumbbell tumor
Duodenum migration, paraesophageal hernia with, 266*f*
DVT. *See* Deep vein thrombosis

E

Ebstein's anomaly
anatomy of, 447
tricuspid valve, repair for, 447
Eccentric pericardiotomy, for AVSDs, 895, 895*f*
ECG. *See* Electrocardiogram
Echocardiographic evaluation, of congenital heart disease, 700–714
ECMO. *See* Extracorporeal membrane oxygenation

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ECMO and ventricular assist devices
CentriMag® console and pump and PediMag® pump, 1160, 1160*f*
chest X-ray demonstrating, 1159*f*
double-lumen, 1159*f*
extracorporeal membrane oxygenation, 1158–1159, 1158*f*, 1159*f*
implantable devices, 1163–1164
paracorporeal devices, 1160–1163
pediatric specific issues, 1164
rationale and indications, 1157–1158
respiratory and heart, failure in pediatric patients, 1157
schematic diagram of, 1158*f*
temporary ventricular assist devices, 1159 1160
ventricular assist device implantation, preparation for, 1159
Ectopia cordis, 1186
historical aspect of, 1187
pathogenesis of, 1188
thoracic, 1189–1190
thoracoabdominal, 1189–1190
types of, 1188*f*
Edwards SAPIEN and SAPIEN XT, 481–482, 481*f*
Electrocardiogram (ECG), 542, 565
Electrocautery incision, 168*f*
Elephant trunk technique, for arch aneurysms, 604, 604*f*
Embolization
congenital heart disease, 366
reduction, neurologic injury prevention, 392
Empyema
chest tubes for, 68
history of, 67
incidence of, 67
open drainage procedure for, 177
pneumonectomy, 67–68
streptokinase for, 315
Endo-ACAB. *See* Endoscope-assisted coronary artery bypass
Endocarditis
of mitral valve, 413
tricuspid valve, 447
Endoscopic Atraumatic Coronary Artery Bypass (Endo-ACAB), 516
Endoscopic endoluminal palliative procedures, for esophageal cancer, 217–218
Endoscopy
advantages of, 2
equipment, 4–6
Endovascular thoracic aortic aneurysm repair, 582–584
End-stage heart failure, 1157
End-to-end anastomosis
aortic coarctation and, 853–854, 853*f*
technique of, 851
Erdheim's cystic medial necrosis, 556
Esophageal cancer
ablative therapies for, 219
brachytherapy for, 219
chemotherapy for, 219
endoscopic endoluminal palliative procedures for, 219
incidence of, 186
laser ablation for, 219
palliation for, 219
photodynamic therapy for, 187, 219
radiation for, 219
stents for, 218–219
Esophageal mobilization, for thoracic diverticula, 193*f*
Esophageal replacement conduits, 158
Esophageal resection, 173, 213
Esophageal shortening, 348
Esophagectomy
left thoracoabdominal approach to, 179
minimally invasive approaches, 190
postoperative management for, 183
thoracic approaches to, 173–183
indications for, 174*t*
surgical technique for, 173
Esophagogastric fat pad, mobilization of, 344*f*
Esophagogastrostomy
hand-sewn, 182, 182*f*
Esophagoscope, dilating, 6*f*
Esophagoscopy, 5–6
procedure for, 7–8
Esophagus, 3–4
assessment, 343–344
European System for Cardiac operative Risk Evaluation score (EuroSCORE), 479
Eventration of diaphragm, 268–272
EVEREST High Risk Registry, 443–444
Excitotoxicity mechanism, of neural cell injury, 391*f*
Extended cervical mediastinoscopy, 23*f*
Extended end-to-end anastomosis, aortic coarctation and, 853–854, 853*f*
Externalized pleuroperitoneal shunt
patency of, 282*f*
placement of, 280*f*
Extracardiac conduits/baffles, cardiac magnetic resonance imaging and, 730–731
Extracardiac vessels, congenital heart disease and, imaging children with, 712
Extracorporeal membrane oxygenation (ECMO), 670, 1157–1165

F

FDG. *See* Fluorodeoxyglucose
FEV. *See* Forced expiratory volume
First thoracic nerve root, from intervertebral foramen, 322*f*
Flexeril. *See* Cyclobenzaprine
Flip over technique, for mitral valve repair, 415*f*
Floppy Collis-Nissen fundoplication, 347*f*
Flow gradients, 746–747
Fluorodeoxyglucose (FDG), 62, 495
FMR. *See* Functional mitral regurgitation
Fontan conversion, 1141–1154
Fontan operation, 1020–1028
early outcome for, 1028*t*
indications for, 1021, 1023–1024
postoperative management after, 1023, 1027
results of, 1027
Fontan suture, in left ventricular reconstruction, 505*f*
Forced expiratory volume (FEV), 41, 1084
Forced vital capacity (FVC), 41
Frailty Index, 480, 480*t*. *See also* Transcatheter aortic valve replacement (TAVR)
Free ventricular wall rupture
clinical presentation of, 542
diagnosis of, 542
incidence of, 542
myocardial infarction complications, 542–544
natural history of, 542
operative technique of, 542–544, 543*f*
operative timing of, 542
pathogenesis of, 542
preoperative treatment of, 542
survival of, 544
Full blood flow rates, calculation methods for, 364*t*
Functional mitral regurgitation (FMR), 432*f*, 519, 524
Functional tricuspid regurgitation, 446–447
Fundoplication
Belsey, 155*f*
completed view of, 156*f*
complications of, 154
gastroplasty and, 152, 153*f*
indications for, 147
laparoscopic, 148–152, 149*f*–152*f*
operative techniques for, 148–154, 148*f*–156*f*
patient preparation for, 148
postoperative care for, 154
results of, 154–155
surgical principles of, 147–148
transthoracic partial, 153–154, 155*f*–156*f*
FVC. *See* Forced vital capacity

G

Gastric mobilization, 158–159, 167, 176*f*, 191, 191*f*
Gastric tube
creation, MIE and, 192*f*
suturing, for MIE, 192*f*
Gastroesophageal (GE) junction, 341, 348
Gastroesophageal migration, paraesophageal hernia with, 266*f*
Gastroesophageal reflux disease (GERD), 147, 186, 200, 207–208, 210–211, 350
Gastroesophageal reflux related motility disorders, 200, 203, 205, 207–208, 210–211
Gastroplasty
collis, 152, 153*f*, 265, 267, 344, 345*f*, 348, 348*f*, 351*f*–352*f*
fundoplication and, 151
GERD. *See* Gastroesophageal reflux disease
Giant paraesophageal hernia, transthoracic repair, 345–347
Glenn shunts. *See* Bidirectional cavopulmonary shunts

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Glucose control neurologic injury prevention and, 393
Graft failure, as post-transplant complications for, pediatric heart transplantation, 1075
Graft surveillance, for heart–lung transplantation, 636
Gustatory sweating, 331

H

Hammock valve, pediatric mitral valve repair and, 1106, 1113*f*
HCA. *See* Hypothermic circulatory arrest
HCFA. *See* Health Care Financing Administration (HCFA)
Health Care Financing Administration (HCFA), 379
Heart–lung transplantation, 629–636
  airway complications with, 636
  background on, 629
  chronic rejection/late complications with, 636
  domino-donor procedures with, 636
  early postoperative clinical management for, 634–635
  graft surveillance for, 636
  immunosuppressive management for, 635
  infection prophylaxis for, 635–636
  long-term results of, 636
  organ procurement preservation for, 629–631
  pediatric, 1085–1097
  postoperative complications with, 636
  recipient operation for, 631–634, 631*f*–635*f*
  recipient selection for, 629
  retransplantation and, 636
HeartMate® II left ventricular assist device, 1163*f*
HeartMate left ventricular assist device, in situ, 538*f*
Heart–single-lung transplant, 636
Heart transplantation, 621–638
  after ventricular assist device, 626–627
  background on, 621
  donor availability/allocation for, 621–622
  donor heart procurement for, 622–623, 623*f*
  donor selection for, 622
  heterotopic, 627, 627*f*
  organ preservation for, 623–624
  orthotopic, 376–377, 624–626, 625*f*
  pediatric, 1074–1082
  postoperative management for, 628
  preoperative evaluation for, 621
  recipient selection for, 621
  results for, 628
HeartWare HVAD pump, 1163*f*
Hemacrit management, on CPB, for neurologic injury prevention, 393
Hemi-arch technique, for arch aneurysms, 601–602
Hemiclamshell incision, superior sulcus tumor resection and, 325, 325*f*
Hemi-Fontan procedure, cavopulmonary shunts and, 1011–1014, 1012–1013*f*
Hemodynamic assessment
  of atrioventricular valve stenosis, or insufficiency, 748
  of mitral valve, 748
  of patients
    with complete mixing lesions, 747
    with functional single ventricle, 747
    with left-to-right shunt lesion, 747
    with right-to-left shunt lesion, 747
  principles of, 746–747
  of pulmonary valve, 747, 750*f*, 752
  of semilunar valve stenosis, or insufficiency, 747
  transcatheter therapy and, for congenital heart disease, 746–760
  of tricuspid valve, 748
Hemorrhage, tracheal resection and, 93*f*
Hemostasis, thymectomy and, 136
Hemothorax, 168, 313, 339
Hepatic segment, of IVC, absence of, 783, 784*f*
Hernia
  of Bochdalek, posterolateral diaphragmatic, 263–265
    repair of, 266*f*
  hiatal, 149, 157, 265–267
  of Morgagni, anterior retrosternal diaphragmatic, 262–263
    repair of, 265*f*
  paraesophageal
    with duodenum migration, 266*f*–267*f*
    with gastroesophageal migration, 266*f*–267*f*
    symptoms of, 210
    treatment for, 210
    view of, 266*f*
Herniated stomach, reduction of, 343*f*
Heterotopic heart transplantation, 627, 627*f*
Hiatal hernia, 149, 157, 265–267, 341
High Reliability Organizations (HROs), 397
High superior vena cava, PAPVR to, 1051–1052
Hilum
  left pulmonary, view of, 54*f*
  right pulmonary, view of, 42*f*
HLHS. *See* Hypoplastic left heart syndrome
Horner's syndrome, 330
HROs. *See* High Reliability Organizations (HROs)
Hybrid coronary artery bypass grafting surgery
  anti-platelet strategy, 517
  current data, 517
  feasibility and efficacy, 502
  limitations, 517
  outcomes, 517*t*
  patient selection, 517, 517*t*
  surgical techniques, 515–517
    operating room, 500*f*
  timing
    "one-" or "two-staged" procedure, 515
Hybrid coronary revascularization (HCR) strategy, 515
Hydrocodone, 338
Hyperhidrosis, 328–329
Hypertrophic cardiomyopathy, 643, 653–661, 1177
  AF and, 643
Hypoplastic left heart syndrome (HLHS), 746, 759*f*, 1030–1040
  anatomy of, 1030–1031
  initial management of, 1031–1032
  pediatric heart transplantation and, 1076–1077, 1076*f*–1077*f*
    donor procurement for, 1076
    recipient operation for, 1076–1077
  presentation of, 1031–1032
  results for, 1038–1040
  stage I reconstructive surgery for, 1032–1034, 1033*f*–1034*f*
  stage II reconstructive surgery for, 1034–1035, 1035*f*–1037*f*
  stage III reconstructive surgery for, 1035–1038, 1038*f*–1039
  surgical therapy for, 1032–1038, 1033*f*–1039*f*
Hypoplastic left ventricle, aortic coarctation and, 858
Hypothermia level, safe circulatory arrest times and, 366*t*
Hypothermic circulatory arrest (HCA), 390, 467, 581, 594, 845*f*–846*f*, 863, 1033*f*

I

Ibuprofen, 338
ICD therapy in children
  introduction to
    common ICD clinical situations, 1175–1176
    defibrillation threshold testing, 1178
    epicardial vs. transvenous considerations, 1176
    follow-ups, 1178
    HRS/American College of Cardiology, recommendations/guidelines, 1174–1175
    long-term complications, 1177–1178
    in patients with and without congenital heart disease, 1176–1177
    special challenges, 1178–1179
    technical considerations, 1177
ICM. *See* Intercostal muscle
ICS. *See* Intercostal space
Idiopathic pulmonary fibrosis (IPF), 231
IMR. *See* Ischemic mitral regurgitation
Infection
  prophylaxis, for heart–lung transplantation, 635
  tracheal resection and, 85, 89, 97
Infective endocarditis, pediatric mitral valve repair and, 1106
Inferior caval vein (IVC)
  anomalies of, 783
  hepatic segment of, absence of, 783, 784*f*
  interruption of, in visceral heterotaxy, 784
  miscellaneous abnormalities of, 784
Inferior pulmonary ligament, 49
  view of, 54*f*
Inferior-type sinus venous atrial septal defect, surgical repair of, 823–824

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Infundibular septum
resection of, 864*f*
VSDs and, 865*f*
Inlet septal defects, 830
repair of, 834
Innominate artery compression
arteriopexy for, 810*f*
positioning/approach for, 810*f*
reduction of, 811*f*
Intercostal arteries, reimplantation of, 581–582
Intercostal muscle (ICM)
harvesting of, 61*f*
Intercostal space (ICS), 112
Internal shunt system
for chylothorax, 279*f*
in position, 280*f*
Interposition graft, aortic coarctation and, 854
Interrupted aortic arch, 861–870
anatomy of, 861
with aortic valve hypoplasia/atresia, 864–865
classification of, 862*f*
clinical presentation of, 861–862
diagnosis of, 861–862
early/late mortality with, 869*t*
neonatal cardiovascular conditions with, 862*t*
operative management of, 862–864
postoperative management for, 867
repair of aortopulmonary window, 842–843
with single ventricle, 866–867
special circumstances of, 864–867
survival for, 867–868
with TGA, 866
with truncus arteriosus, 865, 866*f*
repair of, 915–917
Intervertebral foramen, first thoracic nerve root from, 322*f*
Intracardiac defects, closure procedures for, 756
Intramural hematomas, retrograde extensions of, acute type B aortic dissections, 572
Intraoperative cerebral monitoring, neurologic injury prevention and, 394
Intraoperative transesophageal echocardiography/postoperative imaging, congenital heart disease and, 713–714
Intrathoracic lymphatics
anatomy of, 304
drainage of, 304–307
Iontophoresis, 328
IPF. *See* Idiopathic pulmonary fibrosis
Ischemic mitral regurgitation (IMR), 431–439
Carpentier–McCarthy–Adams IMR etlogix ring for, 435*f*, 436
Carpentier's functional classification of, 432*f*
late outcomes for, 438
mild/moderate, 434
mitral valve exposure and, 435
mitral valve repair and, 435–436
myocardial protection and, 443–435
operative mortality for, 437–438
perioperative considerations for, 434
residual, 437
severe, 434
surgical approaches to, 434
surgical indications, 434
type II
clinical presentation of, 438
diagnosis of, 438
medical/surgical treatment for, 438
pathophysiology of, 438
surgical results for, 438
type IIIb
clinical presentation of, 433
diagnosis of, 433–434
pathophysiology of, 431–433, 433*t*
undersized remodeling ring annuloplasty for, 435–436
Ischemic mitral valve repair, 431–439
Ischiopagus, 1183, 1184*f*
Isolated aneurysms, 590
Isolated ascending aorta replacement, 595. *See also* Ascending aortic aneurysms
Isolated atrial fibrillation, minimally invasive epicardial ablation and, 648
Isolated left superior caval veins, 781–782
IVC. *See* Inferior caval vein

J

Jackson esophagoscopes, 6*f*
Jejunal interposition, 215–216
long-segment, 215
long-segment reconstruction and, 215–216
Juxtaductalcoarctation, 850*f*
J-Wire, 487

K

Ketorolac, 338

L

LAD. *See* Left anterior descending artery
LAO. *See* Left anterior oblique view
Laparoscopic esophagomyotomy
for achalasia, 199–207, 200*f*–206*f*
Dor fundoplication in, 203*f*, 206*f*
trocar port positions, 175*f*
Laparoscopic fundoplication, 186, 148–154
Laparoscopic pyloroplasty, MIE and, 191*f*
Laparoscopic repair
Belsey fundoplication with Collis gastroplasty, suturing sequence, 351*f*
esophageal lengthening procedure, 352
gastroesophageal reflux disease, 350
important aspects, 352
Nissen fundoplication, 350, 352*f*
paraesophageal hernia, 352
transthoracic Nissen fundoplication, 351*f*
Laparoscopic wedge gastroplasty, 346*f*
Laparotomy, 173
Laser ablation, for esophageal cancer, 219
Lateral chest wall resection, polypropylene mesh/methylmethacrylate prosthesis for, 256*f*
LCSG. *See* Lung Cancer Study Group
Leaflet advancement technique, for mitral valve repair, 409–410, 409*f*–410*f*
Left anterior oblique view (LAO), 706*f*, 711*f*–713*f*, 749*f*
Left aortic arch with aberrant right subclavian artery, 804
Left atrial anastomosis, lung transplantation and, 237*f*
Left atrioventricular valve competence, AVSDs and, 898–901
Left atrium, left superior caval veins drainage into, 782
Left inferior pulmonary vein, view of, 65*f*
Left internal mammary artery grafting, coronary artery anomalies and, CABG for, 1058
Left internal mammary artery (LIMA), 375, 403*f*, 494, 498*f*, 500*f*, 515, 1058
Left lower lobectomy, 56–59
sleeve resection, 75
Left lower sleeve, view of, 75*f*
Left main bronchus, left main pulmonary artery and, 55*f*
Left main pulmonary artery, left main bronchus and, 55*f*
Left pulmonary artery
anomalous development of, 802*f*
superior pulmonary artery and, 236*f*
view of, 58*f*
Left pulmonary atrium, right superior caval veins to, 782–784, 784*f*
Left pulmonary hilum, view of, 54*f*
Left-sided endotracheal tube, for right upper lobe sleeve, 73*f*
Left-sided pneumonectomy, surgical procedure for, 64–65
Left-sided pulmonary resections, 53–59
Left-sided resections, 73–75
Left subclavian-to-left coronary artery anastomosis, coronary artery anomalies and, CABG for, 1058
Left superior caval veins
anatomy of, 780
drainage to left atrium, 782
occlusion of, 780
Left superior pulmonary vein, view of, 65*f*
Left thoracoabdominal approach, to esophagectomy, 179
Left thoracotomy, 199, 771*f*, 772, 773*f*, 851*f*–854*f*
esophageal mobilization in, 163, 193*f*
Left-to-right shunt lesion, hemodynamic assessment, of patients, 747
Left upper lobectomy, surgical technique of, 53–58
Left upper lobe sleeve
resection, 73–75
view of, 75*f*
Left ventricular assist devices (LVADs), 532, 1057
preoperative evaluation for, 533–534
Left ventricular end-systolic volume index (LVESVI), 527
Left ventricular function, in normal hearts, 371*f*

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Left ventricular outflow tract obstruction (LVOTO), 869
aortic stenosis and, 871–881
Left ventricular oxygen requirements, of beating nonworking, fibrillating arrested hearts, 371*f*
Left ventricular reconstruction, 519–529
introduction to, 519
preoperative evaluation for, 523–524
LES. *See* Lower esophageal sphincter
Lesions
with lack of valvular tissue, pediatric mitral valve repair and, 1105–1106
Levoatrial cardinal vein
presence of, 782
LIMA. *See* Left internal mammary artery
Limited resections, 40
Linear stapler, wedge resection by, 82*f*
Lipomas, within pericardium, 666
Lobar lung transplantation, pediatric, 1091–1095, 1091*f*–1095*f*
Lobectomy, 109. *See also specific* lobectomy
with VATS, 31, 109
Long-segment jejunal interposition, 215–216
Lower esophageal sphincter (LES), 147, 209
Lower lobe bronchus, pulmonary artery and, retraction of, 59*f*
Lower lobe pulmonary artery, stump of, 51*f*
Low-gradient aortic stenosis, 463
Low superior vena cava/right atrium, PAPVR to, 1049–1051
Lung cancer
preoperative evaluation for, 41
TNM staging system, 15*t*
Lung Cancer Study Group (LCSG), 40
lobectomy study of, 79
Lung/heart–lung transplantation, pediatric, 1085–1097
rejection surveillance/management of, 1096
results of, 1096–1097
Lung implantation
ACT for, 236
preparation for, 234*f*
Lung transplantation, 231–240
anesthetic considerations in, 234
bronchial anastomosis and, 236*f*
bronchus division in, 236*f*
clamshell incision for, 235*f*
CPB and, 236
Duke University immunosuppression protocol, 239*t*
Duke University infection prophylaxis for, 234*t*
indications for, 231
left atrial anastomosis and, 237*f*
lung de-airing during, 237*f*
patient positioning for, 234–236
PGD and, 238
postoperative care for, 238–239
pulmonary artery anastomosis during, 237*f*
recipient procedure for, 234–239
reperfusion in, 237–238
skin incision for, 234–236
survival rates for, 231
LVADs. *See* Left ventricular assist devices
LVESVI. *See* Left ventricular end-systolic volume index
LVOTO. *See* Left ventricular outflow tract obstruction
LVRS. *See* Lung volume reduction surgery
Lymph node(s)
dissection of, 176–177

M

MAC. *See* Mycobacterium avian complex
Magnetic resonance imaging (MRI)
advantages/disadvantages of, 717*t*
for neurologic injury, 395
tumor staging, 307
Major aortopulmonary collaterals (MAPCAs), pulmonary atresia with ventricular septal defect and, 979–988
Malignant pleural mesothelioma (MPM)
diagnosis of, 297–298
invasive staging studies for, 298
multimodal approach to, 298–303
incision for, 299
introduction to, 298–299
lung-sparing procedure for, 299
positioning for, 299
operative techniques for, 297–298
pericardial reconstruction for, 302
with pleural effusion
diagnosis for, 297–298
palliation for, 297–298
with pleural rind, diagnosis for, 298
surgical management of, 296
treatment options for, 297
Malperfusion syndrome, acute type B aortic dissection and, 571*f*
MAPCAs. *See* Major aortopulmonary collaterals
Marfan's syndrome, 333, 338
Marfan syndrome, 593, 1106, 1119
Masaoka incision, superior sulcus tumor resection and, 325, 325*f*
Maximal voluntary ventilation (MVV), 41
Maximum intensity projection (MIP), 717, 718*f*
Maze procedure
for AF, 638–639, 642–643, 645
Cox Maze III modifications and, 639*f*–640*f*
indications for, 642, 644
outcomes for, 644–645
postoperative management for, 644–646
MDRTB. *See* Multidrug-resistant tuberculosis
Mechanical circulatory support (MCS), 533, 1157
Median sternotomy, 35–36
patient positioning for, 35*f*–36*f*
pericardiectomy via, 293, 294*f*
vascular rings requiring, 803
Mediastinal dissection, 404–405
Mediastinal lymph node dissection, 309–311
lefthemithorax and, 292*f*, 367
righthemithorax and, 109, 732*f*
survival of, 308
Mediastinoscopy, 15–20
cervical dissection in, 18*f*
extended cervical, 19
of lymph nodes, 14–20, 22*f*–25*f*
patient position for, 17*f*
technique of, 17–18
Mediastinotomy, anterior, 19–20, 24*f*–25*f*
Medtronic CoreValveReValving System, 482
Merendino. *See* Distal segmental esophagectomy
Metaiodobenzylguanidine (MIBG), 667
Metastases, diagnosis of, 20
Methylmethacrylate reconstruction, multilevel thoracic vertebrectomy/laminectomy and, 322*f*
Metzenbaum scissors, 17, 536
MG. *See* Myasthenia gravis
MIBG. *See* Metaiodobenzylguanidine
MIDCAB. *See* Minimally invasive direct coronary artery bypass
Middle lobe
bronchus, division of, 50*f*
sleeve resection, 73
MIE. *See* Minimally invasive esophagectomy
Mild right ventricular hypoplasia, initial neonatal procedures for, 966
Minimally Invasive Cardiac Surgery (MICS), 515
Minimally Invasive Direct Coronary Artery Bypass (MIDCAB), 494, 515
Minimally invasive esophageal procedures, 186–197
Minimally invasive esophagectomy (MIE), 190–197
completed cervical anastomosis, 196*f*
gastric tube creation and, 192*f*
gastric tube suturing for, 192*f*
laparoscopicpyloroplasty and, 191*f*
results of, 195–197
technique of, 194–195
thoracic esophageal mobilization and, 193*f*
thoracoscopic port placement for, 193*f*
through low transverse cervical incision, 195*f*
Minimally invasive mitral valve surgery, 419–420
patient selection for, 420
MIP. *See* Maximum intensity projection
Mitochondria, neural cell injury and, 391*f*
MitraClip system, 441. *See also* Percutaneous mitral valve repair
schematic view of, 442*f*
transesophageal echocardiographic image
three-dimensional, 443*f*
two-dimensional, 443*f*
Mitral regurgitation (MR), 440–441
Mitral valve 748
annuloplasty, 449–450, 450*f*
anterior/posterior segments of, 408*f*
disease, repair techniques and, 409–410
endocarditis of, 447
exposure, ischemic mitral regurgitation and, 435–436
repair, 407–417
anterior leaflet commissuroplasty for, 413*f*

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daVinciTM S robotic tele-manipulation system, 420*f*
development of techniques for, 407–408
double-orifice technique for, 416*f*
ischemic mitral regurgitation and, 431–438
pediatric, 1105–1115
principles of, 408–409
rheumatic, 408
robotic technology for, 419
variations on, 412–413
re-repair, 417
surgery, Cox Maze procedure and, 638
Mobilized gastric conduit, 191*f*
Modified Bernoulli theorem, ultrasound physics/applications and, 700
Modified Blalock-Taussig shunts
for congenital heart disease, 765–769, 767*t*
takedown of, with hemoclip/division technique, 768*f*
using PTFE graft, 767*f*
Modified cold blood cardioplegia maintenance solution, 372*t*
Modified Dartevelle. *See* Transclavicular approach
Modified thoracosternotomy. *See* Anterior cervicothoracic approach
Molecular markers, for neurologic injury, 394
Monitoring pressure, of cardioplegia, 374
Morphologic left atrium, 954
Morphologic left ventricle, 954
Morphologic right atrium, 780
Morphologic right ventricle, 954
MOTT. *See* Mycobacterium other than tuberculosis
Mountain lymph node map, 16*f,* 304
MPM. *See* Malignant pleural mesothelioma
MR. *See* Mitral regurgitation
MRI. *See* Magnetic resonance imaging
MTB. *See* Mycobacterium tuberculosis
Multidosecardioplegia, for myocardial protection, 373*f*
Multilevel thoracic vertebrectomy/laminectomy, methylmethacrylate reconstruction and, 322*f*
Muscle sparing. *See* Axillary thoracotomy
Muscular ventricular septal defects, 772, 834
MVR. *See* Mitral valve replacement
MVST. *See* Multivessel small thoracotomy
MVV. *See* Maximal voluntary ventilation
Myasthenia gravis (MG), thymectomy and, 134, 137, 199
Myocardial infarction complications
acute ischemic mitral regurgitation and, 548–551
free ventricular wall rupture and, 542–544
pump failure and, 551–554
surgery for, 542–554
ventricularseptal defect and, 544–548
Myocardial metabolic changes, with cardioplegic solution, 372*f*
Myocardial perfusion, cardiac magnetic resonance imaging of, 735
Myocardial protection, 370–378
antegrade/retrograde perfusion for, 371–372
during aortic root replacement, 377
beating heart, during systemic vascular resistance, 378*f*
cold blood cardioplegia for, 370
ischemic mitral regurgitation and, 434
during OPCAB, 394
strategies for, 370
ventricular restoration and, 377
warm blood cardioplegia for, 370
Myocardial tissue/blood tagging, with cardiac magnetic resonance imaging, 735
Myxomas, 664–666
operative approach to, 665*f,* 666

N

Neo-esophagus using linear stapler, 346*f*
NETT. *See* National Emphysema Treatment Trial
Neural cell injury
excitotoxicity mechanism of, 391*f*
mitochondria and, 391*f*
Neurogenic dumbbell tumor, resection of single-stage approach to, 141*f*
Neurologic complications, of CABG, 395
Neurologic injuries
assessing, 394–395
mechanism of, 390
molecular markers for, 394
MRI for, 395
neuropsychologic testing for, 394
spectrum of, 390
Neurologic injury prevention
acid–base management and, 393
after CAB surgery, 394
cerebral perfusion maintenance and, 391
embolization reduction and, 392
glucose control and, 393
hematocrit management, 393
intraoperative cerebral monitoring and, 394
off-pump coronary artery bypass and, 394
pharmacologic intervention for, 393
temperature management and, 392–393
Neuronal nitric oxide synthase (nNOS), 390
Neuropsychologic testing, for neurologic injury, 394
Neurotoxin, NO as, 390
New York State database, 382
Nissen fundoplication, 352*f*
suture placement for, 152*f*
Nitric oxide (NO), as neurotoxin, 390
NMDA receptor. *See N*-methyl -D-aspartate receptor
N-methyl -D-aspartate (NMDA) receptor activation of, 391*f*
NNE database. *See* Northern New England Cardiovascular Study Group database
nNOS. *See* Neuronal nitric oxide synthase
NO. *See* Nitric oxide (NO)
Noncommitted ventricular septal defect, 924, 930–932
DORV with, 924, 930–932
Nonseptic lung disease, 231
Non-small cell lung cancer (NSCLC), 60, 304
Northern New England Cardiovascular Study Group (NNE) database, 380, 382
NSCLC. *See* Non-small cell lung cancer
NTM. *See* Non-tuberculosis Mycobacterium
Nuss procedure, 335–336
illustration of, 335*f*

O

Octreotide, for chylothorax, 276, 277*t*
Off-pump coronary artery bypass surgery (OPCAB), 507–514
anesthesia for, 508
cardiac displacement for, 510–511, 510*f*
clinical outcome after, 512–513
contraindications for, 508*t*
coronary stabilization of, 511
coronary targets for, 510–511, 510*f*
future directions for, 513
grafting sequence for, 510
intraoperative management of, 508
myocardial protection during, 511–512
neurologic injury prevention and, 394
operative plan, 508
postoperative care for, 512
preoperative considerations, 507
prospective randomized studies on, 512
proximal anastomoses and, 512
Off-pump pulmonary valvotomy, with transannular right ventricular outflow tract patch, for neonates, 967
Off-pump transventricular pulmonary valvotomy
for neonates, 966
for pulmonary stenosis, 964
Omni catheter, 485–486
Omphalo-ischiopagus, 1183, 1184*f*
Omphalopagus, 1183, 1183*f*
"One-stage" approach, 515, 516*t*
One-stage complete unifocalization/intracardiac repair, for pulmonary atresia with ventricular septal defect, MAPCAs and, 980–985, 981*f*–988*f*
OPCAB. *See* Off-pump coronary artery bypass surgery
Open distal anastomosis technique, 595–596. *See also* Ascending aortic aneurysms
Open pulmonary valvotomy, for pulmonary stenosis, using cardiopulmonary bypass, 963–964
OPTN. *See* Organ Procurement and Transplantation Network (OPTN)
Organ preservation
for heart–lung transplantation, 613–615, 614*f,* 614*t*
for heart transplantation, 607–608
Organ procurement
for heart–lung transplantation, 629–631, 630*f,* 630*t*
for pediatric lung/heart–lung transplantation, 1088–1089
Organ Procurement and Transplantation Network (OPTN), 231

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Orthotopic heart transplantation, 624–626
alternative techniques for, 626, 626*f*
implantation, 624–626, 625*f*
operative preparation for recipient, 624
Outflow graft-aortic anastomosis, sewing of, 537*f*
Outlet defects, 829–830, 830*f*

P

Pacemaker in children
introduction to
biventricular pacing in pediatrics, 1171
common clinical indications for, 1168
epicardial vs. transvenous considerations, 1169
HRS/American College of Cardiology, recommendations/guidelines, 1168
lead extraction, 1171–1174
pacing for arrhythmias, 1170–1171
pacing in operated congenital heart disease, 1170
subpectoral vs. subcutaneous considerations, 1170
surgical considerations, 1171
PA/IS. *See* Pulmonary atresia with intact septum
Palmar–axillary hyperhidrosis, 329–331
Palmar–plantar hyperhidrosis, 330
Papillary fibroelastomas, 666–667
Papillary muscle-to-commissure fusion, pediatric mitral valve repair and, 1105–1106
PAPVR. *See* Partial anomalous pulmonary venous return
Parachute mitral valve, pediatric mitral valve repair and, 1105, 1114*f*
Paraesophageal hernia
characteristic anatomic defects, 341
with duodenum migration, 266*f*–268*f*
with gastroesophageal migration, 266*f*–268*f*
incidence of, 341
laparoscopic repair, 350–352
mortality rate, 341
surgical approaches
aparoscopic anti-reflux procedures, 342*f*
Bougie position, 345*f*
circular gastric staple line creation, 345*f*
Collis gastroplasty, 348*f*
crura, approximation of, 344–345
esophagogastric fat pad, mobilization of, 344*f*
esophagus assessment, 343–344
excision of, 344*f*
floppy Collis-Nissen fundoplication, 347*f*
fundoplication, 348–350
giant paraesophageal hernia, transthoracic repair, 345–347
herniated stomach, reduction of, 343*f*
laparoscopic wedge gastroplasty, 346*f*
neo-esophagus using linear stapler, 346*f*
operative technique, 347–350
preoperative evaluation, 342–345
standard port placement, 342*f*
symptoms of, 210, 341
transthoracic repair, 352–353
type I, 341
type II, 341
type III, 341
type IV, 341
view of, 265*f*
Parapagus, 1183
Paravertebral fascia, tumor on, 321*f*
Parietal pleura, 116, 141–142, 278, 321*f*
Partial anomalous pulmonary venous return (PAPVR)
definition of, 1049
diagnosis of, 1049
to high superior vena cava, 1051–1052
to low superior vena cava/right atrium, 1049–1051
pathologic anatomy of, 1049
pathophysiology of, 1049
results of, 1052
scimitar syndrome and, 1052
surgical management of, 1049–1052
Partial vascular rings, 803
PARTNER Trial Cohort A and Cohort A, clinical outcomes, 488*t*
Patch aortoplasty, aortic coarctation and, 852–853
Patent ductus arteriosus (PDA), 713, 788–794
diagnosis considerations for, 788
division of, 792*f*
surgical complications for, 793
VATS for, 792–793, 793*f*
Patient blood volume, in CPB, 365*t*
Patient evaluation, for carinal resection, 100
Patient safety, 396
accountability, 400
complexity, 397
high reliability, 399–400
human cognition and, 397–399
learning from others, 397
perception and decision-making, 398*t*
teamwork, 384
PDA. *See* Patent ductus arteriosus (PDA)
PDT. *See* Photodynamic therapy
PE. *See* Pulmonary embolism
Pectus excavatum and carinatum, corrections
clinical presentation, 333
complications, 338–340
early postoperative complications of primary repair, 339*t*
improvement in bar displacement rate, 339*f*
long-term results by length of time bar, 339*f*
post operative management, 338
preoperative evaluation, 333–334
surgical procedure, 334
Nuss procedure, 335–336
Ravitch procedure, 336–338
right-sided thoracoscopic view of, 336*f*
Pediatric abnormal aortic valve, pathology of, 1118–1120, 1120*f*
Pediatric aortic valve repair, 1118–1127
anatomy/function of, 1118
aortic valve stenosis and, 1118–1119
background on, 1118
dilated annulus techniques for, 1126
extended commissurotomy for, 1121, 1121*f*
results for, 1126–1127
surgical intervention timing for, 1120–1122, 1124–1126
surgical valvuloplasty techniques for, 1120–1121
valve regurgitation and, 1121–1122, 1124
Pediatric bilateral sequential lung transplantation, 1089
Pediatric cardiac repair/lung transplantation, for pulmonary atresia, 1091*f*
Pediatric heart–lung transplantation, 1088–1089
Pediatric heart transplantation, 1074–1082
background on, 1074
contraindications for, 1074–1075
donor assessment/management for, 1075
hypoplastic left heart syndromes and, 1076–1077, 1076*f*
indications for, 1074
operative techniques for, 1075–1076
post-transplant complications for, 1075
bleeding as, 1075
graft failure as, 1075
rejection as, 1075
situs inversus and, 1081–1082, 1082*f*
transplant for congenitally corrected transposition of great arteries, 1077–1078, 1078*f*
Pediatric lobar lung transplantation, 1091–1095
donor lobectomy technique of, 1091–1094, 1094*f*–1095*f*
recipient operation of, 1094–1095
Pediatric lung/heart–lung transplantation, 1085–1097
contraindications for, 1086
donor selection for, 1087
indications for, 1085–1086
operative procedure selection for, 1086–1087
organ procurement for, 1087–1088
postoperative management of, 1095–1096
recipient operation for, 1094–1095, 1069*f*
rejection surveillance/management of, 1096
results of, 1096–1097
Pediatric mitral valve repair, 1105–1114
accessory valve tissue and, 1105
according to functional classification, 1109–1112
acquired mitral valve disease and, 1106
anatomy of, 1105
background on, 1105
bypass technique for, 1108
cleft mitral valve and, 1105
congenital anomalies and, 1105
congenital mitral valves and, 1105
echocardiography for, 1107
embryology of, 1105
hammock valve and, 1106
indications for, 1107–1108
infective endocarditis and, 1106
introduction to, 1105

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lesions with lack of valvular tissue and, 1105–1106
papillary muscle-to-commissure fusion and, 1105–1106
parachute mitral valve, 1105
pathology of, 1105–1107
planning for, 1107–1108
regurgitant mitral valves, with congenital cardiac lesions and, 1106
results of, 1112–1113
rheumatic heart disease and, 1106
rheumatic mitral valves and, 1113
supravalvular rings resection and, accessory mitral valve tissue, 1111
surgical technique for, 1108–1112
type I malformation correction and, 1109, 1109*f*
type II malformation correction and, 1109–1110, 1112*f*
type III malformation correction and, 1110–1111, 1113*f*
valvular tags and, 1105
Pediatric single-lung transplantation, 1089
Pedicle muscle flaps, chest wall resections and, for reconstruction, 257*f*
PediMag® pump, 1160, 1160*f*. *See also* ECMO and ventricular assist devices
Peptic esophageal injury, 147
Percent postoperative predicted, 60
Percent postoperative predicted diffusion capacity of lung for carbon monoxide (popDLCO%), 60
Percutaneous mitral valve repair
EVEREST High Risk Registry, 443–444
EVEREST I and II trials, 441–443
historical perceptive
adapted from, 440
direct annular approach, 441
fluoroscopic image, 426*f*
indirect annular approach, 440–441
initial patent drawings, 442*f*
trans-septal approach, 441*f*
MitraClip system, 441
mitral regurgitation, 440
Pericardial effusions
etiology of, 289
pericardial procedures for, 289–292
Pericardial fat
wrapped around anastomosis, 74*f*
Pericardial procedures
for pericardial effusions, 289–292
Pericardial tamponade, physiology of, 289
Pericardial valve, 454, 968
Pericardiectomy, 293
via anterolateral thoracotomy, 294*f*
via median sternotomy, 294*f*
Pericardiocentesis, 290
Pericardium, anatomy of, 289
Perimembranous defects, 829
PGD. *See* Primary graft dysfunction
Phantom sweating, 331
Pharmacologic intervention, for neurologic injury prevention, 393
Phase-encoded velocity mapping, 718, 720, 721*f*, 733–736
Photodynamic therapy (PDT)
for esophageal cancer, 219
Phrenic nerve, diaphragm and, 262*f*
Pleural gases, 116
Pleural space, 116
anatomy, 116
physiology, 116
Pleural surfaces, anatomy of, 117*f*
Pneumonectomy, 60–68. *See also specific* pneumonectomy
arrhythmias after, 66
bronchopleural fistula and, 67–68
chylothorax and, 67
complication management for, 65–68
empyema and, 67–68
history of, 67
indications for, 60–61
pulmonary embolism and, 68
surgical technique for, 60
types of, 60
vocal cord dysfunction and, 65–66
Pneumothorax, 9, 19–20, 116, 329, 338
catamenial, 121
CXR for, 118–119
diagnosis of, 118–119
etiology of, 116–118
management of, 116–121
operative intervention for, 120*t*
operative management for, 120
pathophysiology of, 116–118
spontaneous, 116
treatment for, 119–120
Polyethylene terephthalate cuffs, catheter placement with, 281*f*
Polypropylene mesh
for lateral chest wall resection, 255, 256*f*
Polypropylene mesh/methylmethacrylate prosthesis, for lateral chest wall resection, 258*f*
popDLCO%. *See* Percent postoperative predicted diffusion capacity of lung for carbon monoxide
popFEV1%. *See* Percent postoperative predicted
Porous-diaphragm syndromes, 272
Port placement
for laparoscopic Nissen fundoplication, 148
for minimally invasive esophagectomy (MIE), 191*f*, 193*f*
for paraesophageal hernia repair, 342*f*
for posterior mediastinal tumor, resection of, 141
for transthoracic esophageal resection/reconstruction, 174, 175*f*
for video-assisted thoracic surgery (VATS), 108, 109*f*
Positron emission tomographic (PET) scans, 10–11, 14–16, 297–298, 318, 495
tumor staging and, 22, 40–42
Posterior leaflet
prolapse, 393–394, 393*f*, 394*f*, 406–407, 407*f*
Posterior mediastinal lesions
resection of, 139–145
surgical principles of, 139–141
surgical technique for, 141–145
Posterior mediastinum, anatomy, 139
Posterolateral thoracotomy, 33–34
landmarks for, 33
lateral decubitus position for, 43
Postoperative barium swallow status, after jejunal interposition, 170–171
Postpneumonectomy empyema, 67
Postpneumonectomy pulmonary edema (PPE), 66–67
Postpneumonectomy syndrome, 68
Potts anastomosis, 770, 753
takedown of, with PTFE patch, 772*f*
view of, 771*f*
Potts shunt, 770–771, 771*t*
PPE. *See* Postpneumonectomy pulmonary edema (PPE)
Pre-Fontan management, for single ventricle, 743
Pretracheal fascia, 103
incision of, 18*f*
oblique view of, 20*f*
Pretracheal tunnel, identification of, 21*f*
Primary graft dysfunction (PGD), 231, 238
in lung transplantation, 635
Primary hyperhidrosis, 328. *See also* Hyperhidrosis
Primary malignant cardiac neoplasms, 667–668
Primary sternal tumor, with intended resection lines, 258*f*
Priming volume per tube length/diameter, in CPB, 365*t*
"Prophylactic" aortic valve replacement, 464
Prostar sutures, 486
Prosthetic aortic valve, choice of, 461–462
Proximal anastomoses, OPCAB and, 510
Pseudo-coarctation, acute type B aortic dissection and, 571–572, 571*f*
Pulmonary arteries, 109
anterior aspect of fissure, 56*f*
anterior segmental branch of, 55*f*, 58*f*
coronary artery anomalies from, 1055–1065
identification of, 56*f*
retraction of, 310
Pulmonary artery anastomosis, 237*f*, 625, 627*f*, 627, 636, 764, 767, 767*f*, 957–958, 972, 990, 1010, 1014, 1026*f*, 1077–1079, 1078*f*, 1080*f*, 1082*f*
Pulmonary artery band, 771–775, 773*t*
for congenital heart disease, 709, 747, 755, 759, 759*f*, 764
intraluminal technique of, 775
placement of, 773*f*
sequential tightening of, 774*f*
takedown of
with PTFE patch, 772*f*, 774*f*
with pulmonary artery transaction, 775*f*
Pulmonary artery sling, 730*f*
anatomy of, 806
classification of, 803*t*
clinical presentation of, 806
diagnosis of, 808
surgical anatomy of, 807*f*–808*f*
surgical indications for, 808
surgical technique for, 808, 810

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Pulmonary atresia with intact septum (PA/IS), 964
anatomic considerations for, 965
definitive surgical treatment for
adjustable atrial septal defect and, 969–970, 969*f*
aorta-to-right ventricle shunt, 974
bidirectional cavopulmonary Glenn shunt, 971–972
enlargement of right ventricular cavity and, 968–969
extracardiac Fontan with adjustable fenestration, 973
lateral tunnel Fontan with adjustable atrial septal defect, 972–973
with mild right ventricular hypoplasia, 968–969
moderate right ventricular hypoplasia, 969–971
severe right ventricular hypoplasia, 971–975
tissue valve insertion and, 970
transannular pericardial patch with monocusp valve, 970–971
diagnostic considerations for, 948
initial surgical treatment for, 965–967
for neonates, 967
surgical results after, 975
Pulmonary atresia with ventricular septal defect, MAPCAs and, 962–971
aortopulmonary window for, 980*f*
closure criteria of ventricular septal defect of, 985
follow-up management for, 986
one-stage complete unifocalization/intracardiac repair for, 980–985, 983*f*, 985*f*
postoperative care for, 985–986
Pulmonary embolism (PE), 196, 239, 243, 565, 670
Pulmonary homografts, 454–455, 471, 474–477, 476*f*
Pulmonary resections. *See also specific* pulmonary resections
history of, 40
introduction to, 40–43
postoperative mortality/morbidity of, 50, 52
right-sided, 43
Pulmonary stenosis
diagnostic considerations for, 963
incidence of, 963
off-pump transventricular pulmonary valvotomy for, 964
open pulmonary valvotomy, using cardiopulmonary bypass for, 963
perioperative management for, 964
postoperative care for, 964
surgical complications for, 964
surgical techniques for, 963
surgical treatment for, 963
Pulmonary thromboembolism (PTE), 68, 677–678
Pulmonary valve
hemodynamic assessment of, 747
Pulmonary valve replacement (PVR)
indications and time, 453–454
postoperative outcomes, 456
PV dysfunction pathophysiology factors influence, 453
PV prostheses, selection of
mechanical pulmonary valves, 454
percutaneous PV replacement, 455
pericardial valve, 454
pulmonary homografts, 454–455
surgical technique, 455
tetrology of Fallot (TOF), 453
Pulmonary vascular resistance, VSDs and, 829
Pulmonary veins
cardiac magnetic resonance imaging of, 723
systemic venous connection, 780
Pulmonic valve stenosis/atresia, congenital heart disease and, 749–750
Pump failure, 542
clinical presentation of, 552
diagnosis of, 552
incidence of, 551–552
natural history of, 552
operative technique of, 553–554
operative timing of, 552–553
pathogenesis of, 551–552
preoperative treatment of, 552–553
survival of, 553–554
Pygopagus, 1183
Pyloric drainage, procedure for, 177–178

R

Rachipagus, 1183
Radiation, for esophageal cancer, 219
RAO. *See* Right anterior oblique view
Rastelli operation, on TGA, 944, 946–947, 948*f*, 952*f*
Ravitch procedure, 333, 336–338
modified, illustration of, 337*f*
Receiver operating characteristic (ROC) curve, 381
Recurrent laryngeal nerve injury, 66, 71, 190, 236
Recurrent primary spontaneous pneumothoraces, apical resection for, of subpleural bleb, 117*f*
Re-do sternotomy, orthotopic heart transplantation and, 626
Regurgitant mitral valves, with congenital cardiac lesions, pediatric mitral valve repair and, 1106
Renal cell carcinomas, 668–669
Reoperative cardiac surgery
cannulation, 404–405
cardiac reoperation, 402
cardiopulmonary bypass, 404–405
catheterization, 402–403
computed tomography angiography, 403*f*
incidence and outcomes, 402
incision and discretion, 404
intraoperative preparation, 404
patient selection and assessment, 386
protocol-driven approach, 405
radiographic evaluation, 403–404
Reperfusion, in lung transplantation, 237–238
Residual volume (RV), 41
Resistance calculation, 746
Retransplantation, heart–lung transplantation and, 636
Retroaortic innominate vein, 783
Retrograde cannulas
for cardioplegia, 373, 374*f*
Reverse elephant trunk technique, for arch aneurysms, 604
Rhabdomyoma, 666
Rheumatic heart disease, pediatric mitral valve repair and, 1106
Rheumatic mitral valve repair, 393
pediatric, 1113
Right anterior oblique view (RAO), 702
Right aortic arch, with retroesophageal ligament, 727–728, 797–798
Right inferior pulmonary vein, view of, 62*f*, 97*f*, 179
Right lower lobectomy, 48–49, 109
Right lower lobectomy intercostal muscle on, 62*f*
Right mainstem bronchus intermedius during right upper lobe sleeve, 74*f*
Right pulmonary hilum, view of, 42*f*
Right-sided pneumonectomy, surgical technique for, 61–64
Right-sided pulmonary resections, 40–52
introduction to, 43
surgical technique for, 43–49
Right side sympathicotomy, intraoperative picture, 330*f*
Right stellate ganglion, intraoperative picture, 329*f*
Right superior caval veins
azygos continuation of, 783–784, 784*f*
to left pulmonary atrium, 782–783, 782*f*
Right thoracotomy, 18, 95, 98–99, 173
esophagectomy incisions for, 179*f*
vascular rings requiring, 803
Right-to-left shunt lesion, hemodynamic assessment, of patients, 747
Right transcervical incision, superior sulcus tumor resection and, 323*f*
Right upper lobe bronchus
division of, 48*f*
tumor in, 73*f*
Right upper lobectomy
sleeve resection, 72
Right upper lobe sleeve, for left-sided endotracheal tube, 73*f*
Right ventricular assist devices (RVADs), 532
Robotic arms, 114*f*
Robotic mitral valve surgery, 421
anesthesia for, 421
clinical outcomes of, 421
monitoring, 421
operative techniques, 421
Robotic technology, for mitral valve repair, 419
Root replacement, Ross procedure and, 1131

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Ross procedure, 471–478, 874–875
for aortic valve replacement, 874–875
RV. *See* Residual volume
RVADs. *See* Right ventricular assist devices

S

Safe circulatory arrest times, hypothermia level and, 366*t*
SAM. *See* Systolic anterior motion
SARS. *See* Severe acute respiratory syndrome
Scalene biopsy, 20
Scimitar syndrome
PAPVR and, 1052
surgical repair of, 819
Scleroderma
definition of, 210–211
diagnosis of, 211
etiology of, 211
Secondary hyperhidrosis, 328. *See also* Hyperhidrosis
Secundum-type atrial septal defects
Amplatzer repair of, 820–821
surgical repair of, 819–820
Sedation, conscious, 8, 10
Segmental valve analysis, for type IIIb, ischemic mitral regurgitation, 419
Segmentectomy, 80–82
Semidorsal/semiventral incision view, of superior sulcus tumor resection, 323*f*
Semilunar valve stenosis, hemodynamic assessment of, or insufficiency, 747
Septostomy procedures, for congenital heart disease, 748
Severe acute respiratory syndrome (SARS0), 118
Severe aortic stenosis (AS)
echocardiographic criteria for, 480*t*
Severe right ventricular hypoplasia, initial neonatal procedures for, 971–975
Shaded surface display (SSD), 717
Shunt calculation, 746
"Siamese twins," 1182
Simultaneous antegrade/retrograde cardioplegic perfusion, delivery/monitoring of, 372–374
Single-clamp technique (SCT), 392
Single-lung transplants (SLT), 231
Single venous cannulation, 363*f*
Single ventricle
anatomy of, 1020
tricuspid atresia, 1020–1029
Situs inversus, pediatric heart transplantation and, 1081, 1082*f*
Sleeve lobectomy, 76, 76*t*
Sleeve resection, 72*f*, 73
SLT. *See* Single-lung transplants
Small aortic annulus, 464–466
Society of Th oracic Surgery (STS) PROM score, 479
Somatostatin, for chylothorax, 276, 277*t*
Spontaneous pneumothorax, 117
SSD. *See* Shaded surface display
SSFP. *See* Steady-state free-precession
Steady-state free-precession (SSFP), 716, 718*f*
Stellate ganglion injury, 329*f*, 330
Sternal resections
for chest wall resections, 322
reconstruction of, 258*f*
technique for, 257
Sternotomy. *See also* Thymectomy
anterior mediastinotomy and, 36
median, 35–36, 36*f*
thymectomy via, 128
Streptokinase, for empyema, 315
Structural valve deterioration (SVD), 461–462
STS database. *See* Society of Thoracic Surgeons (STS) database
Subaortic ventricular septal defect
DORV repair, with pulmonary stenosis, 924
double-outlet right ventricles and, 924–933
tunnel repair of, DORV, 925–926, 926*f*
Subclavian artery dissection
Subclavian artery division, 324*f*
superior sulcus tumor resection and, 324
from tumor, 322*f*
Subclavian flap repair, for aortic coarctation, 851–852
Subclavian vein dissection, superior sulcus tumor resection and, 323–324
Subclavian vessel dissection, for superior sulcus tumor resection, 322
Sublobar pulmonary resections, 79–84
Subpleural bleb, of recurrent primary spontaneous pneumothoraces, apical resection for, 117*f*
Subpulmonary obstruction repair, for ccTGA, 956–957
Subpulmonary ventricular septal defect
DORV with, anatomic repair of, 927–928, 928*f*
double-outlet right ventricles and, 924–933
Subxiphoid pericardial window, 290–291, 291*f*
Superior caval system, aneurysmal dilation of, 783
Superior caval veins (SVCs)
anomalies of, 780–784
isolated left, 781–782
left, 782
Superior mediastinum, 43, 53
Superior pulmonary arterial branch, dissection of, 235*f*
Superior pulmonary artery, left pulmonary artery and, 236*f*
Superior sulcus tumor
anatomical considerations for, 318
anterior approaches for, 323–324
anterior scalene muscle insertion and, 324*f*
brachial plexus dissection for, 321 322
chest wall closure and, 325
chest wall resections for, 320–321, 324–325
clinical considerations for, 318
diagnostic considerations for, 318–319
hemiclamshell incision and, 324, 324*f*
indications for, 319–320
investigations for, 319
Masaoka incision and, 325, 325*f*
operative technique for, 320–325
perinduction MRI of, 319*f*
perioperative patient management for, 320
posterolateral approach of, 320–323
postoperative care and, 325
postoperative complications and, 325
pulmonary resection, 322–323
pulmonary resection and, 325
resection, 318–326
right transcervical incision and, 323*f*
semidorsal/semiventral incision view of, 323*f*
subclavian artery dissection and, 324
subclavian artery division and, 324*f*
subclavian vein dissection and, 323–324
subclavian vessel dissection for, 322
transclavicular approach to, 323–325
vertebral body resection for, 322
Superior-type sinus venous atrial septal defect, surgical repair of, 821–823
Supravalvar aortic stenosis, 878–881
Supraventricular tachycardia (SVT), 66
Surgical treatment of hypertrophic cardiomyopathy
anatomic subtypes, 656*f*
clinical presentation and natural history, 654–656
indications for operation, 655–656
intracardiac pressures, 658*f*
left ventricular pressure–volume loops, 661*f*
oblique aortotomy, 657*f*
operative risk of isolated septal myectomy, 659*f*
outcomes of myectomy, 657–660
overall survival of patients, 660*f*
pathology and pathophysiology, 653–654
risk of
hypertrophic cardiomyopathy-related death, 655*f*
progression to severe symptoms, 655*f*
septal excision, 657*f*
special considerations, 660–661
survival free from all-cause mortality, 659*f*
techniques, 656–657
transapical ventriculotomy, 661*f*
SVCs. *See* Superior caval veins
SVD. *See* Structural valve deterioration
SVR. *See* Surgical ventricular reconstruction
SVT. *See* Supraventricular tachycardia
Sympathicotomy. *See* Thoracic sympathectomy
Synthetic material choices, for chest wall resections, 255–256
Systemic pulmonary veins, congenital heart disease and, 755–756
Systemic vascular resistance, myocardial protection during, beating heart, 378*f*
Systemic veins, cardiac magnetic resonance imaging of, 732
Systemic venous atrium, bilateral superior caval veins drainage to, 780–781
Systemic venous connection
to pulmonary veins, 784
total anomalous, 784–785
Systemic venous drainage
anomalies of, 780–787
Systolic anterior motion, 314–417

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T

Table-mounted retractor, for transhiatal esophagectomy, 158
Tachycardia-induced cardiomyopathy, 642, 644, 648
Takeuchi repair, modified, for coronary artery anomalies, 1061–1062
TAPVR. *See* Total anomalous pulmonary venous return
Target vessel exposure in, 510
TCD. *See* Transcranial Doppler
TECAB. *See* Totally endoscopic coronary artery bypass
TEE. *See* Transesophageal echocardiography
Tetralogy of Fallot (TOF), 453, 643, 690, 697, 707, 710, 711*f*, 718, 721–722, 724. *See also* Pulmonary valve replacement (PVR)
  with absent pulmonary valve, 710, 730, 889
  background on, 990
  incidence of, 990
TGA. *See* Transposition of great arteries
Thoracic cage defect, 1190
Thoracic diverticula, 226–229
  clinical presentation of, 226
  diagnosis of, 226
  esophageal mobilization for, 227*f*
  postoperative care for, 229
  surgical complications for, 228
  surgical indications for, 227–228
  surgical procedure for, 227–229
Thoracic duct
  anatomy of, 274
  mediastinal structures and, 278*f*
  physiology of, 274
  thoracoscopic approach to, 278*f*
Thoracic ectopia cordis, 1190
Thoracic esophageal mobilization, MIE and, 193*f*
Thoracic incisions, 178
Thoracic outlet syndromes, 242–243
Thoracic sympathectomy
  anatomy, 328
  hyperhidrosis patient
    nonsurgical management, 328–329
    preoperative evaluation, 328
  indications, 331
  surgical management, 329
    complications and side effects, 330–331
    surgical technique, 330
Thoracic sympathetic chain, 328
  anatomic drawing of, 329*f*
Thoracic/thoracoabdominal aneurysms, 575–587
  anesthesia/monitoring for, 577
  aortic graft selection and, 581
  Crawford classification of, 575, 576*f*
  diagnostic considerations of, 575–576
  hemodynamic instability control, during aortic cross-clamping, 578–579
  incision/operative exposure in, 577–578
  operation completion for, 582
  spinal cord preservation and, 579–580
Thoracoabdominal incision, 38, 38*f*, 174, 600
Thoraco-omphalopagus, 1183, 1184*f*
Thoracopagus, 1183, 1183*f*
Thoracosternotomy, 36–37, 1088–1089
  incision for, 38*f*
Thoracotomy
  anterior, 35–36, 35*f*
  for chylothorax, 277*f*
  left, 199, 227
  posterolateral, 33–35, 141
  right, 18, 90, 95
Thymectomy, 30–34, 134–138
  dissection in, 134
  introduction to, 126
  MG and, 126
  postoperative recovery of, 97, 128
  preoperative preparation for, 134
  transcervica, 134–138
Thymus gland
  inferior/superior horns of, 34
Tissue transfer options, for chest wall resections, 256–257, 256*t*
TLC. *See* Total lung capacity
TNM staging system, 14
  for lung cancer, 15
Toradol. *See* Ketorolac
Total anomalous pulmonary venous return (TAPVR)
  anatomic defects of, 1041–1042
  angiocardiography for, 1026
  cardiac catheterization for, 1026
  chest X-ray for, 1043, 1044*f*
  classification of, 1041–1042
  diagnosis of, 1043–1044
  echocardiography for, 1043–1044
  embryology of, 1041
  extracorporeal circulation management for
    cardiac type, 1047
    infracardiac type, 1048
    mixed type, 1048
    supracardiac type, 1045–1046
  historical aspects of, 1041
  pathophysiology of, 1042–1043
  postoperative management for, 1048
  preoperative management for, 1044–1045
  results for, 1048–1049
  surgical intervention timing for, 1044–1045
  surgical technique for, 1045, 1047–1048
Total anomalous systemic venous connection, 784–785
Total lung capacity (TLC), 41
Totally Endoscopic Coronary Artery Bypass Graft (TECAB), 516
Trabecular defects, 830
Trachea
  anatomy of, 85
  bronchoscopic view of, 3*f*
Tracheal compression, anomalous innominate artery with, 804–805
Tracheal lesions
  clinical presentation of, 86–87
  diagnostic studies of, 86–87
  etiology of, 85–86
Tracheal reconstruction
  anastomotic complications of, 97–98
  postoperative management of, 96–97
Tracheal resection
  anastomotic complications of, 97–98
  hemorrhage and, 93*f*
  postoperative management for, 96–97
Tracheobronchial angle, exposure of, 57*f*
Tracheobronchial tree, 2, 4*f*
Transapical ventriculotomy, 661, 661*f*
Transatrial closure, of VSDs, 833*f*
Transaxillary approach, dorsal sympathetic chain, stellate ganglion and, 320
Transcatheter aortic valve replacement, 385, 479–491
Transcatheter aortic valve replacement (TAVR)
  annulus sizing criteria, 481*t*
  current transcatheter heart valve technology, 481–482
  Frailty Index, 480, 480*t*
  pathophysiology, 479
  patient selection and screening, 479–480
  preoperative evaluation, 480–481
  risk stratification, 479–480
  surgical technique
    anesthesia, 482
    heart valve team approach, 482
    preparations, 482–483
  transfemoral approach
    balloon aortic valvuloplasty, 484–485, 487
    clinical outcomes, 487–489
    closure, 487
    PARTNER Trial Cohort A clinical outcomes, 488*t*
    representative transesophageal images, 469*f*
    transapical and vascular access, 486–487
    transapical approach, 486
    valve deployment, 485, 487
    valve positioning and deployment., 468*t*
    vascular access, 483–484
    vascular access site closure, 485–486
Transcatheter heart valve (THV) replacement, 479
Transcatheter therapy
  for congenital heart disease, 748–760
  hemodynamic assessment and, for congenital heart disease, 746–760
Transcervical thymectomy (TCT), 134–138
Transclavicular approach, 323–325
Transcranial Doppler (TCD), 394, 584, 802
Transdiaphragmatic fenestration, diaphragmatic surface and, 118*f*
Transesophageal echocardiography (TEE), 234, 367, 375, 392, 412, 416, 434, 447, 455, 475, 481, 535, 550–551, 577, 612, 713–714, 873–875, 878, 973
Transhiatal esophagectomy, 157–171
  cervical phase of, 160–161
  introduction to, 157
  operative technique for, 157–168
  postoperative care for, 169–170

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postoperative complications for, 169–170
preoperative assessment for, 156, 169
preparation for, 157–158
results of, 168–170
Transplant for congenitally corrected transposition of great arteries
pediatric heart transplantation and, 1074–1082
Transposition of great arteries (TGA), 710, 861, 866
anatomy of, 937–938
arterial repair and, 940–944
atrial repair, 940
background on, 937
clinical features of, 939
management of, 939
operative intervention of, 940
palliative operations of, 940
pathophysiology of, 938
Rastelli operation on, 944, 946–947
surgical correction for, 940–944
surgical repair history, 939–940
surgical repair results, 930–936
Transthoracic esophageal resection/reconstruction principles of, 173–176
surgical technique for, 173
Transthoracic Nissen fundoplication, 351*f*
Transthoracic partial fundoplication, 153–154
Transthoracic repair, 352–353
Transthoracic total fundoplication, 152–153
Transventricular closure, of conoventricular defect, 834*f*
Tricuspid atresia, single ventricle and, 708–709
Tricuspid regurgitation evaluation/operative indications, 446–447
functional, 446–447
Tricuspid valve, 447–452
anatomy of, 447–448
annuloplasty, 449–450
carcinoid heart disease, 446–447
Ebstein's anomaly, 447, 448*f*
introduction to, 447
operative setup for, 448
repair, for Ebstein's anomaly, 451–452
replacement, 450–451
Tricuspid valve replacement, pediatric, operative technique for, 1102
Truncal valve repair/replacement, truncus arteriosus with, 917–918
Truncus arteriosus, 711
alternative repair, 914–915
background on, 910
diagnosis of, 911
interrupted aortic arch with, 915–917
pathophysiology of, 910–911
postoperative care for, 919, 921
surgical technique for, 911–919
truncal valve repair/replacement with, 900–902
van Praagh classification, 894*t*
Tube graft placement, 581
Tumor(s). *See also specific* cancers; *specific* neoplasms*; specific* tumors
cardiac, 664–669
of diaphragm, 272
on paravertebral fascia, 321
primary, 43
subclavian artery dissection from, 322
Tunnel repair, of DORV, with subaortic ventricular septal defect, 925–926
"Two-staged" approach, 515, 516*t*
Type II paraesophageal hernias, 341
Type III paraesophageal hernias, 341
Type IV paraesophageal hernias, 341

U

Ultrasound frequencies, ultrasound physics/applications and, 700
Ultrasound physics/applications
Doppler principle and, 700–701
modified Bernoulli theorem and, 701
principles of, 700–701
ultrasound frequencies and, 700
Undersized remodeling ring annuloplasty, for type IIIb, ischemic mitral regurgitation, 435–436
Unidirectional cavopulmonary shunts, 1008–1009, 1008*f*, 1009*f*

V

VA database. *See* Veterans Affairs database
VADs. *See* Ventricular assist devices
Valsalva sinus aneurysm 886–888
clinical presentation of, 886
morphologic considerations of, 886
surgical indications for, 886–887
surgical results for, 887–888
surgical technique for, 886–887
Valvar aortic stenosis
in neonate/infant, 871–875, 872*f*
in older child, 872–873
Valve regurgitation, pediatric aortic valve repair and, 1119–1120
Valvular tags, pediatric mitral valve repair and, 1105
Van Praagh classification, for truncus arteriosus, 711–712
Vascular connections, closure procedures for, 756–758
Vascular rings
classification of, 797–810, 803*t*
complete, 797–804
anatomy of, 797
embryonic development of, 799–800
requiring median sternotomy, 803
requiring right thoracotomy, 803
VATS. *See* Video-assisted thoracic surgery
Velocity mapping, with cardiac magnetic resonance imaging, 703*f*, 721*f*
Venae cavae, 693–694
Venous connections
cardiac magnetic resonance imaging and, preoperative/postoperative, 732–733, 732*f*
Venous embryology, schematic of, 781*f*
Venous sinus valves, anomalies of, 785–786
Ventricular assist devices (VADs), 532–540, 1157–1165
background on, 532–533
patient selection for, 533–534
postoperative care for 540
potential complications for, 539–540
preoperative evaluation for, 533–534
surgical procedure for, 535–538
Ventricular septal defects (VSDs), 829–836
aortic coarctation and, 857
cardiac/noncardiac pathologic conditions associated with, 831
characteristics of, 830
classification of, 829–830
clinical presentation of, 544
closure, for ccTGA, 955–956
common incisions for, 832*f*
infundibular septum and, 710*f*
myocardial infarction complications, 544–548
natural history of, 544
operative technique for, 545–548
operative timing of, 544–545
pathogenesis of, 544
patient characteristics of, 830
pulmonary vascular resistance and, 830–831
surgical results for, 947–953
survival of, 548
transatrial closure of, 833
view of, 840*f*
Ventricular septum, 694–698, 697*f*, 707–708
Ventricular tachycardia (VT), 66
Ventriculoarterial alignments, 694*f*
Vertebral body resection
of superior sulcus tumor, 322
for superior sulcus tumor resection, 321*f*, 322, 322*f*
Veterans Affairs (VA) databas, 380
Viacor's PTMA coronary sinus device, 441. *See also* Percutaneous mitral valve repair
Vicodin. *See* Hydrocodone
Vicryl suture, 104, 170*f*
Video-assisted thoracic surgery (VATS), 108, 110, 111
Video-assisted thoracoscopic pulmonary resections, 108–114
contraindications for, 108, 110
indications for, 108, 112
morbidity/mortality after, 110
outcomes of, 110
wedge resection with, 109–110
Vigorous achalasia, 199, 202*f*
Vocal cord dysfunction, pneumonectomy and, 65–66
Volume-rendered object (VRT), 717
VRT. *See* Volume-rendered object
VSDs. *See* Ventricular septal defects
VT. *See* Ventricular tachycardia

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W

Warm blood cardioplegia
  for myocardial protection, 370
  solution contents, 373*f*
Waterston anastomosis
  construction of, 770*f*
  preparation of, 769*f*
  takedown of, with cardiopulmonary bypass/PTFE patch, 770*f*
Waterston/Cooley shunt, for congenital heart disease, 769–770, 769*t*, 769*f*, 770*f*
Wedge resection
  of apical blebs, 121*f*
  using linear stapler, 82*f*
  with VATS, 82

X

Xiphoid process, 337
Xiphopagus, 1183, 1184*f*

Z

Zenker's diverticulum, 222–226, 223*f*, 224*f*
  barium esophagram of, 223*f*
  clinical presentation of, 222
  diagnosis of, 222
  endoscopic surgical technique for, 223–224
  preoperative management of, 223
  surgical results for, 225–226

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